AMYOTROPHIC LATERALS SCLEROSIS (ALS)

Etiology
- heterogeneous etiology
- most are sporadic
- 5-10% are hereditary, both dominant and recessive inheritance has been described:
  - one hereditary form is linked to chromosome 2q33-q35, another is Cu/Zn superoxide dismutase (SOD1) mutation on chromosome 21q22
- degeneration of both upper and lower motor neurons

Clinical features
- incidence around 2/100,000
- usually onset in the older age groups around 60 years but may present earlier
- males>females
- painless weakness
- upper limb muscles affected more frequently than lower limb muscles
- bulbar muscles may be involved; slurring of speech, dysphagia,
- extracervical muscles and anal or urethral sphincters are rarely clinically affected
- upper motor neuron signs, increased tendon reflexes and positive Babinski sign, often present
- average survival after diagnosis 3 years, in patients presenting with bulbar symptoms shorter survival

Strategy
- confirm neurogenic EMG findings in muscles in three regions of the body (the regions are (1) brainstem, (2) brachial, (3) thoracic, (4) trunk and (5) crural
- exclude polyneuropathy
- differentiate from multifocal motor neuropathy with conduction block (MMN)
- differentiate from spinal hereditary motor neuronopathies (spinal muscular atrophies)
- differentiate from previous polio

Diagnostic criteria
- The diagnosis of ALS requires the presence of: (1) lower motor neuron signs (LMS), (2) upper motor neuron signs (UMS) and (3) progression of the disorder
  1. definite ALS: LMS in three regions and UMS
  2. probable ALS: LMS in two regions and UMS
  3. possible ALS: LMS in one region and UMS
  4. suspected ALS: LMS in two or three regions

Expected abnormal findings

EMG
- subacute neurogenic muscle findings in at least 3 regions. Preferentially the findings should be asymmetric without any definite proximal or distal predominance.
- Neurography
  - MCS often show reduced amplitudes
  - If the AMP is reduced significantly the conduction velocity may be reduced (loss of fastest conducting axons)
- Central motor conduction time
  - often abnormal

Expected normal findings

Neurography
- SCS: if AMP is normal or only moderately reduced, CV should be normal
- no motor conduction block
- unusual number of F-waves in mildly affected muscles (indicating lack of proximal conduction block)

Evoked potentials
- VEP
- BAEP
- SEP (may rarely be abnormal)

Procedure

EMG (if normal findings on one side examine both sides!)
- m.orbicularis oris/m.masseter/m.genioglossus (insert electrode below chin)
- m.trapezius
- interosseus dorsalis I / m.biceps brachii / m.deltoides
- m.tibialis anterior / m.gastrocnemius caput mediale / m.vastus lateralis
- if the patient presents with dysarthria and there are no other EMG abnormalities, test m.chricothyreoideus

Neurography MCS (bilaterally)
- n.medianus
- n.lateralis (also including supraclavicular stimulation)
- n.peroneus
- n.tibialis

Neurography SCS (bilaterally)
- n.suralis
- n.radialis

References
Polio, Acute

Etiology
- following infection by poliovirus most patients will have fever and gastrointestinal symptoms, but 1-2 % develop paralysis
- due to vaccination polio has been eradicated from developed countries
- attenuated virus used for vaccination may cause paralysis, estimated risk is 1 per million
- sometimes coxackie and echo virus may cause a similar paralysis

Clinical features
- around 1-2 weeks after an episode of febrile illness 1-2 % of patients with a polio virus infection will develop a paralytic disorder
- the risk of paralysis increases with age, adults are about 10 times as likely to develop paralysis than children
- any group of motor neurons may be affected, the distribution of weakness varies
- after the initial paralysis a varying degree of restitution takes place
- some patients with polio perceive after a stable period of more than 15 years new loss of muscle function (often called post-polio syndrome, see Polio, sequale following previous infection

Strategy
- show acute neurogenic involvement of several muscles usually in several regions of the body
- assess distribution of abnormalities and degree of involvement
- differentiate from polyradiculitis

Expected abnormal findings
EMG
- acute neurogenic EMG findings in muscles
- distribution not confined to one segment or nerve; if local - reconsider diagnosis

Neurography
- MCS: reduced amplitudes in weak muscles
- if the amplitude is severely reduced the MCV may be reduced

Expected normal findings
Neurography
- SCS

Procedure
EMG
- weak muscles should be studied

Neurography
The following motor nerves should be studied bilaterally
- n.medianus
- n.ulnaris
- n.peroneus
- n.tibialis
The following sensory nerves should be tested
- n.suralis
- n.radialis

Note
- although vaccination has largely eradicated acute polio from countries with a good vaccination program, polio may still occur in some parts of the world. In industrialized countries there are minorities that are negative to vaccinations and refugees that may still lack vaccination against polio.
POLIO, SEQUELA FOLLOWING PREVIOUS PARALYSIS

Etiology
- previously suffered from polio, the patient has recovered from the acute episode to various degrees
- the causes of sequela vary considerably: (1) normal aging with loss of contractile strength contributes loss of motor units with aging, (2) pain from joints and tendons and (3) psycho-social factors
- there are no well defined biological parameters that define this disorder

Clinical features
- after a stable period of more than 15 years after the initial paralysis some patients develop subjective new loss of muscle function (often called post-polio syndrome)
- the distribution of weakness varies considerably and depends on the distribution and severity of the initial affection

Strategy
- show inactive neurogenic involvement of several muscles usually in several regions of the body
- make sure the polio diagnosis is correct
- assess distribution of abnormalities and degree of involvement
- beware of additional diseases: radiculopathy, CTS, polynuropathy, polymyositis

Expected abnormal findings
EMG
- inactive neurogenic findings in weak and often also in strong muscles depending on involvement
- distribution not confined to one segment or nerve (if findings are focal - reconsider diagnosis)

Neurography
- MCS may show reduced amplitudes and if the amplitude is severely reduced the MCV may be reduced

Expected normal findings
Neurography
- SCS

Procedure
- the muscles studied depends entirely on the clinical symptoms...
- if the diagnosis is obvious, neurography is not necessary, unless the patient has a clinical problem that warrants neurography.
- Macro EMG for quantitation of motor unit size
- motor unit counting for quantification of number of motor units

Note
- neurophysiological findings or other laboratory findings do not differentiate between patients that are stable and those that experience new loss of muscle function (post-polio syndrome)

References
- Rodríguez AA, Agre JC. Correlation of motor units with strength and spectral characteristics in polio survivors and controls. Muscle Nerve. 1991;14:429-34
- Stålberg E, Grimby G: Dynamic electromyography and biopsy changes in a 4 year follow up: study of patients with history of polio. Muscle Nerve 1995; 699-707
- Wiechers DO, Hubbell SL: Late changes in the motor unit after acute poliomyelitis. Muscle Nerve 1981; 4:524-528

SPINAL MUSCULAR ATROPHY 1, WERDNIG-HOFFMAN

Etiology
- autosomal recessive inheritance
- genetic defect localized to chromosome 5q11-q13
- deletion in a region that codes the SMN (the survival motor neuron gene) and the NAIP (neuronal apoptosis inhibitory protein)
- good correlation between the size of the deletion and the severity of the phenotype
- incidence of deletions of both SMN and NAIP is 62% in SMA type I, 8.8% in type II and 12% in type III
- degeneration of the anterior horn cells in the spinal cord

Clinical features
- second most common lethal autosomal recessive disorder
onset in utero or before the age of 3 months
hypotonia and weakness, often the child is in a frog posture
respiratory problems, diaphragmatic breathing, costal recession
absent tendon reflexes
intellectually normal
poor prognosis; most die within the first year
CK normal or moderately elevated
Incidence is around 1/10000 (1/67000-1/25000 has been reported)
carrier rate of the gene in the general population is around 1/80

Strategy
* demonstrate acute or subacute neurogenic EMG findings of limb muscles
* demonstrate normal sensory neurography

Expected abnormal findings
EMG
* acute or subacute neurogenic EMG findings in several muscles (proximal muscles tend to be more involved than distal muscles)

Neurography
* MCS: reduced AMPL, CV may be reduced if AMPL is very low

Expected normal findings
Neurography
* SCS

Procedure
EMG (unilaterally)
* m.deltoideus/m.biceps brachii
* m.interosseus dorsalis
* m.vastus lateralis
* m.tibialis anterior

Neurography
* one motor and sensory nerve in the upper and lower extremities

Note
* sometimes the muscles shows typical continuous involuntary motor unit activity

References
* Ignatius J. The natural history of severe spinal muscular atrophy - further evidence for clinical subtypes (letter). Neuromusc Disord 1994;4:527-8
* Thomas NH, Dubowitz V. The natural history of type I (severe) spinal muscular atrophy. Neuromusc Disord 1994;4:497-502
* Werdnig G. Die frühkindliche progressive spinal Amyotrophie. Archive für Psychiatrie und Nervenheilkunde 1894;1894:706-744

Modified
* 26.3.1997 BF, ES 3.4.97, 8.5.1997 BF

SPINAL MUSCULAR ATROPHY 2, INTERMEDIATE

Etiology
* autosomal recessive inheritance
* genetic defect localized to chromosome 5q11-q13
* deletion in a region that codes the SMN (the survival motor neuron gene) and the NAIP (neuronal apoptosis inhibitory protein).
* degeneration of the anterior horn cells in the spinal cord
* good correlation between the size of the deletion and the severity of the phenotype
* incidence of deletions of both SMN and NAIP is 62% in SMA type I, 8.8% in type II and 12% in type III.
* degeneration of the anterior horn cells in the spinal cord

Clinical features
* onset between 6 and 12 months,
* symmetric weakness of proximal weakness, more in the legs than arms
* never learn to walk (in contrast to patients with SMA 3 who will learn to walk)
* tendon reflexes absent
* facial muscles are clinically spared
* scoliosis
* normal intellect
* slowly progressive
* CK: normal or moderately elevated

Strategy
* demonstrate chronic neurogenic EMG findings of limb muscles (proximal muscles more involved than distal muscles)
* demonstrate normal sensory neurography

Expected abnormal findings
EMG
* acute or subacute neurogenic EMG findings in several muscles

Neurography
* MCS: reduced AMPL, CV may be reduced if AMPL is very low
**Expected normal findings**

**Neurography**
- SCS

**Procedure**
- EMG (unilaterally)
  - m.deltoides/m.biceps brachii
  - m.interosseus dorsalis
  - m.vastus lateralis
  - m.tibialis anterior

**Expected abnormal findings**

**EMG**
- acute or subacute neurogenic EMG findings in several muscles

**Neurography**
- MCS: reduced AMPL, CV may be reduced if AMPL is very low

**References**
- Dubowitz V. Infantile muscular atrophy -- prospective study with particular reference to a slowly progressive variety. Brain 1964;87:707-18
- Dubowitz V. Chaos in classification of the spinal muscular atrophies of childhood. Neuromusc Disord 1991;1:77-80

**Modified**
- 26.3.1997 BF ES 3.4.97

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**SPINAL MUSCULAR ATROPHY 3, KUGELBERG-WELANDER**

**Etiology**
- autosomal recessive inheritance
- genetic defect localized to chromosome 5q11-q13
- deletion in a region that codes the SMN (the survival motor neuron gene) and the NAIP (neuronal apoptosis inhibitory protein).
- degeneration of the anterior horn cells in the spinal cord
- good correlation between the size of the deletion and the severity of the phenotype
- incidence of deletions of both SMN and NAIP is 62% in SMA type I, 8.8% in type II and 12% in type III.
- degeneration of the anterior horn cells in the spinal cord

**Clinical features**
- onset between 1-20 years of age
- learn to walk but loose later the ability to walk
- present with difficulties with walking, running climbing or jumping
- Gower’s sign
- proximal weakness
- legs weaker than arms
- hypermobility of joints
- CK normal or moderately elevated
- slow progression

**Strategy**
- demonstrate symmetric chronic neurogenic EMG findings of limb muscles (proximal muscles more involved than distal muscles)
- demonstrate normal sensory neurography

**Expected abnormal findings**

**EMG**
- acute or subacute neurogenic EMG findings in several muscles

**Neurography**
- ECS: reduced AMPL, CV may be reduced if AMPL is very low

**References**
**X-LINKED BULBOSPINAL HEREDITARY NEURONOPATHY (KENNEDY SYNDROME)**

**Etiology**
- X-linked recessive inheritance
- Xq21-22, mutation consists of expansion of CAG trinucleotide repeats (disease severity correlated with number of CAG repeats)
- gene product: androgen receptor
- spinobulbar motor neuronopathy

**Clinical features**
- onset at the age 30 to 50 years, sometimes as young as 15 years
- muscle cramps often initial symptom
- proximal muscle weakness, legs > arms
- facial and bulbar muscles affected
- distribution of affected muscles in the beginning varies somewhat, patients often complain about focal weakness.
- gynecomastia, impotence, infertility
- absent of depressed tendon reflexes
- normal life expectancy
- CK normal or mildly elevated (up to five times normal)

**Strategy**
- demonstrate subacute neurogenic abnormalities with a proximal and symmetric distribution
- differentiate from sensory-motor polyneuropathy
- differentiate from ALS
- differentiate from previous polio

**Expected abnormal findings**

**EMG**
- symmetric subacute neurogenic abnormalities with proximal distribution, fasciculations common

**Neurography**
- mild sensory abnormalities with low sensory amplitudes

**Procedure**
- EMG (unilaterally)
  - m. deltoides/m. biceps brachii
  - m. interosseus dorsalis
  - m. vastus lateralis
  - m. tibialis anterior

**Neurography**
- one motor and sensory nerve in the upper and lower extremities

**References**

**Modified**
- 26.3.1997 BF 2.4.1997 BF 3.4.97 ES

**DISTAL SPINAL MUSCULAR ATROPHY**

**Etiology**
- genetically heterogeneous
- there are several autosomal dominant forms and recessive forms
- degeneration of spinal anterior horn cells

**Clinical features**
- patients with dominant form of the disease develop symptoms during the first decade, usually before 20 years of age
- distal weakness and wasting of muscles
- often pes cavus
- legs more affected than arms
- tendon reflexes normal or depressed

**Strategy**
- demonstrate subacute neurogenic abnormalities in distal limb muscles
- differentiate from sensory-motor polyneuropathy and ALS
**Expected abnormal findings**

**EMG**
* subacute neurogenic abnormalities in distal muscles bilaterally
* proximal muscles less affected

**Neurography**
* MCS low AMPL

**Expected normal findings**

**Neurography**
* normal SCS

**Procedure**

**EMG (unilaterally)**
* m.deltoideus/m.biceps brachii
* m.interosseus dorsalis
* m.vastus lateralis
* m.tibialis anterior

**Neurography (unilaterally)**
* MCS: n.medianus, n.peroneus
* SCS: n.suralis, n.radialis

**References**

**Modified**
* 26.3.1997 BF 3.4.97 ES

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**MONOMELIC SPINAL MUSCULAR ATROPHY**

**Etiology**
* unknown
* most are sporadic
* autosomal dominant inheritance has been described

**Clinical features**
* onset usually at the age of around 20 years, mostly before the age of 30
* weakness and wasting of intrinsic hand muscles, forearm muscles may be affected
* progression lasts usually 1-2 years and then the process is arrested
* cases affecting proximal leg muscles have been described

**Strategy**
* demonstrate focal subacute neurogenic abnormalities within a few myotomes, usually C7-C8-Th1
* findings are usually bilateral
* differentiates from ALS and MMN

**Expected abnormal findings**

**EMG**
* subacute neurogenic abnormalities in C7-C8-Th1 muscles

**Neurography**
* MCS low AMPL in ulnar, median and radial nerves

**Expected normal findings**

**Neurography**
* normal SCS

**EMG**
* normal findings in leg muscles and muscles innervated by cranial nerves

**Procedure**

**EMG**
* m.deltoideus
* m.biceps
* m.triceps, bilaterally
* m.extensor digitorum communis, bilaterally
* m.interosseus dorsalis, bilaterally
* m opponens pollicis/m.abductor pollicis brevis, bilaterally
* m.vastus lateralis
* m.tibialis anterior
* m.trapezius

**Neurography**
* n.medianus, MCS and SCS
* n.ulnaris, MCS and SCS
* n.radialis, SCS
* n.suralis, SCS
* n.peroneus, MCS

**References**
* De Visser M, de Visser BW, Verbeeten B. Electromyographic and computed tomographic findings in five patients with monomelic spinal muscular atrophy. Eur Neurol 1985; 26:135-138
BULBAR HEREDITARY MOTOR NEURONOPATHY (FAZIO-LONDE’S DISEASE)

Etiology
- unknown motor neuronopathy of the bulbar motor neurons

Clinical features
- onset usually at 2 to 3 years age, latest 12 years
- dysphagia
- facial weakness
- extraocular palsies
- limb muscles may be involved
- most patients die within two years of onset

Strategy
- demonstrate motor neurogenic EMG findings of muscles innervated by the cranial nerves
- differentiate from SMA

Expected abnormal findings
- subacute neurogenic abnormalities in muscles innervated by the cranial nerves
- limb muscles may be involved

Neurography
- MCS low AMPL in n.facialis

Expected normal findings
- normal SCS

Procedure
- EMG
  - m.orbicularis oris
  - m.masseter
  - m.trapezius
  - m.deltoides/m.biceps
  - m.interosseus dorsalis/m.opponens pollicis/m.abductor pollicis brevis
  - m.vastus lateralis
  - m.tibialis anterior

- Neurography
  - n.medianus, MCS and SCS
  - n.suralis, SCS
  - n.peroneus, MCS

References

2. POLYNEUROPATHIES

AXONAL SENSORY-MOTOR POLYNEUROPATHY

Etiology
- almost 200 different metabolic, toxic and genetic causes are known
- most common cause in the developed countries is diabetes
- other common causes are renal insufficiency and chronic alcoholism

Clinical features
- the sensory and motor symptoms vary considerably and depend to some extent on the etiology
- weakness of distal muscles, legs affected more than arms
- sensory loss starting in the distal part of the legs and spreading proximally
- paresthesias, dysesthesias in the distal parts of the extremities usually worse at rest
- muscle cramps

Strategy
- demonstrate generalized axonal dysfunction of peripheral sensory and motor nerves
- often also autonomic nerves are affected to varying degrees.
- are the findings compatible with a specific axonal polyneuropathy?

Assess
- type (motor/sensory/autonomic)
- time course (acute, subacute, inactive)
- severity
pathophysiology (axonal/demyelinating/conduction block)
- distribution: diffuse (distal/proximal) or multifocal

**Expected abnormal findings**

**Neurography**
- SCS ampl reduced. Distal leg nerves are more affected than arm nerves.
- in moderate to severe axonal polyneuropathies also motor amplitudes are reduced
- CV is slow normal or slightly reduced (not more than 30% of reference values) in axonal polyneuropathies
- F waves are delayed and the number of F waves may be reduced
- A-waves are often seen

**EMG**
- limb muscles show varying degrees of neurogenic involvement. Distal muscles are more affected than proximal muscles.
- Leg muscles are more affected than arm muscles

**Autonomic testing**
- often abnormal

**Sensory thresholds**
- often abnormal

**Procedure**

**Neurography**
- MCS: n.medianus, n.ulnaris, n.tibialis, n.peroneus unilaterally
- SCS: n.medianus and n.ulnaris bilaterally and n.radialis, n.suralis unilaterally.

**EMG (optional)**
- one distal and proximal muscle in the lower extremities.

**Autonomic tests (optional)**
- RR-interval
- SSR
- plethysmography

**Sensory thresholds (optional)**
- temperature and vibration thresholds

**Demyelinating Sensory-Motor Polyneuropathy**

**Etiology**
- various causes have been described. Demyelinating polyneuropathies are much less common than axonal polyneuropathies
- diphtheria
- monoclonal gammopathies and paraneoplastic
- inflammatory: acute polyradiculitis, chronic polyradiculitis
- hereditary: HMSN1, HMSN3, HMSN4, HNPP

**Clinical features**
- the sensory and motor symptoms vary considerably and depend to some extent on the etiology
- weakness of distal muscles, legs affected more than arms
- sensory loss starting in the distal part of the legs and spreading proximally
- paresthesias, dysesthesias in the distal parts of the extremities usually worse at rest
- muscle cramps

**Strategy**
- demonstrate generalized demyelinating dysfunction of peripheral sensory and motor nerves.
- are the findings compatible with a specific demyelinating polyneuropathy? (polyradiculitis, HMSN1, HNPP)

**Assess**
- type (motor/sensory/autonomic)
- time course (acute, subacute, inactive)
- severity
- pathophysiology (axonal/demyelinating/conduction block)
- distribution: diffuse (distal/proximal) or multifocal

**Expected abnormal findings**

**Neurography**
- sensory and motor nerve conduction velocities are reduced, usually by more than 30%
- distal motor latencies often > 7 ms
- the amplitude of the sensory nerve action potentials are reduced. Distal leg nerves are more affected than arm nerves.
- in severe demyelinating axonal polyneuropathies also motor amplitudes are reduced due to secondary axonal involvement
- F wave latencies are delayed and the number of F waves may be reduced
- A-waves may be seen

**EMG**
- limb muscles show varying degrees of neurogenic involvement. distal muscles are more affected than proximal muscles. leg muscles are more affected than arm muscles

**Autonomic testing**
- often abnormal

**Sensory thresholds**
- often abnormal

**Procedure**

**Neurography**
- MCS: n.medianus, n.ulnaris, n.tibialis, n.peroneus unilaterally
- SCS: n.medianus and n.ulnaris bilaterally and n.radialis, n.suralis unilaterally.

**EMG (optional)**
- one distal and proximal muscle in the lower extremities.

**Autonomic tests (optional)**
- RR-interval
- SSR
- plethysmography

**Sensory thresholds (optional)**
**Temperature and vibration**

**Note**
- Hereditary demyelinating polyneuropathies usually do not display conduction block
- Acquired demyelinating polyneuropathies tend to have conduction block

**Acute Polyradiculitis (Guillain-Barré Syndrome, GBS, Acute Inflammatory Demyelinating Polyneuropathy, AIDP)**

**Etiology**
- Probably autoimmune reaction against peripheral nerves sometimes triggered by preceding infection (especially Campylobacter jejuni) trauma, operation or childbirth.

**Clinical features**
- Incidence 1-2/100000 per year in people younger than 45 years, in 70-75 year old the incidence is 4-6/100000
- Male : female ratio 1.5:1
- Occurs in all age groups, peak 50-70 years
- Acute onset within days
- At the onset 80 % have paresthesias and 60 % have weakness
- Paresthesias precede the weakness by a few days
- Typically patients notice weakness in the legs before the arms
- The presentation of weakness is often ascending (50-60%) from legs to arms and cranial muscles
- The presentation may be descending in a portion of patients
- Pain is not uncommon; 15-50 % of patients have pain
- Facial nerve is involved in 50 %
- The autonomic nervous system may be affected, especially in patients with severe motor deficits
- Tendon reflexes are decreased or absent
- Most patients worsen over 1-2 weeks, some for up to 4 weeks
- Mortality 2-5 %
- After plateau subsequent recovery over 6-12 months
- Patients that worsen for more than 8 weeks probably have chronic polyradiculitis
- Classically it has been thought that polyradiculitis is primarily a demyelinating sensory and motor polyneuropathy, currently it is accepted that there is an primarily axonal form of polyradiculitis, sometimes affecting only motor nerves (acute motor axonal neuropathy AMAN)

**Strategy**
- Demonstrate acute motor and sensory neuropathy
- In the acute stage the motor nerves are more affected than sensory nerves
- The neuropathy is typically demyelinating with conduction block, the most prominent findings are often in the proximal parts of the nerves
- Sometimes the neuropathy is predominantly axonal, especially if associated with Campylobacter jejuni
- Assess: Severity, pathology, distribution

**Expected abnormal findings**

**Neurography, MCS**
- Conduction block (dist/prox amplitude >30 % in upper extremities and > 50 % in lower extremities without considerable increase in dispersion)
- F waves delayed and few due to conduction block
- DL prolonged
- Reduced MCV, sometimes normal initially
- Distal amplitude may be initially normal - low amplitude with normal DL indicates severe axonal involvement

**Neurography SCS**
- Reduced SCV is not necessarily seen during the first weeks
- Reduced amplitude is seen in the presence of axonal loss

**EMG**
- < 10-18 days from onset of symptoms: only reduced interference pattern
- > 10-18 days from onset: signs of acute neurogenic EMG findings

**Autonomic tests**
- Often abnormal

**Procedure**

**Neurography**
- MCS: n.medianus, n.ulnaris, n.tibialis, n.peroneus unilaterally
- SCS: n.radialis, n.suralis unilaterally.

**EMG**
- < 10 days from onset of symptoms not necessarily informative, but may reveal earlier onset and confirm peripheral cause of weakness
- > 10 days from onset, should be done to demonstrate degree of axonal involvement.

**Autonomic tests (optional)**
- RR-interval
- SSR
- Plethysmography

**Sensory thresholds (optional)**
- Temperature and vibration

**References**
ACUTE AXONAL MOTOR NEUROPATHY (AMAN)

**Etiology**
- immune reaction against motor axons, probably triggered by preceding infection, especially Campylobacter jejuni, trauma, operation or childbirth.

**Clinical features**
- most patients described come from northern China and Mexico
- usually triggered by Campylobacter jejuni infection
- most patients are children or young adults
- most cases occur during summer months, peak in August
- rapidly developing symmetric flaccid paralysis, may progress to tetraparesis and respiratory failure
- extraocular muscles rarely involved
- mortality around 5%
- CSF shows elevation of proteins during second or third week from onset
- prognosis is usually favourable in spite of the axonal involvement, it is thought that the axonal involvement is distal
- the prognosis does not differ from the classical demyelinating sensory-motor type of polyradiculitis

**Strategy**
- demonstrate acute motor axonal neuropathy
- differentiate from polio, which tends to be asymmetric and pathcy
- assess: severity, pathology, distribution

**Expected abnormal findings**

* Neurography, MCS
  - DL prolonged
  - mildly reduced MCV, sometimes normal initially
  - M wave amplitude reduced
  - F waves absent or number reduced

* EMG
  - < 10-18 days from onset of symptoms: only reduced interference pattern
  - > 10-18 days from onset: signs of acute neurogenic EMG findings

* Expected normal findings*
  - Neurography, SCS
    - all sensory nerves should be normal

* Sensory thresholds
  - temperature and vibration

* Procedure*
  - Neurography
    - MCS: n.medianus, n.ulnaris, n.tibialis, n.peroneus unilaterally
    - SCS: n.radialis, n.suralis unilaterally.
  - EMG
    - < 10 days from onset of symptoms not necessarily informative, but may reveal earlier onset and confirm peripheral cause of weakness
    - > 10 days from onset, should be done to demonstrate degree of axonal involvement.

* Autonomic tests (optional)*
  - RR-interval
  - SSR
  - plethysmography

* Sensory thresholds (optional)*
  - temperature and vibration

**References**
**MILLER-FISHER SYNDROME**

**Etiology**
- Probably autoimmune reaction against peripheral nerves sometimes triggered by preceding infection (particularly Campylobacter jejuni) trauma, operation or childbirth
- Considered to be a variant of acute polyradiculitis

**Clinical features**
- Characterized by external ophthalmoplegia, ataxia and areflexia
- Patients experience diplopia and unsteadiness of gait, either of these symptoms may be the initial sign
- Other muscles innervated by cranial nerves may show weakness; dysarthria, facial palsy, ptosis, tongue weakness, palatal palsy
- Acute onset within days, peak of symptoms is reached around one to two weeks from onset
- Mild weakness and sensory symptoms may be present
- Usually the course is monophasic and relatively benign, recurrences have been described
- CSF protein is elevated

**Strategy**
- Demonstrate acute motor and sensory neuropathy
- In the limbs the neuropathy is predominantly sensory
- Assess: severity, pathology, distribution

**Expected abnormal findings**

**Neurography, MCS**
- Mild slowing of motor CV
- F waves delayed and few due to conduction block
- DL may be prolonged
- Mildly reduced MCV, sometimes normal initially
- Distal amplitude reduced, especially in facial muscles

**Neurography SCS**
- Reduced amplitude dominates over reduced CV

**EMG**
- < 10-18 days from onset of symptoms: only reduced interference pattern
- > 10-18 days from onset: signs of acute neurogenic EMG findings

**Autonomic tests**
- Often abnormal

**Blink reflex**
- Absent or mildly prolonged R1 components
- R2 may be absent, but latency is usually normal

**Procedure**

**Neurography**
- MCS: n.medianus, n.ulnaris, n.tibialis, n.peroneus unilaterally
- MCS: the branches of n.facialis bilaterally
- SCS: n.radialis, n.suralis unilaterally.

**EMG**
- < 10 days from onset of symptoms not necessarily informative, but may reveal earlier onset and confirm peripheral cause of weakness
- > 10 days from onset, should be done to demonstrate degree of axonal involvement.
- Facial muscles should be studied

**Blink reflex**
- If supraorbital nerve stimulation gives normal findings, consider doing also the infraorbital nerve

**Autonomic tests (optional)**
- RR-interval
- SSR
- Plethysmography

**Sensory thresholds (optional)**
- Temperature and vibration

**References**
**CHRONIC POLYRADICULITIS (CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY, CIDP)**

**Etiology**
* probably prolonged autoimmune reaction against peripheral nerves.

**Clinical features**
* occurs in all age groups from 2 years
* may begin as a typical acute polyradiculitis but may start with subacute symptoms as well
* either chronic progressive (slowly or stepwise) course or relapsing
* progression > 6 weeks followed by episodes of improvement and worsening for at least 3 months or slow progression for > 6 months
* bilateral relatively symmetric weakness
* paresthesias in toes and fingers
* facial muscles may be affected (10-15% of patients)
* elevated CSF protein concentration during deterioration
* areflexia at ankles and general hyporeflexia for at least 1 month
* patients with a relapsing course often resolve after a few years and they tend to have a better prognosis than those with chronic progressive course
* MRI shows increased signals on T2 weighted images at the sites of conduction blocks. MRI of the brachial plexus or other regions with suspected conduction blocks may be a useful accessory investigation in the diagnosis

**Strategy**
* demonstrate subacute motor and sensory neuropathy
  * the neuropathy is typically demyelinating with conduction block, the most prominent findings are often in the proximal parts of the nerves
  * sometimes the neuropathy is predominantly axonal
  * assess: severity, pathology, distribution

**Expected abnormal findings**
**Neurography, MCS**
* reduced MCV
* conduction block (dist/prox amplitude >30 % in upper extremities and > 50 % in lower extremities without considerable increase in dispersion)
* F waves delayed and few due to conduction block
* dist lat prolonged
* distal ampl may be initially normal - low amplitude with normal dist.lat. indicates severe axonal involvement

**Neurography SCS**
* reduced SCV
* reduced amplitude is seen in the presence of axonal loss

**EMG**
* depends on severity of disorder, in mild cases distal muscles show subacute or inactive neurogenic findings in more severe cases also proximal muscles are involved

**Autonomic nervous system tests**
* RR-interval often abnormal
* SSR may be abnormal

**Procedure**
**Neurography**
* MCS: n.medianus, n.ulnaris, n.tibialis, n.peroneus unilaterally
* SCS: n.radialis, n.suralis unilaterally.

**EMG**
* one proximal and distal muscle in the upper and lower extremities

**Autonomic tests (optional)**
* RR-interval
* SSR
* pletysmography

**Sensory thresholds (optional)**
* temperature and vibration

**References**
* Feasby TE. Axonal CIDP. A premature concept. Muscle Nerve 1996;372-374

**Modified**
* 25.6.1997 BF, 2.4.1997 BF, 3.4.97 ES

**MULTIFOCAL MOTOR NEUROPATHY WITH CONDUCTION BLOCK (MMN)**

**Etiology**
* unknown, possibly an autoimmune reaction against gangliosides (GM1) in some patients
* regarded by many as a variant of CIDP

**Clinical features**
slowly progressive weakness, usually distributed within one peripheral nerve (in ALS the distribution follows spinal myotomes)
progression usually slow over years and decades
weakness is often distally accentuated, but may be proximal in some patients
muscle atrophy of some weak muscles is less pronounced than would be expected (weakness may be due to conduction block)
fasciculations, cramps and myokymia may be seen
MMN is predominantly a motor neuropathy, but mild sensory symptoms and findings may be seen
areflexia may be seen in clinically not affected muscles
no signs of upper motor neuron lesion
very rarely involvement of cranial nerves
m.diaphragma is rarely affected
the resemblance of MMN to ALS; clinically it may sometimes be difficult to distinguish them
differentiation of ALS and MMN can readily be done with EMG and nerve conduction studies
MRI shows increased signals on T2 weighted images at the sites of conduction blocks. MRI of the brachial plexus or other regions with suspected conduction blocks may be a useful accessory investigation in the diagnosis of MMN.

Strategy
- confirm subacute neurogenic EMG findings of muscles in several parts of the body
- demonstrate significant conduction block in motor nerves
- exclude sensory motor polyneuropathy
- differentiate from ALS
- differentiate from spinal hereditary motor neuronopathies (spinal muscular atrophies)
- differentiate from previous polio

Expected abnormal findings

EMG
- subacute or chronic neurogenic muscle findings in several nerves (the weakness and EMG findings are often related with individual nerves rather than myotomes)

Neurography
- motor nerve show conduction block (amplitude and area decay, reduced number of F-waves)
- MCS often show reduced amplitudes
- motor conduction velocity may be reduced, especially if amplitude is small

Expected normal findings

Neurography
- SCS normal
- Central motor conduction time
- normal

Procedure

EMG
The muscles should be chosen based on the clinical muscle weakness; if weakness is widespread, the following muscles are recommended
- m.interosseus dorsalis I
- m.biceps brachii/m.deltoides
- m.tibialis anterior/m.gastrocnemius caput mediale
- m.vastus lateralis
- m.trapezius/orbicularis oris/m.genioglossus (insert electrode below chin)

Neurography
The following motor nerves should be studied bilaterally
- n.medianus
- n.ulnaris (also including supraclavicular stimulation)
- n.peroneus
- n.tibialis
The following sensory nerves should be tested
- n.suralis
- n.radialis

Note
- patients with MMN do not have upper motor neuron signs
- muscles innervated by cranial nerves are rarely affected

References

Modified

**HEREDITARY MOTOR AND SENSORY NEUROPATHY TYPE I (HMSN1, CHARCOT-MARIE-TOOTH)**

**Etiology**
- several independent subtypes have been described
  - 1a is most common (70-95%). Autosomal dominant inheritance, linked to chromosome 17p11.2. Gene product peripheral myelin protein 22 (PMP-22)
  - 1b is less common. Autosomal dominant inheritance, linked to chromosome 1q21-23. Gene product peripheral myelin protein P0 (PNPO)
  - 1c, other autosomal loci have been described
  - X-linked dominant (X1), linked to Xq13, gene product connexin
  - X-linked recessive (X2) have been described

**Clinical features**
- a relatively common polyneuropathy
- slowly developing sensory and motor polyneuropathy
- first symptoms usually during the second decade but may start earlier or considerably later
- peroneal muscle weakness first symptom, later distal hand muscle weakness
- peroneal muscles are affected much more than calf muscles
- the foot often has a typical pes cavus deformity and clawed toes
- parasthesias are uncommon, if prominent, challenge the diagnosis
- distal neuropathic pain is uncommon, but the foot deformity often causes pain when the patient walks
- essential tremor is sometimes present; previously such patients were designated as Roussy-Lévy syndrome
- usually does not affect life expectancy
- the severity of the disorder even within each group depends on the exact location of the mutation

**Strategy**
- demonstrate generalized demyelinating dysfunction of peripheral sensory and motor nerves. Often also autonomic nerves are affected to varying degrees.
- differentiate from acquired demyelinating polyneuropathies

**Expected abnormal findings**

**Neurography**
- sensory and motor nerve conduction velocities are reduced, usually by more than 30%
- median motor CV in most patients <38 m/s
- distal motor latencies often > 7 ms
- conduction block is uncommon
- the amplitude of the sensory nerve action potentials are reduced. Distal leg nerves are more affected than arm nerves.
- F wave latencies are delayed and the number of F waves may be reduced
- A-waves maybe seen

**EMG**
- limb muscles show varying degrees of neurogenic involvement
- distal muscles are more affected than proximal muscles
- leg muscles are more affected than arm muscles

**Autonomic testing**
- may be abnormal

**Sensory thresholds**
- often abnormal

**Procedure**

**Neurography**
- MCS: n.medianus, n.ulnaris, n.tibialis, n.peroneus unilaterally
- SCS: n.medianus and n.ulnaris bilaterally and n.radialis, n.suralis unilaterally.

**EMG (optional)**
- one distal and proximal muscle in the lower extremities.

**Autonomic tests (optional)**
- RR-interval
- SSR
- plethysmography

**Sensory thresholds (optional)**
- temperature and vibration

**References**
HEREDITARY MOTOR AND SENSOR NEUROPATHY TYPE 2 (HMSN2, CHARCOT-MARIE-TOOTH)

Etiology
- genetically heterogeneous, several subtypes have been described
- most follow an autosomal dominant inheritance
- so far two types have been identified: one type is linked to chromosome 1p35-p36 and another to 3q13-22

Clinical features
- slowly developing sensory and motor neuropathy
  - first symptoms usually during the second decade but may start earlier or considerably later
  - peroneal muscle weakness first symptom later distal hand muscle weakness
  - peroneal muscles are affected much more than calf muscles
  - the foot often has a typical pes cavus deformity and clawed toes
  - paresthesias are uncommon, if prominent, challenge the diagnosis
  - distal neuropathic pain is uncommon, but the foot deformity causes often pain when the patient walks
  - essential tremor is sometimes present, previously such patients were designated as Roussy-Lévy syndrome
  - usually does not affect life expectancy

Strategy
- demonstrate generalized axonal dysfunction of peripheral sensory and motor nerves. Often also autonomic nerves are affected to varying degrees.
- differentiate from acquired demyelinating polyneuropathies

Expected abnormal findings

Neurography
- SCS amplitudes are reduced, distal leg nerves are more affected than arm nerves.
- CV is slow normal or slightly reduced (not more than 30% of reference values) in axonal polyneuropathies
- median MCV >38 m/s
- F waves are delayed and the number of F waves may be reduced
- A-waves are often seen

EMG
- limb muscles show varying degrees of neurogenic involvement.
- distal muscles are more affected than proximal muscles.
- leg muscles are more affected than arm muscles

Autonomic testing
- may be abnormal

Sensory thresholds
- often abnormal

Procedure

Neurography
- MCS: n.medianus, n.ulnaris, n.tibialis, n.peroneus unilaterally
- SCS: n.medianus and n.ulnaris bilaterally and n.radialis, n.suralis unilaterally.

EMG (optional)
- one distal and proximal muscle in the lower extremities.

Autonomic tests (optional)
- RR-interval
- SSR
- plethysmography

Sensory thresholds (optional)
- temperature and vibration

References
- Harding AE, Thomas PK. The clinical features of hereditary motor and sensory neuropathy type I and II. Brain 1980;103:259-80
HEREDITARY MOTOR AND SENSORY NEUROPATHY TYPE 3 (HMSN3, DEJERINE-SOTTAS)

**Etiology**
- genetically heterogeneous
- autosomal dominant and recessive inheritance, may be sporadic
- linkage to chromosomes 17p11.2, 1q22 and 8qter have been described

**Clinical features**
- starts neonatally or in infancy
- debilitating sensory and motor polyneuropathy
- patients usually confined to wheelchair in adulthood

**Strategy**
- demonstrate generalized demyelinating dysfunction of peripheral sensory and motor nerves. Often also autonomic nerves are affected to varying degrees.
- differentiate from acquired demyelinating polyneuropathies

**Expected abnormal findings**

**Neurography**
- sensory and motor nerve conduction velocities are usually severely reduced (CV = 5-15 m/s)
- the amplitude of the sensory nerve action potentials are reduced. Distal leg nerves are more affected than arm nerves.
- in severe demyelinating axonal polyneuropathies also motor amplitudes are reduced due to secondary axonal involvement
- F wave latencies are delayed and the number of F waves may be reduced
- A-waves maybe seen

**EMG**
- limb muscles show varying degrees of neurogenic involvement.
- distal muscles are more affected than proximal muscles.
- leg muscles are more affected than arm muscles

**Autonomic testing**
- often abnormal

**Sensory thresholds**
- often abnormal

**Procedure**

**Neurography**
- MCS: n.medianus, n.ulnaris, n.tibialis, n.peroneus unilaterally
- SCS: n.medianus and n.ulnaris bilaterally and n.radialis, n.suralis unilaterally.

**EMG (optional)**
- one distal and proximal muscle in the lower extremities.

**Autonomic tests (optional)**
- RR-interval
- SSR
- plethysmography

**Sensory thresholds (optional)**
- temperature and vibration

**References**
- Ouvrier R. Correlation between the histopathologic, genotypic, and phenotypic features of hereditary peripheral neuropathies of childhood. J Child Neurol 1996;11:133-146

**Modified**
- 31.3.1997 BF, 3.4.97 ES

HEREDITARY MOTOR AND SENSORY NEUROPATHY TYPE 4 (HMSN4, MB REFSUM)

**Etiology**
- phytanic acid storage disease, patients show almost no oxidation of phytanic acid
- deficiency of the peroxisomal enzyme alpha-hydroxy acid oxidase
- autosomal recessive inheritance
Clinical features
- onset from childhood to third decade
- pigmentary retinal degeneration, night blindness
- sensory-motor neuropathy, peripheral nerves are often palpably enlarged
- visual dysfunction precedes polyneuropathy
- cerebellar symptoms: ataxia, nystagmus and intention tremor
- pes cavus
- often ichthyosis of the skin, anosmia, cardiomyopathy and sensorineural hearing loss

Strategy
- demonstrate demyelinating polyneuropathy
- demonstrate pigmentary retinopathy

Expected abnormal findings

Neurography
- sensory and motor nerve conduction velocities are moderately reduced, usually by more than 30%
- the amplitude of the sensory nerve action potentials are reduced. Distal leg nerves are more affected than arm nerves.
- F wave latencies are prolonged and the number of F waves may be reduced
- A-waves maybe seen

EMG
- limb muscles show varying degrees of neurogenic involvement. distal muscles are more affected than proximal muscles
- leg muscles are more affected than arm muscles

Electroretinography (ERG)
- absence of both cone and rod responses

BAEP (brainstem auditory evoked potentials)
- abnormal

Autonomic testing
- often abnormal

Sensory thresholds
- often abnormal

Procedure

Neurography
- MCS: n.medianus, n.ulnaris, n.tibialis, n.peroneus unilaterally
- SCS: n.medianus and n.ulnaris bilaterally and n.radialis, n.suralis unilaterally.

EMG (optional)
- one distal and proximal muscle in the lower extremities.

Autonomic tests (optional)
- RR-interval
- SSR
- plethysmography

Sensory thresholds (optional)
- temperature and vibration

Electroretinography (ERG)
- absence of both cone and rod responses

References

Modified
- 2.4.1997, ES 3.4.97

HEREDITARY NEUROPATHY WITH LIABILITY TO PRESSURE PALSYES (HNPP)

Etiology
- autosomal dominant inheritance, linked to chromosome 17p11.2
- gene product peripheral myelin protein 22 (PNP-22)
- locus of deletion is identical to HMSN 1a which has a duplication at the same site

Clinical features
- liability to develop local nerve lesions following compression or trauma that usually would not cause a nerve lesion
- presents often in the late teens or young adult age, sometimes seen also in young children
- some patients may have several nerve lesions
- usually the recovery of the local nerve lesions is good
- many carriers of the gene do not have any local nerve lesions
- before the age of 30 neurological examination is normal except for the local nerve lesions
- in old age there are distinct clinical signs of polyneuropathy, decreased tendon reflexes and peroneal muscle weakness

Strategy
- demonstrate a mild generalized demyelinating dysfunction of peripheral sensory and motor nerves
- most clearcut findings will be found at common entrapment sites (n.medianus at the wrist, n.ulnaris at the elbow)
- most patients will present with mononeuropathies, most commonly radial nerve lesions, peroneal lesions, median nerve lesions
- differentiate from acquired demyelinating polyneuropathies

Differential diagnosis
- other polyneuropathies
- HMSN1, usually has more slowing of nerve conduction
- hereditary recurrent
**Expected abnormal findings**

**Neurography**
- sensory and motor nerve conduction velocities are reduced to varying degrees
- some nerves may have CVs in the low normal range
- n.radialis and suralis SCV is usually reduced by 20-30%
- the conduction velocities at common entrapment sites are often considerably reduced
- median nerve motor distal latencies can be prolonged up to 5-7 ms without symptoms of CTS
- the amplitude of the sensory nerve action potentials are reduced in middle-aged and older subjects

**EMG**
- findings depend on the type of local nerve lesions
- middle-aged and older patients show signs of neurogenic involvement in distal leg muscles due to the polyneuropathy (sometimes it may difficult to tell whether the findings in the leg muscles are due to mild repeated peroneal nerve lesions or polyneuropathy)

**Sensory thresholds**
- vibration thresholds and thermal thresholds are normal in young subjects but show abnormalities in middle-aged or older subjects

**Procedure**

**Neurography**
- MCS bilateral: n.medianus, n.ulnaris (fractionated across the elbow) and peroneal nerves (fractionated across the fibular head),
- SCS: n.medianus and n.ulnaris bilaterally and n.radialis, n.suralis unilaterally.

**EMG (optional)**
- one distal and proximal muscle in the lower extremities.

**Autonomic tests (optional)**
- RR-interval
- SSR
- plethysmography

**Sensory thresholds (optional)**
- temperature and vibration

**References**
- Davies DM. Recurrent peripheral-nerve palsies in a family. Lancet II 1954;266-268

**Modified**
- 31.3.1997 BF, 3.4.97 ES

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**INHERITED RECURRENT BRACHIAL PLEXUS NEUROPATHY**

**Etiology**
- autosomal dominant inheritance
- linked to chromosome 17q24

**Clinical features**
- acute, usually painful episodes of local neuropathies
- most nerve lesions are confined to plexus brachialis and the upper extremities, but any nerve may be affected
- a distinctive feature is that nerve lesions appear often during pregnancy and puerperium
- infections, surgery, immunization and strenuous exercise may trigger episodes
- the episodes resemble sporadic episodes of “neuralgic amyotrophy”
- onset varies considerably, often first episode in early childhood
- prognosis of individual nerve lesions is generally good
- many reports describe mild facial dysmorphic features, in our experience they are not essential

**Strategy**
- demonstrate local or regional neuropathy
- exclude generalized polyneuropathy

**Expected abnormal findings**

**Neurography**
- depends on the nerve affected

**EMG**
- muscles chosen in relation to the affected nerve or plexus structure

**Expected normal findings**

**EMG**
- normal outside affected nerves

**Neurography**
- normal outside affected nerves

**Autonomic tests**
- RR-interval
- SSR
- plethysmography
**Procedure**

**Neurography**
- MCS bilateral: n.medianus, n.ulnaris and n.peroneus
- SCS: n.medianus, n.ulnaris, n.radialis and n.suralis bilaterally.

**EMG**
- one distal and proximal muscle in the lower extremities.

**Sensory thresholds (optional)**
- temperature and vibration

**Autonomic tests (optional)**
- RR-interval
- SSR
- plethysmography

**References**
- Parsonage MJ, Turner JWA. Neuralgic amyotrophy. The shoulder-girdle syndrome. Lancet 1948;i:973-8
- Taylor RA. Heredofamilial mononeuritis multiplex with brachial predilection. Brain 1960;83:113

**Modified**
- 31.3.1997 BF, 3.4. 97 ES

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**Critical Illness Polyneuropathy**

**Etiology**
- exact mechanism of neuropathy is not clear
- occur during intensive care in association with sepsis and multiple organ failure
- axonal polyneuropathy

**Clinical features**
- neuropathy becomes apparent after the sepsis and multiple organ failure has been controlled
- often the neuropathy is recognized when attempts to wean the patient from the respirator are made
- the patient is awake and often unable to maintain spontaneous breathing

**Strategy**
- demonstrate acute sensory and motor axonal polyneuropathy

**Expected abnormal findings**

**Neurography**
- SCS: AMPL reduced or missing responses
- SCS: CV may be reduced
- MCS: low amplitudes
- MCS: slight CV reduction may be present
- F waves: reduced number of F waves, related to low AMPL
- Autonomic test usually show abnormalities

**Expected normal findings**
- none

**Procedure**

**Neurography**
- MCS: n.medianus, n.ulnaris and n.peroneus and n.tibialis unilaterally
- SCS: n.radialis and n.suralis bilaterally.

**EMG**
- one distal and proximal muscle in the lower extremities
- one distal and proximal muscle in the upper extremities

**Sensory thresholds (optional)**
- temperature and vibration

**Autonomic tests (optional)**
- RR-interval
- SSR
- plethysmography

**References**

**Modified**
- 2.4.1997, 3.4.97 ES
SENSORY AXONAL POLYNEUROPATHY

Etiology
- paraneoplastic, often related to small cell lung cancer
- Sjögren's syndrome

Clinical features
- loss of sensory modalities with a distal distribution
- paresthesias

Strategy
- demonstrate generalized dysfunction of peripheral sensory nerves.
- differentiate from sensory-motor axonal polyneuropathies

Expected abnormal findings

Neurography
- SCS: AMPL reduced
- SCS: CV may be reduced

Sensory thresholds
- vibration thresholds and thermal thresholds are abnormal

Expected normal findings

Neurography
- MVC

EMG

Autonomic tests
- RR-interval
- SSR
- plethysmography

Procedure

Neurography
- MCS bilateral: n.medianus, n.ulnaris and n.peroneus
- SCS: n.medianus, n.ulnaris, n.radialis and n.suralis bilaterally.

EMG
- one distal and proximal muscle in the lower extremities.

Sensory thresholds (optional)
- temperature and vibration

Autonomic tests (optional)
- RR-interval
- SSR
- plethysmography

References

Modified
- 2.4.1997, 3.4.97 ES

HEREDITARY SENSORY POLYNEUROPATHY TYPE I (HEREDITARY SENSORY NEUROPATHY OF DENNY-BROWN)

Etiology
- autosomal dominant inheritance
- mapped to chromosome 9q22.1-q22.3 in one family

Clinical features
- onset during second decade
- progressive distal extremity sensory loss
- pain and temperature more affected than touch and pressure
- mutilation of feet
- autonomic function usually preserved except sweating distally

Strategy
- demonstrate generalized dysfunction of peripheral sensory nerves
- differentiate from sensory-motor axonal polyneuropathies

Expected abnormal findings

Neurography
- the amplitude of the sensory nerve action potentials are reduced in young patients
- no sensory nerve action potentials obtainable in older patients

Sensory thresholds
- vibration thresholds and thermal thresholds are abnormal

Expected normal findings

Neurography
- MVC

EMG
- all muscles

Autonomic tests
- RR-interval
- SSR
- plethysmography

Procedure
Neurography
* MCS bilateral: n.medianus, n.ulnaris and n.peroneus
* SCS: n.medianus, n.ulnaris, n.radialis and n.suralis bilaterally.

EMG
* one distal and proximal muscle in the lower extremities.

Sensory thresholds (optional)
* temperature and vibration

Autonomic tests (optional)
* RR-interval
* SSR
* plethysmography

References
* Hicks EP. Hereditary perforating ulcer of the foot. Lancet 1922;319-321

Modified
* 1.4.1997 BF, 3.4.97 ES

HEREDITARY SENSORY POLYNEUROPATHY TYPE 2

Etiology
* autosomal recessive inheritance

Clinical features
* rare
* onset in childhood or at birth
* all sensory modalities are affected, touch and pressure earlier than pain and temperature
* hands and feet mutilated

Strategy
* demonstrate generalized dysfunction of peripheral sensory nerves
* differentiate from sensory-motor axonal polyneuropathies

Expected abnormal findings
Neurography
* unobtainable sensory nerve action potentials

Sensory thresholds
* vibration thresholds and thermal thresholds are abnormal

Expected normal findings
Neurography
* MVC

EMG
* RR-interval
* SSR
* plethysmography

Procedure
Neurography
* MCS bilateral: n.medianus, n.ulnaris and n.peroneus
* SCS: n.medianus, n.ulnaris, n.radialis and n.suralis bilaterally.

EMG
* one distal and proximal muscle in the lower extremities.

Sensory thresholds (optional)
* temperature and vibration

Autonomic tests (optional)
* RR-interval
* SSR
* plethysmography

References

Modified
* 1.4.1997 BF, 3.4.97 ES

HEREDITARY SENSORY POLYNEUROPATHY TYPE 3 (FAMILIAL DYSAUTONOMIA, RILEY-DAY SYNDROME)

Etiology
* autosomal recessive inheritance, majority of patients are Jewish
* linked to chromosome 9q31-33
* very rare

Clinical symptoms
* symptoms present from birth
* autonomic dysfunction very prominent
* absent lacrimation, corneal ulceration
* labile sweating, blood pressure and temperature
* diminution of pain and temperature sensation
* touch and pressure preserved
* mutilation unusual
* intelligence normal
* scoliosis common
* decreased life expectancy

**Strategy**
* demonstrate generalized dysfunction of peripheral sensory nerves
* demonstrate generalized dysfunction of the autonomic nervous system
* differentiate from sensory-motor axonal polyneuropathies

**Expected abnormal findings**

**Neurography**
* the amplitude of the sensory nerve action potentials are reduced

**Sensory thresholds**
* vibration thresholds and thermal thresholds are abnormal

**Autonomic function**
* RR-interval
* SSR
* pletysmography

**Expected normal findings**

**Neurography**
* MVC
* EMG
* all muscles

**Procedure**

**Neurography**
* MCS bilateral: n.medianus, n.ulnaris and n.peroneus
* SCS: n.medianus, n.ulnaris, n.radialis and n.suralis bilaterally.

**EMG**
* one distal and proximal muscle in the lower extremities.

**Sensory thresholds (optional)**
* temperature and vibration

**Autonomic tests (optional)**
* RR-interval
* SSR
* pletysmography

**Note**
* A prompt miosis of the pupil in response to 2.5% metacholine is a characteristic

**References**

**Modified**
* 1.4.1997, 3.4.97 ES

**HEREDITARY SENSORY POLYNEUROPATHY TYPE 4 (CONGENITAL SENSORY NEUROPATHY WITH ANHIDROSIS)**

**Etiology**
* autosomal recessive inheritance

**Clinical features**
* symptoms present from birth
* widespread absence of pain and temperature sensation
* preserved touch and pressure sensation
* episodic fever
* self mutilation common
* mental retardation
* short stature

**Strategy**
* demonstrate generalized dysfunction of the autonomic nervous system
* demonstrate dysfunction of unmyelinated and thin myelinated sensory axons
* differentiate from sensory-motor axonal polyneuropathies

**Expected abnormal findings**

**Sensory thresholds**
* thermal thresholds are abnormal

**Autonomic function**
* RR-interval
**Expected normal findings**

**Neurography**
- MVC
- SCV

**EMG**
- all muscles

**Sensory thresholds**
- vibration thresholds

**Procedure**

**Neurography**
- MCS bilateral: n.medianus, n.ulnaris and n.peroneus
- SCS: n.medianus, n.ulnaris, n.radialis and n.suralis bilaterally.

**EMG**
- one distal and proximal muscle in the lower extremities.

**Sensory thresholds (optional)**
- temperature and vibration

**Autonomic tests (optional)**
- RR-interval
- SSR
- plethysmography

**References**
- Rosemberg S, Marie SKN, Kliemann S. Congenital insensitivity to pain with anhidrosis (hereditary sensory and autonomic neuropathy type IV). Pediatr Neurol 1994;11:50-56

**Modified**
- 1.4.1997 BF, 3.4.97 ES

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**Tangier Disease**

**Etiology**
- autosomal recessive inheritance
- hereditary high-density lipoprotein deficiency and widespread tissue storage of cholesteryl esters

**Clinical features**
- onset is mostly after the age of 20 years
- progressive weakness of proximal limb muscles
- pseudo-syringomyelic picture with a dissociated sensory loss

**Strategy**
- demonstrate evidence of an axonal neuropathy
- differentiate from sensory-motor axonal polyneuropathies

**Expected abnormal findings**

**Neurography**
- MVC: normal or slight abnormalities compatible with axonal neuropathy
- SCV: reduced amplitude, normal or slightly reduced CV

**EMG**
- Findings compatible with chronic neuropathy

**Sensory thresholds (vibration thresholds)**
- thermal thresholds are abnormal

**Autonomic function**
- RR-interval
- SSR
- plethysmography

**Expected normal findings**

**Neurography**
- sometimes normal MCS

**EMG**
- sometimes normal findings

**Procedure**

**Neurography**
- MCS bilateral: n.medianus, n.ulnaris and n.peroneus
- SCS: n.medianus, n.ulnaris, n.radialis and n.suralis bilaterally.

**EMG**
- one distal and proximal muscle in the lower extremities.

**Sensory thresholds (optional)**
- temperature and vibration

**Autonomic tests (optional)**
- RR-interval
- SSR
- plethysmography

**References**
**Diphtheria**

**Etiology**
- exotoxin produced by Corynebacterium diphtheriae
- the exact mechanism of peripheral neuropathy is uncertain

**Clinical features**
- incubation period 2-6 days
- fever, sore throat, and characteristic membranous pharyngitis
- 2-4 weeks following the initial infection paralysis occurs in 10-20% of patients
- major attack is on cranial and peripheral nerves
- most frequent manifestation is palatal paralysis
- sometimes peripheral neuropathy resembling polyradiculitis develops 3-8 weeks after the infection
- palatal weakness is the only weakness before 2 weeks after onset
- during 3 to 5 weeks there is pharyngeal paresthesia and paralysis of extracranial muscles,
- during 5-7 weeks weakness of the larynx and diaphragm occur
- sensory-motor polyneuropathy develops 2-3 months after the onset

**Strategy**
- demonstrate sensory-motor polyneuropathy
- differentiate from polyradiculitis

**Expected abnormal findings**

**EMG**
- neurogenic EMG findings in muscles

**Neurography**
- SCS: reduced CV and ampl
- MCS: reduced CV and amplitude

**Procedure**

**EMG**
- m.deltoideus
- m.interosseus
- m.vastus lateralis
- m.tibialis anterior

**Neurography**
- SCS: n.suralis, n.radialis bilaterally
- MCS: n.medianus and n.peroneus bilaterally

**References**

**Leprosy (Hansen’s Disease)**

**Etiology**
- infection by Mycobacterium leprae
- although rarely seen in industrialized countries, it is most common neuropathy in the world
- transmitted through nasal secretions and cutaneous contact
- only a small minority of exposed subjects will be infected
- manifestations depend on the immunological reaction of the host

**Clinical features**

**Lepromatous leprosy**
- low host immune reaction, abundant bacilli
- skin in cooler areas (fingers, toes, pinnae of the ears and nose) of the body become infiltrated with bacilli
- damage to to unmyelinated nerves initially
- characteristically loss of pain and temperature sensation
- loss of pain sensation results in mutilation
- peripheral nerves are thickened
- ulnar nerve at the wrist and elbow and n.peroneus at the knee may be affected leading to motor deficits

**Tuberculoid leprosy**
- strong cell mediated immune reaction leading to localized reaction with tissue damage
- cutaneous depigmented lesions that are anhidrotic and anesthetic
- distribution of skin changes is asymmetric
- most often affected nerves are n.facialis, n.medianus, n.ulnaris and n.peroneus

**Borderline leprosy**
- intermediate form between these two

**Strategy**
- demonstrate affection of distal sensory nerves
- variable affection of n.ulnaris, n.medianus n.peroneus, motor axons may be affected
**Expected abnormal findings**

* EMG
  * distal hand muscles and the peroneal muscles if the nerves are locally affected
* Neurography
  * SCS in the distal parts of the limbs, especially the digital nerves

**Procedure**

* EMG
  * Depends on the region affected
* Neurography
  * SCS: n.suralis, n.radialis, n.peroneus superficialis, n.medianus, n.ulnaris bilaterally
  * MCS: n.medianus, n.ulnaris and n.peroneus bilaterally

**References**

* Ridley DS, Jopling WH: Classification of leprosy according to immunity: A five-group system. Int J Lepr Other Mycobact Dis 1966;34:255-258
* Sabin TD: Neurologic features of lepromatous leprosy. Am Fam Phys 1971;4:84-87

**Modified**

* 15.4.1997

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### 3. NEUROMUSCULAR TRANSMISSION DISORDERS

#### MYASTHENIA GRAVIS (MG)

**Etiology**

* humoral autoimmune response against acetylcholine receptors

**Clinical features**

* incidence 2-10/1000000 per year
  * prevalence 25-125/1000000
  * female preponderance, female to male ratio is 6:4
  * female incidence peaks around 30 years, male around 60-70 years
  * abnormal fatigability and weakness of some or all voluntary muscles
  * muscles innervated by cranial nerves and proximal muscles are more affected than distal muscles
  * bilateral or unilateral ptosis and ocular palsies are often the initial symptom
  * weakness increases during repeated or sustained exertion
  * symptoms are aggravated by heat and often improved in cold
  * spontaneous remissions may occur for varying periods, complete remissions are rare

**Strategy**

* demonstrate postsynaptic neuro-muscular transmission defect
  * exclude other causes of fatigue

**Expected abnormal findings**

* Repetitive stimulation
  * abnormal decrement m.deltoidus/m.nasalis/m.trapezius/m.anconeus (abnormal in 65-80% of patients with generalized MG)

* SFEMG
  * increased jitter

* EMG
  * increased “jiggle”
  * a few fibrillations and positive sharp waves may be seen, rarely moderate amounts
  * MUPs may be brief

* Neurography
  * MCS ampl may be reduced in severe MG

**Expected normal findings**

* Neurography
  * SCS
  * MCS

**Procedure**

<table>
<thead>
<tr>
<th>Rep.stim in a proximal muscle</th>
<th>decrement+ moderate facilitation</th>
<th>EMG: normal or shows jiggle, and decreased duration and a few fibs</th>
<th>MG</th>
</tr>
</thead>
<tbody>
<tr>
<td>↓</td>
<td></td>
<td>as above</td>
<td>LEMS</td>
</tr>
<tr>
<td>if normal then</td>
<td>decrement+ low ampl + pronounced facilitation</td>
<td>as above</td>
<td>MG</td>
</tr>
<tr>
<td>Rep.stim in another weak muscle</td>
<td>decrement+ moderate facilitation</td>
<td>as above</td>
<td></td>
</tr>
</tbody>
</table>
if normal then decrement+ low ampl + pronounced facilitation ➔ as above ➔ LEMS

SFEMG in a weak muscle ➔ increased jitter ➔ as above ➔ MG

if normal then

SFEMG in another weak muscle ➔ increased jitter ➔ as above ➔ MG

if normal then

consider other alternatives ➔ EMG, neurography ➔ other disorders

---

**Note**

* distal muscles show decrement only in 15-20 % of patients with generalized MG

**References**

- Oh SJ; Kim DE; Kuruoglu R; Bradley RJ; Dwyer D. Diagnostic sensitivity of the laboratory tests in myasthenia gravis. Muscle Nerve 1992;15:720-724
- Rivero A; Crovetto L; Lopez L; Maselli R; Nogues M. Single fiber electromyography of extraocular muscles: a sensitive method for the diagnosis of ocular myasthenia gravis. Muscle Nerve 1995;18:943-947
- Rouseev R; Ashby P; Basinski A; Sharpe JA. Single fiber EMG in the frontalis muscle in ocular myasthenia: specificity and sensitivity. Muscle Nerve 1992;15:399-403
- Sanders DB. The electrodiagnosis of myasthenia gravis. Ann NY Acad Sci 1987;505:539-556

**Modified**

* 2.4.1997 BF, 3.4.97 ES

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**NEONATAL MYASTHENIA GRAVIS**

**Etiology**

* occurs in neonates whose mother have myasthenia gravis
* antibodies against acetylcholine receptors diffuse across the placenta from the mother
* neonatal myasthenia develops in 10-15 % of children born to mothers with myasthenia gravis

**Clinical features**

* symptoms appear within the first hours after birth
* generalized weakness, feeding and respiratory weakness
* lasts usually for 3-4 weeks
**Strategy**
- suspect disorder if the mother has myasthenia gravis
- demonstrate postsynaptic neuro-muscular transmission defect

**Expected abnormal findings**
- **Repetitive stimulation**
  - increased decrement in m.deltoides/m.nasalis/m.trapezius/m.anconeus
- **SFEMG**
  - increased jitter
  - MUPs may be brief

**Referenced findings**
- **Neurography**
  - SCS
  - MCS

**Procedure**

1. **confirm MG in the mother**
   - mother has MG
     - Repetitive stimulation in a weak muscle
       - decrement
         - neonatal MG
       - no decrement
         - stimulated SFEMG
           - increased jitter
             - neonatal MG
           - normal jitter
             - consider other disorders

**References**

**MYASTHENIC SYNDROME (LAMBERT-EATON MYASTHENIC SYNDROME, LEMS)**

**Etiology**
- humoral autoimmune response against presynaptic calcium channels
- disturbed neuromuscular transmission

**Clinical features**
- rare
- male preponderance male to female ratio 4.7:1
- 70% of males and 40% of females have an associated malignancy, small cell carcinoma of the lung is most common
- weakness and fatigueability of limb and truncal muscles
- symptoms due to dysfunction of the autonomic nervous system are common: reduced lacrimation, dryness of the mouth, impotence and orthostatism

**Strategy**
- demonstrate presynaptic neuro-muscular transmission defect
- exclude other causes of fatigue

**Expected abnormal findings**
- **Repetitive stimulation**
  - increased decrement (even distal muscles are often affected in LEMS)
  - abnormal facilitation following exercise
  - small CMAP amplitude
SFEMG
  " increased jitter
  EMG
  " increased “jiggle”
  " a few fibrillations and positive sharp waves may be seen, rarely moderate amounts
  " MUPs may be brief

Neurography
  " MCS amplitude is often reduced

**Expected normal findings**

Neurography
  " SCS
  " MCS, except amplitude

**Procedure**

Rep.stim in a distal muscle → decrement+ moderate facilitation → EMG: normal or shows jiggle, and decreased duration and a few fibs → MG

↓

if normal then

↓

Rep.stim in another weak muscle → decrement+ moderate facilitation → as above → LEMS

↓

if normal then

↓

SFEMG in a weak muscle → increased jitter → as above → MG

↓

if normal then

↓

SFEMG in another weak muscle → increased jitter → as above → MG

↓

if normal then

↓

Consider other alternatives → EMG, neurography → other disorders

**Note**

- in contrast to MG, LEMS shows decrement also in distal muscles

**References**

- Schwartz MS, Stålberg E. Myasthenic syndrome studied with single fiber electromyography. Arch Neurol 1975;32:815-817

**Modified**

_ 2.4.1997 BF, 3.4.97_
SLOW CHANNEL SYNDROME

Etiology
- Autosomal dominant inheritance with complete penetrance
- Variable expression; sporadic cases do occur
- MEPP and EPP durations are prolonged
- MEPP amplitude decreased in severely affected muscles
- Quantal content normal
- Prolonged open time of AchR ion channels probably the cause of abnormalities (slow channels)
- On electron microscopy junctional folds are abnormal; the number acetylcholine receptors is reduced

Clinical features
- Onset may be from infancy to adulthood
- Severity and progression varies considerably
- Severe weakness and fatigability of cervical, scapular and finger extensor muscles
- The fatigability and weakness fluctuates but much less than in myasthenia gravis
- Mild involvement of eyelid and extracocular muscles
- Leg muscles are less affected
- Reduced tendon reflexes
- Acetylcholinesterase inhibitors have no effect

Strategy
- Demonstrate neuromuscular transmission defect
- Demonstrate at single stimuli repetitive motor responses

Expected abnormal findings
- EMG: unstable MUP shape in weak muscles
- SF-EMG: increased jitter
- Repetitive stimulation
  - Abnormal decrement at 2-3 Hz stimulation
  - Postactivation exhaustion initially improves decrement then within a minute increased decrement
  - At single stimuli there are repetitive motor responses (seen as abnormal shape of the CMAP) that disappear at stimulus frequencies above 0.2 Hz

Expected normal findings
- Neurography
  - SCS
  - MCS
- Procedure
  - EMG
    - M.deltoides/m.biceps/m.trapezius
    - M.interosseus/m.extensor digitorum communis
    - M.vastus lateralis
    - M.tibialis anterior
  - Neurography
    - SCS: n.suralis, n.radialis unilaterally
    - MCS: n.medianus and n.peroneus unilaterally
  - Repetitive stimulation
    - M.trapezius/m.deltoides
    - Thenar muscles
    - Check for repetitive responses at low stimulus frequencies (present in slow channel syndrome and congenital AChE deficiency)

Note
- M-response shape may be abnormal with irregular terminal part due to extra discharges.

References

Modified
- 30.3.1997 BF, 3.4.97 ES

CONGENITAL ACETYLCHOLINERECPTOR (ACR) DEFICIENCY

Etiology
- Probably autosomal recessive inheritance, but it is more common in men than women
- Number of AcCR reduced
- Small MEPPs
- Poorly developed junctional folds

Clinical features
- Onset at birth or before 2 years
- Ptosis and bulbar muscle involvement
- Mild to moderate fatiguable weakness
- Generally benign course, persists into adult life
- No atrophy or myopathy
- Acetylcholine esterase inhibitors improve symptoms

Strategy
FAMILIAL INFANTILE MYASTHENIA

Etiology
- autosomal recessive inheritance
- miniature end-plate potential amplitude decreases during prolonged activity
- probably progressive decrease of acetylcholine content in synaptic vessels during prolonged stimulation (defect in acetylcholine re-uptake or synthesis)

Clinical features
- presents in infancy or early childhood
- fluctuating ptosis
- involvement of bulbar muscles, weakness of sucking and breathing
- course worsened often by infections and fever
- apneas from respiratory muscle weakness may cause sudden death
- during childhood patients appear normal between crises or have mild weakness
- after 10 years of age patients have easy fatiguability
- responds to acetylcholinesterase inhibitors
- normal tendon reflexes, no muscle atrophy

Strategy
- demonstrate neuromuscular transmission defect

Expected abnormal findings
EMG
- unstable MUP shape in weak muscles
SF-EMG
- increased jitter
Repetitive stimulation
- abnormal decrement at 2-3 Hz stimulation
- postactivation exhaustion initially improves decrement then within a minute increased decrement

Expected normal findings
Neurography
- SCS
- MCS

Procedure
EMG
- m.deltoides/m.biceps/m.trapezius
- m.interosseus/m.extensor digitorum communis
- m.vastus lateralis
- m.tibialis anterior

Neurography
- SCS: n.suralis, n.radialis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

References

Modified
- 30.3.1997 BF, 3.4.97
**CONGENITAL ENDPLATE ACETYLCHOLINE ESTERASE DEFICIENCY (LIMB-GIRDLE MYASTHENIA)**

**Etiology**
- autosomal recessive inheritance
- lack of acetylcholine esterase at the neuromuscular junctions
- MEPP has a normal amplitude but decay the phase is prolonged
- EPP has a reduced amplitude because the quantal content is decreased

**Clinical features**
- onset from birth or before two years
- selective involvement of axial muscles leading to scoliosis in older patients
- bulbar muscles affected
- motor milestones delayed
- the symptoms remain relatively static until the end of the first decade after that progression of symptoms
- acetylcholinesterase inhibitors do not improve symptoms, some patients may get worse
- no muscle atrophy

**Strategy**
- demonstrate neuromuscular transmission defect

**Expected abnormal findings**

**EMG**
- unstable MUP shape in weak muscles

**SF-EMG**
- increased jitter

**Repetitive stimulation**
- abnormally prolonged decay at 2-3 Hz stimulation
- postactivation exhaustion initially improves decrement then within a minute increased decrement
- at single stimuli there are repetitive motor responses that disappear at stimulus frequencies above 0.2 Hz

**Expected normal findings**

**Neurography**
- SCS
- MCS

**Procedure**

**EMG**
- m.deltoides/m.biceps/m.trapezius
- m.interosseus/m.extensor digitorum communis
- m.vastus lateralis
- m.tibialis anterior

**Neurography**
- SCS: n.suralis, n.radialis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

**Repetitive stimulation**
- m.trapezius/m.deltoideus
- thenar muscles
- check for repetitive responses at low stimulus frequencies (present in slow channel syndrome and congenital AChE deficiency)

**Note**
- M-response shape may be abnormal with irregular terminal part due to extra discharges

**References**

**Modified**
- 30.3.1997 BF, 3.4.97 ES

**BOTULINUM INTOXICATION**

**Etiology**
- seven different types neurotoxins A-G. produced by Clostridium botulinum
- the toxin is taken up by peripheral cholinergic nerve terminals where it initially blocks acetylcholine release and subsequently causes reversible denervation of muscle fibers
Clinical features
- infection is acquired when improperly processed foods are eaten
- wound botulism has been described
- in infants the spores of Clostridium botulinum may colonize the gut
- the toxin is destroyed by heat
- after an asymptomatic 12-36 hour (extremes 2 hours to 2 weeks) incubation period nonspecific symptoms of nausea, diarrhea
- ophtalmoplegia, bulbar palsy, respiratory paralysis, paralysis of limb muscles
- pupils dilated
- autonomic symptoms, bradycardia, hypotension, hyperhidrosis
- sensation normal

Strategy
- demonstrate neuromuscular transmission defect and neurogenic EMG findings in muscles

Expected abnormal findings
EMG
- fibrillations and positive sharp waves 7-10 days following the onset
- small polyphasic MUPs
- jiggle

Neurography
- MCS: reduced amplitude

Repetitive stimulation
- decrement at 2-3 Hz
- facilitation after short voluntary activation or at high stimulation frequencies

Expected normal findings

Neurography
- SCS

Procedure
EMG
- m.deltoides/m.biceps/m.trapezius
- m.interosseus/m.extensor digitorum communis
- m.vastus lateralis
- m.tibialis anterior

Neurography
- SCS: n.suralis, n.radialis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

Repetitive stimulation
- m.trapezius/m.deltoides
- thenar muscles

References

Modified
- 2.4.1997, 3.4.97 ES
MYOPATHIES

4 MYOPATHIES

4.1 INFLAMMATORY MYOPATHIES

POLYMYOSITIS

Etiology
- cell mediated autoimmune response against muscle fibers

Clinical features
- incidence 1/9-1000000
- more common in women than men
- usually begins after the age of 20 years
- muscle weakness develops subacutely or insidiously
- proximal limb and neck flexor muscles involved more than other muscles
- muscle pain is sometimes present, but it is not as prominent as one would expect
- dysphagia is common in polymyositis
- CK is usually moderately elevated

Strategy
- demonstrate myopathy
- assess: severity (mild, moderate, severe) and activity (stationary, active)

Expected abnormal findings

EMG
- fibrillations in the acute stage, they disappear in remission
- small, brief MUPs in the acute stage
- later long duration polyphasic MUPs in the chronic stage

Neurography
- if distal muscles are severely involved MCS amp is reduced

Expected normal findings

Neurography
- SCS
- MCS

Procedure

EMG
- m.vastus lateralis/m.vastus medialis
- m.deltoides/m.biceps
- m.interosseus dorsalis I/m.abductor digiti minimi
- m.tibialis anterior
- paravertebral muscles in the low thoracic region should be studied if limb muscles do not show distinct abnormalities

Neurography
- MCS: n.medianus and n.peroneus unilaterally
- SCS: n.radialis and n.suralis unilaterally

NOTE
- in chronic PM, there may be a mixed pattern of small and large MUPs
- it is not unusual for patients with PM to have concurrent PNP, especially if they have malabsorption

References
- Askanas V; Engel WK; Mirabella M. Idiopathic inflammatory myopathies: inclusion-body myositis, polymyositis, and dermatomyositis. Curr Opin Neurol 1994;7:448-56
- Chow WH; Gridley G; Mellemkjaer L; Olsen JH; Fraumeni JF Jr. Cancer risk following polymyositis and dermatomyositis: a nationwide cohort study in Denmark. Cancer Causes Control 1995; 6; 9-13
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- Targoff IN. Diagnosis and treatment of polymyositis and dermatomyositis. Compr Ther 1990; 16: 16-24
- Tymms KE; Beller EM; Webb J; Schrieber L; Buchanan WW. Correlation between tests of muscle involvement and clinical muscle weakness in polymyositis and dermatomyositis. Clin Rheumatol 1990; 9: 523-9

Modified
- 1.4.1997, 3.4.97 ES
**DERMATOMYOSITIS**

**Etiology**

humoral mediated autoimmune response against arterioles in muscles and skin

**Clinical features**

- misery
- muscle weakness: limb, bulbar and respiratory develops subacutely or insidiously
- proximal limb and neck flexor muscles involved more than other muscles
- muscle pain and tenderness are sometimes present
- arthralgia
- skin rash, typically around the eyelids (violaceous)
- skin rash over joints of fingers, knees and ankles
- vasculitis/skin ulcers
- later in the disease calcinocis of the skin
- CK is usually moderately elevated
- course of the disease is variable

**Strategy**

- demonstrate myopathy with spontaneous activity
- assess: severity (mild, moderate, severe) and activity (stationary, active)

**Expected abnormal findings**

**EMG**

- fibrillations in the acute stage, these disappear in remission
- small, brief MUPs in the acute stage
- later long duration, polyphasic MUPs in the chronic stage

**Neurography**

- if distal muscles are severely involved MCS ampl are reduced

**Expected normal findings**

**Neurography**

- SCS
- MCS

**Procedure**

**EMG**

- m.vastus lateralis/m.vastus medialis
- m.deltoides/m.biceps
- m.interosseus dorsalis l/m.abductor digitii minimi
- m.tibialis anterior
- paravertebral muscles in the low thoracic region

**Neurography**

- MCS: n.medianus and n.peroneus unilaterally
- SCS: n.radialis and n.suralis unilaterally

**References**

- Askanas V; Engel WK; Mirabella M. Idiopathic inflammatory myopathies: inclusion-body myositis, polymyositis, and dermatomyositis. Curr Opin Neurol 1994;7:448-56
- Barohn RJ; Amato AA; Sahenk Z; Kissel JT; Mendell JR. Inclusion body myositis: explanation for poor response to immunsuppressive therapy. Neurology 1995; 45: 1302-4
- Chow WH; Gridley G; Mellemkjaer L; McLaughlin JK; Olsen JH; Fraumeni JF Jr. Cancer risk following polymyositis and dermatomyositis: a nationwide cohort study in Denmark. Cancer Causes Control 1995 ; 6: 9-13
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**Modified**

2.4.1997, 3.4.97 ES

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**INCLUSION BODY MYOSITIS AND INCLUSION BODY MYOPATHIES (IBM)**

**Etiology**

IBM has multiple different causes which manifest in a similar pattern

-most patients are considered to be sporadic

-familial cases with dominant and recessive inheritance have been described

-in autosomal recessive for several families with different ethnic backgrounds have bee mapped to chromosome 9p1-q1

-possibly autoimmune T cell-mediated mechanism

**Clinical features**

-onset age > 30 years, usually after 50 years

-duration of weakness >6 months

-muscle weakness affects proximal and distal muscles

-the patient must have at least one of the following:

1. finger flexor weakness
2. wrist flexor weakness > wrist extensor weakness
3. quadriceps weakness >4 MRC scale
* dysphagia occurs in one third of patients
* muscles innervated by cranial nerve are mostly unaffected, but m.orbicularis oculi may be affected
* CK >12 times normal
* in sporadic IBM there is abnormal accumulation of beta amyloid protein and ubiquitin
* biopsy shows invasion of nonnecrotic muscle fibers by mononuclear cells, vacuolated muscle fibers, amyloid deposits or 15-
18 nm tubofilaments

**Strategy**
* demonstrate myopathy with spontaneous activity
* assess: severity: (mild, moderate, severe) and activity: (stationary, active)

**Expected abnormal findings**

**EMG**
* fibrillations
* small, brief MUPs

**Neurography**
* if distal muscles are severely involved MCS ampl are reduced

**Expected normal findings**

**Neurography**
* SCS
* MCS

**Procedure**

**EMG**
* m.flexor carpi radialis/m.flexor digitorum profundus
* m.vastus lateralis/m.vastus medialis
* m.deltoides/m.biceps
* m.interosseus dorsalis l/m.abductor digitii minimi
* m.tibialis anterior

**Neurography**
* MCS: n.medianus and n.peroneus unilaterally
* SCS: n.radialis and n.suralis unilaterally

**Note**
* MRI of the forearm muscles may be helpful in defining the pattern of affected flexor muscles, especially m.flexor digitorum profundus is affected

**References**
* Askanas V; Alvarez RB; Engel WK. Beta-Amyloid precursor epitopes in muscle fibers of inclusion body myositis. Ann Neurol 1993;34:551-60
* Askanas V; Engel WK. New advances in inclusion-body myositis. Curr Opin Rheumatol 1993;5:732-41
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* Lindberg C; Persson LI; Bjorkander J; Oldfors A. Inclusion body myositis: clinical, morphological, physiological and laboratory findings in 18 cases. Acta Neurol Scand 1994;89:123-31
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**Modified**
* 26.6.1997 BF, 2.4.1997 BF, 3.4.97 ES

### 4.2 Muscular Dystrophies (DMD)

**Etiology**
* X-linked recessive inheritance
* mutation located at Xp21.2
* out of frame deletion of the dystrophin gene
37

- 1/3 of the patients are new mutations
- 2/3 of the mothers to Duchenne boys are carriers of the gene

**Clinical features**
- incidence 1/3400 boys
- onset within the first 5 years
- delay in walking, abnormal gait, difficulty in climbing stairs
- proximal weakness, legs more than arms
- hypertrophy of calf muscles
- mild intellectual retardation
- loss of ambulation at 8-12 years
- life expectancy teens to 20 years
- CK very high

**Strategy**
- demonstrate myopathy with a proximal predominance

**Expected abnormal findings**
- EMG, myopathic usually with fibrillations
  - m.deltoides/m.biceps
  - m.vastus lateralis
  - m.tibialis anterior

**Expected normal findings**

**Neurography**
- MCS
- SCS

**Procedure**
- EMG
  - m.deltoides/m.biceps/m.trapezius
  - m.interosseus/m.extensor digitorum communis
  - m.vastus lateralis
  - m.tibialis anterior

**References**

**Modified**
- 30.3.1997, 3.4.97 ES

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**BECKER MUSCULAR DYSTROPHY (BMD)**

**Etiology**
- X-linked recessive inheritance
- mutation located at Xp21.2
- in frame deletion of the dystrophin gene

**Clinical features**
- onset variable, usually after 5 years may at 40 years or even later
- difficulty in running and climbing stairs
- proximal muscle weakness, initially only lower limb muscles
- quadriceps atrophy may be the only manifestation for a long time
- may present with cardiomyopathy
- calf hypertrophy
- slow progression
- ambulation after 16 years
- CK grossly elevated

**Strategy**
- demonstrate myopathy with a proximal predominance

**Expected abnormal findings**
- EMG, myopathic with fibrillations
  - m.deltoides/m.biceps
  - m.vastus lateralis
  - m.tibialis anterior

**Expected normal findings**

**Neurography**
- MCS
- SCS

**Procedure**
- EMG
  - m.deltoides/m.biceps/m.trapezius
  - m.interosseus/m.extensor digitorum communis
  - m.vastus lateralis
  - m.tibialis anterior

**Neurography**
SCS: n.suralis, n.radialis unilaterally
MCS: n.medianus and n.peroneus unilaterally

References

Modified
* 30.3.1997 BF, 3.4.97 ES

FACIO-SCAPULO-HUMERAL MUSCULAR DYSTROPHY (FSH)

Etiology
* autosomal dominant heredity
* linked to chromosome 4q35-4qter
* there is a deletion of an integral number of tandem repeats, there being a maximum of 8 of the original 12-96

Clinical features
* variable onset from childhood to adult age, 90 % will manifest before the age of 20
* an infantile variety has been described
* presenting symptom weakness of facial or shoulder muscles
* facial weakness is present in >50% of affected family members, particularly eye closure is affected
* scapular muscles are affected, particularly m.pectoralis major, however, m.deltoides is spared for a long time
* asymmetry of the muscles is a rule, usually the right side is first affected
* some patients have weakness of the pelvic girdle
* course is variable, some may be mild with normal lifespan, some may loose ambulation in adult life
* cardiomyopathy is not a part of FSH
* CK normal or slightly elevated

Strategy
* demonstrate myopathic changes with facio-scapulo-humeralperoneal distribution

Expected abnormal findings
EMG, myopathic findings in
* m.trapezius
* m.infraspinatus
* m.tibialis anterior
* m.orbicularis oculi

Expected normal findings
Neurography
* SCS
* MCS

Procedure
EMG
* m.biceps brachii/m.triceps brachii
* m.infraspinatus/m.trapezius
* m.interosseus/m.extensor digitorum communis
* m.vastus lateralis
* m.tibialis anterior

Neurography
* SCS: n.suralis, n.radialis unilaterally
* MCS: n.medianus and n.peroneus unilaterally

Note
* m.deltoides may be normal in the early stages of FSH

References

Modified
* 30.3.1997 BF, 3.4.97 ES

EMERY-DREIFUSS MUSCULAR DYSTROPHY

Etiology
* X-linked recessive inheritance also autosomal dominant inheritance has been described with very similar clinical presentation
in X-linked form the gene location is Xq27-28
- mutation in a gene called STA, which encodes a 254 amino acid protein, known as emerin. Emerin is expressed in most tissues, can be demonstrated from skin and leukocytes

**Clinical features**
- onset with pros.
  - late childhood, adolescence, onset after 20 years is rare
  - mild weakness
  - focal wasting of m.biceps brachii and m.triceps brachii or m.gastrocnemii and the peroneal muscles
  - rigidity of spine
  - contractures of the elbow and the Achilles tendon develop before significant weakness
  - equinus of the feet
  - cardiac conduction defect, cardiomyopathy, cardiac arrhythmia life-threatening in early adult life
  - slowly progressive
  - CK moderately elevated but may be normal

**Strategy**
- demonstrate myopathic changes with humero-peroneal distribution, m.biceps brachii is particularly affected

**Expected abnormal findings**
**EMG, a mixture of small and large MUPs**
- m.biceps
- m.tibialis anterior
- paravertebral muscles in the lumbar and cervical region

**Expected normal findings**
**Neurography**
- SCS
- MCS

**Procedure**
**EMG**
- m.deltoides/m.biceps/m.trapezius
- m.interosseus/m.extensor digitorum communis
- m.vastus lateralis
- m.tibialis anterior

**Neurography**
- SCS: n.suralis, n.radialis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

**Note**
- contractures of joints often precede the weakness, especially in the elbow
- altered expression of emerin in patients and carriers can be tested with monoclonal antibodies from skin and leukocytes

**References**
- Emery AEH. X-linked muscular dystrophy with early contractures and cardiomyopathy (Emery-Dreifuss type). Clin Genet 1987,32:360-367
- Manilala, S, Sewry CA, Nguyen thi Man, Muntoni F, Morris GE. Diagnosis of x-linked Emery-Dreifuss dystrophy by protein analysis of leucocytes and skin with monoclonal. Neuromusc Disord

**Modified**

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**Limb-girdle muscular dystrophy**

**Etiology**
- heterogeneous
- most patients are autosomal recessive, two different forms have been found so far, one linked to chromosome 2p and another linked to chromosome 15q15
- calpain-3 deficiency and maps to 15q15
- autosomal dominant: gene located 5q22.1-31.3

**Clinical features**
- onset from early childhood to adult life
- difficulty with gait, running and climbing stairs
- onset with proximal muscle weakness in the pelvic or shoulder girdle muscles or both
- calf hypertrophy is common
- progression usually slow, but may sometimes be rapid
- CK elevated

**Strategy**
- demonstrate myopathic changes with limb-girdle distribution

**Expected abnormal findings**
**EMG**
- m.trapezius
- m.deltoides
- m.vastus lateralis

**Neurography**
- MCS may show reduced amplitude in advanced cases

**Expected normal findings**
**Neurography**
- SCS
- MCS

**Procedure**

**EMG**
- m.deltoides/m.biceps/m.trapezius
- m.interosseus/m.extensor digitorum communis
- m.vastus lateralis
- m.tibialis anterior

**Neurography**
- SCS: n.suralis, n.radialis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

**Note**
Sometimes, m.extensor digitorum brevis is hypertrophic and may show high amplitude MUPs

**References**

**Modified**
- 30.3.1997 BF, 3.4.97 ES

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**CONGENITAL MUSCULAR DYSTROPHIES**

- a group of myopathies often associated with central nervous system abnormalities, currently the group includes the following disorders:
  1. Fukuyama congenital muscular dystrophy
  2. Walker-Warburg syndrome
  3. Muscle eye brain disease (MEB)
  4. Congenital muscular dystrophy with merosin deficiency
  5. Congenital muscular dystrophy with normal merosin
- Each of the disorders will be dealt with under its own heading.

**FUKUYAMA CONGENITAL MUSCULAR DYSTROPHY**

**Etiology**
- autosomal recessive inheritance, gene defect on chromosome 9q31-q33

**Clinical features**
- onset at birth, contractures, mean age of death around 8-10 years, most die by 18 years
- CNS involved, mental retardation, seizures
- no retinal abnormalities
- severe hypotonia, weakness and wasting of muscles
- seizures, mental retardation
- CK moderately high

**Strategy**
- demonstrate myopathic abnormalities
- differentiate from SMA1

**Expected abnormal findings**

**EMG**
- myopathic findings in muscles, proximal muscles tend to be more involved

**Neurography**
- MCS amplitudes may be low

**Expected normal findings**

**Neurography**
- SCS
- MCS: CV and DLAT, F-responses

**Procedure**

**EMG**
- m.interosseus dorsalis I
- m.tibialis anterior
- m.deltoides
- m.vastus lateralis

**Neurography**
- SCS: n.radialis and n.suralis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

**References**
**Walker-Warburg Syndrome**

**Etiology**
- autosomal recessive inheritance
- deficiency of laminin β2 chain, linked to chromosome 3p21

**Clinical features**
- onset at birth
- mental retardation, type II lissencephaly, cerebellar and ocular malformations
- form of lissencephaly (type 2) with agyria
- an absent cortical layer
- absent or small corpus callosum and septum pellucidum
- small, dysplastic cerebellum and brain stem with absence of the posterior vermis
- CT scan will show a cobblestone appearance of the cortex and diffusely abnormal (cystic) white matter, as well as a marked area of translucency in the white matter around the ventricles.
- hydrocephalus has been variously attributed to aqueduct stenosis, Dandy-Walker malformation and herniation of the cerebellar tonsils
- retinal abnormalities
- CK variable
- death in infancy

**Strategy**
- demonstrate myopathic abnormalities
- differentiate from SMA1

**Expected abnormal findings**

**EMG**
- myopathic findings in muscles, proximal muscles tend to be more involved

**Neurography**
- MCS amplitudes may be low

**Expected normal findings**

**Neurography**
- SCS
- MCS: CV and DLAT, F-responses

**Procedure**

**EMG**
- m.interosseus dorsalis I
- m.tibialis anterior
- m.deltoides
- m.vastus lateralis

**Neurography**
- SCS: n.radialis and n.suralis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

**References**

**Modified**
- 30.3.1997 BF, 3.4.97 ES

**Muscle Eye Brain Disease (MEB)**

**Etiology**
- recessive

**Clinical features**
- onset during the first six months of life
- slow motor development, severe mental retardation, hydrocephalus
nystagmus and uncontrolled eye movements, anterior chamber defects with glaucoma, myopia, retinal dystrophy, cataracts and occasionally microspherophakia
epilepsy common
CK slightly elevated
death by 6 to 16 years

**Strategy**

- demonstrate myopathic abnormalities
- differentiate from SMA1

**Expected abnormal findings**

**EMG**

- myopathic findings in muscles, proximal muscles tend to be more involved

**Neurography**

- MCS amplitudes may be low

**Expected normal findings**

**Neurography**

- SCS: CV and DLAT, F-responses

**Procedure**

**EMG**

- m.interosseus dorsalis I
- m.tibialis anterior
- m.deltoides
- m.vastus lateralis

**Neurography**

- SCS: n.radialis and n.suralis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

**References**


**Modified**

- 30.3.1997 BF, 3.4.97 ES

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**CONGENITAL MUSCULAR DYSTROPHY WITH MEROsin DEFIciency**

**Etiology**

- autosomal recessive inheritance, mutation of merosin (laminin alpa-2 chain) gene on chromosome 6q22-23

**Clinical features**

- neonatal hypotonia and muscle weakness
- never learn to walk
- MRI shows significant developmental CNS abnormalities
- intelligence often normal in spite of MRI abnormalities in the brain
- muscle histochemistry shows absence of merosin staining

**Strategy**

- demonstrate myopathic abnormalities
- differentiate from SMA1

**Expected abnormal findings**

**EMG**

- myopathic findings in muscles, proximal muscles tend to be more involved

**Neurography**

- MCS amplitudes may be low

**Expected normal findings**

**Neurography**

- SCS: CV and DLAT, F-responses

**Procedure**

**EMG**

- m.interosseus dorsalis I
- m.tibialis anterior
- m.deltoides
- m.vastus lateralis

**Neurography**

- SCS: n.radialis and n.suralis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

**References**

- Dubowitz V. 22nd ENMC sponsored workshop on congenital muscular dystrophy held in Baarn, The Netherlands, 14-16 May 1993. Neuromusc Disord 1994,4:75-82
**CONGENITAL MUSCULAR DYSTROPHY WITH NORMAL MEROSIN**

**Etiology**
- autosomal recessive inheritance

**Clinical features**
- neonatal hypotonia and muscle weakness
- non-progressive course
- mild disability, learn to walk in contrast to patients with merosin deficiency
- intelligence normal
- contractures
- CK variable, normal to moderately high
- normal merosin on muscle biopsy
- MRI normal CNS

**Strategy**
- demonstrate myopathic abnormalities
- differentiate from SMA1

**Expected abnormal findings**

**EMG**
- myopathic findings in muscles, proximal muscles tend to be more involved

**Neurography**
- MCS amplitudes may be low

**Expected normal findings**

**Neurography**
- SCS, MCS: CV and DLAT, F-responses

**Procedure**
- EMG
  - m.interosseus dorsalis I
  - m.tibialis anterior
  - m.deltoides
  - m.vastus lateralis
- Neurography
  - SCS: n.radialis and n.suralis unilaterally
  - MCS: n.medianus and n.peroneus unilaterally

**References**
- Dubowitz V. 22nd ENMC sponsored workshop on congenital muscular dystrophy held in Baarn, The Netherlands, 14-16 May 1993. Neuromusc Disord 1994,4:75-82
- Topaloglu H, Kale G, Yalnizoglu D, et al. Analysis of "pure" congenital muscular dystrophies in thirty eight cases. How different is the classical type 1 from the occidental type cerebromuscular dystrophy?. Neuropediatrics 1994,25:94-100

**Modified**
- 30.3.1997 BF, 3.4.97

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**OCULOPHARYNGEAL MUSCULAR DYSTROPHY**

**Etiology**
- autosomal dominant inheritance with complete penetration
- located to chromosome 14q11.2

**Clinical features**
- onset during fourth to sixth decades
- ptosis, variable extraocular eye muscle weakness
- dysphagia
- mild proximal limb weakness
- CK normal or mildly elevated

**Strategy**
- demonstrate myopathic abnormalities in muscles innervated by cranial nerves
- differentiate from progressive external ophthalmoplegia (mitochondrial myopathy)

**Expected abnormal findings**

**EMG**
- myopathic abnormalities

**Neurography**
- MCS amplitude may be reduced
**EXPECTED NORMAL FINDINGS**

**Neurography**
- SCS

**Procedure**
- EMG
  - m. orbicularis oculi, m. levator palpebrae
  - m. deltoideus/m. biceps brachii
  - m. interosseus dorsalis l/m. abductor pollicis brevis/m. extensor digitorum communis/m. flexor carpi radialis
  - m. vastus lateralis/m. vastus medialis/m. tensor fascia latae
  - m. tibialis anterior

**Neurography**
- SCS: n. suralis and n. radialis on one side
- MCS: n. peroneus and n. medianus on one side

**References**

**Modified**
- 30.3.1997 BF, 3.4.97 ES

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**PRIMARY ADHALINOPATHY (α-SARCOCYLGACONAPATHY)**

**Etiology**
- autosomal recessive inheritance with complete penetration
- mapped to chromosome 17q12-q21 and in some patients to 13q12
- different subtypes of primary adhalinopathy have been described
- also secondary adhalinopathies exist

**Clinical features**
- clinically similar to Duchenne dystrophy
- onset during average onset around 8 years, ranging from 3 to 15 years
- weakness of proximal muscles most in the pelvic girdle muscles
- distal muscle involvement in the early stages is minimal, m. tibialis anterior predominates
- trunk extensor more affected than abdominal muscles
- neck muscles spared
- calf hypertrophy common
- CK elevated

**Strategy**
- demonstrate myopathic abnormalities mainly in proximal limb and trunk muscles

**EXPECTED ABNORMAL FINDINGS**

**EMG**
- myopathic abnormalities

**Neurography**
- MCS amplitude may be reduced

**EXPECTED NORMAL FINDINGS**

**Neurography**
- SCS

**Procedure**
- EMG
  - m. orbicularis oculi, m. levator palpebrae
  - m. deltoideus/m. biceps brachii
  - m. interosseus dorsalis l/m. abductor pollicis brevis/m. extensor digitorum communis/m. flexor carpi radialis
  - m. vastus lateralis/m. vastus medialis/m. tensor fascia latae
  - m. tibialis anterior

**Neurography**
- SCS: n. suralis and n. radialis on one side
- MCS: n. peroneus and n. medianus on one side

**References**

**Modified**
- 26.6.1997 BF

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**4.3 DISTAL MYOPATHIES**

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**LATE ONSET DISTAL MYOPATHY TYPE 1 (WELANDER TYPE)**

**Etiology**
- autosomal dominant heredity

**Clinical features**
onset around 45-50 years (range 20 to 80 years)
distal hand muscle weakness, especially extension of the thumb and index finger
distal leg muscles are affected later (in contrast to tibial muscle dystrophy which starts in the legs)
slow progression, normal life expectancy
distal tendon reflexes are often absent
CK either normal or mildly elevated
most patients described have Swedish ancestry
in Sweden this myopathy is common around 100 km north from Stockholm in a village called Hedesunda. Local people call this distal myopathy "Hedesunda disease"

Strategy
- demonstrate myopathic changes with distal predominance in arms and legs

Expected abnormal findings
EMG
- m.interosseus dorsalis
- m.extensor digitorum communis
- m.tibialis anterior

Expected normal findings
EMG
- m.biceps brachii
- m.vastus lateralis/m.vastus medialis

Neurography
- SCS
- MCS may show reduced amplitudes

Procedure
EMG
- m.biceps
- m.interosseus
- m.extensor digitorum communis
- m.vastus lateralis
- m.tibialis anterior

Neurography
- SCS: n.suralis, n.radialis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

Note
- EMG often shows fibrillation potentials and complex repetitive discharges. In severely affected muscles reduced interference pattern may be seen.

References

Modified
- 1.4.1997 BF, 3.4.97 ES

LATE ONSET DISTAL MYOPATHY TYPE 2 (MARKESBURY)

Etiology
- autosomal dominant heredity

Clinical features
- starts with weakness of distal leg muscles and spreads later to hand muscles
- late in the course also proximal limb muscles are involved
- cardiomyopathy with congestive heart failure and tachyarrytmias may occur
- CK normal or slightly elevated

Strategy
- demonstrate myopathic changes with distal predominance
- legs affected more than arms

Expected abnormal findings
EMG
- m.tibialis anterior
- m.interosseus dorsalis
- m.extensor digitorum communis
- m.deltoides/m.biceps (later in the disease)
- m.vastus lateralis (later in the disease)

Expected normal findings
EMG (in the early stages of the disease)
- m.biceps brachii
- m.vastus lateralis/m.vastus medialis

Neurography
- SCS
- MCS may show reduced amplitudes

Procedure
EMG
- m.biceps
- m.interosseus
- m.extensor digitorum communis
EARLY ADULT ONSET DISTAL MYOPATHY TYPE 1

**Etiology**

- autosomal recessive or sporadic
- some cases may be identical with hereditary inclusion body myopathy

**Clinical features**

- onset with weakness of the muscles in the anterior compartment of the leg
- slow progression
- hand muscles affected
- proximal limb muscles may be affected later in the disease

**Strategy**

- demonstrate myopathic changes with distal predominance
- legs affected more than arms

**Expected abnormal findings**

**EMG**

- m.tibialis anterior
- m.interosseus dorsalis
- m.extensor digitorum communis
- m.deltoideus/m.biceps (later in the disease)
- m.vastus lateralis (later in the disease)

**Expected normal findings**

**EMG** (in the early stages of the disease)

- m.biceps brachii
- m.vastus lateralis/m.vastus medialis

**Neurography**

- SCS
- MCS may show reduced amplitudes

**Procedure**

**EMG**

- m.biceps
- m.interosseus
- m.extensor digitorum communis
- m.vastus lateralis
- m.tibialis anterior

**Neurography**

- SCS: n.suralis, n.radialis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

**References**


**Modified**

* 1.4.1997, 3.4.97 ES

EARLY ADULT ONSET DISTAL MYOPATHY TYPE 2 (MIYOSHI)

**Etiology**

- autosomal recessive or sporadic
- linked to chromosome 2p12-14

**Clinical features**

- onset second or third decade
- onset of weakness and atrophy of the calf muscles
- usually sparing of intrinsic foot muscles
- severe progressive disorder, results in loss of ambulation
- CK moderately to severely elevated (10 to 150 times normal)

**Strategy**

- demonstrate myopathic changes with distal predominance
- legs affected more than arms

**Expected abnormal findings**

**EMG**

- m.gastrocnemius > m.tibialis anterior
- m.vastus lateralis (later in the disease)

**Expected normal findings**

**Neurography**

- SCS
- MCS may show reduced amplitudes

**Procedure**
EMG
* m.biceps
* m.interosseus
* m.extensor digitorum communis
* m.vastus lateralis
* m.tibialis anterior
Neurography
* SCS: n.suralis, n.radialis unilaterally
* MCS: n.medianus and n.peroneus unilaterally

References
* Barohn RJ, Miller RG, Griggs RC. Autosomal recessive distal dystrophy. Neurology 1991;41:1365-1370

Modified
* 1.4.1997, 3.4.97 ES

TIBIAL MUSCULAR DYSTROPHY

Etiology
* autosomal dominant heredity

Clinical features
* peroneal weakness, starting at the age 35 or later
* hand muscles appear clinically normal
* the muscle abnormalities are most prominent in the muscles peroneal muscles of the leg, however m.extensor digitorum brevis in the foot is spared
* hamstring muscles and calf muscles may be mildly affected
* CK is normal or mildly elevated
* slow progression

Strategy
* demonstrate myopathic changes with distal predominance in the lower extremeties

Expected abnormal findings
EMG
* tibialis anterior, myopathic

Expected normal findings
EMG
* m.biceps brachii/m.deltoideus
* m.vastus lateralis/m.vastus medialis
* m.extensor digitorum brevis shows normal age related abnormalities

Neurography
* all nerves, also n.peroneus (m.extensor digitorum brevis is spared).

Procedure
EMG
* m.interosseus dorsalis I
* m.tibialis anterior
* m.extensor digitorum brevis
* m.deltoideus/m.biceps brachii
* m.vastus lateralis

Neurography
* SCS: n.radialis and n.suralis unilaterally
* MCS: n.medianus and n.peroneus unilaterally

Note
* may show EMG complex repetitive discharges. in severely affected muscles reduced interference pattern may be seen.

References

Modified
* 30.3.1997, 3.4.97 ES

4.4 MYOTONIAS

MYOTONIC DYSTROPHY

Etiology
* autosomal dominant inheritance
* gene location 19q13.2-q13.3
* there is an expanded CGT trinucleotide repeat. In normal subjects the length is up to 30 repeats in patients with myotonic dystrophy up to 2000 repeats can be found. Severity is related with the number of repeats
Gene product myotonin

Clinical features
- myotonic dystrophy is the most common adult myopathy, prevalence is around 5/100,000
- presentation and onset age varies considerably
- onset usually in the early teens with distal muscle weakness in arms and legs
- sometimes myotonic dystrophy presents in a congenital form with hypotonia, bilateral facial weakness, mental retardation, neonatal respiratory distress and talipes; the mother is almost invariably a carrier of the myotonic dystrophy gene in congenital myotonic dystrophy
- facial muscles and sternocleidomastoid muscles are also weak, mild ptosis
- myotonia is usually present and can be demonstrated in the hand muscles
- patients very rarely complain about myotonia (unlike patients with myotonia congenita)
- myotonia is reduced after exercise "warm-up" effect like myotonia congenita.
- myotonic dystrophy affects many other organ systems:
  1. cataracts, retinal dysfunction
  2. CNS, mild mental dysfunction
  3. premature balding
  4. mild polyneuropathy is sometimes seen
  5. endocrine abnormalities
  6. smooth muscle dysfunction

Strategy
- demonstrate myotonic discharges
- demonstrate myopathic abnormalities with a distal distribution

Expected abnormal findings

EMG
- in distal muscles usually abundant myotonic discharges in adults
- trains of positive waves, sometimes difficult to differentiate from myotonic discharges
- short duration, polyphasic MUPs in distal muscles
- typically dense interference pattern, maybe reduced in advanced stages

Neurography
- MCV mostly normal CV, in 10% mild to moderate reduced CV, sometimes low amplitude
- SCS normal

Procedure

EMG
- m.interosseus dorsalis I
- m.tibialis anterior
- m.extensor digitorum communis
- m.masseter/m.orbicularis oris/m. sternocleidomastoideus
- m.deltoides/m.vastus lateralis

Neurography
- SCS: n.radialis and n.suralis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

Note
- some adults (very rarely) with myotonic dystrophy do not have myotonic discharges
- generally children with myotonic dystrophy do not show myotonic discharges
- children with congenital myotonic dystrophy do not show myotonic discharges or any significant abnormalities on EMG
- if congenital myotonic dystrophy is suspected, study also the mother, in almost all children with the congenital form the mother is affected
- around 10% of patients with myotonic dystrophy have a mild polyneuropathy

References

Modified
- 28.3.1997 BF, 3.4.97 ES

Proximal Myotonic Myopathy (PROMM)

Etiology
- autosomal dominant inheritance

Clinical features
- onset of symptoms at the age 20 to 60 years
- myotonia varies over time, may be absent on many days, myotonia has a warm-up effect
- electrical myotonia decreases with cold and increases after heating
- mild proximal weakness especially in the legs
- clinical weakness tends to be proximal but electrophysiological abnormalities are more pronounced distally
- muscle pain is common often disabling
- often polychromatic posterior cataracts
diabetes is common
clinical course is mild

**Strategy**
- demonstrate myotonia, myopathic MUP abnormalities with a proximal preponderance
- differentiate from myotonic dystrophy

**Expected abnormal findings**

**EMG**
- myotonic discharges, especially in distal muscles
- MUP analysis normal or mild myopathic abnormalities, especially in distal muscles

**Expected normal findings**

**Neurography**
- MCS
- SCS

**Procedure**

**EMG**
- m.deltoides/m.biceps brachii
- m.interosseus dorsalis l/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor facia latae
- m.tibialis anterior

**Neurography**
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

**References**


**Revisions**


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**MYOTONIA CONGENITA (THOMSEN´S FORM)**

**Etiology**
- autosomal dominant inheritance
- gene location 7q35
- gene product: muscle Cl⁻ channel, which are formed of tetrameres; the mutation interferes with tetramer formation
- the chloride conductance in the muscle fibre membrane is reduced resulting in accumulation of K⁺ ions in the t-tubules
- the membrane depolarization is prolonged giving rise to spontaneous repeated action potentials

**Clinical features**
- onset in infancy
- myotonia is detected in childhood and persists throughout life
- presentation varies considerably even within families, penetrance around 90%
- myotonia is usually mild
- myotonia decreases after repeated movements, “warm-up”
- muscle strength normal
- often muscles appear hypertrophic
- CK normal or occasionally borderline
- myotonia may aggravated by β₂ agonist drugs

**Strategy**
- demonstrate myotonic discharges without other myopathic features in muscles

**Expected abnormal findings**

**EMG**
- myotonic discharges, particularly distally (sometimes after provocation such as percussion, needle movements, cooling)
- warming up effect - less myotonia after a period of maximal contraction

**Repetitive nerve stimulation**
- decrementing response, especially at high stimulation frequencies (30 Hz) without intratetanic facilitation
- following exercise the amplitude is decreased, reverse to facilitation, cooling does not affect this in contrast to paramyotonia congenita

**Expected normal findings**

**Neurography**
- normal

**EMG**
- MUPs are usually normal

**Procedure**

**EMG**
- m.interosseus dorsalis l
- m.tibialis posterior
- m.extensor digitorum communis
- m.masserter/m.orbicularis oris
MYOTONIA CONGENITA (BECKER’S FORM)

Etiology
* autosomal recessive inheritance
* gene location chromosome 7q35
* gene product: muscle chloride channel

Clinical features
* myotonia manifests at 7-14 years of age, sometimes later and persists throughout life
* myotonia in Becker’s form is more severe than Thomsen’s form
* muscle strength is normal, on exercise strength decreases
* leg and gluteal muscles are usually hypertrophic
* weakness is more prominent in the arms and stiffness in the legs
* lordosis is common
* neck, shoulder and arm muscles appear atrophic
* myotonia decreases after repeated movements, “warm-up”
* male carriers are said to have myotonia, but not females
* CK normal or mildly elevated (2-3 times normal)

Strategy
* demonstrate myotonic discharges, in many patients EMG shows distally mild myopathic features

Expected abnormal findings
EMG
* myotonic discharges, particularly distally
* MUP analysis often shows mild myopathic abnormalities
* warming up effect - less myotonia after a period of maximal contraction

Repetitive nerve stimulation
* decrementing response, especially at high stimulation frequencies (30 Hz)
* following exercise the amplitude is decreased, reverse to facilitation
* cooling does not affect this in contrast to paramyotonia congenita

Expected normal findings
Neurography
* normal

Procedure
EMG
* m.interosseus dorsalis I
* m.tibialis anterior
* m.extensor digitorum communis
* m.masseter/m.orbicularis oris
* m.deltoides/m.vastus lateralis

Neurography
* SCS: n.radialis and n.suralis unilaterally
* MCS: n.medianus and n.peroneus unilaterally

Note
* male carriers show myotonic discharges on EMG, but not female carriers

References

Modified
* 27.3.1997, 3.4.97 ES
MYOTONIA FLUCTUANS

Etiology
- autosomal dominant inheritance
- gene location chromosome 17q13.1
- mutation of muscle alpha-subunit of the sodium channel
- genetic defect allelic to hyperkalemic periodic paralysis and paramyotonia congenita

Clinical features
- myotonia is precipitated by exercise, develops during rest within 30 minutes and lasts for about one hour
- at rest there is no myotonia
- normal muscle strength
- cold does not precipitate myotonia like in paramyotonia congenita
- potassium loading aggravates myotonia
- depolarizing muscle relaxants (e.g. suxamethonium) exacerbates myotonia

Strategy
- demonstrate myotonic discharges at rest with lack of structural myopathic abnormalities
- differentiate from paramyotonia congenita, myotonia congenita and hyperkalemic periodic paralysis

Expected abnormal findings
- EMG
  - myotonic discharges
  - there may fibrillation like spontaneous activity at room temperature
  - following exercise the amount myotonic discharges increase

Expected normal findings
- EMG
  - normal
  - MUP analysis

Neurography
- MCS
  - SCS

Procedure
- EMG
  - m.deltoideus/m.biceps brachii
  - m.interosseus dorsalis I/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
  - m.vastus lateralis/m.vastus medialis/m.tensor faciae latae
  - m.tibialis anterior

- Neurography
  - SCS: n.suralis and n.radialis on one side
  - MCS: n.peroneus and n.medianus on one side

References

Modified
- 28.3.1997, 3.4.97 ES

PARAMYOTONIA CONGENITA

Etiology
- autosomal dominant inheritance
- gene location chromosome 17q13.1
- mutation of muscle alpha-subunit of the sodium channel
- genetic defect allelic to hyperkalemic periodic paralysis and myotonia fluctuans

Clinical features
- symptoms present from birth and persist throughout life
- myotonia is paradoxical, it increases during exercise
- severe worsening of exercise induced myotonia in cold
- predilection of face, neck and distal upper extremity muscles
- weakness after prolonged exercise and cold in most cases
- CK elevated up to 5-10 times normal

Strategy
- demonstrate myotonic discharges, in many patients EMG shows distally mild myopathic features

Expected abnormal findings
- EMG
  - myotonic discharges, much less prominent than in other myotonias
  - cooling will initially induce repetitive spontaneous motor unit discharges, some authors describe fibrillation like activity in cooled muscles
  - with increased cooling myotonia disappears with complete muscle depolarization and paralysis

- Neurography
  - MCS amplitude is reduced in cooled muscles

Expected normal findings
- Neurography
  - normal

Procedure
- EMG
  - m.interosseus dorsalis I
  - m.tibialis anterior
**CHONDRODYSTROPHIC MYOTONIA (SCHWARTZ-JAMPEL SYNDROME)**

**Etiology**
- autosomal recessive inheritance
- localized to 1p34-p36

**Clinical features**
- small stature, multiple skeletal deformities and myopia
- kyphoscoliosis, lumbar lordosis, pectus carinatum, bowing of the long bones, pes planus, a valgus deformity of the ankles and wide metaphyses.
- radiographs show platyspondyly, coronal clefts of the vertebral bodies, and an epiphyseal dysplasia, especially around the hip
- muscle stiffness
- symptoms appear within the first 3 years
- thigh muscles appear hypertrophic, shoulder girdle muscles atrophic
- non-progressive

**Strategy**
- demonstrate continuous spontaneous activity with little modulation of amplitude and frequency (complex repetitive discharges) some authors also describe myotonic discharges. Sometimes this is best seen in m. orbicularis oris.

**Expected abnormal findings**
- myotonic discharges or complex repetitive present from age 7 months onwards

**Expected normal findings**

**Neurography**
- normal SCS
- normal MCS

**Procedure**

**EMG**
- m.interosseus dorsalis I
- m.tibialis anterior
- m.extensor digitorum communis
- m.masseter/m.orbicularis oris
- one proximal muscle: m.deltoideus or m.quadriceps femoris (may show only little abnormalities)

**Neurography**
- SCS: n.radialis and n.suralis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

**Note**
- the children have arthrogryposis, kyfoscoliosis, pectus carinatum and short stature

**References**

**Modified**
- 12.5.1977 BF, 28.3.1997 BF, 3.4.97 ES
4.5 CONGENITAL MYOPATHIES

CENTRAL CORE DISEASE

**Etiology**
- autosomal dominant inheritance
- gene location chromosome 19q12-13.1 (allelic to malignant hyperthermia)
- mutation of ryanodine receptor

**Clinical features**
- onset in infancy, rarely later
- hypotonia, proximal muscles > distal, legs>arms
- extraocular muscles spared, facial muscles little involved
- slowly progressive
- kyphoscoliosis, congenital hip dislocation common, pes cavus
- characteristic histological findings
- CK: high
- slowly progressive

**Strategy**
- demonstrate myopathic abnormalities
- differentiate from SMA1

**Expected abnormal findings**

**EMG**
- myopathic findings in muscles, proximal muscles tend to be more involved

**Expected normal findings**

**Neurography**
- SCS
- MCS

**Procedure**

**EMG**
- m.interosseus dorsalis I
- m.tibialis anterior
- m.deltoides
- m.vastus lateralis

**Neurography**
- SCS: n.radialis and n.suralis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

**References**

**Modified**
- 1.4.1997 BF, 3.4.97 ES

NEMALINE MYOPATHY

**Etiology**
- genetically heterogeneous
- autosomal dominant inheritance common: gene defect localized to chromosome 1q21-23 (mild form)
- autosomal recessive inheritance: chromosome 2q21.2-q22 (severe form)

**Clinical features**
- onset in infancy, hypotonia, proximal muscles > distal
- the weakest muscles were the facial muscles, flexors of the neck and trunk, dorsiflexors of the feet and the extensors of the toes
- extraocular muscles spared
- proximal and distal muscles affected
- no arthrogryposis
- pectus excavatum, kyphoscoliosis, talipes equinovarus
- no cardiac involvement
- nemaline bodies on Gomori trichrome staining
- CK usually normal or slightly elevated

**Strategy**
- demonstrate myopathic abnormalities
- differentiate from SMA1

**Expected abnormal findings**

**EMG**
- myopathic findings in muscles, proximal muscles tend to be more involved, EMG may be normal

**Expected normal findings**

**Neurography**
- SCS
- MCS
**Procedure**

**EMG**
- m.interosseus dorsalis I
- m.tibialis anterior
- m.deltoides
- m.vastus lateralis

**Neurography**
- SCS: n.radialis and n.suralis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

**References**

**Modified**
- 1.4.1997, BF, 3.4.97

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**CENTRONUCLEAR MYOPATHY (MYOTUBULAR MYOPATHY) NEONATAL FORM**

**Etiology**
- X-linked recessive inheritance
- gene location Xq28

**Clinical features**
- neonatal onset
- polyhydramnion
- narrow face and long fingers
- severe hypotonia and weakness
- respiratory insufficiency
- swallowing difficulties
- ophthalmoplegia and ptosis
- fatal outcome, mean life expectancy 5 months
- normal or mild elevation of CK

**Strategy**
- demonstrate myopathic abnormalities

**Expected abnormal findings**

**EMG**
- fibrillations and positive sharp waves, complex repetitive discharges may be seen
- MUP analysis shows myopathic abnormalities

**Neurography**
- MCS may show reduced amplitude

**Expected normal findings**

**Neurography**
- SCS
- MCS normal CV, DLAT, decay and F Waves

**Procedure**

**EMG**
- m.deltoides/m.biceps brachii
- m.interosseus dorsalis/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor facia latae
- m.tibialis anterior

**Neurography**
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

**References**
CENTRONUCLEAR MYOPATHY (MYOTUBULAR MYOPATHY) LATE INFANTILE-LATE CHILDHOOD FORM

Etiology
* autosomal recessive inheritance

Clinical features
* onset late infancy to childhood
* hypotonia
* ptosis, ophthalmoplegia
* skeletal abnormalities
* areflexia
* slowly progressive
* normal or mild elevation of CK

Strategy
* demonstrate myopathic abnormalities

Expected abnormal findings

EMG
* fibrillations and positive sharp waves, complex repetitive discharges may be seen
* MUP analysis shows myopathic abnormalities

Neurography
* MCS may show reduced amplitude

Expected normal findings

Neurography
* SCS
  * MCS normal CV, DLAT, decay and F Waves

Procedure

EMG
  * m.deltoideus/m.biceps brachii
  * m.interosseus dorsalis I/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpII radialis
  * m.vastus lateralis/m.vastus medialis/m.tensor faciae latae
  * m.tibialis anterior

Neurography
  * SCS: n.suralis and n.radialis on one side
  * MCS: n.peroneus and n.medianus on one side

References


Modified
* 1.4.1997 BF, 3.4.97 ES

CENTRONUCLEAR MYOPATHY (MYOTUBULAR MYOPATHY) LATE CHILDHOOD-ADULT TYPE

Etiology
* autosomal dominant inheritance

Clinical features
* onset late childhood to adult age
* mild limb-girdle weakness
* facial and external ocular muscles may be affected
* normal or mild elevation of CK
* slowly progressive

Strategy
* demonstrate myopathic abnormalities

Expected abnormal findings

EMG
* fibrillations and positive sharp waves, complex repetitive discharges may be seen
* MUP analysis shows myopathic abnormalities

Neurography
* MCS may show reduced amplitude

References


Modified
* 1.4.1997 BF, 3.4.97 ES
**Expected normal findings**

**Neurography**
- SCS
- MCS normal CV, DLAT, decay and F-waves

**Procedure**

**EMG**
- m.deltoides/m.biceps brachii
- m.interosseus dorsalis l/m. abductor pollicis brevis/m. extensor digitorum communis/m. flexor carpi radialis
- m.vastus lateralis/m. vastus medialis/m. tensor facia latae
- m.tibialis anterior

**Neurography**
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

**References**

Modified
- 1.4.1997 BF, 3.4.97 ES

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**MULTICORE DISEASE**

**Etiology**
- probably autosomal dominant inheritance

**Clinical features**
- delayed motor milestones
- proximal muscle weakness
- rarely ptosis and ophthalmoplegia
- skeletal abnormalities (slender body, high arched plate, clubfeet, scoliosis)
- predisposition to malignant hypertermia
- non-progressive
- CK normal

**Strategy**
- demonstrate myopathic abnormalities
- EMG findings not specific

**Expected abnormal findings**

**EMG**
- variable degrees of myopathic abnormalities

**Neurography**
- MCS may show reduced amplitude

**Expected normal findings**

**Neurography**
- SCV
- MCV: nerve conduction velocity, decay and F-waves

**Procedure**

**EMG**
- m.deltoides/m.biceps brachii
- m.interosseus dorsalis l/m. abductor pollicis brevis/m. extensor digitorum communis/m. flexor carpi radialis
- m.vastus lateralis/m. vastus medialis/m. tensor facia latae
- m.tibialis anterior

**Neurography**
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

**References**

**Modified**
- 1.4.1997, 3.4.97 ES

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**MINI CORE DISEASE**

**Etiology**
- sporadic or autosomal recessive

**Clinical features**
- onset in infancy rarely later
- onset in infancy, hypotonia, proximal muscles > distal
- ptosis, extraocular muscles weak
- cardiac involvement
- non progressive
"distinctive electron microscopy findings

**Strategy**
- demonstrate myopathic abnormalities, sometimes EMG may be normal
- differentiate from SMA1

**Expected abnormal findings**

**EMG**
- myopathic findings in muscles, proximal muscles tend to be more involved

**Expected normal findings**

**Neurography**
- SCS
- MCS

**Procedure**

**EMG**
- m.interosseus dorsalis I
- m.tibialis anterior
- m.deltoides
- m.vastus lateralis

**Neurography**
- SCS: n.radialis and n.suralis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

**References**

**FINGERPRINT BODY MYOPATHY**

**Etiology**
- pathogenesis is unknown
- rare
- not certain that this is a separate entity, fingerprint like structures can be seen in other myopathies

**Clinical features**
- hypotonia in infancy
- truncal and extremity weakness
- cranial muscles spared
- non-progressive or slowly progressive
- mental retardation
- electron microscopy shows typical fingerprint patterns

**Strategy**
- demonstrate myopathic abnormalities
- differentiate from SMA1

**Expected abnormal findings**

**EMG**
- myopathic findings

**Expected normal findings**

**Neurography**
- SCS
- MCS

**Procedure**

**EMG**
- m.interosseus dorsalis I
- m.tibialis anterior
- m.deltoides
- m.vastus lateralis

**Neurography**
- SCS: n.radialis and n.suralis unilaterally
- MCS: n.medianus and n.peroneus unilaterally

**References**

**HYALINE BODY MYOPATHY**

**Etiology**
- sporadic occurrence
- etiology not known

**Clinical features**
- hypotonia
- delayed milestones
- weakness of proximal muscles

**References**
slow progression or stationary
CK normal

**Strategy**
* demonstrate myopathic abnormalities in more severe proximal than in distal muscles

**Expected abnormal findings**
**EMG**
* myopathic abnormalities
**Neurography**
* MCS may show reduced amplitudes

**Expected normal findings**
**Neurography**
* MCS

**Procedure**
**EMG**
* m.deltoidus/m.biceps brachii
* m.interosseus dorsalis l/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
* m.vastus lateralis/m.vastus medialis/m.tensor facia latae
* m.tibialis anterior

**Neurography**
* SCS: n.suralis and n.radialis on one side
* MCS: n.peroneus and n.medianus on one side

**References**
* Barohn RJ, Brumback RA, Mendell JR. Hyaline body myopathy. Neuromusc Disord 1994;4:257-262

**Modified**
* 1.4.1997 BF, 3.4.97 ES

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**4.6 MITOCHONDRIAL MYOPATHIES**

**MYOCLONIC EPILEPSY AND RAGGED RED FIBERS (MERFF)**

**Etiology**
* maternal inheritance
* point mutation of mitochondrial DNA, an adenine to guanine substitution at nucleotide 8344 in the transfer-RNA gene for lysine
* the mitochondrial DNA population is heteroplasmic, there is mutant and wild type DNA is varying proportions

**Clinical features**
* early development is usually normal
* onset of symptoms varies from childhood to adult age (5 to 50 years)
* myopathy with proximal weakness
* myoclonus
* generalized seizures
* ataxia
* hearing loss and deafness
* intellectual deterioration
* CK normal or mildly elevated
* muscle biopsy shows ragged red fibers on Gomori trichrome staining
* electron microscopy shows abnormal mitochondria
* some patients have neuropathy

**Strategy**
* demonstrate myopathic abnormalities in proximal muscles
* EEG shows generalized slowing and generalized spike and wave discharges

**Expected abnormal findings**
**EMG**
* MUP analysis shows myopathic abnormalities
**Neurography**
* may show slight abnormalities

**Expected normal findings**
**Neurography**
* may be normal

**Procedure**
**EMG**
* m.frontalis
* m.deltoidus/m.biceps brachii
* m.interosseus dorsalis l/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
* m.vastus lateralis/m.vastus medialis/m.tensor facia latae
* m.tibialis anterior

**Neurography**
* SCS: n.suralis and n.radialis on one side
* MCS: n.peroneus and n.medianus on one side

**References**
MITOCHONDRIAL MYOPATHY, ENCEPHALOPATHY, LACTIC ACIDOSIS AND STROKELIKE EPISODES (MELAS)

**Etiology**
- may be familial, but far less often than MERRF
- point mutation of mitochondrial DNA, an adenine to guanine substitution occurs in the tRNA for leucine at nucleotide 3243.
  - the mutant DNA coexists with normal DNA

**Clinical features**
- early development normal
- begins usually between 3 and 11 years, before 20 years
- stroke like episodes
- seizures
- short stature
- episodic vomiting
- hearing loss
- intellectual deterioration
- myopathy
- CK may be elevated
- lactate, especially CSF lactate elevated

**Strategy**
- evaluate presence of myopathic abnormalities

**Expected abnormal findings**

**EMG**
- may show mild myopathic abnormalities

**EEG**
- generalized slowing
- focal abnormalities may be seen
- focal and generalized epileptiform discharges

**Expected normal findings**

**EMG**
- may be normal

**Neurography**
- SCS
- MCS

**Procedure**

**EMG**
- m.frontalis
- m.deltoides/m.biceps brachii
- m.interosseus dorsalis l/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor faciae latae
- m.tibialis anterior

**Neurography**
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

**EEG**

**References**

**Succinate dehydrogenase deficiency (complex II deficiency)**

**Etiology**
- Succinate dehydrogenase is coded by nuclear DNA, autosomal recessive inheritance has been described

**Clinical Features**
- presents in childhood with exercise intolerance
- rhabdomyolysis and myoglobinuria may be present
- between attacks normal neurologic examination
in relation to attacks elevated CK and abnormal EMG
* cardiac muscle spared

**Strategy**
* usually normal EMG between attacks
* demonstrate abnormal EMG in relation to attacks

**Expected abnormal findings in relation to attacks**

**EMG**
* fibrillations and positive sharp waves
* MUP analysis myopathic

**Neurography**
* reduced MCS amplitude may be present in severe cases

**Expected normal findings between attacks**

**EMG**

**Neurography**

**Procedure**

**EMG**
* m.frontalis
* m.deltoides/m.biceps brachii
* m.interosseous dorsalis l/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
* m.vastus lateralis/m.vastus medialis/m.tensor facia latae
* m.tibialis anterior

**Neurography**
* SCS: n.suralis and n.radialis on one side
* MCS: n.peroneus and n.medianus on one side

**References**

Modified
* 1.4.1997, 3.4.97 ES

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**FATAL INFANTILE MYOPATHY WITH CYTOCHROME C OXIDASE DEFICIENCY (COMPLEX IV DEFICIENCY)**

**Etiology**
* occurs as sporadic and autosomal recessive forms
* genetic defect nor known
* selective absence of subunits VII a, b of cytochrome C oxidase

**Clinical features**
* normal at birth
* hypotonia and weakness develops during first weeks
* nearly all patients have renal tubular defect leading to aminoaciduria
* death by 1 year

**Strategy**
* demonstrate myopathic abnormalities

**Expected abnormal findings**

**EMG**
* myopathic abnormalities

**Neurography**
* MCS may show reduced amplitudes

**Expected normal findings**

**Neurography**
* SCS

**Procedure**

**EMG**
* m.frontalis
* m.deltoides/m.biceps brachii
* m.interosseous dorsalis l/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
* m.vastus lateralis/m.vastus medialis/m.tensor facia latae
* m.tibialis anterior

**Neurography**
* SCS: n.suralis and n.radialis on one side
* MCS: n.peroneus and n.medianus on one side

**References**

Modified
* 1.4.1997, 3.4.97 ES
**Benign Infantile Myopathy with Cytochrome C Oxidase Deficiency (Complex IV Deficiency)**

**Etiology**
- Sporadic
- Genetic defect not known
- Selective absence of subunits VIIa, b and II of cytochrome C oxidase

**Clinical features**
- Normal at birth
- Hypotonia and weakness develops during first weeks
- In spite of severe weakness often requiring ventilatory assistance the children recover and become normal by 2-3 years of age.
- CK is elevated, lactic acidosis

**Strategy**
- Demonstrate myopathic abnormalities

**Expected abnormal findings**
- EMG: Myopathic abnormalities
- Neurography: MCS may show reduced amplitudes

**Expected normal findings**
- Neurography: SCS
- EMG: m. frontalis, m. deltoideus/m. biceps brachii, m. intersseus dorsalis/ m. abductor pollicis brevis, m. extensor digitorum communis, m. flexor carpi radialis, m. vastus lateralis/m. vastus medialis/m. tensor facia latae, m. tibialis anterior
- Neurography: SCS: n. suralis and n. radialis on one side, MCS: n. peroneus and n. medianus on one side

**References**

**Leigh’s Syndrome**

**Etiology**
- Selective absence of all subunits of cytochrome c oxidase
- Genetically heterogeneous
- Autosomal recessive inheritance, X-linked inheritance and mitochondrial inheritance have been described

**Clinical features**
- Onset usually during first year of life, sometimes in the neonatal period, rarely in adults
- First stage between 8-12 months is mostly with vomiting and failure to thrive
- Second stage in infancy by motor regression, eye signs and altered breathing
- Third stage between 2-10 years is of hypotonia and dysphagia
- Some patients present with dystonia
- Hypotonia
- Episodic vomiting and feeding problems
- Sphincter
- Hearing and visual loss
- Loss of motor and verbal skills
- No muscular weakness although there is absence of all subunits of cytochrome c oxidase
- Muscle biopsy can be used for the diagnosis

**Strategy**
- Demonstrate normal EMG and neurography findings
- EEG shows abnormal findings

**Expected normal findings**
- EMG
- Neurography: SCS: n. suralis and n. radialis on one side, MCS: n. peroneus and n. medianus on one side
- EEG

**References**
KEARNS-SAYRE SYNDROME

Etiology
- mitochondrial DNA shows deletions, usually there is a mixture of abnormal DNA with deletions and normal wild type DNA, the ratio determines the severity
- sporadic, very rarely transmitted

Clinical features
- onset before the age of 20
- progressive external ophthalmoplegia, ptosis
- myopathy
- pigmentary degeneration of the retina
- heart block
- ataxia
- hearing loss
- dementia
- peripheral neuropathy
- elevated spinal fluid proteins
- hypothyroidism

Strategy
- demonstrate myopathic abnormalities with proximal preponderance
- demonstrated sensory motor polyneuropathy

Expected abnormal findings

EMG
- mild myopathic abnormalities in muscles

Neurography
- MCS shows normal or slightly reduced CV, abnormal F-waves
- SCS shows reduced amplitudes and slight reduction of CV

EEG
- nonspecific generalized slowing

Expected normal findings

EMG
- in mild forms the EMG may be normal

Procedure

EMG
- m.frontalis
- m.deltoides/m.biceps brachii
- m.interosseus dorsalis l/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor facia latae
- m.tibialis anterior

Neurography
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

EEG

References

FAMILIAL PROGRESSIVE EXTERNAL OPHTALMOPLEGIA (PEO) SYNDROME

Etiology
- genetically heterogeneous
- maternal inheritance, autosomal dominant inheritance and autosomal recessive inheritance has been described
in the form with maternal inheritance mitochondrial DNA deletions have not been found
in the dominant for multiple deletions are found

**Clinical features**
- progressive external ophthalmoplegia, ptosis
- proximal muscle weakness
- in the maternally inherited form no systemic effects like heart block or CNS symptoms like in Kearns-Sayre
- in the dominantly inherited form there is ataxia, hearing loss and peripheral sensory motor polyneuropathy (the dominant form may be clinically similar to Kearns-Sayre
- CK is normal or slightly elevated, lactate and pyruvate may be elevated

**Strategy**
- demonstrate myopathic abnormalities with proximal preponderance
- in the dominant form there is also sensory motor polyneuropathy

**Expected abnormal findings**

**EMG**
- mild myopathic abnormalities in muscles, particularly in facial muscles

**Neurography**
- signs of axonal sensory motor polyneuropathy seen in dominant for of PEO

**Expected normal findings**

**EMG**
- in mild forms the EMG may be normal

**Neurography**
- in the maternally inherited form normal findings

**Procedure**

**EMG**
- m.frontalis
- m.deltoides/m.biceps brachii
- m.interosseus dorsalis l/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpii radialis
- m.vastus lateralis/m.vastus medialis/m.tensor facia latae
- m.tibialis anterior

**Neurography**
- MCS: n.suralis and n.radialis on one side
- MCS: n.peroneous and n.medianus on one side

**References**

**Modified**
- 1.4.1997, 3.4.97 ES

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**LATE ONSET MITOCHONDRIAL MYOPATHY**

**Etiology**
- focal accumulation of deleted mitochondrial DNA in and altered energy status in muscles
- the mitochondrial alterations may due to exaggerated form of changes seen with normal aging in mitochondria
- histology shows ragged red fibers, fibers negative for cytochrome c oxidase (COX)

**Clinical features**
- onset after 65 years of age
- proximal muscle weakness and fatiguability
- ptosis in some patients

**Strategy**
- demonstrate mild myopathic changes in muscles

**Expected abnormal findings**

**EMG**
- mild myopathic changes, especially in proximal muscles

**Expected normal findings**

**EMG**
- in some described patients EMG has been normal

**Neurography**
- MCS
- SCS

**Procedure**

**Neurography**
- MCS: n.medianus/n. ulnaris and n.peroneous/tibialis on one side
- SCS: n.radialis and n.suralis on one side.

**EMG**
- one distal and proximal muscle in the lower extremities
- one distal and proximal muscle in the upper extremities
- paravertebral muscle in the thoracic region

**References**

**Modified**
- 2.4.1997, 3.4.97 ES Björn, var kommer Torbergsens avhandling in, maternell mitokondriesjukdom med multiborgan-defekter
4.7 METABOLIC MYOPATHIES

ACID MALTASE DEFICIENCY (POMPE’S DISEASE, GLYCOGENOSIS TYPE 2)

**Etiology**
- autosomal recessive inheritance
- gene localized to chromosome 17q21-23
- at least six different mutations have been described
- three clinical forms: infantile, childhood and adult

**Clinical features**
- **infantile**: floppy infants, progressive muscle weakness, heart, liver and CNS involved, death usually before age of 2
- **childhood**: delayed motor milestones and progressive proximal muscle weakness
- **adult**: progressive proximal weakness, resembles limb-girdle dystrophy
- specific diagnosis depends on demonstration of acid maltase deficiency in fibroblasts or lymphocytes

**Strategy**
- demonstrate myopathic abnormalities, especially in proximal muscles

**Expected abnormal findings**

**EMG**
- EMG shows fibrillations and positive sharp waves, especially in paravertebral muscles
- MUP analysis shows typical myopathic abnormalities

**Neurography**
- MCS may show reduced amplitude

**Expected normal findings**

**Neurography**
- SCS

**Procedure**

**EMG**
- m.deltoides/m.biceps brachii
- m.interosseous dorsalis/lm.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpii radialis
- m.vastus lateralis/m.vastus medialis/m.tensor facia latae
- m.tibialis anterior

**Neurography**
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

**References**

**Modified**
- 1.4.1997 BF, 3.4.97 ES

DEBRANCHING ENZYME DEFICIENCY (GLYCOGENOSIS TYPE 3)

**Etiology**
- autosomal recessive inheritance
- gene has been cloned and is localized to chromosome 1
- defect of either debranching enzyme transferase or debranching enzyme glucosidases (either one or both)

**Clinical features**
- may present in childhood or as adults
- liver is often affected, hepatomegaly
in adults proximal and sometimes also distal weakness, in children hypotonia
stiffness and cramping in exercising and myoglobinuria are nor common
CK elevated 2-20 times normal
final diagnosis is based on biochemical assay of muscle tissue

**Strategy**

- demonstrate myopathic abnormalities in muscles

**Expected abnormal findings**

**EMG**
- weak muscles show abundant fibrillations, complex repetitive discharges
- MUP analysis: typical myopathic abnormalities

**Neurography**
- in some patients mild abnormalities

**Expected normal findings**

**Neurography**
- most patients show normal findings

**Procedure**

**EMG**
- m.deltoideus/m.biceps brachii
- m.interosseus dorsalis l/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor facia latae
- m.tibialis anterior

**Neurography**
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

**References**

**BRANCHING ENZYME DEFICIENCY (GLYCOGENOSIS TYPE 4)**

**Etiology**
- autosomal recessive inheritance
- deficiency of branching enzyme

**Clinical features**
- onset in infancy with failure to thrive and liver failure
- later hypotonia and muscle weakness
- death occurs in second year
- definite diagnosis is based on demonstration of deficiency of branching enzyme in fibroblasts

**Strategy**

**Expected abnormal findings**

**EMG**
- EMG findings have nor been described in the literature

**Neurography**
- not known

**Procedure**

**EMG**
- m.deltoideus/m.biceps brachii
- m.interosseus dorsalis l/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor facia latae
- m.tibialis anterior

**Neurography**
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

**References**

**Modified**
- 1.4.1997, 3.4.97 ES

**MYOPHOSPHORYLASE DEFICIENCY (MCARDLE’S DISEASE, GLYCOGENOSIS TYPE 5))**

**Etiology**
- usually autosomal recessive inheritance, sometimes autosomal dominant inheritance
- gene location: 11q13
- myophosphorylase deficiency

**Clinical features**
- exercise intolerance and easy fatiguability
stiffness and cramping in exercising
myalgia
myoglobinuria
mild proximal weakness in one third of the patients, increases with age
can clinically variable from very severe to very mild

Strategy
- demonstrate myopathic abnormalities
- exercise induced painful cramps are electrically silent unlike common cramps
- high frequency repetitive stimulation shows decrement
- definite diagnosis is based on histochemical demonstration of lack of myophosphorylase or DNA analysis

Expected abnormal findings

EMG
- the EMG may show fibrillations and positive sharp waves
- MUP analysis shows typical myopathic abnormalities
- exercise induced cramps are electrically silent

Neurography
- MCS may show reduced amplitude

Decrement
- hypothenar muscles with 20 Hz stimulation for 50 sec shows more than 25 % decrement

Expected normal findings

Neurography
- normal SCS

Procedure
EMG
- m.deltoides/m.biceps brachii
- m.interosseus dorsalis /m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor facia latae
- m.tibialis anterior

Neurography
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

Decrement
- hypothenar muscles with 20 Hz stimulation for 50 sec

Exercise induced cramp
- electrical silence

References
- Chui LA, Munsat TL. Dominant inheritance of McArdle syndrome. Arch Neurol 1976;33:636-41

Modified
- 1.4.1997, 3.4.97 ES

PHOSPHOFRUCTOKINASE DEFICIENCY (TARUI’S DISEASE, GLYCOGENOSIS TYPE 7)

Etiology
- may be genetically heterogeneous
- autosomal recessive inheritance
- gene location: 1cen32 and 12q have been described
- lack of phosphofructokinase
- male preponderance, male: female ratio 9:1 is not understood

Clinical features
- onset in childhood
- exercise intolerance and easy fatigability
- stiffness and cramping in exercising
- myalgia
- myoglobinuria
- episodes of hemolysis and jaundice may occur, often mild hemolytic anemia
- can clinically be very variable from very severe to very mild
- CK normal or mildly elevated

Strategy
- demonstrate myopathic abnormalities
- high frequency repetitive stimulation shows decrement
- definite diagnosis is based on histochemical demonstration of lack of phosphofructokinase

Expected abnormal findings

EMG
the EMG may show fibrillations and positive sharp waves
- MUP analysis shows typical myopathic abnormalities
- exercise induced cramps are electrically silent

**Neurography**
- MCS may show reduced amplitude

**Decrement**
- hypothenar muscles with 20 Hz stimulation for 50 sec shows more than 25 % decrement

**Expected normal findings**

**Neurography**
- normal SCS

**Procedure**

**EMG**
- m.deltoides/m.biceps brachii
- m.interosseus dorsalis/lm.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor facia latae
- m.tibialis anterior

**Neurography**
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

**References**

**Modified**
- 1.4.1997, 3.4.97 ES

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**PHOSPHOGLYCEERATE KINASE DEFICIENCY (GLYCOGENOSIS TYPE 9)**

**Etiology**
- X-linked recessive inheritance
- gene location 1q13
- phosphoglycerate kinase deficiency

**Clinical features**
- most patients have CNS symptoms, mental retardation and seizures
- some may have a purely myopathic syndrome with progressive proximal muscle weakness and episodes of myoglobinuria
- exercise intolerance, easy fatigability
- CK elevated
- forearm lactate test - no elevation of lactate

**Strategy**
- EMG is usually normal

**Expected normal findings**

**EMG**
- m.deltoides/m.biceps brachii
- m.interosseus dorsalis/lm.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor facia latae
- m.tibialis anterior

**Neurography**
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

**References**

**Modified**
- 1.4.1997, 3.4.97 ES

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**PHOSPHOGLYCEERATE MUTASE DEFICIENCY (GLYCOGENOSIS TYPE 10)**

**Etiology**
- autosomal recessive inheritance
- gene location 7p12-p13
- phosphoglycerate mutase deficiency

**Clinical features**
- most patients have CNS symptoms, mental retardation and seizures
- some may have a purely myopathic syndrome with progressive proximal muscle weakness and episodes of myoglobinuria
- exercise intolerance, easy fatigability
- CK elevated
- forearm lactate test - subnormal elevation of lactate

**References**

**Modified**
- 1.4.1997, 3.4.97 ES
Strategy
* demonstrate normal EMG between attacks

**Expected normal findings**

**EMG**

**Neurography**

**Procedure**

**EMG**
* m.deltoideus/m.biceps brachii
* m.interosseus dorsalis I/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
* m.vastus lateralis/m.vastus medialis/m.tensor facia latae
* m.tibialis anterior

**Neurography**
* SCS: n.suralis and n.radialis on one side
* MCS: n.peroneus and n.medianus on one side

**References**

**Modified**
* 1.4.1997, 3.4.97 ES

**LACTATE DEHYDROGENASE DEFICIENCY (GLYCOGENOSIS TYPE II)**

**Etiology**
* autosomal recessive inheritance
* localized to chromosome 11p15.4

**Clinical features**
* excessive fatigue
* exercise intolerance
* myalgia
* myoglobinuria
* normal muscle strength
* often skin rash
* forearm exercise test does not increase lactate

**Strategy**
* demonstrate normal EMG findings between attacks

**Expected normal findings**

**EMG**

**Neurography**

**Procedure**

**EMG**
* m.deltoideus/m.biceps brachii
* m.interosseus dorsalis I/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
* m.vastus lateralis/m.vastus medialis/m.tensor facia latae
* m.tibialis anterior

**Neurography**
* SCS: n.suralis and n.radialis on one side
* MCS: n.peroneus and n.medianus on one side

**References**

**Modified**
* 1.4.1997 BF, 3.4.97 ES

**CARNITINE DEFICIENCY**

**Etiology**
* autosomal recessive inheritance or acquired in the secondary form
* carnitine deficiency

**Clinical features**
* *myopathic form*: progressive, painless proximal muscle weakness in childhood or early adult life (infantile form has been described)
* *systemic form*: presents in early childhood with progressive cardiomyopathy and attacks resembling Reye’s syndrome. Proximal muscle weakness. Death often occurs during the attacks before the age of 30
* *secondary form*: may be caused by renal failure, pregnancy, malnutrition, valproate therapy, liver failure, Reye’s syndrome, myopathies, mitochondrial disorders
* CK normal in 50, others elevated

**Strategy**
* demonstrate myopathic abnormalities

**Expected abnormal findings**
EMG
- some fibrillations and positive sharp waves may occur
- MUP analysis shows myopathic abnormalities

Neurography
- MSC may show reduced amplitude

Expected normal findings

Neurography
- SCS
- MCS

Procedure
EMG
- m.deltoideus/m.biceps brachii
- m.interosseus dorsalis/l.m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor fascia latae
- m.tibialis anterior

Neurography
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

References
* Brengstall GN. Carnitine deficiency syndromes. Pediatr Neurol 1990;6:75-81

Modified
* 1.4.1997, 3.4.97 ES

CARNITINE PALMITYL TRANSFERASE DEFICIENCY

Etiology
- autosomal recessive inheritance
  - CPT gene is localized to chromosome 11p11-p13
  - deficiency of carnitine palmitoyl transferase
  - male:female ratio 5:1
  - autosomal dominant inheritance has been described

Clinical features
- episodes of myalgia, cramps and myalgia following prolonged exercise (>40 min) or fasting.
  - between attacks muscle weakness is uncommon
  - episodes can be triggered by general anesthesia, cold, emotional stress, low intake of carbohydrates, lack of sleep and high intake of lipids
  - most common cause for myoglobinuria
  - diagnosis is based on biochemical analysis of muscle tissue

Strategy
- EMG is usually normal between episodes

Expected normal findings between attacks

EMG

Neurography

Procedure
EMG
- m.deltoideus/m.biceps brachii
- m.interosseus dorsalis/l.m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor fascia latae
- m.tibialis anterior

Neurography
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

References
* DiMauro S, Melis di Mauro PM. Muscle carnitine palmitoyltransferase deficiency and myoglobinuria. Science 1973;182:929-931

Modified
* 1.4.1997, 3.4.97 ES

MYOADENYLATE DEAMINASE DEFICIENCY

Etiology
- the clinical significance of myoadenylate deaminase deficiency is not clear, it is a benign state, symptom free athletes with myoadenylate deaminase deficiency have been described

References

Modified
* 1.4.1997, 3.4.97 ES
myoadenylate deaminase deficiency
autosomal recessive inheritance

Clinical features
* exercise intolerance
* myalgia
* symptoms develop during second to fourth decades
* CK may be mildly elevated
* diagnosis is based on histochemical analysis of muscle biopsy specimens

Strategy
* exclude other neuromuscular disorders
* normal findings or mild non-specific EMG abnormalities

Expected abnormal findings
EMG
* mild non-specific abnormalities

Expected normal findings
EMG

Neurography

Procedure
EMG
* m.deltoideus/m.biceps brachii
* m.interosseus dorsalis I/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
* m.vastus lateralis/m.vastus medialis/m.tensor facia latae
* m.tibialis anterior

Neurography
* SCS: n.suralis and n.radialis on one side
* MCS: n.peroneus and n.medianus on one side

References

Modified
* 2.4.1997 BF, 3.4.97 ES

4.8 PRIMARY PERIODIC PARALYSIS

Hypokalemic periodic paralysis

Etiology
* autosomal dominant inheritance
* gene location: chromosome 1q31-q32
* mutation of the calcium channel = dihydropyridine (DHP)-receptor alpha 1 subunit

Clinical features
* first attacks within the second decade, invariably before the age of 30, more than half present before age of 16 years
* attacks last usually more than two hours often more than 24 hours
* attacks are precipitated by carbohydrate ingestion or strenuous exercise
* during attacks serum potassium is decreased
* CK normal between attacks and after attacks mildly elevated

Strategy
* during attack demonstrate reduced number of motor units and myopathic abnormalities
* between attacks patients with fixed weakness show myopathic abnormalities on EMG
* demonstrate abnormal exercise test between attacks

Expected abnormal findings during attack
Neurography
* MCS: reduced amplitude of the M wave
EMG
* insertion activity is reduced
* reduced interference pattern
* reduction in duration and amplitude of MUPs

Decrement studies
* facilitation of responses up to 400 %

Expected normal findings during attack
* SCS

Expected abnormal findings between attacks
EMG
* in patients with permanent weakness reduction in duration and amplitude of MUPs
Exercise test
* abnormal initial increase in M amplitude and successively abnormal decrease

Expected normal findings between attacks
EMG
* in patients without permanent weakness normal EMG
Neurography
**Procedure**

**Neurography**
- MCS: n.medianus and n.peroneus unilaterally

**EMG**
- m.deltoideus
- m.interosseus dorsalis/m.opponens pollicis
- m.vastus lateralis
- m.tibialis anterior

**Decrement (during attacks)**
- hypothenar muscles
- Exercise test (between attacks)
  - hypotenar muscles; M wave amplitude: for 2-5 minute maximal voluntary exercise 15 sec and 5 sec rests
  - in controls M amplitude increases immediately after exercise less than 27 % and decreases less than 30 % 5 min after the exercise test
  - in periodic paralysis increase after exercise >35% and successive drop in amplitude >27%

**Note**
- specific diagnosis can be obtained with genetic molecular methods
- muscle biopsy shows distinctive vacuoles
- provocative tests: glucose loading can be used

**References**

**HYPERKALEMIC PERIODIC PARALYSIS**

**Etiology**
- autosomal dominant inheritance
- gene location chromosome 17q13.1
- mutation of muscle alpha-subunit of the sodium channel
- this disorder is more heterogeneous than hypokalemic periodic paralysis
- hyperkalemic periodic paralysis is allelic to paramyotonia congenita and myotonia fluctuans

**Clinical features**
- first attacks during the first decade
- initially attacks are rare but they become more frequent
- duration of attacks 15 min to 4 hours
- cold, emotional stress, glucocorticoids and pregnancy aggravate attacks
- after strenuous exercise weakness can follow within minutes, sustained exercise may prevent weakness
- attacks precipitated by fasting or exercise
- the serum potassium level reaches 5-6 mmol/l and rarely reaches cardiotoxic levels

**Stratagy**
- during attack demonstrate reduced number of motor units and myopathic abnormalities
- between attacks patients with fixed weakness show myopathic abnormalities on EMG
- demonstrate abnormal exercise test between attacks

**Expected abnormal findings during attack**

**Neurography**
- MCS: reduced amplitude of the M wave

**EMG**
- reduced interference pattern
- reduction in duration and amplitude of MUPs
- myotonic discharges tend to diminish during attacks

**Decrement studies**
- facilitation of responses up to 400 %

**Expected normal findings during attack**
- SCS

**Expected abnormal findings between attacks**

**EMG**
- in patients with permanent weakness reduction in duration and amplitude of MUPs
- myotonic discharges

**Exercise test**
- abnormal initial increase in M amplitude and successively abnormal decrease

**Expected normal findings between attacks**

**EMG**
- in patients without permanent weakness normal EMG

**Neurography**
- SCS
- MCS

**Procedure**
Neurography
- MCS: n.medianus and n.peroneus unilaterally
- EMG
  - m.deltoides
  - m.interosseus dorsalis/m.opponens pollicis
  - m.vastus lateralis
  - m.tibialis anterior

Decrement (during attacks)
- hypothenar muscles

Exercise test (between attacks)
- hypothenar muscles: M wave amplitude: for 2-5 minute maximal voluntary exercise 15 sec and 5 sec rests
- in controls M amplitude increases immediately after exercise less than 27% and decreases less than 30% 5 min after the exercise test
- in periodic paralysis increase after exercise >35% and successive drop in amplitude >27%

Note
- specific diagnosis can be obtained with genetic molecular methods
- hyperkalemic periodic paralysis can be differentiated from paramyotonia congenita using cold provocation: in paramyotonia congenita cooling of the muscle to 20 C eliminates myotonic discharges and voluntary activity
- muscle biopsy shows vacuoles that are smaller than in hypokalemic periodic paralysis
- provocative tests: potassium loading can be used

References

Modified
- 1.4.1997 BF, 3.4.97 ES

4.9 MISCELLANEOUS MYOPATHIES

BENT SPINE SYNDROME (ISOLATED NECK EXTENSOR MYOPATHY)

Etiology
- heterogenous
- some hereditary forms have been described

Clinical features
- onset in old age most described patients are above 65 years of age
- weakness of paraspinal muscles on standing, resulting in a bent spine or inability to maintain the head erect (dropped head syndrome)

Strategy
- demonstrate myopathy in the paraspinal muscles, especially in the cervical and thoracic region
- differentiate from ALS, more widespread myopathies, especially polymyositis

Expected abnormal findings

EMG
- active myopathic findings in paraspinal muscles

Expected normal findings

EMG
- limb muscles

Neurography
- MCS
- SCS

Procedure

Neurography
- MCS n.medianus, n.ulnaris and n.peroneus on one side
- SCS: n.radialis and n.suralis on one side.

EMG
- Paraspinal muscles in the cervical, thoracic and lumbar region
- one distal and proximal muscle in the lower extremities.

References

Modified
- 2.4.1997, 3.4.97 ES

MALIGNANT HYPERThERMIA

Etiology
genetic basis is incompletely understood
autosomal dominant inheritance is most common
gene location: chromosome 19q13.1 in a portion of the patients
mutation of the ryanodine receptor gene
patients with central core disease (gene mutation on the same allele as above) has a susceptibility to malignant hyperthermia

Clinical features
- malignant hyperthermia is triggered by volatile anesthetic agent
- onset is often more abrupt if succinycholine is used
- once malignant hyperthermia is induced the temperature rises and may exceed 43 C
- 75 % show muscle rigidity
- may proceed to rigor and death
- serum potassium, CK, ionized calcium, myoglobin and sodium is elevated
- pH falls below 7.00
- treatment by intravenous dantrolene sodium and cooling

Clinical features
- malignant hyperthermia is triggered by volatile anesthetic agent
- onset is often more abrupt if succinycholine is used
- once malignant hyperthermia is induced the temperature rises and may exceed 43 C
- 75 % show muscle rigidity
- may proceed to rigor and death
- serum potassium, CK, ionized calcium, myoglobin and sodium is elevated
- pH falls below 7.00
- treatment by intravenous dantrolene sodium and cooling

Strategy
- between attacks no EMG abnormalities

Expected normal findings
Neurography
- SCS
- MCS
EMG
- MCS: n.radialis and n.peroneus unilaterally
EMG
- m.deltoides
- m.interosseus dorsalis/m opponens pollicis
- m.vastus lateralis
- m.tibialis anterior

References

Modified
- 1.4.1997, 3.4.97 ES

NEUROLEPTIC MALIGNANT SYNDROME

Etiology
- not related with malignant hypertermia
- sporadic
- less than 1% of persons taking neuroleptics, especially haloperidol and fluphenazine develop neuroleptic malignant syndrome

Clinical features
- occurs especially in young men on neuroleptic drug treatment
- fever, rigidity, confusion, tachycardia
- rhabdomyolysis
- myoglobinuria

Strategy
- EMG is usually normal between episodes

Expected normal findings between attacks
EMG
Neurography

Procedure
EMG
- m.deltoides/m.biceps brachii
- m.interosseus dorsalis l/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor facia latae
- m.tibialis anterior
Neurography
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

References

Modified
- 1.4.1997, 3.4.97 ES

RIPPLING MUSCLE DISEASE

Etiology
autosomal dominant, chromosome 1q41
may be genetically heterogeneous

Clinical features
- onset of symptoms in teens
- symptoms worst at age of onset, subsequently symptoms decrease
- muscle cramps and stiffness, especially with exercise
- stiffness most marked in proximal muscles
- muscle hypertrophy
- mechanical tapping of the muscle induces local mounding and rippling muscle contractions
- CK is mildly or moderately elevated (up to 17 times normal)

Strategy
- demonstrate that there is no electric muscle activity related with the rippling muscle contractions

Expected abnormal findings
EMG
- mechanical tapping induces electrically silent muscle contractions

Expected normal findings
EMG
- insertional activity
- MUP analysis
Neurography
- SCS
- MCS

Procedure
EMG
- m.deltoides/m.biceps brachii
- m.interosseous dorsalis /m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor fascia latae
- m.tibialis anterior
Neurography
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus, n.tibialis, n.ulnaris and n.medianus on one side

References
- Ansevin CF; Agamanolis DP. Rippling muscles and myasthenia gravis with rippling muscles. Arch-Neurol 1996;53:197-199
- Burns RJ; Bretag AH; Blumbergs PC; Harbord MG. Benign familial disease with muscle mounding and rippling. J Neurol Neurosurg Psychiatry 1994;57: 344-347
- Ricker K; Moxley RT; Rohkamm R. Rippling muscle disease. Arch Neurol 1989: 46: 405-408

Modified
- 1.4.1997, 3.4.97 ES

5. LOCAL NERVE LESIONS

5.1 SHOULDER, NECK AND UPPER EXTREMITIES

CARPAL TUNNEL SYNDROME (CTS)

Etiology
- chronic compression of the median nerve in the carpal tunnel

Clinical features
- by far the most common local nerve lesion
- paresthesias of digits 1-4, especially during the night or following use of hands
- in early cases only intermittent numbness in more severe entrapments constant numbness
- affects women more often than men (ratio 4:1)
- most patients are 40-60 years of age
- younger onset of symptoms is not unusual, occurs rarely in children
- in severe cases atrophy of the thenar muscles
- onset is insidious in most patients
- pain in the wrist and forearm, often also in the shoulder region
- predisposing factors: obesity, heavy manual work, pregnancy, wrist fractures, rheumatoid arthritis, diabetes, tenosynovitis of finger flexor tendons, hypothyreosis, amyloidosis, acromegaly

Strategy
- confirm local lesion of median nerve in the carpal tunnel
Assess
- severity: mild - moderate -severe - total
- pathophysiology: conduction block - demyelination -axonal degeneration

Expected abnormal findings
Neurography
- reduced median nerve SCV or prolonged sensory latency at the wrist
- in moderate and severe cases prolonged distal motor latency of the median nerve
- in moderate to severe cases reduced SCS amplitude
- in severe cases reduced MCS amplitude
EMG
Expected normal findings

- **SCS**: ulnar and radial nerves
- **MCS**: ulnar nerve

**Procedure**

**Neurography (always bilaterally)**
- SCS, median nerve digit 3 and one other digit to wrist, (palm to wrist may be helpful)
- SCS, ulnar nerve digit 5 to wrist, (palm to wrist may be helpful)
- sometimes comparison of the median nerve SCS with radial nerve forearm to thumb may be helpful
- MCS median nerve

**EMG (optional: if it is necessary to do EMG to exclude a proximal lesion):**
- m.opponentis/m.abductor pollicis brevis
- m.interosseus dorsalis I
- m.flexor carpi radialis/flexor pollicis longus
- m.extensor indicis proprius
- m.triceps

**References**

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- Brain WR, Wright AD, Wilkinson M. Spontaneous compression of both median nerves in the carpal tunnel. Lancet 1947;1:277-282
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- Kimura J. The carpal tunnel syndrome: localization of conduction abnormalities within the distal segment of the median nerve. Brain 1979;102:619-635
Clinical features

- loss of sensation and paresthesia of digits 1-4

Etiology

- trauma, especially with knife and glass

Revised

BF 12.4.1997
weakness and/or atrophy of the thenar muscles
pain is variable
trauma caused by sharp objects, especially knives and glass may cut only some fascicles; this may case limited symptoms

Strategy
- confirm local lesion of median nerve above the wrist
- differentiate from carpal tunnel syndrome

Assess
- severity: mild - moderate - severe - total
- pathophysiology: conduction block - demyelination - axonal degeneration

Expected abnormal findings

Neurography
- reduced n.medianus SCV in nerve lesions caused by compression
- in moderate and severe cases prolonged distal motor latency of the median nerve
- reduced n.medianus SCS amplitude, especially in partial lesions due to cutting with sharp objects
- in moderate or severe cases reduced MCS amplitude

EMG
- in moderate to severe cases m.abductor pollicis brevis show neurogenic findings

Expected normal findings
- SCS: ulnar and radial nerves
- MCS: ulnar nerve

Procedure

Neurography
- SCS, median nerve digit 3 and one other digit to wrist crease and 50 mm above wrist crease
- SCS, ulnar nerve digit 5 to wrist
- MCS median nerve bilaterally

EMG
- m. opponens pollicis/m. abductor pollicis brevis
the following muscles are optional, required only if it is necessary to do EMG to exclude a proximal lesion
- m.interosseus dorsalis I
- m.flexor carpi radialis/flexor pollicis longus
- m.extensor indicis proprius
- m.triceps
- m.biceps
- m.deltoides

ANTERIOR INTEROSSEUS NERVE LESION

Etiology
- acute idiopathic mononeuropathy (neuralgic amyotrophy) by far most common cause of n.interosseus anterior neuropathy
- traumatic injury in association with elbow luxation and other injuries
- often described in textbooks as an entrapment neuropathy (anterior interosseus syndrome), we have not encountered a real entrapment at this site and are doubtful as to whether such an entrapment really exists

Clinical features
- usually acute onset within hours
- often severe pain in the anterior part of the forearm, pain subsides after a few days or weeks
- flexion weakness of the distal phalanx of the thumb and forefinger
- no sensory loss
- prognosis with conservative treatment is usually good

Strategy
- demonstrate neurogenic EMG findings in muscles in muscles innervated by n.interosseus anterior
- normal findings in other muscles innervated by n.medianus

Expected abnormal findings

EMG:
- neurogenic findings in m.pronator quadratus and m.flexor pollicis longus

Neurography
- MCS latency to pronator quadratus prolonged

Expected normal findings

EMG
- m.flexor carpi radialis
- m.abductor pollicis brevis
- m.pronator teres

Neurography
- MCS: n.medianus
- SCS: n.medianus

Procedure

EMG:
- m. pronator quadratus (n.interosseus anterior, distal part)
- m.flexor pollicis longus (n.interosseus anterior, proximal part)
- m.flexor carpi radialis/m.pronator teres (n.medianus, proximal part)
- m.opponens pollicis/m.abductor pollicis brevis (n.medianus, distal part)
- m.interosseus dorsalis (n.ulnaris)
- m.extensor digitorum communis (n.radialis)

Neurography
- MCS latency elbow pronator quadratus bilaterally
- n.medianus MCS in the forearm to m.abductor pollicis brevis
- n.medianus SCS to digit 3
the following muscles are optional, required only if it is necessary to do EMG to exclude a proximal lesion
* m.triceps
* m.biceps
* m.deltoides

Note
* we discourage the use of the term anterior interosseus syndrome which alludes to an entrapment neuropathy

References
* O’Brien MD, Upton ARM. Anterior interosseous nerve syndrome. J Neurol Neurosurg Psychiatry 1972;35:531-536

Revised
* BF 1.2.1997, 12.4.1997

MEDIAN NERVE LESION AROUND THE ELBOW

Etiology
* acute idiopathic mononeuropathy (neuralgic amyotrophy) by far most common cause of n.interosseus anterior neuropathy
* traumatic injury in association with elbow luxation, supracondylar humerus fractures and other injuries
* often described in textbooks as an entrapment neuropathy (pronator syndrome) in the region where the median nerve passes thorough the pronator teres muscle; we have never encountered a real entrapment at this site and are doubtful as to whether such an entrapment really exists

Clinical features
* paresthesia of fingers 1-4
* in moderate or severe cases weakness of thumb abduction, opposition and flexion, wrist flexion and finger flexion

Strategy
* demonstrate neurogenic EMG findings in muscles innervated by the median nerve distal to the pronator muscle

Expected abnormal findings

EMG
* m.flexor pollicis longus
* m.opponens pollicis/abductor pollicis brevis
* m.flexor digitorum profundus and superficialis

Neurography
* reduced CV and/or conduction block of median nerve in the proximal part of the forearm

Expected normal findings

EMG
* m.pronator teres and m.flexor carpi radialis (innervated proximal to the lesion)
* muscles innervated by the ulnar and radial nerve

Neurography
* normal ulnar and radial nerve MCS and SCS

Procedure

EMG:
* m.flexor pollicis longus (n.interosseus anterior)
* m.flexor carp radialis/m.pronator teres (n.medianus, proximal part)
* m.abductor pollicis brevis (n.medianus, distal part)
* m.interosseus dorsalisis (n.ulnaris)
* m.extensor digitorum communis (n.radialis)

Neurography
* MCS latency elbow pronator quadratus
* n.medianus MCS in the forearm to thenar muscles
* n.medianus SCS to digit 3

Note
* we discourage the use of the term pronator syndrome which alludes to an entrapment neuropathy

References
* Bolton CF, Driedger AA, Lindsay RM. Ischaemic neuropathy in uraemic patients caused by bovine arteriovenous shunt. J Neurol Neurosurg Psychiatry 1979;42:810-814


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Revised

6.4.1997

MEDIAN NERVE LESION ABOVE THE ELBOW (AT STRUTHER'S LIGAMENT)

Etiology

- described in the literature as an entrapment neuropathy in lower part of the upper arm, the entrapment is due to a ligament from supracondylar process to the medial epicondyle. We have never encountered a real entrapment at this site and are doubtful as to whether such an entrapment really exists.

Clinical features

- paresthesia of fingers 1-4
- in moderate or severe cases weakness of thumb abduction, opposition and flexion, wrist flexion and finger flexion
- plain X-rays show Struther's ligament ligament from supracondylar process to the medial epicondyle

Strategy

- demonstrate neurogenic EMG findings in muscles innervated by the median nerve in the forearm
- differentiate from other peripheral nerve lesions.

Expected abnormal findings

EMG

- neurogenic EMG findings in muscles innervated by the median nerve.

Neurography

- reduced CV and/or conduction block of median nerve in distal part of the upper arm.

Expected normal findings

EMG

- muscles innervated by the n.ulnaris and n.radialis nerve.

Neurography

- n.ulnaris and n.radialis MCS and SCS.

Procedure

EMG:

- m.flexor pollicis longus (n.interosseus anterior)
- m.flexor carpi radialis/m.pronator teres (n.medianus, proximal part)
- m.opponens pollicis/abductor pollicis brevis (n.medianus, distal part)
- m.interosseus dorsalis (n.ulnaris)
- m.extensor digitorum communis (n.radialis)

Neurography

- MCS latency elbow pronator quadratus
- n.medianus MCS in the forearm to thenar muscles
- n.medianus SCS to digit 3

References
RADIAL NERVE LESION IN THE UPPER ARM

**Etiology**
- temporary compression of the radial nerve in radial groove, often during sleep following heavy drinking
- secondary to fracture of the humerus
- tourniquet paralysis
- erroneous injections

**Clinical features**
- wrist drop in severe cases
- weakness of finger extension, wrist extension, and elbow flexion (may be compensated by m.biceps brachii)
- tendon reflex of m.brachioradialis reduced or absent
- no weakness of elbow extension and m.triceps tendon reflex normal
- inconsistent sensation loss over the dorsal side of the hand between metacarpal bones 1 and 2

**Strategy**
- demonstrate neurogenic EMG findings in muscles innervated by the radial nerve below the radial groove
- assess severity and pathophysiology (axonal lesion/conduction block) to be able to give a prognosis

**Expected abnormal findings**

**EMG**
- denervation of m.brachioradialis
- denervation of wrist extensors
- denervation of finger extensors

**Neurography**
- radial nerve MCS: conduction block in the radial groove (not commonly assessed)
- radial nerve MCS: reduced amplitude
- radial nerve SCS: reduced amplitude (normal if it is a pure conduction block)

**Expected normal findings**

**EMG**
- m.triceps
- muscles innervated by n.medianus and n.ulnaris

**Neurography**
- n.ulnaris and n.medianus

**Procedure**

**Neurography**
- SCS: n.radialis, n.medianus, n.ulnaris
- MCS: n.radialis (fractionated across the radial groove)(optional)

**EMG**
- m.biceps brachii
- m.triceps
- m.brachioradialis/m.extensor carpi radialis
- m.extensor digitorum communis/m.extensor indicis
- m.opponens pollicis/m.abductor pollicis brevis
- m.interosseus dorsalis

**References**
- Marinacci AA. The value of the electromyogram in the diagnosis of pressure neuropathy from "hanging arm." Electromyography 1967;1:5-10

**Revised**
- 6.4.1997 BF, 7.5.1997 BF

RADIAL NERVE LESION IN THE AXILLA

**Etiology**
- compression by crutches
- trauma to the shoulder

**Clinical features**
- wrist drop in severe cases
- weakness of finger extension, wrist extension, elbow flexion and elbow extension
- m.triceps tendon reflex absent
- inconsistent sensation loss over the dorsal side of the hand between metacarpal bones 1 and 2

**Strategy**
- demonstrate neurogenic EMG findings in muscles innervated by the radial nerve
- assess severity and pathophysiology (axonal lesion/conduction block) to be able to give a prognosis

**Expected abnormal findings**
EMG
  * m.triceps
  * m.brachioradialis/m.extensor carpi radialis
  * extensor digitorum communis

Neurography
  * n.radialis MCS: conduction block in the axilla (not commonly assessed)
  * n.radialis MCS: reduced amplitude
  * n.radialis SCS: reduced amplitude (normal if it is a pure conduction block)

**Expected normal findings**

EMG
  * n.axillaris, n.ulnaris and n.medianus innervated muscles

Neurography
  * ulnar and median nerves

Procedure

Neurography
  * SCS: n.radialis, n.medianus, n.ulnaris
  * MCS: n.radialis (fractionated across the radial groove)(optional)

EMG
  * m.deltoideus (posterior fascicle)
  * m.biceps
  * m.triceps
  * m.brachioradialis
  * m.extensor digitorum communis/extensor indicis
  * m.opponens pollicis/m.abductor pollicis brevis (n.medianus)
  * m.interosseus doralis (n.ulnaris)

References

Revised
  * BF 1.2.1997

**POSTERIOR INTEROSSEUS NERVE LESION**

**Etiology**
  * acute idiopathic mononeuropathy (acute neuralgic amyotrophy)
  * trauma to the forearm, laceration
  * Monteggia-fracture
  * entrapment is extremely rare, if at all existent

**Clinical features**
  * weakness of finger extension
  * no weakness of wrist extension, elbow flexion or elbow extension and m.triceps tendon reflex normal
  * no sensory abnormalities
  * usually painless but there may be local pain 5-8 cm distal to the lateral epicondyle

**Strategy**
  * demonstrate neurogenic EMG findings in muscle innervated by the deep branch of the radial nerve

**Expected abnormal findings**

EMG
  * m.extensor digitorum communis/m.extensor indicis proprius

Neurography
  * n.radialis MCS (elbow to forearm) abnormal (CV reduced or conduction block)

**Expected normal findings**

EMG
  * n.axillaris, n.medianus, n.ulnaris

Neurography
  * SCS: n.radialis

Procedure

Neurography
  * SCS: n.radialis, n.medianus, n.ulnaris
  * MCS: n.radialis (fractionated across the arcade of Frohse)

EMG
  * n.axillaris, n.medianus, n.ulnaris

Neurography
  * SCS: n.radialis, n.medianus, n.ulnaris
  * MCS: n.radialis (fractionated across the arcade of Frohse)

EMG
  * m.triceps (n.radialis proximal to lesion)
  * m.brachioradialis/m.extensor carpi radialis longus (n.radialis proximal to lesion)
  * m.extensor digitorum communis/m.extensor indicis
  * m.opponens pollicis/m.abductor pollicis brevis (n.medianus)
  * m.interosseus doralis (n.ulnaris)

References
LESION OF THE SENSORY RADIAL NERVE THE FOREARM (WARTENBERG’S SYNDROME)

Etiology
- temporary compression of the superficial radial nerve in the forearm by tight handcuffs or wrist watch
- surgery for de Quervain's tenosynovitis
- intravenous catethers in the radial aspect of the forearm

Clinical features
- numbness and/or loss of sensation over the dorsal side of the hand between metacarpal bones 1 and 2, sometimes on the dorsal side of fingers 1-3
- sometimes when the lesion is caused by a sharp cutting object only one branch of n.radialis is damaged

Strategy
- demonstrate lesion of the superficial radial nerve or one of its branches

Expected abnormal findings

Neurography
- SCS: n.radialis low amplitude and/or reduced CV

Expected normal findings

Neurography
- SCS: n.medianus, n.ulnaris

EMG
- m.extensor digitorum communis
- m.interosseus dorsalis I
- m.abductor pollicis brevis

Procedure

Neurography
- SCS: n.radialis, n.medianus, n.ulnaris
- it may be necessary to study separately the n.radialis branches to the thumb, forefinger and long finger

EMG
- m.extensor digitorum communis
- m.opponens pollicis/m.abductor pollicis brevis
- m.interosseus dorsalis

References
- Dorfman LV, Jayaram AR. Handcuff neuropathy. JAMA 1978;239:957
- Trojaborg W. Rate of recovery in motor and sensory fibres of the radial nerve: clinical and electrophysiological aspects. J Neurol Neurosurg Psychiatry 1970;33:625-638

ULNAR NERVE LESION IN THE CUBITAL TUNNEL (CUBITAL TUNNEL SYNDROME)

Etiology
- entrapment of the ulnar nerve at the flexor retinaculum of the m.flexor carpi ulnaris 1-2 cm distal to the medial epicondyde

Clinical features
- paresthesia of fingers 4 and 5
- weakness of spreading of the fingers
- in severe cases atrophy of the intrinsic hand muscles except the thenar muscles
- pain sometimes present over the elbow region, pain is not at all as prominent as in carpal tunnel syndrome
- the elbow does not usually show clinically or radiologically deformity

Strategy
demonstrate nerve conduction abnormality just distal to the medial epicondyle
• demonstrate neurogenic EMG findings in muscles innervated by the ulnar nerve
• exclude proximal nerve lesions (lower part of plexus and C8 radiculopathy)

**Expected abnormal findings**

**EMG**
• m.interosseus dorsalis/m.abductor digitii minimi
• m.flexor carpi ulnaris

**Neurography**
• MCS, n.ulnaris: reduced CV and/or conduction block immediately distal to the medial epicondyle
• SCS, n.ulnaris digits 4 and 5 and n.ulnaris ramus dorsalis reduced amplitude
• ulnar nerve inching: abnormal finding 10-20 mm distal to the medial epicondyle

**Expected normal findings**

**EMG**
• m.extensor indicis proprius (C7-C8)
• m.abductor pollicis brevis (may be innervated from n.ulnaris and show abnormality)
• paraverternal muscles

**Neurography**
• MCS and SCS n.medianus and n.radialis
• SCS, n.cutaneus antebrachii medialis

**Procedure**

**Neurography**
• MCS n.ulnaris, fractionated across the elbow
• MCS, SCS n.ulnaris
• SCS, n.ulnaris ramus dorsalis
• SCS n.medianus
• inching across the elbow: n.ulnaris

**EMG**
• m.interosseus dorsalis I (distal n.ulnaris)
• m.opponens pollicis/m.abductor pollicis brevis (n.medianus)
• m.flexor carpi ulnaris (proximal n.ulnaris)
• m.extensor indicis proprius (C8 and inferior trunk)

**References**

• Campbell WW; Pridgeon RM, Sahni KS. Short segment incremental studies in the evaluation of ulnar neuropathy at the elbow. Muscle Nerve 1992;15:1050-1054
• Gilliatt RW, Thomas PK. Changes in nerve conduction with ulnar lesions at the elbow. J Neurol Neurosurg Psychiatry 1960;23:312-320
• Miller RG. The cubital tunnel syndrome: Diagnosis and precise localization, Ann Neurol 1979;6:56-59
• Odusote K, Eisen A. An electrophysiological quantification of the cubital tunnel syndrome. Can J Neurol Sci 1979;6:403-410

**Revised**
• 1.7.1997 BF, 3.4.1997 BF

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**Ulnar Nerve Lesion at the Medial Epicondyle (Ulnar Sulcus)**

**Etiology**
• entrapment in the ulnar sulcus at the medial epicondyle or just proximal to it
• temporary compression during sleep (often following alcohol consumption) or anesthesia
• trauma to the elbow

**Clinical features**
• paresthesia of fingers 4 and 5
• weakness of spreading of the fingers
• in severe cases atrophy of the intrinsic hand muscles except the thenar muscles
• pain sometimes present over the elbow region, pain is not at all as prominent as in carpal tunnel syndrome
• if the lesion is an entrapment the elbow often shows clinically or radiologically deformity (limited extension of the elbow)

**Strategy**
• demonstrate nerve conduction abnormality at the medial epicondyle
• demonstrate neurogenic EMG findings in muscles innervated by the ulnar nerve
• exclude proximal nerve lesions (lower part of plexus and C8 radiculopathy)

**Expected abnormal findings**

**EMG**
• m.interosseus dorsalis/m.abductor digitii minimi
• m.flexor carpi ulnaris

**Neurography**
"SCS, n.ulnaris: reduced CV and/or conduction block immediately distal to the medial epicondyle
"SCS, n.ulnaris digits 4 and 5 and n.ulnaris ramus dorsalis reduced amplitude
"ulnar nerve inching: abnormal finding at the medial epicondyle or just proximal to it

**Expected normal findings**

**EMG**
"m.extensor indicis proprius (C7-C8)
"m.opponens pollicis/m.abductor pollicis brevis (may be innervated from n.ulnaris and show abnormality)
"paravertebral muscles

**Neurography**
"SCS and MCS n.mediansus and n.radialis
"SCS: n.cutaneus antebrachii medialis

**Procedure**

**Neurography**
"MCS n.ulnaris, fractionated across the elbow
"MCS, SCS n.ulnaris
"SCS, n.ulnaris ramus dorsalis
"SCS n.medianus
"inching across the elbow: n. ulnaris

**EMG**
"m.interosseus dorsalis I (distal n.medianus)
"m.abductor pollicis brevis (n.medianus)
"m.flexor carpi ulnaris (proximal n.ulnaris)
"m.extensor indicis proprius (C8 and inferior trunk)

**References**

- Ashenhurst EM. Anatomical factors in etiology of ulnar neuropathy. Can Med Assoc 1962;87:159-163
- Campbell WW, Pridgeon RM, Salmi KS. Entrapment neuropathy of the ulnar nerve at its point of exit from the flexor carpi ulnaris muscle. Muscle Nerve 1986;1:467-470
- Eisen A. Early diagnosis of ulnar nerve palsy. Neurology 1974;24:256-262
- Gilliatt RW, Thomas PK. Changes in nerve conduction with ulnar lesions at the elbow. J Neurol Neurosurg Psychiatry 1960; 23312-320
- Jabre JF. Ulnar nerve lesions at the wrist: new technique for recording from the sensory dorsal branch of the ulnar nerve. Neurology 1980;30:873-876
- Pechan J, Julis 1. The pressure measurement in the ulnar nerve: a contribution to the pathophysiology of the cubital tunnel syndrome. J Biomech 1975;8:75-77
- Stewart JD. The variable clinical manifestations of ulnar neuropathies at the elbow. J Neurol Neurosurg Psychiatry 1987;50:252-258
Ulnar Nerve Lesion at the Wrist

Etiology
- entrapment in the canal of Guyon (usually due to a lipoma, ganglion or aneurysm), rare
- temporary or chronic compression to the hypothenar region (due to bicycle bars, crutches)
- trauma, especially sharp cutting objects

Clinical features
- paresthesia and/or loss of sensation of fingers 4-5
- no alteration of the sensation over the dorsal side of the hand innervated by r.dorsalis n.ulnaris
- weakness of ulnar innervated hand muscles

Strategy
- demonstrate lesion of the ulnar nerve at the wrist
- assess severity and pathophysiology

Expected abnormal findings

EMG
- neurogenic EMG findings in m.interosseus dorsalis/m.abductor digitii minimi

Neurography
- ulnar MCS reduced amplitude and prolonged DLAT
- ulnar SCS reduced CV at the wrist and reduced AMPL

Expected normal findings

EMG
- m.flexor carpi ulnaris
- m.extensor indicis proprius

Neurography
- median and radial nerves
- n.ulnaris ramus dorsalis

Procedure

Neurography
- MCS n.ulnaris, fractionated across the elbow
- MCS, SCS n.ulnaris
- SCS, n.ulnaris ramus dorsalis
- SCS n.medianus
- inching across the elbow: n.ulnaris

EMG
- m.interosseus dorsalis I (distal n.ulnaris)
- m.abductor pollicis brevis (n.medianus)
- m.flexor carpi ulnaris (proximal n.ulnaris)
- m.extensor indicis proprius (C8 and truncus inferior)

References
- Hankey GJ, Gubbay SS. Compressive mononeuropathy of the deep palmar branch of the ulnar nerve in cyclists. J Neurol Neurosurg Psychiatry 1998;51:1588-1590
- Kornberg M, Aulicino PL, DuPuy TE. Laceration of the ulnar nerve with a closed fracture of the distal radius and ulnar. Orthopedics 1983;6:729-731
- Olney RK, Hanson M. Ulnar neuropathy at or distal to the wrist. Muscle Nerve 1988;1:1828-832

Revised
- 1.7.1997 BF, 12.4.1997 BF
DEEP MOTOR BRANCH OF THE ULNAR NERVE LESION AT THE WRIST

**Etiology**
- entrapment in the canal of Guyon (usually due to a lipoma, ganglion or aneurysm)
- temporary or chronic compression to the hypothenar region (due to bicycle bars, crutches)
- trauma, especially sharp cutting objects

**Clinical features**
- weakness and atrophy of intrinsic hand muscles, except thenar muscles
- no sensory abnormality

**Strategy**
- demonstrate lesion of the deep motor branch of the ulnar nerve at the wrist
- assess severity and pathophysiology

**Expected abnormal findings**

**EMG**
- neurogenic EMG findings in m.interosseus dorsalis/m.abductor digiti minimi

**Neurography**
- ulnar MCS reduced amplitude and prolonged DLAT

**Expected normal findings**

**EMG**
- m.flexor carpi ulnaris
- m.extensor indicis proprius

**Neurography**
- SCV: n.ulnaris, n.medianus and n.radialis

**Procedure**

**Neurography**
- MCS n.ulnaris, fractionated across the elbow
- MCS, SCS n.ulnaris
- SCS, n.ulnaris ramus dorsalis
- SCS n.medianus
- inching across the elbow: n.ulnaris

**EMG**
- m.interosseus dorsalis I/m.abductor digiti minimi (distal n.ulnaris)
- m.abductor pollicis brevis (n.medianus)
- m.flexor carpi ulnaris (proximal n.ulnaris)
- m.extensor indicis proprius (C8 and truncus inferior)

**References**
- Hankey GJ, Gubbay SS. Compressive mononeuropathy of the deep palmar branch of the ulnar nerve in cyclists. J Neurol Neurosurg Psychiatry 1988;51:1588-1590
- Komborg M, Aulicino PL, DuPuy TE. Laceration of the ulnar nerve with a closed fracture of the distal radius and ulnar. Orthopedics 1983;6:729-731
- Olney RK, Hanson M. Ulnar neuropathy at or distal to the wrist. Muscle Nerve 1988; 11:828-832

**Revised**
- 12.4.1997 BF, 15.4.1997 BF

SUPERFICIAL SENSORY BRANCH OF THE ULNAR NERVE LESION AT THE WRIST

**Etiology**
- entrapment in the canal of Guyon (usually due to a lipoma, ganglion or aneurysm)
- temporary or chronic compression to the hypothenar region (due to bicycle bars, crutches)
- trauma, especially sharp cutting objects

**Clinical features**
- paresthesia and/or loss of sensation of fingers 4-5
**Strategy**
- demonstrate lesion of the superficial sensory branch of the ulnar nerve at the wrist
- assess severity and pathophysiology

**Expected abnormal findings**

**Neurography**
- ulnar SCS reduced CV at the wrist and amplitude

**Expected normal findings**

**EMG**
- m.abductor digiti minimi/m.interosseus dorsalis I
- m.flexor carpi ulnaris
- m.extensor indicis proprius

**Neurography**
- MCS: n.medianus and n.radialis, n.ulnaris ramus dorsalis
- SCS: n.ulnaris, n.medianus

**Procedure**

**Neurography**
- MCS n.ulnaris, fractionated across the elbow
- MCS, SCS n.ulnaris
- SCS, n.ulnaris ramus dorsalis
- SCS n.medianus
- inching across the elbow: n.ulnaris

**EMG**
- m.interosseus dorsalis I (distal n.ulnaris)
- m.abductor pollicis brevis (n.medianus)
- m.flexor carpi ulnaris (proximal n.ulnaris)
- m.extensor indicis proprius (C8 and truncus inferior)

**References**


Hoyt CS. Ulnar neuropathy in bicycle riders. Arch Neurol 1976;33:372

Kombarg M, Aulicino PL, DuPuy TE. Laceration of the ulnar nerve with a closed fracture of the distal radius and ulnar. Orthopedica 1983;6:729-731


Olney RK, Hanson M. Ulnar neuropathy at or distal to the wrist. Muscle Nerve 1988;1:1828-832


**Revised**


**LATERAL CUTANEOUS NERVE OF THE FOREARM LESION**

**Etiology**
- isolated lesions rare
- acute idiopathic mononeuropathy (neuralgic amyotrophy)
- traumatic injuries due to stabbing or bullets
- trauma in association with venipuncture of v.cephalica
- surgery in the antecubital fossa
- fractures of the elbow and proximal part of forearm

**Clinical features**
- loss of sensation over the anterior lateral part of the forearm (n.cutaneus antebrachii lateralis)

**Strategy**
- demonstrate sensory abnormality limited to n.cutaneus antebrachii lateralis

**Expected abnormal findings**

**Neurography**
- SCS: n.cutaneus antebrachii lateralis

**Expected normal findings**

**EMG**
- m.biceps brachii
- m.brachialis/m.coracobrachialis
- m.deltoideus
- m.brachioradialis
- m.triceps brachii
**Procedure**

**EMG**
- m.biceps brachii (n.musculocutaneus)
- m.brachioradialis (C6, plexus brachialis: upper trunk)
- m.flexor carpi radialis (plexus brachialis: lateral fascicle)
- m.triceps (posterior fascicle)

**Neurography**
- n.cutaneus antebrachii lateralis bilaterally
- n.radialis

**References**
- Trojaborg W. Motor and sensory conduction in the musculocutaneous nerve. J Neurol Neurosurg Psychiatry 1976;39:890-899

**Revised**
- 1.7.1997 BF, 23.4.1997 BF

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**MEDIAL CUTANEOUS NERVE OF THE FOREARM LESION**

**Etiology**
- isolated lesions rare
- acute idiopathic mononeuropathy (neuralgic amyotrophy)
- traumatic injuries due to stabbing or bullets
- trauma in association with venipuncture of v.basilica
- surgery in the antecubital fossa
- fractures of the elbow and proximal part of forearm

**Clinical features**
- loss of sensation over the anterior medial part of the forearm (n.cutaneus antebrachii medialis)

**Strategy**
- demonstrate sensory abnormality limited to n.cutaneus antebrachii medialis
- differentiate from Th1 radiculopathy

**Expected abnormal findings**

**Neurography**
- SCS: n.cutaneus antebrachii medialis

**Expected normal findings**

**EMG**
- m.abductor pollicis brevis/m.opponens pollicis
- m.flexor carpi ulnaris
- m.interosseus dorsalis I/m.abductor digiti minimi

**Procedure**

**EMG**
- m.abductor pollicis brevis/m.opponens pollicis (truncus inferior)
- m.flexor carpi ulnaris (truncus inferior)
- m.interosseus dorsalis I/m.abductor digiti minimi

**Neurography**
- SCS: n.cutaneus antebrachii medialis bilaterally, n.cutaneus antebrachii lateralis

**References**
- Revised
- 1.7.1997 BF, 12.5.1997 BF

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**MUSCULOCUTANEUS NERVE LESION**

**Etiology**
- isolated lesions very rare
- acute idiopathic mononeuropathy (neuralgic amyotrophy)
- traumatic injuries due to stabbing or bullets
- shoulder luxation

**Clinical features**
- weakness of elbow flexion
- loss biceps tendon reflex
- loss of sensation over the anterior lateral part of the forearm (n.cutaneus antebrachii lateralis)

**Strategy**
- demonstrate neurogenic EMG findings in the muscles innervated by n.musculocutaneus
- exclude other C5 or C6 radiculopathy, plexus brachialis lesion: lateral fascicle and plexus brachialis: upper trunk

**Expected abnormal findings**

**EMG**
- m.biceps brachii
- m.brachialis/m.coracobrachialis

**Neurography**
- SCS: n.cutaneus antebrachii lateralis

**Expected normal findings**

**EMG**
**AXILLARY NERVE LESION**

**Etiology**
- luxation of the humerus
- fracture of the surgical neck of humerus
- traumatic injuries due to stabbing or bullets
- acute idiopathic mononeuropathy (neuralgic amyotrophy)
- an entrapment neuropathy "quadrilateral space syndrome " has been described, we are doubtful about the existence of an entrapment

**Clinical features**
- weakness of shoulder abduction
- loss of sensation over the upper lateral part of the upper arm

**Strategy**
- demonstrate neurogenic EMG findings in the muscles innervated by the axillary nerve
- exclude other nerve lesions in the axilla, especially the suprascapular nerve

**Expected abnormal findings**
- m.deltoideus/m.teres minor

**Expected normal findings**
- m.supraspinatus
- m.biceps
- m.triceps

**SUPRASCAPULAR NERVE LESION**

**Etiology**
- acute idiopathic mononeuropathy (neuralgic amyotrophy)
- entrapment at the incisura scapulae
- entrapment at the spinoglenoid notch
- fractures of the scapula
- traumatic injuries to the shoulder

**Clinical features**
- weakness of upper arm abduction
- weakness of upper arm external rotation
- atrophy of m.supraspinatus and m.infraspinatus
**LONG THORACIC NERVE LESION**

**Etiology**
- acute idiopathic mononeuropathy (neuralgic amyotrophy)
- resection of first rib
- traumatic injuries, especially due to stabbing or bullets, sometimes blunt injuries

**Clinical features**
- winging of the scapula

**Strategy**
- show neurogenic EMG findings in m.serratus anterior

**Expected abnormal findings**

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<tr>
<th>Procedure</th>
<th>EMG</th>
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<td>m.trapezius</td>
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<td>m.serratus anterior</td>
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<td>m.deltoides</td>
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<td>m.infraspinatus</td>
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<td>m.rhomboideus</td>
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<tr>
<td>m.biceps brachii</td>
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</table>

**References**
- Kaplan PE. Electrodagnostic confirmation of long thoracic nerve palsy. J Neurol Neurosurg Psychiatry 1980;43:50-52
- Petrera JE, Trojaborg W. Conduction studies along the long thoracic nerve in serratus anterior palsy of different etiology. Neurology 1984;149:160-163

* Revised
* BF 15.4.1997
DORSAL SCAPULAR NERVE LESION

**Etiology**
- isolated lesions very rare
- acute idiopathic mononeuropathy (neuraltic amyotrophy)
- traumatic injuries due to stabbing or bullets

**Clinical features**
- slight winging of the scapula
- the medial margin of the scapula is displaced laterally
- the inferior angle of the scapula is rotated outwards

**Strategy**
- show neurogenic EMG findings in m.levator scapulae and m.rhomboides major and minor
- exclude C5 radiculopathy and plexus brachialis lesion: upper trunk

**Expected abnormal findings**
- EMG
  - m.rhomboides
  - m.levator scapulae

**Expected normal findings**
- EMG
  - m.supraspinatus
  - m.deltoides

**Procedure**
- EMG
  - m.levator scapulae
  - m.rhomboides
  - m.deltoides
  - m.supraspinatus
  - m.biceps brachii
  - m.triceps brachii

- Revised
  - 23.4.1997 BF

THORACODORSAL NERVE LESION

**Etiology**
- isolated lesions very rare
- acute idiopathic mononeuropathy (neuraltic amyotrophy)
- traumatic injuries due to stabbing or bullets

**Clinical features**
- weakness of shoulder adduction and inward rotation

**Strategy**
- show neurogenic EMG findings in m.latissimus dorsi
- exclude C7 radiculopathy and plexus brachialis lesion: middle trunk

**Expected abnormal findings**
- EMG
  - neurogenic EMG findings in m.latissimus dorsi

**Expected normal findings**
- EMG
  - m.latissimus dorsi

**Procedure**
- EMG
  - m.latissimus dorsi
  - m.deltoides
  - m.triceps
  - m.biceps brachii
  - m.supraspinatus

- Revised
  - 23.4.1997 BF

SUBSCAPULAR NERVE LESION

**Etiology**
- isolated lesions very rare
- acute idiopathic mononeuropathy (neuraltic amyotrophy)
- traumatic injuries due to stabbing or bullets

**Clinical features**
- weakness of inward rotation o0f the upper arm

**Strategy**
- show neurogenic EMG findings in m .subscapularis and m.teres major
- exclude C5 radiculopathy and plexus brachialis lesion: upper trunk

**Expected abnormal findings**
- EMG
  - m.subscapularis
  - m.teres major

**Expected normal findings**
- EMG
BRACHIAL PLEXUS LESIONS

Etiology

Trauma
- traffic accidents, these injuries are normally combined with lesions of other proximal structures (other trunks, root avulsions)
- in contact sports blow to the head or shoulder may cause a “burner” or “stinger”, which may be due to a plexus lesions or radiculopathy
- stab and bullet wounds
- birth injuries especially in large babies there may be a disproportion between the shoulders of the baby and the pelvis of the mother. Erb’s palsy affects the upper part of the plexus and Klumpke’s palsy affects the lower part of the plexus

Temporary compression
- during coronary by-pass surgery 5-7 % of patients get a plexus lesion, usually the lower trunk is affected
- during general anesthesia if the arm is supinated and abducted more than 90 degrees

Inflammatory or unknown causes
- acute idiopathic brachial plexus neuropathy (neuraltic amyotrophy, "neuritis"). There is often acute onset with pain lasting a few days to a few weeks. Any part of the plexus, including spinal roots may be affected. This may be triggered by infections, immunization, surgery, pregnancy, childbirth or trauma. Diabetes predisposes to this. There is a rare autosomal dominant hereditary disorder, "hereditary acute brachial plexus neuropathy" where this occurs.

Related with cancer or cancer treatment
- tumors (especially lung cancer and metastasis of the lymph nodes in the axilla)
- radiotherapy for cancer

Chronic compression (often misleadingly called thoracic outlet syndrome, TOS)
- anomalous cervical rib
- following fracture of the clavicle due to deformity compression
- there are several poorly defined syndromes that have been suggested: (1) pectoralis minor syndrome and (2) scalenus anticus syndrome; the evidence is not convincing

Iatrogenic
- complication of plexus anesthesia
- hematoma from transaxillary percutaneous angiograms
- complications of surgery for thoracic outlet syndrome

Clinical features
- weakness and sensory abnormalities depend on the part of the plexus that is affected
- depending on the etiology pain may be present

Upper trunk
- weakness and/or atrophy of shoulder abduction, elbow flexion, upper arm outward rotation
- loss of sensation over the lateral side of the upper arm and the thumb

Middle trunk
- weakness of elbow extension, wrist flexion and extension
- numbness and/or loss of sensation in fingers 2-3

Lower trunk
- weakness of elbow extension, wrist flexion and extension
- numbness and/or loss of sensation in fingers 2-3

Posterior cord
- weakness and atrophy of muscles innervated by n.axillaris and n.radialis
- loss of sensation on the dorsal side of the hand between digits I and II

Lateral cord
- weakness and atrophy of muscles innervated by n.medianus and n.musculocutaneus
- numbness and/or loss of sensation of fingers 1-3

Medial cord
- weakness of muscles innervated by n.ulnaris and n.medianus
- numbness and/or loss of sensation in fingers 4-5

Strategy
- demonstrate neurogenic EMG findings in affected part of the plexus
- demonstrate abnormal SCS responses in affected parts
- exclude radiculopathy

Expected abnormal and normal findings

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<tr>
<th>Muscle</th>
<th>superior trunk</th>
<th>medial trunk</th>
<th>inferior trunk</th>
<th>lateral fascicle</th>
<th>posterior r fascicle</th>
<th>medial fascicle</th>
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<td>n.radialis to digit I</td>
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<td>H-reflex to m.flexor carpi rad.</td>
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**Procedure**

**EMG**

* paravertebral muscles C5-Th1
* m.rhomboideus
* m.infraspinatus/m.supraspinatus
* m.deltoides
* m.biceps brachii
* m.triceps
* m.pronator teres/m.flexor carpi radialis
* m.extensor indicis proprius
* m.interosseus dorsalis I
* m.opponens pollicis/m.abductor pollicis brevis

**Neurography**

**SCS**

* n.medianus to digit I
* n.medianus to digits II and III
* n.ulnaris to digit V
* n.radialis
n.cutaneus antebrachii lateralis
n.cutaneus antebrachii medialis

MCS
n.medianus
n.ulnaris, fractionated (including plexus stimulation)
n.medianus: H-reflex to m.flexor carpi radialis

Note
Traumatic lesions of superior trunk may be combined with avulsion of cervical roots C7 - T1 (sometimes also C6), giving normal SNAP amplitudes in median, ulnar, and radial nerves (with loss of sensory function and cervical/cortical SEP) combined with loss of motor responses in the same nerves (lesions proximal to the sensory ganglion)

References
Kline DG. Civilian gunshot wounds to the brachial plexus. J Neurosurg 1989;70:166-174
Kwast O. Electrophysiological assessment of maturation of regenerating motor nerve fibres in infants with brachial plexus palsy. Dev Med Child Neurol 1989;31:56-65
Stewart JD. Electrodagnostic techniques in the evaluation of nerve compressions and injuries in the upper limb. Hand Clin 1986;2:677-687
Trojaborg W. Electrophysiological findings in pressure palsy of the brachial plexus. J Neurol Neurosurg Psychiatry. 1977;40:1160-1167
Yannickas C, Shahani BT, Young RR. The investigation of traumatic lesions of the brachial plexus by electromyography and short latency somatosensory evoked by stimulation of multiple peripheral nerves. J Neurol Neurosurg Psychiatry 1983:46:1014-1022

Revised
23.4.1997 BF

Phrenic nerve lesions

Etiology
- acute idiopathic mononeuropathy
- surgery in the neck, especially sympathectomy
- as a complication of plexus anesthesia
- trauma
**Clinical features**
- dyspnea
- weakness of respiratory muscles

**Strategy**
- demonstrate neurogenic EMG findings in m.diaphragma
- rule out C3 or C4 radiculopathy and affection of plexus cervicalis and plexus brachialis: upper trunk

**Expected abnormal findings**

**EMG**
- neurogenic EMG findings in m.diaphragma

**Neurography**
- MCS: n.phrenicus: reduced amplitude and prolonged latency

**Expected normal findings**

**EMG**
- m.trapezius (n.accessorius and plexus cervicalis)
- m.deltoides/m.infraspinatus/m.supraspinatus (C5 and superior trunk)
- paravertebral muscles C3 and C4 (radiculopathy)

**Procedure**

**EMG**
- m.diaphragma
- m.trapezius (n.accessorius and plexus cervicalis)
- m.deltoides/m.infraspinatus/m.supraspinatus (C5 and superior trunk)

**Neurography**
- MCS: n.phrenicus: reduced amplitude and prolonged latency

**References**
- Knoblache GE. The incidence and etiology of phrenic nerve blockade associated with supraclavicular brachial plexus block. Anaesth Intens Care 1979;7:346-349

**5.2 Cranial Nerves**

**Oculomotor Nerve Lesion (III)**

**Etiology**
- acute idiopathic mononeuropathy
- head trauma
- congenital defects (Möbius syndrome)
- trauma during delivery to upper brachial plexus and cervical plexus
- neoplasms involving the brainstem nuclei, intracranial portion of the nerve (acoustic neuromas, meningeomas)
- vascular (especially in diabetes)
- aneurysm
- infections (meningitis, borreliosis, tuberculosis, herpes zoster, AIDS)
- sarcoidosis

**Clinical features**
- diplopia
- paralysis of external ocular muscles

**Strategy**
- demonstrate neurogenic EMG findings in muscles innervated by the oculomotor nerve
- exclude possibility of other cranial nerve lesions
- exclude ocular myopathy

**Expected abnormal findings**

**EMG**
- m.rectus superior
- m.rectus inferior
- m.rectus medialis
- m.obliquus inferior
- m.levator palpebrae superioris

**Expected normal findings**

**EMG**
- m.obliquus superior
- m.rectus lateralis

**Procedure**

**EMG**
- m.rectus lateralis (abducesens nerve)
- m.obliquus superior (trochlear nerve)
- m.rectus superior/m.rectus inferior/m.rectus medialis/m.obliquus inferior (oculomotor nerve)
- m.levator palpebrae superior (oculomotor nerve)

**References**
TROCHLEAR NERVE LESION (IV)

**Etiology**
- acute idiopathic mononeuropathy
- head trauma
- neoplasms involving the brainstem nuclei, intracranial portion of the nerve (acoustic neuromas, meningeomas)
- vascular (especially in diabetes)
- aneurysm
- infections (meningitis, borreliosis, tuberculosis, herpes zoster, AIDS)
- sarcoidosis

**Clinical features**
- vertical diplopia combined with image tilting

**Strategy**
- demonstrate neurogenic EMG findings in muscles innervated by the oculomotor nerve
- exclude possibility of other cranial nerve lesions
- exclude ocular myopathy

**Expected abnormal findings**
**EMG**
- m. obliquus superior

**Expected normal findings**
**EMG**
- m. rectus lateralis
- m. rectus superior
- m. rectus inferior
- m. rectus medialis
- m. obliquus inferior
- m. levator palpebrae superioris

**Procedure**
**EMG**
- m. rectus lateralis (abducens nerve)
- m. obliquus superior (trochlear nerve)
- m. rectus superior/m. rectus inferior/m. rectus medialis/m. obliquus inferior (oculomotor nerve)
- m. levator palpebrae superior (oculomotor nerve)

**References**

TRIGEMINAL NERVE LESIONS (V)

**Etiology**
- Sjögren's syndrome, scleroderma, mixed connective tissue disease, systemic lupus erythematosus, rheumatoid arthritis
- Wegener's granulomatosis
- tumors: Intracranial or extracranial, metastatic or primary, menigioma, schwannoma, epidermoid, chordoma
- trauma (250)
- diabetes mellitus
- sinusitis
- herpes zoster (255)
- amyloidosis
- trauma
- surgery
- acute idiopathic mononeuropathy (idiopathic trigeminal neuropathy)
- neurinomas
- congenital trigeminal defects
- vascular (especially in diabetes)
- aneurysm
- infections (meningitis, borreliosis, tuberculosis, herpes zoster, AIDS)
- sarcoidosis
- facelifting surger may damage individual branches of n.trigeminus

**Clinical features**
- numbness and/or loss of sensation over the face
- weakness of closing of the jaw

**Strategy**
- demonstrate neurogenic EMG findings in muscles innervated by the trigeminal nerve
- demonstrate affection of the sensory branches of the trigeminal nerve
- exclude affection of other cranial nerves

**Expected abnormal findings**
**EMG**
- m. masseter/m.temporalis/m.tensor veli palatini

**Neurography**
- SCS: n.alveolaris inferior
**Blink reflex**
- n.supraorbitalis
- n.infraorbitalis
- n.mentalis

**Masseter reflex**

**Expected normal findings**

**EMG**
- m.orbicularis oris

**Procedure**

**EMG**
- m.masseter/m.temporalis/m.tensor veli palatini
- m.orbicularis oris

**Neurography**
- SCS: n.alveolaris inferior (optional)

**Blink reflex**
- n.supraorbitalis
- ninfraorbitalis
- n.mentalis

**Masseter reflex**

**References**
- Garg RK, Agrawal A, Nag D, Jha S: Herpes zoster oticus associated with facial, auditory and trigeminal involvement. JAPI 1992;49:45-
- Rizzo M, Bosch EP, Gross CE: Trigeminal sensory neuropathy due to dural external carotid cavernous sinus fistula. Neurology 1982;32:89-
- Schecter AD, Anziska B: Isolated complete post-traumatic trigeminal neuropathy. Neurology 1990;40:1634-

**Revised**
- 23.4.1997 BF

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**INFERIOR ALVEOLAR NERVE LESION (V)**

**Etiology**
- extraction of tooth from lower jaw (usually wisdom’s tooth)
- sagittal osteotomy
- during intraosseal tooth implantations
- tumors of the mandible

**Clinical features**
- numbness and/or loss of sensation of the skin below the lower lip

**Strategy**
- demonstrated conduction abnormality in the inferior alveolar nerve

**Expected abnormal findings**

**Neurography**
- inferior alveolar nerve SCS

**Blink reflex**
- abnormal finding with stimulation of the mental nerve

**Expected normal findings**

**EMG**
- m.masseter/m.temporalis

**Blink reflex**
- normal finding with stimulation of the infraorbital and supraorbital branches

**Procedure**

**EMG**
- m.masseter

**Neurography**
- n.alveolaris inferior bilaterally with near nerve needle electrodes

**References**

**Revised**
-
LINGUAL NERVE LESION (V)

**Etiology**
- extraction of 2. or 3. molar teeth
- sagittal osteotomy
- tumors of the mandible

**Clinical features**
- loss of sensation and paresthesia over the tongue, floor of the mouth and medial side of gingiva,
- loss of taste over the anterior two thirds of the tongue

**Strategy**
- demonstrated conduction abnormality in the inferior alveolar nerve

**Expected abnormal findings**

**Neurography**
- inferior alveolar nerve SCS

**Blink reflex**
- abnormal finding with stimulation of the mental nerve

**Expected normal findings**

**EMG**
- m.masseter/m.temporalis

**Blink reflex**
- normal finding with stimulation of the infraorbital and supraorbital branches

**Procedure**

**EMG**
- m.masseter

**Neurography**
- n.alveolaris inferior bilaterally with near nerve needle electrodes

**References**

ABDUCENS NERVE LESION (VI)

**Etiology**
- acute idiopathic mononeuropathy (especially in diabetes)
- vascular lesions in the nerve
- head trauma
- neoplasms involving the brainstem nuclei, intracranial portion of the nerve (acoustic neuromas, meningeomas)
- vascular lesions in the brainstem
- aneurysm
- infections (meningitis, borreliosis, tuberculosis, herpes zoster, AIDS)
- sarcoidosis
- Möbius syndrome (congenital weakness in muscles innervated by n.abducens and n.facialis, probably due to congenital agenesis of the brainstem nuclei)

**Clinical features**
- diplopia
- weakness of abduction lateral gaze)of the eye

**Strategy**
- demonstrate neurogenic EMG findings in muscles innervated by the oculomotor nerve
- exclude possibility of other cranial nerve lesions
- exclude ocular myopathy

**Expected abnormal findings**

**EMG**
- m.rectus lateralis

**Expected normal findings**

**EMG**
- m.obliqus superior
- m.rectus superior
- m.rectus inferior
- m.rectus medialis
- m.obliqus inferior
- m.levator palpebrae superior

**Procedure**

**EMG**
- m.rectus lateralis (abduces nerve)
- m.obliqus superior (trochlear nerve)
- m.rectus superior/m.rectus inferior/m.rectus medialis/m.obliqus inferior (oculomotor nerve)
- m.levator palpebrae superior (oculomotor nerve)

**References**
- Rush JA, Younge BR: Paralysis of cranial nerves III, IV, and IV: Cause and prognosis in 1000 cases. Arch Ophthalmol 1981;99:76-

**Modified**
- 23.4.1997 BF
**FACIAL NERVE LESION (VII)**

**Etiology**
- **Bell’s palsy**
- **polyradiculitis**
- **trauma**
- **herpes zoster, (Ramsay Hunt syndrome)**
- **borreliosis**
- **Möbius syndrome** (congenital weakness in muscles innervated by n.abducens and n.facialis, probably due to congenital agenesis of the brainstem nuclei)
- **tumors of the parotid gland**
- **surgery in the middle ear** (cholesteatoma, stapedectomy, tympanoplasty etc), acustic neuroma, glomus jugulare tumours, parotid gland tumours, facielifting operations

**Clinical features**
- weakness of the facial muscles
- inability close eyelids
- depending on the level of the lesion there may be loss of taste over the tongue
- numbness and loss of sensation of n.intermedius behind and around the ear

**Strategy**
- demonstrate neurogenic EMG findings in muscles innervated by the facial nerve
- assess pathophysiology (neurapraxia, axonal degeneration) and severity

**Expected abnormal findings**

**EMG**
- m.orbicularis oculi/m.frontalis
- m.oribicularis oris/m.zygomaticus major

**Neurography**
- facial nerve MCS: reduced amplitude

**Blink reflex**
- delayed efferent path of the blink reflex

**Expected normal findings**

**EMG**
- m.masseter

**Procedure**

**EMG**
- m.orbicularis oculi/m.frontalis
- m.oribicularis oris/m.zygomaticus major

**Neurography**
- facial nerve MCS:
  - r.temporalis: m.frontalis
  - r.zygomaticus: m.nasalis
  - r.buccalis: m.orbicularis oris
  - r.mandibularis: m.depressor anguli oris
  - r.cervicalis: m.platysma

**Blink reflex**

**References**
- Adour KK: Diagnosis and management of facial paralysis. N Engl J Med 1982;307:348-
- Deshpande AD: Recurrent Bell's palsy in pregnancy. J Laryngol Otol 1990;104:713-
- George MD, Pahor AL: Sarcoidosis: A cause for bilateral facial palsy. Ear Nose Throat J 1991;70:492-
- Hoffmann DF, May M, Kubal W: Slowly progressive facial paralysis due to vascular malformation of the brain stem. Am J Otol 1190:90:1:357-
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- King TT, Morrison AW: Primary facial nerve tumors within the skull. J Neurosurg 1990;72:1-
- McKennan KX, Chole RA: Facial paralysis is temporal bone trauma. Am J Otol 1992;13:167-
- Monkhouse WS: The anatomy of the facial nerve. Ear Nose Throat J 1990;69:677-
- Nielsen VK, Jannetta PJ: Pathophysiology of hemifacial spasm, part III. Effects of facial nerve decompression. Neurology 1984;34:891-
GLOSSOPHARYNGEAL NERVE LESION (IX)

**Etiology**
- processes around the jugular foramen (the vagus nerve and spinal accessory nerves are also affected)
- acute idiopathic mononeuropathy (neuralgic amyotrophy)
- isolated glossopharyngeal nerve lesions are very uncommon

**Clinical features**
- difficulties in swallowing
- numbness and loss of sensation of the pharynx

**Strategy**
- demonstrate neurogenic EMG findings in muscles innervated by the glossopharyngeal nerve

**Expected abnormal findings**
- m.stylopharyngeus

**Expected normal findings**
- m.genioglossus/m.hyoglossus
- m.trapezius
- m.cricothyroideus

**Procedure**
- m.stylopharyngeus
- m.cricothyroideus (n.laryngeus superior)
- m.trapezius/m.sternocleidomastoideus (n.accessorius)
- m.genioglossus/m.hyoglossus (m.hypoglossus)

**References**

VAGUS NERVE LESION (X)

**Etiology**
- surgery around the thyroid gland
- acute idiopathic mononeuropathy (neuralgic amyotrophy)
- vascular lesions
- processes around the jugular foramen n. glossopharyngeus and n. accessorius are also affected

**Clinical features**
- numbness and loss of sensation of the external ear
- dysphagia
- dysarthria

**Strategy**
- demonstrate neurogenic EMG findings in muscles innervated by the vagus nerve

**Expected abnormal findings**
- m.vocalis/m.cricoarytenoideus
- m.cricothyroideus

Revised
- 23.4.1997, 12.5.1997 BF
**m.palatoglossus/m.palatopharyngeus**

**Expected normal findings**

**EMG**
- m.trapezius/m.sternocleidomastoideus
- m.genioglossus/m.hyoglossus

**Procedure**

**EMG**
- m.vocalis/m.cricoarytenoideus (intrinsic muscles of the larynx)
- m.cricothyroideus (n.laryngeus superior)
- m.palatoglossus/m.palatopharyngeus (n.vagus)
- m.trapezius/m.sternocleidomastoideus (n.accessorius)
- m.genioglossus/m.hyoglossus (m.hypoglossus)

**References**

- Berry H: Isolated vagus nerve palsy and vagal mononeuritis. Arch Otolaryngol 1980;106:333-
- Pierre PA, Laterre CE, Van Den Bergh PY: Neuralgic amyotrophy with involvement of cranial nerves IX, X, XI, and XII. Muscle Nerve 1990;13:704-

**Revised**
- 23.4.1997

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**RECURRENT NERVE LESION (X)**

**Etiology**
- surgery around the thyroid gland, carotid endarteractomy
- acute idiopathic mononeuropathy
- in association with systemic diseases: LED, AIDS

**Clinical features**
- dysarthria

**Strategy**
- demonstrate neurogenic EMG findings in muscles innervated by the inferior laryngeal nerve

**Expected abnormal findings**

**EMG**
- m.vocalis/m.cricoarytenoideus (intrinsic muscles of the larynx)

**Expected normal findings**

**EMG**
- m.cricothyroideus (innervated by n.laryngeus superior)

**Procedure**

- m.cricothyroideus
- m.vocalis/m.cricoarytenoideus
- m.palatoglossus/m.palatopharyngeus
  if clinically indicated
- m.sternocleidomastoideus
- m.genioglossus/m.hyoglossus (glossopharyngeal nerve)

**References**

- Pierre PA, Laterre CE, Van Den Bergh PY: Neuralgic amyotrophy with involvement of cranial nerves IX, X, XI, and XII. Muscle Nerve 1990;13:704-

**Revised**
- 22.7.1997

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**ACCESSORY NERVE LESION (XI)**

**Etiology**
- lymph node biopsy, or excision
- surgical trauma, radical neck dissection, l
- blunt or penetrating trauma
- jugular vein cannulation
- cervical internal carotid artery dissection
- carotid endarterectomy
- radiation therapy to head and neck
Clinical features

- Drooping of the shoulder
- Winging of the scapula
- Weakness of shoulder elevation and head rotation to the contralateral side

Strategy

- Demonstrate neurogenic EMG findings in muscles innervated by the accessory nerve
- Localize lesion (trapezius with or without sternocleidomastoideus lesion)
- Exclude other reason for atrophy

Expected abnormal findings

- EMG
  - M. trapezius
  - M. sternocleidomastoideus is involved in proximal lesions (not denervated in accessory nerve lesions due to lymph node biopsies)

Related by |

- Neurography
  - MCS: n.accessorius reduced amplitude

Expected normal findings

- EMG
  - M. masseter
  - M. deltoideus
  - M. orbicularis oris

Procedure

- EMG
  - M. trapezius (upper, middle and lower portion should be examined separately)
  - M. sternocleidomastoideus
  - M. hyoglossus/m. genioglossus
  - M. masseter
  - M. orbicularis oris/m. zygomaticus major

Neurography (optional)

- MCS: n.accessorius

References

- Dellon AL, Campbell JN, Comblath D: Stretch palsy of the spinal accessory nerve: Case report. J Neurosurg 1990;72:500-
- Eisen A, Bertrand G: Isolated accessory nerve palsy of spontaneous origin: A clinical and electromyographic study. Arch Neurol 1972;27:496-
- Hoffman JC: Permanent paralysis of the accessory nerve after cannulation of the internal jugular vein. Anesthesiology 1983;58:583-
- Petrera JE, Trojaborg W: Conduction studies along the accessory nerve and follow-up of patients with trapezius palsy. J Neurol Neurosurg Psychiatry 1984;47:630-
- Pierre PA, Laterre CE, Van Den Bergh PY: Neuralgic amyotrophy with involvement of cranial nerves IX, X, XI, and XII. Muscle Nerve 1990;13: 704-
- Vastamaki M, Solonen KA: Accessory nerve injury. Acta Orthop Scand 1984;55.296-

Revised

- 23.4.1997

Hypoglossal Nerve Lesion (XII)

Etiology

- Acute idiopathic mononeuropathy (neuralgic amyotrophy)
- Fracture of the skull base
- Tumors of the skull base and neck
- Aneurysms of the carotid artery
- Carotid endarterectomy
- Clivus chordoma
- Glomus jugulare tumor
- Metastases to base of skull
- Carotid artery aneurysm at base of skull
- Atlantoaxial subluxation
Clinical features
* isolated hypoglossal nerve lesions are very rare
* weakness and atrophy of the tongue

Strategy
* demonstrate neurogenic EMG findings in muscles innervated by n.hypoglossus (mainly muscles of the tongue)
* differentiate from bulbar paralysis involving other cranial nerves

Expected abnormal findings
EMG
* m.hypoglossus/m.genioglossus

Neurography
* MCS: m.hypoglossus reduced amplitude

Expected normal findings
EMG
* m.masseter
* m.orbicularis oris

Procedure
EMG
* m.hypoglossus/m.genioglossus
* m.masseter
* m.orbicularis oris/m.zygomaticus major

Neurography (optional, measurement requires special electrode)
* MCS: n.hypoglossus

References
* DeCock M. Nervus hypoglossus (n. XII). Acta Otorhinolaryngol Belg 1986;40:260-
* Greenberg HS, Deck MDF, Vikram B et al: Metastasis to the base of the skull: Clinical findings in 43 patients. Neurology 1981;31:530-
* Jabourian A, Mikaelian DO: Hypoglossal nerve paralysis. Trans Pa Acad Ophthalmol Otolaryngol 1989;41:87-
* Pierre PA, Laterre CE, Van Den Bergh PY: Neuralgic amyotrophy with involvement of cranial nerves IX, X, XI, and XII. Muscle Nerve 1990;13: 70-

Revised
* 23.4.1997 BF

5.3 LOWER EXTREMITIES

Sciatic nerve lesion

Etiology
* surgery for hip joint replacement
* gluteal injection
* gluteal contusion
* nerve infarction
* fractures of the femur
* hip fracture and/or dislocation
* metastatic lesions
* radiation therapy

Clinical features
* weakness of muscles innervated by n.ischiadicus
* usually n.peroneus is more affected than n.tibialis
* numbness and/or loss of sensation in the foot
* pain may be present

Strategy
* demonstrate neurogenic EMG findings in muscles innervated by n.ischiadicus
* exclude radiculopathy or other proximal lesion
* define level of lesion if possible

Expected abnormal findings
EMG:
* neurogenic findings in 1-5 depending on the level of the lesion (examine one from each set of muscles)
  * 1. biceps femoris caput brevis
  * 2. semitendinosus/semimembranosus
  * 3. tibialis anterior/peroneus longus/extensor hallucis longus
  * 4. gastrocnemius caput meduale or laterale.
  * 5. intrinsic foot muscles

Neurography:
* low M-amplitude in m.extensor digitorum brevis, abductor hallucis
* F-wave prolonged or absent in n.peroneus or n.tibialis
* n.suralis SNAP amplitude reduced
* n.peroneus superficialis SNAP amplitude reduced
* H-reflex absent or prolonged

Expected normal findings
EMG
* paravertebral muscles
* gluteus maximus (n.gluteus inferior)
LUMBOSACRAL PLEXUS LESION

**Etiology**
- often the lumbar and the sacral plexus are separately affected but sometimes both may be affected
- traumatic lesions in the hip region
- surgery for hip joint replacement
- injection into erroneous site
- tumors and metastatic lesions
- radiation therapy
- acute idiopathic mononeuropathy (neuralgic amyotrophy) (in diabetes patients often called diabetic amyotrophy)
- pelvic fractures
- aortic aneurysm
- pregnancy
- retroperitoneal hematoma

**Clinical features**
- weakness of muscles innervated by n.ischiadicus
- weakness of muscles innervated by n.femoralis and n.obturatorius
- usually n.peroneus is more affected than n.tibialis
- numbness and/or loss of sensation in the thigh, leg and foot
- pain may be present

**Strategy**
- demonstrate neurogenic EMG findings in muscles innervated by the lumbosacral plexus
- exclude radiculopathy

**Expected abnormal findings**

**EMG**
- neurogenic findings in muscles innervated by the lumbosacral plexus
  - gluteus maximus (n.gluteus inferior)
  - tensor fascia latae/gluteus medius (n.gluteus superior)
  - biceps femoris caput brevis
  - semitendinosus/semimebranosus
  - tibialis anterior/peroneus longus/extensor hallucis longus
  - gastrocnemius caput medial or laterale.
  - quadriceps femoris
  - adductor magnus
  - intrinsic foot muscles

**Neurography**
- low M-amplitude in m.extensor digitorum brevis, m.abductor hallucis
- F-wave prolonged or absent in n.peroneus or n.tibialis
- n.suralis SNAP amplitude reduced
- n.saphenus SNAP amplitude reduced
- H-reflex absent or prolonged

**Expected normal findings**

**EMG**
- paravertebral muscles

**Procedure**
- EMG:
  - examine one from each set of muscles
    - m.gluteus maximus (n.gluteus inferior)
    - m.tensor fascia latae/m.gluteus medius (n.gluteus superior)
    - m.biceps femoris caput brevis
    - m.semitendinosus/m.semimebranosus
    - m.tibialis anterior/m.peroneus longus/m.extensor hallucis longus
    - m.gastrocnemius caput medial or laterale.
    - m.quadriceps femoris/m.iiliopsoas
    - m.adductor magnus

**Neurography**
- MCS: n.peroneus, n.tibialis
- SCS: n.suralis, n.peroneus superficialis, n.saphenus
- H-reflex

**References**
**LUMBAR PLEXUS LESION**

**Etiology**
- acute idiopathic mononeuropathy (neuralgic amyotrophy) (in diabetes patients often called diabetic amyotrophy)
- trauma
- pelvic fractures
- aortic aneurysm
- pregnancy
- retroperitoneal hematoma
- tumors
- radiation

**Clinical features**
- weakness of knee extension, thigh abduction and flexion
- numbness and/or loss of sensation over the anterior and medial side of the thigh
- loss of patellar tendon reflex

**Strategy**
- demonstrate neurogenic EMG findings in muscles innervated by the lumbar plexus
- exclude radiculopathy

**Expected abnormal findings**

**EMG**
- neurogenic findings in:
  - m.quadriceps femoris (m.vastus lateralis/m.vastus medialis or m.recuts femoris)
  - tibialis anterior
  - iliopsoas
  - adductor magnus

**Neurography**
- n.saphenus SNAP amplitude reduced

**Expected normal findings**

**EMG**
- paravertebral muscles
- m.gluteus maximus (n.gluteus inferior)
- m.tensor fascia latae/gluteus medius (n.gluteus superior)
- m.biceps femoris caput brevis
- m.semimembranosus/m.semimembranosus
- m.gastrocnemius caput mediale or laterale
- intrinsic foot muscles

**Neurography**
- n.tibialis and n.peroneus to m.extensor digitorum brevis normal
- n.suralis normal
- H-reflex normal

**Procedure**

**EMG:**
- examine one from each set of muscles
  - m.tensor fascia latae/gluteus medius (n.gluteus superior)
  - m.tibialis anterior/peroneus longus/extensor hallucis longus
  - m.gastrocnemius caput mediale or laterale.
  - m.quadriceps femoris
  - m.adductor magnus
  - m iliopsoas

**Neurography**
- MCS: n.peroneus, n.tibialis
- SCS: n.suralis, n.peroneus superficialis, n.saphenus
- H-reflex

**References**

**Revised**
- 8.5.1997 BF

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**SACRAL PLEXUS LESION**

**Etiology**
- traumatic lesions in the hip region
- surgery for hip joint replacement
- injection into erroneous site
- tumors and metastatic lesions
- radiation therapy
**Acute idiopathic mononeuropathy (neuralgic amyotrophy)**

- aortic aneurysm
- pregnancy

**Clinical features**

- weakness of muscles innervated by n. ischiadicus
- usually n. peroneus is more affected than n. tibialis
- numbness and/or loss of sensation in the foot
- pain may be present

**Strategy**

- demonstrate neurogenic EMG findings in muscles innervated by the sacral plexus
- exclude radiculopathy

**Expected abnormal findings**

**EMG**

- neurogenic findings in muscles innervated by the lumbosacral plexus
  - m. gluteus maximus (n. gluteus inferior)
  - m. tensor fascia latae/m. gluteus medius (n. gluteus superior)
  - m. biceps femoris caput brevis
  - m. semitendinosus/m. semimembranosus
  - m. tibialis anterior/m. peroneus longus/m. extensor hallucis longus
  - m. gastrocnemius caput mediale or laterale.
  - m. quadriiceps femoris
  - (m. adductor magnus) this muscle may also be innervated from n. ischiadicus
- intrinsic foot muscles

**Neurography**

- low M-amplitude in EDB, abductor hallucis
- F-wave prolonged or absent in n. peroneus or n. tibialis
- n. suralis SNAP amplitude reduced
- H-reflex absent or prolonged

**Expected normal findings**

**EMG**

- paravertebral muscles

**Neurography**

- SCS: n. saphenus

**Procedure**

**EMG:**

- examine one from each set of muscles
  - m. gluteus maximus (n. gluteus inferior)
  - m. tensor fascia latae/m. gluteus medius (n. gluteus superior)
  - m. biceps femoris caput brevis
  - m. semitendinosus/m. semimembranosus
  - m. tibialis anterior/m. peroneus longus/m. extensor hallucis longus
  - m. gastrocnemius caput mediale or laterale.
  - m. quadriiceps femoris
  - m. adductor magnus

**Neurography**

- MCS: n. peroneus, n. tibialis
- SCS: n. suralis, n. peroneus superficialis, n. saphenus
- H-reflex

**References**


**Revised**

- 8.5.1997

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**Pudendal Nerve Lesion**

**Etiology**

- pelvic tumors
- pelvic fractures and surgery for pelvic fractures
- surgery
- perineal branches may be compressed during cycling

**Clinical features**

- urinary bladder dysfunction, mainly weakness of the bladder contractility
- anal incontinence, weakness of the sphincter ani externus
- impotence
- numbness and/or loss of sensation around the perianal region and in the penis

**Strategy**

- demonstrate neurogenic EMG findings in muscles innervated by the pudendal nerve

**Expected abnormal findings**
LESION OF THE LATERAL CUTANEOUS NERVE OF THE THIGH (MERALGIA PARESTHETICA)

**Etiology**
- entrapment at the anterior superior iliac spine
- surgery to the anterior iliac crest (bone transplantation)
- surgery around the anterior iliac spine

**Clinical features**
- numbness and/or loss of sensation over the anterior-lateral part of the thigh
- sometimes pain

**Strategy**
- demonstrate abnormality of the lateral cutaneous nerve of the thigh at the superior anterior iliac spine

**Expected abnormal findings**

**Neurography**
- lateral cutaneous nerve of the thigh SCS abnormal at the anterior superior iliac spine

**Expected normal findings**

**Procedure**

**Neurography**
- SCS: n.cutaneus femoris lateralis, ortodromically with near nerve needle electrodes

**References**
- Beresford HR. Meralgia paresthetica after seat-belt trauma. J Trauma 1971;1:629-630
- Ecker AD, Woltman HW. Meralgia paresthetica: a report of one hundred and fifty cases JAMA 1938;110:1650-1652
- Nathan H. Gangliform enlargement on the lateral cutaneous nerve of the thigh. J Neurosurg 1960;17:843450
- Stevens H. Meralgia paresthetica. Arch Neurol Psychiatry 1957;77:557-574
- Stokey B. Meralgia paresthetica: etiology and surgical treatment. JAMA 1928;90:1705-1707
**ILIOPINGUAL NERVE LESION**

**Etiology**
- retroperitoneal tumors
- surgery, especially herniorrhaphy and kidney operations
- acute idiopathic mononeuropathy ("neuralgic amyotrophy, neuritis")
- entrapment has been described as the nerve emerges through the fascia lata

**Clinical features**
- weakness of the lower abdominal muscles, bulging of the abdomen
- numbness and/or loss of sensation over the symphysis, labia majora (in women), base of the penis and scrotum (in men)

**Strategy**
- demonstrate neurogenic EMG findings in m.obliqus internus abdominis
- differentiate from ilioinguinal nerve and genitofemoral nerve lesions

**Expected abnormal findings**

**EMG**
- m.obliqus internus abdominis

**SEP**
- SEP by stimulation of the sensory areas innervated by the nerve

**Expected normal findings**

**EMG**
- m.iliopsoas
- m.rectus abdominis

**Procedure**

**EMG**
- m.obliqus internus abdominis
- m.iliopsoas (femoral nerve, L2-L4 radiculopathy)

**Neurography**
- sometimes it is possible to record SCS with needle electrodes

**SEP**
- SEP by stimulation of the sensory areas innervated by the nerve

**References**

**ILIOLYPOGASTRIC NERVE LESION**

**Etiology**
- retroperitoneal tumors
- surgery for inguinal hernia, especially kidney
- acute idiopathic mononeuropathy ("neuralgic amyotrophy, neuritis")
- the anterior sensory branch can be damaged by Pfannenstiel’s incision

**Clinical features**
- weakness of the lower abdominal muscles, bulging of the abdomen
- numbness and/or loss of sensation below and behind the spinous process of the ilium

**Strategy**
- demonstrate neurogenic EMG findings in m.obliqus internus abdominis
- differentiate from ilioinguinal nerve and genitofemoral nerve lesions

**Expected abnormal findings**

**EMG**
- m.obliqus internus abdominis

**SEP**
- SEP by stimulation of the sensory areas innervated by the nerve: lateral cutaneous branch and anterior cutaneous branch

**Expected normal findings**

**EMG**
- m.iliopsoas
- m.rectus abdominis

**Neurography**
- SCS: n.ilioinguinalis

**Procedure**

**EMG**
- m.obliqus internus abdominis
- m.iliopsoas (femoral nerve, L2-L4 radiculopathy)

**SEP**
- SEP by stimulation of the sensory areas innervated by the nerve: lateral cutaneous branch and anterior cutaneous branch
NEUROGRAPHY

SCS: n.ilioinguinalis

REFERENCES


Revised

6.4.1997

GENITOFEMORAL NERVE LESION

ETIOLOGY
hemorrhaphy and other operations in the lateral pelvis or inguinal ligament
acute idiopathic mononeuropathy (*neuralgic amyotrophy, neuritis*)

CLINICAL FEATURES
numbness and/or loss of sensation medially below the inguinal ligament
abnormal cremaster reflex

STRATEGY
there are no simple reliable nerve conduction methods to study this nerve
exclude ilioinguinal nerve lesion and iliohypogastric lesion

EXPECTED ABNORMAL FINDINGS

SPECIAL STUDIES
abnormal cremaster reflex
n.genitofemoralis

EXPECTED NORMAL FINDINGS

EMG
transverse abdominal muscles

NEUROGRAPHY
SCV: n.ilioinguinalis
SEP
n.ilioinguinalis
n.iliohypogastricus

PROCEDURE
SPECIAL STUDIES
abnormal cremaster reflex

EMG
transverse abdominal muscles

NEUROGRAPHY
SCV: n.ilioinguinalis
SEP
n.genitofemoralis
n.ilioinguinalis
n.iliohypogastricus

REFERENCES

Revised

6.4.1997

FEMORAL NERVE LESION

ETIOLOGY
trauma in the inguinal region
surgery: appendectomy, hip operations, hemorrhaphy, hysterectomy through Pfannenstiel’s incision
arteriography through the femoral artery
femoral artery reconstruction
hematoma in the groin, retroperitoneal hematoma
metastasis in the retroperitoneal region
retroperitoneal abscess
aneurysm of the femoral artery
acute idiopathic mononeuropathy (neuralgic amyotrophy)

CLINICAL FEATURES
weakness of knee extension
atrophy of m.quadriceps femoris
loss patellar tendon reflex
numbness and/or loss of sensation over the anterior part of the thigh and anterior-medial part of the leg

STRATEGY
demonstrate neurogenic EMG findings in muscles innervated by the femoral nerve and abnormal SNC findings in n.saphenus and r.cutaneus femoris anterior
differentiate from lumbar plexus lesion and L2-L4 radiculopathy

EXPECTED ABNORMAL FINDINGS

EMG
m.quadriceps femoris
m.iliopectas if the lesion is above the inguinal ligament

NEUROGRAPHY
abnormal SNC in n.saphenus
abnormal SNC in r.cutaneus femoris anterior

**Expected normal findings**

**EMG**
- m.tibialis anterior
- m.adductor magnus

**Neurography**
- SCS: n.suralis

**Procedure**

**EMG**
- m.iiilopsoas
- m.vastus lateralis/m.vastus medialis
- m.adductor magnus (obturator nerve, L2-L3 radiculopathy)
- m.tibialis anterior (peroneal nerve, L4 radiculopathy)
- paravertebral muscles L2-L4 (radiculopathy)

**Neurography**
- SNC: n.saphenus
- SNC: r.cutaneus femoris anterior (optional)

**References**


**Revised**
- 7.4.1997 BF

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**SAPHENOUS NERVE LESION**

**Etiology**
- iatrogenic lesion following surgery for varicose veins

**Clinical features**
- loss of sensation over the anterior-medial part of the thigh

**Strategy**
- demonstrate abnormality of n.saphenus
- exclude proximal nerve lesions, radiculopathy L3 or L4, and lumbar plexus lesions

**Expected normal findings**

**Neurography**
- saphenous nerve SCS

**Expected normal findings**

**EMG**
- m.tibialis anterior
- m.vastus lateralis/medialis
- m.adductor magnus

**References**

- Garnjobs W. Injuries to the saphenous nerve following operations for varicose veins. Surg Gynecol Obstet 1964;119:359-361

**Revised**
- 6.4.1997 BF

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**OBTURATOR NERVE LESION**

**Etiology**
- acute idiopathic mononeuropathy (neuralgic amyotrophy)
- pelvic fractures
- complication of hip replacement
- obturator hernia
- metastasis of the bones around the obturator foramen

**Clinical features**
- weakness of thigh adduction
- loss of sensation or numbness on the inner side of the thigh

**Strategy**
- demonstrate neurogenic EMG findings in muscles innervated by the obturator nerve
- differentiate from L2-L4 radiculopathy and lumbar plexus lesion

**Expected abnormal findings**

**EMG**
- m.adductor magnus/m.adductor longus/m.gracilis
- SEP
- n.obturatorius
Expected normal findings

EMG
- m.vastus lateralis/m.vastus medialis
- m iliopsoas
- m.tibialis anterior
- paravertebral muscles L2-L4

Procedure

EMG
- m.adductor magnus/m.adductor longus
- m.vastus lateralis/m.vastus medialis
- m iliopsoas
- m.tibialis anterior
- m.tensor fascia latae
- paravertebral muscles L2-L4

SEP
- stimulation of the skin area innervated by the cutaneous branch of the obturator nerve

Note
- m.adductor magnus may be innervated also from the sciatic nerve
- m.adductor longus may be innervated from the femoral nerve

References

Revised
* 16.4.1997 BF

POSTERIOR CUTANEOUS FEMORAL NERVE LESION

Etiology
- lacerations over the posterior part of the thigh
- acute idiopathic mononeuropathy (neuralgic amyotrophy)
- injections into the buttock
- pelvic fractures
- metastatic tumors

Clinical features
- numbness and/or loss of sensation over the dorsal side of the thigh

Strategy
- demonstrate abnormal findings in n.cutaneus femoris posterior
- differentiate from sciatic nerve lesions

Expected abnormal findings

Neurography
- SCS: n.cutaneus femoris posterior

Expected normal findings

EMG
- m.vastus lateralis/m.vastus medialis
- m.tibialis anterior
- paravertebral muscles L2-L4

Neurography
- SCS: n.suralis
- MCS: n.peroneus profundus, n.tibialis

Procedure

EMG
- m.vastus lateralis/m.vastus medialis
- m.tibialis anterior
- m.gastrocnemius
- m.biceps femoris
- paravertebral muscles L4-S1

Neurography
- SCS: n.cutaneus femoris posterior, n.suralis
- MCS: n.tibialis, n.peroneus

References

Revised
* 16.4.1997

COMMON PERONEAL NERVE LESION AT THE KNEE

Etiology
- trauma to the lateral side of the knee
- prolonged kneeling, when picking berries
- inversion distorsion of the ankle
- prolonged pressure to the fibular head, e.g. when sitting with knees crossed
" surgery around the knee

**Clinical features**
- weakness of ankle dorsiflexion and extension of toes
- loss of sensation or numbness over the dorsal side of the foot and antero-lateral part of the leg

**Strategy**
- demonstrate neurogenic EMG findings in muscles innervated by the common peroneal nerve
- demonstrate conduction abnormality across fibular head
- exclude proximal lesion

**Expected abnormal findings**

**EMG**
- m.tibialis anterior
- m.extensor hallucis longus
- m.peroneus longus
- m.peroneus brevis

**Neurography**
- n.peroneus profundus MCS across fibular head abnormal: conduction block or conduction velocity reduced
- n.peroneus superficialis SNAP amplitude reduced

**Expected normal findings**

**EMG**
- m.tibialis posterior
- m.flexor hallucis longus
- m.biceps femoris caput brevis
- m.semitendinosus
- m.gluteus medius
- m.tensor fascia latae
- paravertebral muscles L5

**Neurography**
- SCS: n.suralis
- MCS: n.tibialis

**Procedure**

**EMG**
- m.tibialis anterior/m.extensor hallucis longus (n.peroneus profundus)
- m.peroneus longus/m.peroneus brevis neurogenic (n.peroneus superficialis)
- m.tibialis posterior/m.flexor hallucis longus (n.tibialis, L5)
- m.gluteus medius/m.tensor fascia latae (L5)
- m.biceps femoris caput brevis (optional, sciatic nerve)

**Neurography**
- MCS: n.peroneus profundus across fibular head and below the knee (if no response or very small response is recorded from m.extensor digitorum brevis the measurement should be made with surface electrodes over m.tibialis anterior)
- SCS: n.peroneus superficialis

**Note**
- In most people m.extensor digitorum brevis is innervated solely by n.peroneus profundus. In a few persons the muscle receives innervation from a branch of n.peroneus superficialis (n.peroneus accessorius)

**References**
ANTERIOR TARSAL TUNNEL SYNDROME

**Etiology**
- compression of the distal part of n.peroneus profundus at the ankle

**Clinical features**
- loss of sensation over the dorsal side of toes I and II
- atrophy of m.extensor digitorum brevis

**Strategy**
- demonstrate lesion of the distal part of n.peroneus profundus
- exclude proximal lesion of the peroneal nerve

**Expected abnormal findings**

**Neurography**
- abnormal SCS of n.peroneus profundus (toes to ankle)

**EMG**
- m.extensor digitorum brevis

**Expected normal findings**

**Neurography**
- SCS: n.suralis
- SCS: n.peroneus superficialis

**EMG**
- m.tibialis anterior/m.extensor hallucis longus

**Strategy**

**EMG**
- m.tibialis anterior/m.extensor hallucis longus
- m.extensor digitorum brevis
- m.peroneus longus/m.peroneus brevis

**Neurography**
- SCS: n.peroneus superficialis both branches
- SCS: n.suralis
- SCS: n.peroneus profundus
- MCS: n.peroneus

**References**
- Krause KH, Witt T, Ross A: The anterior tarsal tunnel syndrome. J Neurol 1977;217:67-
- Marinacchi AA: Neurological syndromes of the tarsal tunnels. Bull Los Angeles Neurol Soc 1968;33:90-

**Revised**
- 6.4.1997 BF, 8.5.1997 BF

SUPERFICIAL PERONEAL NERVE LESION IN THE FOOT

**Etiology**
- severe distorsion of the ankle
- surgery to the forefoot or lower part of the leg
- traumatic herniation of muscle trough fascia
- usually a lesion of the laterally located branch of the terminal sensory branches

**Clinical features**
- loss of sensation on the dorsal side of the foot pain in the forefoot

**Strategy**
- demonstrate lesion of the distal superficial peroneal nerve branch

**References**

**Revised**
- 3.4.1997
Exclude proximal lesion of the peroneal nerve

**Expected abnormal findings**

**Neurography**

- abnormal SCS of the lateral branch of n.peroneus superficialis

**Expected normal findings**

**Neurography**

- SCS: n.suralis
- SCS: medial branch of n.peroneus superficialis

**EMG**

- m.tibialis anterior

**Strategy**

**EMG**

- m.tibialis anterior/m.extensor hallucis longus

**Neurography**

- SCS: n.peroneus superficialis both branches
- SCS: n.suralis
- MCS: n.peroneus

**References**


**Revised**

- BF 8.5.1997

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**TARSA L TUNNEL SYNDROME**

**Etiology**

- Entrapment neuropathy of the tibial nerve or its branches in the area of the medial malleolus under the flexor retinaculum
- Usually due to rheumatoid arthritis, tenosynovitis or malleolar fractures

**Clinical features**

- Very rare
- Numbness and pain over the sole of the foot
- Pain around the medial malleolus

**Strategy**

- Demonstrate neurogenic EMG findings in intrinsic foot muscles innervated by the lateral and medial plantar nerve
- Demonstrate reduced CV in the tibial nerve across the tarsal tunnel
- Differentiate from proximal tibial nerve lesion

**Expected abnormal findings**

**EMG**

- m.abductor hallucis
- m.abductor digiti minimi

**Neurography**

- MCS: reduced CV or prolonged DL in n.tibialis across the tarsal tunnel
- SCS: reduced CV in n.tibialis across the tarsal tunnel

**Expected normal findings**

**EMG**

- m.gastrocnemius c.mediale/laterale
- m.tibialis anterior

**Neurography**

- SCS: n.suralis
- MCS: n.peroneus

**Procedure**

**EMG**

- m.abductor hallucis
- m.abductor digiti minimi
- m.extensor digitorum brevis
- m.gastrocnemius c.mediale/laterale
- m.tibialis anterior

**Neurography**

- MCS: n.tibialis across the tarsal tunnel (use near nerve needles for stimulation)
- SCS: n.tibialis across the tarsal tunnel (use near nerve needles for recording)

**Note**

- Although this is often suspected this is a very rare disorder, we have never seen an idiopathic tarsal tunnel syndrome without deformity of the ankle around the medial malleolus
- The intrinsic foot muscles show mild to moderate neurogenic abnormalities even in healthy subjects, compare findings with the opposite foot

**References**

- Dellon AL, Mackinnon SE. Tibial nerve branching in the tarsal tunnel. Arch Neurol 1984;41:645-646
MORTON’S METATARSALGIA

Etiology
* entrapment of the plantar digital nerves between the distal metatarsal heads
* usually the digital nerves II and III, between the II/III and III/IV metatarsal heads, are affected

Clinical features
* a fairly common disorder in 50-70 year old women, sometimes seen in younger persons
* pain in the forefoot when walking, often symptoms are alleviated if shoes are taken off
* clinically often distinct painful area between affected metatarsal heads
* associated with hallux valgus and rheumatoid arthritis
* plantar digital nerve to interspaces II/III and III/IV commonly affected, in most patients both interspaces affected

Strategy
* demonstrate lesion of the plantar digital nerves

Expected abnormal findings
Neurography
* SCS: reduced CV of the affected plantar digital nerves
* SCS: reduced sensory AMPL of the affected plantar digital nerves

Procedure
* orthodromic SCS with near nerve needle electrodes in all four interspaces (I/II, II/III, III/IV and IV/V)
* either lateral or medial sides of toes can be studied
* in borderline cases it may be helpful to study both lateral and medial plantar digital branches

Note
* in most patients the digital nerves II (between II/III metatarsal heads) and III (between III/IV metatarsal heads) are affected

References

SURAL NERVE LESION AT THE KNEE

Etiology
* Baker's cyst
* surgery, arthroscopy of the knee
* knee trauma

Clinical features
* loss of sensation over the lateral side of the foot
* sometimes pain

Strategy
* demonstrate lesion of n.suralis
* exclude proximal lesion, especially S1 radiculopathy unless the lesion is obvious

Expected abnormal findings
Neurography
* SCS: n.suralis

Expected normal findings
EMG
* m.gastrocnemius
* n.tibialis innervated intrinsic muscles of the foot

Neurography
* n.peroneus superficialis

Procedure
EMG
* m.gastrocnemius c. mediale
* optional : m.abductor digiti minimi, adductor hallucis

Neurography
* SCS: n.suralis, n.peroneus superficialis
* MCS: n.tibialis

References

Modified
* 6.4.1997 BF

SURAL NERVE LESION IN THE CALF

Etiology
* compression by ski boot or tight socks
surgery, varicose veins

Clinical features
- loss of sensation over the lateral side of the foot
- sometimes pain

Strategy
- demonstrate lesion of n.suralis
- exclude proximal lesion, especially S1 radiculopathy unless the lesion is obvious

Expected abnormal findings

Neurography
- SCS: n.suralis

Expected normal findings

EMG
- m.gastrocnemius
- n.tibialis innervated intrinsic muscles of the foot

Neurography
- n.peroneus superficialis

Procedure

EMG
- m.gastrocnemius c. mediale
- optional: m.abductor digiti minimi, adductor hallucis

Neurography
- SCS: n.suralis, n.peroneus superficialis
- MCS: n.tibialis

References

Modified
- 6.4.1997 BF

SURAL NERVE LESION AT THE MALLEOLUS

Etiology
- bimalleolar fracture
- compression by ski boot
- ankle sprain
- cysts
- posttraumatic fibrosis
- surgery around the ankle
- the sural nerve is often used for nerve biopsy and nerve grafting

Clinical features
- loss of sensation over the lateral side of the foot
- sometimes pain

Strategy
- demonstrate lesion of n.suralis
- exclude proximal lesion, especially S1 radiculopathy unless the lesion is obvious

Expected abnormal findings

Neurography
- SCS: n.suralis

Expected normal findings

EMG
- m.gastrocnemius
- n.tibialis innervated intrinsic muscles of the foot

Neurography
- n.peroneus superficialis

Procedure

EMG
- m.gastrocnemius c. mediale
- optional: m.abductor digiti minimi, adductor hallucis

Neurography
- SCS: n.suralis (bilaterally), n.peroneus superficialis
- MCS: n.tibialis

References

Modified
- 6.4.1997 BF

6. RADICULOPATHIES

General strategy
- demonstrate neurogenic EMG findings in muscles innervated by specific myotomes
- the EMG finding is definite if muscles innervated by the ventral ramus and dorsal ramus are affected. Therefore neurogenic abnormalities should be demonstrated in the limb muscles and paravertebral muscles.
if paravertebral muscles are normal, examine two leg muscles innervated by different nerves
examine muscles innervated by the adjacent roots, above and below, to determine whether it is an isolated or multiple radiculopathy

**C1 RADICULOPATHY**

**Etiology**
- extremely rare and probably difficult, if not impossible, to diagnose neurophysiologically
- herniated intervertebral disc
- spondylarthrosis
- very rarely neuroma, unless the patient has Mb von Recklinghausen

**Clinical features**
- acute, subacute or chronic neck pain

**Strategy**
- demonstrate neurogenic EMG findings in C1 innervated muscles
- examine muscles innervated by the adjacent roots

**Expected abnormal findings**
EMG, neurogenic findings in:
- paravertebral muscles C1

**Expected normal findings**
EMG
- paravertebral muscles from C3 downwards

**Procedure**
EMG
- m.rhomboideus major/m.deltoides/m.infraspinatus
- m.sternocleidomastoideus
- m.diahragma
- m.trapezius
- paravertebral muscles C1-C3

**Revised**
- BF 1996-11-07, 8.5.1997 BF

**C2 RADICULOPATHY**

**Etiology**
- extremely rare and probably difficult, if not impossible, to diagnose neurophysiologically
- herniated intervertebral disc
- spondylarthrosis
- very rarely neuroma, unless the patient has Mb von Recklinghausen

**Clinical features**
- acute, subacute or chronic neck pain

**Strategy**
- demonstrate neurogenic EMG findings in C2 innervated muscles
- examine muscles innervated by the adjacent roots

**Expected abnormal findings**
EMG, neurogenic findings in:
- paravertebral muscles C2

**Expected normal findings**
EMG
- m.rhomboideus major/m.deltoides/m.infraspinatus
- m.sternocleidomastoideus
- m.diahragma
- m.trapezius
- paravertebral muscles C4

**Procedure**
EMG
- m.rhomboideus major/m.deltoides/m.infraspinatus
- m.sternocleidomastoideus
- m.diahragma
- m.trapezius
- paravertebral muscles C1-C4

**Revised**
- 8.5.1997 BF, 7.11.1996 BF

**C3 RADICULOPATHY**

**Etiology**
- rare and probably difficult to diagnose neurophysiologically
- herniated intervertebral disc
- spondylarthrosis
- very rarely neuroma, unless the patient has Mb von Recklinghausen

**Clinical features**
- acute, subacute or chronic neck pain

**Strategy**
- demonstrate neurogenic EMG findings in C3 innervated muscles
- examine muscles innervated by the adjacent roots
Expected abnormal findings

EMG
- m.sternocleidomastoideus
- paravertebral muscles C3
- m.diaphragma

Expected normal findings

EMG
- m.rhomboideus major/m.deltoideus/m.infraspinatus
- m.brachioradialis

Procedure

EMG
- m.rhomboideus major/m.deltoideus/m.infraspinatus
- m.sternocleidomastoideus
- m.trapezius
- paravertebral muscles C2-C4
- m.diaphragma (optional, examine only if necessary)

Modified
* 8.5.1997 BF, 7.11.1997 BF

C4 radiculopathy

Etiology
- herniated intervertebral disc
- spondylarthrosis
- very rarely neuroma, unless the patient has Mb von Recklinghausen

Clinical features
- acute, subacute or chronic neck pain radiating into the shoulder

Strategy
- demonstrate neurogenic EMG findings in C4 innervated muscles
- examine muscles innervated by the adjacent roots

Expected abnormal findings

EMG
- m.trapezius
- m.sternocleidomastoideus
- m.diaphragma
- paravertebral muscles C4

Expected normal findings

EMG
- m.rhomboideus
- m.deltoideus

Procedure

EMG
- m.rhomboideus major/m.deltoideus/m.infraspinatus
- m.sternocleidomastoideus
- m.trapezius
- m.triceps
- paravertebral muscles C3-C5
- m.diaphragma (optional, examine only if necessary)

References
* Yiannikas C, Shahani BT, Young RR: Short-latency somatosensory-evoked potentials from radial, median, ulnar, and peroneal nerve stimulation in the assessment of cervical spondylosis: Comparison with conventional electromyography. Arch Neurol 1986;43:1264—1271

Modified
* BF 1996-11-07
C5 RADICULOPATHY

Etiology
- herniated intervertebral disc between C4/5
- spondylarthrosis
- very rarely neuroma, unless the patient has Mb von Recklinghausen

Clinical features
- 6% of cervical intervertebral disc herniations occur at this level
- acute, subacute or chronic neck pain radiating into the shoulder
- weakness of upper arm abduction and external rotation
- numbness over the lateral part of the upper arm (not often present)

Strategy
- demonstrate neurogenic EMG findings in C5 innervated muscles
- examine muscles innervated by the adjacent roots

Expected abnormal findings
EMG
- m.rhomboideus major
- m.deltoideus
- m.infraspinatus
- paravertebral muscles C5

Expected normal findings
EMG
- m.trapezius
- m.brachioradialis
- m.pronator teres
- m.triceps

Procedure
EMG
- m.rhomboideus major
- m.deltoideus/m.infraspinatus
- m.trapezius
- m.brachioradialis/m.pronator teres
- m.triceps
- paravertebral muscles C4-C6

References

Modified
- BF 1996-11-07

C6 RADICULOPATHY

Etiology
- herniated intervertebral disc at the C5/6 level
- spondylarthrosis
- very rarely neuroma, unless the patient has Mb von Recklinghausen

Clinical features
- acute, subacute or chronic neck pain radiating into the shoulder and arm
- weakness of elbow flexion
- decreased or absent biceps and brachioradialis tendon reflexes
- numbness over the radial side of the forearm and thumb

Strategy
- 25% of cervical intervertebral disc herniations occur at this level
- demonstrate neurogenic EMG findings in C6 innervated muscles
- demonstrate conduction abnormality in the proximal part of the C6 segment
- examine muscles innervated by the adjacent roots

References

Modified
- BF 1996-11-07
**Expected abnormal findings**

**EMG, neurogenic findings in:**
- paravertebral muscles C6
- m.biceps brachii
- m.brachioradialis
- m.pronator teres

**Expected normal findings**

**EMG**
- m.rhomboideus
- m.triceps

**Neurography**
- SCS n.medianus to digit I
- SCS n.radialis to digit I
- SCS n.cutaneus antebrachii lateralis

**Procedure**

**EMG**
- m.rhomboideus major
- m.deltoideus/m.infraspinatus
- m.brachioradialis/m.pronator teres
- m.triceps
- m.interosseus dorsalis
- paravertebral muscles C5-C7

**Neurography**
- SCS: n.radialis
- SCS n.medianus
- SCS: n.ulnaris
- CSC n.cutaneus antebrachii lateralis

**References**

- Yiannikas C, Shahani BT, Young RR: Short-latency somatosensory-evoked potentials from radial, median, ulnar, and peroneal nerve stimulation in the assessment of cervical spondylosis: Comparison with conventional electromyography. Arch Neurol 1986;43:1264—1271
- Yiannikas C, Shahani BT, Young RR: The investigation of traumatic lesions of the brachial plexus by electromyography and short-latency somatosensory potentials evoked by stimulation of multiple peripheral nerves. J Neurol Neurosurg Psychiatry 1983;46:1014—1022

**Modified**
- BF 1996-11-07 BF 1997-04-16

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**C7 radiculopathy**

**Etiology**
- herniated intervertebral disc at the C6/7 level
- spondylarthrosis
- very rarely neuroma, unless the patient has Mb von Recklinghausen

**Clinical features**
- 60 % of cervical intervertebral disc herniations occur at this level
- acute, subacute or chronic neck pain radiating into the shoulder and arm
- weakness of elbow extension and wrist flexion
- decreased or absent triceps tendon reflex
- numbness over the forefinger and middle finger

**Strategy**
- demonstrate neurogenic EMG findings in C7 innervated muscles
- demonstrate conduction abnormality in the proximal part of the C7 segment
- examine muscles innervated by the adjacent roots

**Expected abnormal findings**

**EMG, neurogenic findings in:**
- m.triceps brachii
- m.flexor carpi radialis
- m.latissimus dorsi/m.pectoralis major
- m.extensor digitorum communis
- paravertebral muscles C7
**Neurography**
* H-reflex to m.flexor carpi radialis

**Expected normal findings**

**EMG**
* m.biceps brachii
* m.abductor digit minimi/m.interosseus dorsalis I

**Neurography**
* SCS n.medianus dig II and III

**Procedure**

**EMG**
* m.biceps/m.brachioradialis
* m.flexor carpi radialis
* m.triceps
* m.interosseus dorsalis I
* m.abductor pollicis brevis
* paravertebral muscles C6-C8

**Neurography**
* SCS: n.radialis
* SCS n.medianus
* SCS: n.ulnaris
* MCS: n.medianus; H-reflex to m.flexor carpi radialis

**References**

**C8 radiculopathy**

**Etiology**
* herniated intervertebral disc
* spondylarthrosis
* very rarely neuroma, unless the patient has Mb von Recklinghausen

**Clinical features**
* acute, subacute or chronic neck pain radiating into the shoulder and arm
* weakness of the intrinsic hand muscles
* numbness of the index and little fingers
* may be accompanied by Horner’s syndrome

**Strategy**
* demonstrate neurogenic EMG findings in C8 innervated muscles
* demonstrate conduction abnormality in the proximal part of the C8 segment
* examine muscles innervated by the adjacent roots

**Expected abnormal findings**

**EMG, neurogenic findings in:**
* m.extensor indicis proprius
* m.interosseus dorsalis I/abductor digit minimi
* m.abductor pollicis brevis
* paravertebral muscles C8

**Expected normal findings**

**EMG**
* m.pronator teres
* m.triceps

**Neurography**
* SCS from dig V normal

**Procedure**

**EMG**
**TH1 RADICULOPATHY**

**Etiology**
- herniated intervertebral disc
- spondylarthrosis
- very rarely neuroma, unless the patient has Mb von Recklinghausen

**Clinical features**
- acute, subacute or chronic neck pain radiating into the shoulder arm
- weakness of thumb
- numbness over the ulnar side of the forearm

**Strategy**
- demonstrate neurogenic EMG findings in Th1 innervated muscles
- examine muscles innervated by the adjacent roots

**Expected abnormal findings**

**EMG, neurogenic findings in:**
- m.opponens pollicis/m.abductor pollicis brevis
- m.interosseus dorsalis
- paravertebral muscles Th1

**Expected normal findings**

**EMG**
- m.triceps/m.extensor digitorum communis

**Neurography**
- SCS normal in cutaneous antebrachii medialis

**Procedure**

**EMG**
- m.brachioradialis/m.pronator teres
- m.triceps
- m.interosseus dorsalis I
- m.extensor indicis
- m.opponens pollicis/m.abductor pollicis brevis
- paravertebral muscles C8-Th1

**Neurography**
- SCS: n.radialis
- SCS n.medians
- SCS: n.ulnaris

**References**

**TH2-Th10 RADICULOPATHY**

**Etiology**
- herniated intervertebral disc
- spondylarthrosis
- very rarely neuroma, unless the patient has Mb von Recklinghausen

**Clinical features**
- acute, subacute or chronic pain radiating unilaterally over the of thorax

**Strategy**
- demonstrate neurogenic EMG findings in paravertebral muscles and intercostal muscles at the respective level

**Expected abnormal findings**
- EMG, neurogenic findings in:
  - appropriate paravertebral muscles
  - appropriate intercostal muscles
  - at levels Th7-Th12 m.rectus abdominis

**Procedure**
- EMG
  - appropriate paravertebral muscles
  - appropriate intercostal muscles
  - m.rectus abdominis, the upper part is innervated from Th7 to Th9 and the lower part from Th9 to Th12

**Modified**
- BF 1996-11-07

**TH11 RADICULOPATHY**

**Etiology**
- herniated intervertebral disc
- spondylarthrosis
- very rarely neuroma, unless the patient has Mb von Recklinghausen

**Clinical features**
- pain radiating unilaterally over the abdomen

**Strategy**
- demonstrate neurogenic EMG findings in Th11 innervated muscles
- examine muscles innervated by the adjacent roots

**Expected abnormal findings**
- EMG, neurogenic findings in:
  - paravertebral muscles Th11
  - m.rectus abdominis pars inferior

**Procedure**
- EMG
  - paravertebral muscles Th10-12
  - m.rectus abdominis

**Modified**
- 8.5.1997 BF, 7.11.1996 BF

**TH12 RADICULOPATHY**

**Etiology**
- herniated intervertebral disc
- spondylarthrosis
- very rarely neuroma, unless the patient has Mb von Recklinghausen

**Strategy**
* demonstrate neurogenic EMG findings in Th11 innervated muscles
* examine muscles innervated by the adjacent roots

**Expected abnormal findings**

**EMG, neurogenic findings in**
* paravertebral muscles Th12
* m.rectus abdominis pars inferior

**Expected normal findings**

**EMG**
* m.iliopsoas

**Procedure**

**EMG**
* paravertebral muscles Th11-L1
* m.rectus abdominis pars inferior
* m.transversus abdominis
* m.iliopsoas

**References**

**Modified**
* BF 1996-11-07

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**L1 RADICULOPATHY**

**Etiology**
* herniated intervertebral disc
* spondylarthrosis
* very rarely neuroma, unless the patient has Mb von Recklinghausen

**Clinical features**
* acute, subacute or chronic pain radiating unilaterally over the abdomen

**Strategy**
* demonstrate neurogenic EMG findings in L1 innervated muscles
* examine muscles innervated by the adjacent roots

**Expected abnormal findings**

**EMG, neurogenic in**
* m.rectus abdominis pars inferior
* paravertebral muscles L1

**Expected normal findings**

**EMG**
* m.adductor magnus

**Procedure**

**EMG**
* paravertebral muscles Th12-L2
* m.rectus abdominis
* m.iliopsoas
* m.adductor magnus

**References**

**Modified**
* BF 1996-11-07

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**L2 RADICULOPATHY**

**Etiology**
* herniated intervertebral disc
* spondylarthrosis
* very rarely neuroma, unless the patient has Mb von Recklinghausen

**Clinical features**
* acute, subacute or chronic pain radiating unilaterally into the thigh
* weakness of the thigh flexion
* numbness over the anterior part of the thigh

**Strategy**
* demonstrate neurogenic EMG findings in L2 innervated muscles
* examine muscles innervated by the adjacent roots

**Expected abnormal findings**

**EMG, neurogenic in**
* m.iliopsoas
* m.adductor magnus
* m.vastus lateralis/m.vastus medialis
* paravertebral muscles L2

**Expected normal findings**

**EMG**
* m.tibialis anterior

**Neurography**
* saphenus SCS normal

**Procedure**

**EMG**
* paravertebral muscles L1-L3
* m.iliopsoas
L3 RADICULOPATHY

**Etiology**
- herniated intervertebral disc
- spondylarthrosis
- spinal anesthesia
- very rarely neuroma, unless the patient has Mb von Recklinghausen

**Clinical features**
- acute, subacute or chronic pain radiating unilaterally into the thigh
- weakness of the thigh flexion and knee extension
- numbness over the anterior part of the thigh and knee

**Strategy**
- demonstrate neurogenic EMG findings in L3 innervated muscles
- examine muscles innervated by the adjacent roots

**Expected abnormal findings**
- EMG neurogenic in:
  - m.adductor magnus
  - m.iliopsoas
  - m.vastus lateralis/m.vastus medialis
  - paravertebral muscles L3

**Expected normal findings**
- EMG
  - m.tibialis anterior

**Neurography**
- saphenus SCS normal

**Procedure**
- EMG
  - paravertebral muscles L2-L4
  - m.iliopsoas
  - m.adductor magnus
  - m.vastus lateralis/m.vastus medialis
  - m.tibialis anterior

**References**
- Revised
  - 8.5.1997 BF

L4 RADICULOPATHY

**Etiology**
- herniated intervertebral disc
- spondylarthrosis
- spinal anesthesia
- very rarely neuroma, unless the patient has Mb von Recklinghausen

**Clinical features**
- acute, subacute or chronic back pain radiating into the leg
- numbness of the anterior part of the leg
- loss of patellar reflex
- weakness of knee extension

**Strategy**
- demonstrate neurogenic EMG findings in L4 innervated muscles
- examine muscles innervated by the adjacent roots

**Expected abnormal findings**
- EMG neurogenic in:
  - m.vastus lateralis/m.vastus medialis
  - m.tibialis anterior
  - paravertebral muscles L4

**Expected normal findings**
- EMG
  - m.peroneus longus/m.extensor hallucis longus

**Neurography**
- SCS n. saphenus

**Procedure**
- EMG
paravertebral muscles L3-L5
m. iliopsoas
m. adductor magnus
m. vastus lateralis/m. vastus medialis
m. tibialis anterior
m. peroneus longus/m. extensor hallucis longus
m. gastrocnemius caput medialis/m. gastrocnemius caput laterale

Neurography
- SCS: n. saphenus
- MCS: n. peroneus

References
- Khati BO, Baruah J, McQuillen MP. Correlation of electromyography with computed tomography in evaluation of lower back pain. Arch Neurol 1984;41:594-597
- Young WB. The clinical diagnosis of lumbar radiculopathy. Semin Ultrasound CT MR 1993;14:385-388

Revised

L5 RADICULOPATHY

Etiology
- herniated intervertebral disc
- spondylarthrosis
- spinal anesthesia
- very rarely neuroma, unless the patient has Mb von Recklinghausen

Clinical features
- acute, subacute or chronic back pain radiating into the leg
- numbness over the dorsal side of the foot
- decreased toe and ankle dorsal flexion

Strategy
- demonstrate neurogenic EMG findings in L5 innervated muscles
- demonstrate conduction abnormality in the proximal part of the L5 segment
- examine muscles innervated by the adjacent roots

Expected abnormal findings
EMG, neurogenic
- paravertebral muscles L3-L5
- m. peroneus longus/Extensor hallucis longus/tibialis anterior/flexor hallucis longus/ tibialis posterior
- tensor fascia latae/semitendinosus

Neurography
- M-wave amplitude may be low in m. extensor digitorum brevis and other muscles innervated by n. peroneus
- F-waves in peroneal nerve may be abnormal

Expected normal findings
EMG
**S1 RADICULOPATHY**

**Etiology**
- herniated intervertebral disc
- spondylarthrosis
- spinal anesthesia
- very rarely neurona, unless the patient has Mb von Recklinghausen

**Clinical features**
- usually acute or subacute lumbosacral pain radiating in the leg
- numbness on the lateral side of the foot
- Achilles’s reflex reduced or absent

**Strategy**
- demonstrate neurogenic EMG findings in S1 innervated muscles
- demonstrate conduction abnormality in the proximal part of the S1 segment
- examine muscles innervated by the adjacent roots

**Expected abnormal findings**

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**Procedure**

**EMG**
- paravertebral muscles L4-S1
- m.vastus lateralis/m.vastus medialis (L4)
- m.tibialis anterior/m.peroneus longus/m.extensor hallucis longus (distal L5)
- m.tensor fascia latae/m.gluteus medius/m.semitendinosus (proximal L5)
- m.gluteus maximus/m.biceps femoris (proximal S1)
- m.gluteus maximus caput medialis/m.gastrocnemius caput laterale (distal S1)

**Neurography**
- SCS: n.suralis
- MCS: n.peroneus
- MCS: n.tibialis
- MCS: n.tibialis H-reflex to m.soleus

**References**

- Khatri BO, Baruah J, McQuillen MP. Correlation of electromyography with computed tomography in evaluation of lower back pain. Arch Neurol 1984;41:594-597
- Mixter VJ, Barr JS. Rupture of the intervertebral disk with involvement of the spinal canal. N Eng J Med 1934;211:210-218
- Young WB. The clinical diagnosis of lumbar radiculopathy. Semin Ultrasound CT MR 1993;14:385-388
EMG, neurogenic findings in
- paravertebral muscles S1
- m.gluteus maximus/m.biops femoris caput brevis
- m.gastrocnemius caput mediale/m.gastrocnemius caput laterale/m.soleus

Neurography
- H-reflex abnormal
- F-waves to calf muscles abnormal

Expected normal findings
EMG
- peroneus longus/flexor hallucis longus/extensor hallucis longus
- tensor fascia latae

Neurography
- n.suralis

Procedure
EMG
- paravertebral muscles L5-S1
- m.vastus lateralis/m.vastus medialis (L4)
- m.tibialis anterior/m.peroneus longus/m.extensor hallucis longus (distal L5)
- m.tensor fascia latae/m.gluteus medius/m.semitendinosus (proximal L5)
- m.gluteus maximus/m.biceps femoris (proximal S1)
- m.gastrocnemius caput mediale/m.gastrocnemius caput laterale (distal S1)

Neurography
- SCS: n.suralis
- MCS: n.peroneus
- MCS: n.tibialis
- MCS: n.tibialis H-reflex to m.soleus

References
- Khatri BO, Baruah J, McQuillen MP. Correlation of electromyography with computed tomography in evaluation of lower back pain. Arch Neurol 1984;41:594-597
- Mixter VJ, Barr JS.- Rupture of the intervertebral disk with involvement of the spinal canal. N Eng J Med 1934;211:210-218
- Young WB. The clinical diagnosis of lumbar radiculopathy. Semin Ultrasound CT MR 1993;14:385-388

Revised

LOW BACK PAIN WITH NO RADICULOPATHY

Strategy
- normal findings in clinically suspected roots.
- usually it is sufficient to examine L4, L5 and S1 roots.
- if there are clinically symptoms or signs of L1-L3 involvement; check also those.

Expected normal findings
EMG in one muscle from each of the following five groups and the corresponding paravertebral muscles
- L4: m.vastus lateralis/m.vastus medialis
- L5, proximal muscles. m.tensor fascia latae/m.gluteus medius/m.semitendinosus/m.semimembranosus
L5, distal muscles: m.peroneus longus/m.extensor hallucis longus/m.tibialis anterior or m.flexor hallucis longus
S1, proximal muscles: m.gluteus maximus/m.biceps femoris caput brevis
S1, distal muscles: m.gastrocnemius caput laterale/m.gastrocnemius caput medialis/m.soleus

Neurography
H-wave from soleus
n.tibialis F-wave
n.peroneus F-wave

Procedure
EMG
paravertebral muscles
m.vastus lateralis/m.vastus medialis (L4)
m.tibialis anterior/m.peroneus longus/m.extensor hallucis longus (distal L5)
m.tensor fascia latae/m.gluteus medius/m.semitendinosus (proximal L5)
m.gluteus maximus/m.biceps femoris (proximal S1)
m.gastrocnemius caput medialis/m.gastrocnemius caput laterale (distal S1)

Neurography
SCS: n.saphenus
SCS: n.peroneus superficialis
SCS: n.suralis
MCS: n.peroneus
MCS: n.tibialis
MCS: n.tibialis H-reflex to m.soleus

NECK PAIN WITH NO RADICULOPATHY

Strategy
* normal findings in clinically suspected roots. Usually it is sufficient to examine C5, C6, C7, C8 and Th1 roots.
* if there are signs of involvement in neighboring areas check also those.

Expected normal findings
EMG in one muscle from each of the following five groups and the corresponding paravertebral muscles
* m.rhomboideus/m.deltoideus
* m.biceps brachii/m.brachioradialis
* m.triceps brachii/m.flexor carpi radialis
* m.extensor indicis proprius/m.interosseus dorsalis I
* m.abductor pollicis brevis/m.interosseus dorsalis I
* paravertebral muscles C5-Th1

Neurography
H-wave from m.flexor carpi radialis
n.medianus and n.ulnaris F-waves
SCS: n.suralis and n.medianus

Procedure
EMG
M.deltoideus/m.infraspinatus (C5)
m.biceps/m.brachioradialis/m.pronator teres (C6)
m.triceps /m.flexor carpi radialis (C7)
m.interosseus dorsalis (C8)
m.opponens pollicis/m.abductor pollicis brevis (Th1)
* paravertebral muscles

Neurography
SCS: n.radialis
SCS n.medianus
SCS: n.ulnaris
MCS: n.medianus: H-reflex to m.flexor carpi radialis

7. DISORDERS OF THE SPINAL CHORD AND CENTRAL NERVOUS SYSTEM WITH NEUROMUSCULAR ABNORMALITIES

CERVICAL SYRINGOMYELIA

Etiology
* syringomyelia is a chronic disorder involving the spinal cord or medulla or both.
* it is characterized by the development of cavitation and gliosis within the spinal chord or medulla
* intraspinal dilatation of cavity is due to developmental, vascular or traumatic causes

Clinical features
* onset usually between 20 and 40 years, but may be seen both in younger and older age groups
* dissociated sensory loss; loss of pain and temperature, but preservation of touch
* muscle weakness and atrophy
* impairment of long tract functions
* syringomyelia is considered slowly progressive and degenerative, but there are indications that progression may be arrested by appropriate treatment.

Strategy
* demonstrate neurogenic abnormalities in muscles innervated from affected myotomes
* exclude peripheral nerve lesions (ulnar nerve lesions, lower part of plexus and C8 radiculopathy)
* differentiate from monomelic spina muscular atrophy, motor neuropathy with multifocal conduction block and ALS
**Expected abnormal findings**

**EMG**
- m.interosseus dorsalis/m.abductor digiti minimi
- m.flexor carpi ulnaris
- m.extensor indicis/m.extensor digitorum communis

**Neurography**
- MCS, AMPL may be reduced, in the presence of low AMPL CV may be reduced

**Expected normal findings**

**Neurography**
- SCS

**Procedure**

**Neurography (bilaterally)**
- MCS n.medianus, n.ulnaris (fractionated across the elbow)
- SCS n.ulnaris, n.medianus, n.radialis, n.cutaneus antebrachii lateralis, n.cutaneus antebrachii medialis

**EMG (bilaterally)**
- m.interosseus dorsalis I (n.ulnaris)
- m.opponens pollicis/m.abductor pollicis brevis (n.medianus)
- m.extensor indicis proprius (C8 and inferior trunk)
- m.triceps brachii
- m.biceps brachii/m.deltoidus
- m.trapezius (to evaluate brainstem involvement, syringobulbia)
- m.hyoglossus/m.hyoglossus (to evaluate brainstem involvement, syringobulbia)

**Evoked potentials**
- MEP and SEP may be helpful in the evaluation of long tract involvement

**References**

* Revised

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**FRIEDREICH’S ATAXIA**

**Etiology**
- autosomal recessive inheritance
- gene location chromosome 9cen-q21
- the mutation consists of an unstable expansion of GAA repeats in the first intron of the frataxin gene on chromosome 9, which encodes a protein of unknown function

**Clinical features**
- prevalence 1-2 per 100 000
- onset of symptoms usually between 8-15 (range 2-50 years)
- staggering gait often first symptom, sometimes clumsiness
- ataxia
- loss of tendon reflexes, sometimes preserved quite late
- dysarthria
- scoliosis is common
- high foot arches
- eventually the patients become bedridden
- decreased longevity

**Strategy**
- demonstrate abnormal SCS amplitude, CV is normal or only slightly reduced
- EMG and MCS normal
- differentiate from polyneuropathy

**Expected abnormal findings**

**Neurography**
- SCS: reduced SCS amplitude

**Expected normal findings**

**EMG**
- MCS

**Procedure**

**EMG**
- m.deltoides/m.biceps brachii
- m.interosseus dorsalis I/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor fascia latae
- m.tibialis anterior

**Neurography**
- SCS: n.suralis and n.radialis, n.medianus and n.ulnaris bilaterally
- MCS: n.peroneus and n.medianus on one side

**References**


**Modified**
- October 21st, 1996, BF

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**MACHADO-JOSEPH DISEASE,**

**Etiology**
autosomal dominant inheritance

gene maps to long arm of chromosome 14q24.3-32.1

the mutation consists of an unstable expansion of CAG repeats in the 3’ end of the coding region

the variability of the clinical severity depends on the number of trinucleotide repeats

Clinical features

Four subtypes have been suggested:

I. The age of onset is in the mid-20s. It is noted for its relative lack of cerebellar findings. This syndrome is characterized by the pyramidal and extrapyramidal signs of spasticity, hyperreflexia, Babinski's sign, clonus, rigidity, dystonia. There is also the onset of facial and lingual fasciculations, nystagmus, and PEO.

II. Onset in the fourth through sixth decades. The second subtype includes primarily cerebellar and extrapyramidal features. There is truncal ataxia in association with a lurching gait. There is progression to dysarthria and the appearance of dystonia and occasionally chorea.

III. The onset here is in the fifth through seventh decades. The third subtype is characterized by the cerebellar signs in association with a distal polyneuropathy. There is distal sensory loss and amyotrophy with hyporeflexia.

IV. The fourth subtype is dominated by neuropathy and parkinsonian-like features.

The diagnosis is made by the positive family history, the cerebellar syndrome, and the pyramidal and extrapyramidal findings. These in combination with bulging eyes, lid retraction, and the PEO are unique to this syndrome.

Strategy

demonstrate abnormal findings in sensory nerves

muscles show neuropathic changes in many patients, especially in the later stages

CNS also affected

Expected abnormal findings

Neurography

* SCS: reduced or absent SCS responses

* neuropathic EMG abnormalities with fasciculations

* abnormal VEP, SEP and BAEP

Expected normal findings

EMG

Neurography

Procedure

EMG

* m.deltoideus/m.biceps brachii

* m.interosseus dorsalis /m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis

* m.vastus lateralis/m.vastus medialis/m.tensor fascia latae

* m.tibialis anterior

Neurography

* SCS: n.suralis and n.radialis, n.medianus and n.ulnaris bilaterally

* MCS: n.peroneus and n.medianus on one side

Evoked potentials

* BAEP

* SEP

* VEP

References


Modified

October 21st 1996, BF

METACHROMATIC LEUKODYSTROPHY

Etiology

deficiency of arylsuphatase a

autosomal recessive inheritance

linked to chromosome 22q-22ter13

the metachromatic leukodystrophies are a group of genetically determined disorders affecting the nervous system

CNS and PNS is affected

accumulation of galactosyl sulfatide and other sulfatide lipids

Clinical features

late infantile form:

* onset between 1-4 years

* initial symptom walking difficulty

* mental retardation

* later ataxia an tetraplegia

* blindness and deafness

* Juvenile and adult onset form

* similar to infantile form but later onset

Strategy

* in late infantile form demonstrate demyelinating sensory motor polyneuropathy

* in juvenile adult form the demyelinating polyneuropathy is not as prominent

Expected abnormal findings

Neurography

* in the late infantile form reduced CV, often in the range of 10-20 m/s, SCS absent sensory responses reduced in amplitude or absent

* in the juvenile adult form the CV is slightly reduced

EMG
GALACTOSYLCERAMIDE LIPIDOSIS (KRABBE DISEASE, GLOBOID CELL LEUKODYSTROPHY)

Etiology
* the gene has been mapped to chromosome 14q24-q32
* galactocerebroside β-galactosidase deficiency
* autosomal recessive inheritance

Clinical features
* normal neonatal development
* onset at the age of 3 to 6 months (cases with later onset and slower progression have been described)
* mental retardation
* hypertonicity of limbs
* aggravated reflexes
* children become blind and deaf
* CSF protein elevated
* death by 2 years of age
* prenatal diagnosis is available

Strategy
* demonstrate severe demyelinating polyneuropathy

Expected abnormal findings

Neurography
* SCS: CV reduced by 50 %, reduced SCS amplitude
* MCS: CV reduced by 50 %

EMG
* Neurogenic abnormalities

Procedure

EMG
* m.deltoideus/m.biceps brachii
* m.interosseus dorsalis l/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpii radialis
* m.vastus lateralis/m.vastus medialis/m.tensor fascia latae
* m.tibialis anterior

Neurography
* SCS: n.suralis and n.radialis bilaterally
* MCS: n.peroneus and n.medianus bilaterally

References

Modified
* 25.3.1997 BF.

ADRENOLEUKODYSTROPHY/ADRENOMYELONEUROPATHY

Etiology
* X-linked recessive inheritance
* deficient δ-oxygenation of very long chain fatty acids
* accumulation of very long chain (25-26 carbon chain fatty acids)

Clinical features
* onset of symptoms varies markedly, may begin in childhood or adulthood
* progressive cerebral degeneration
* cerebellar ataxia, pyramidal tract symptoms
* cortical blindness, deafness, spasticity, seizures
* peripheral neuropathy
* sometimes adrenal failure

Strategy

References

Modified
* 26.3.1997 BF
**Expected abnormal findings**
*EMG*
- mild neurogenic abnormalities
*Neurography*
- SCS: reduced CV and amplitude

**Procedure**
*EMG*
- m.deltoideus/m.biceps brachii
- m.interosseus dorsalis I/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor fascia latae
- m.tibialis anterior
*Neurography*
- SCS: n.suralis and n.radialis bilaterally
- MCS: n.peroneus and n.medianus bilaterally

**References**

**Modified**
- 25.3.1997 BF

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**8. MISCELLANEOUS**

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**BENIGN FASCICULATION**

**Etiology**
- unknown
- sporadic

**Clinical features**
- fasciculations in muscles
- onset may be at any age
- most persons seeking medical advice for this disorder are medical personnel, especially medical students

**Strategy**
- demonstrate fasciculations in muscles but no signs of denervation or reinnervation
- differentiate from ALS, MNM, SMA and other neurogenic disorders

**Expected abnormal findings**
*EMG*
- fasciculations

**Expected normal findings**
*EMG*
- no fibrillations or positive sharp waves
- MUP analysis
*Neurography*
- MCS
- SCS

**Procedure**
*EMG*
- m.deltoideus/m.biceps brachii
- m.interosseus dorsalis I/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpi radialis
- m.vastus lateralis/m.vastus medialis/m.tensor fascia latae
- m.tibialis anterior
*Neurography*
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus and n.medianus on one side

**References**

**Modified**
- 25.3.1997 BF

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**NEUROMYOTONIA (ISAAC’S SYNDROME, CONTINOUS MUSCLE FIBER ACTIVITY)**

**Etiology**
- probably autoimmune disorder
- antibodies against voltage gated potassium channels
- often occurs sporadically without an apparent cause
- may be caused by penicillamine
- sometimes a paraneoplastic feature, especially related with lung cancer or thymoma, plasmacytoma and IgM paraproteinememia

**Clinical features**
- muscle stiffness and rippling of the muscles
usually responds well to phenytoin or carbamazepine, plasmapheresis is also effective

**Strategy**
- demonstrate neuromyotonic activity
- differentiate from myotonia

**Expected abnormal findings**

**EMG**
- abundant neuromyotonia, especially in distal muscles

**Neurography**
- MCS may sometimes be abnormal, reduced CV and amplitude

**Expected normal findings**

**EMG**
- MUP analysis normal

**Neurography**
- MCS, sometimes abnormal
- SCS

**Procedure**

**EMG**
- m.deltoides/m.biceps brachii
- m.interosseus dorsalis l/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpii radialis
- m.vastus lateralis/m.vastus medialis/m.tensor fascia latae
- m.tibialis anterior

**Neurography**
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus, n.tibialis, n.ulnaris and n.medianus on one side

**References**

**STIFF MAN SYNDROME**

**Etiology**
- probably autoimmune disorder
- antibodies against glutamic acid decarboxylase (GAD)

**Clinical features**
- onset age 8-76, average around 40 years
- men and women equally affected
- initially tightness and aching of axial muscles
- progression to symmetric stiffness of trunk and limb muscles
- sudden stimuli, auditory and sensory but not visual, painful muscle spasms (spasmodic myoclonia)
- hyperlordosis and elevation of shoulders
- muscles stiffness disappears during sleep and with benzodiazepine treatment
- tendons reflexes normal or increased
- often associated with type 1 diabetes (30-50% of the patients) and other immune mediated disorders (myasthenia gravis, vitiligo, pernicious anemia, thyroiditis, etc.)
- antibodies against GAD

**Strategy**
- demonstrate abundant ongoing EMG activity in axial muscles and limb muscles
- differentiate from myotonia

**Expected abnormal findings**

**EMG**
- abundant EMG activity, especially in axial muscles

**Expected normal findings**

**EMG**
- normal insertional activity, if it can be evaluated
- MUP analysis normal

**Neurography**
- MCS
- SCS

**Procedure**

**EMG**
- m.deltoides/m.biceps brachii
- m.interosseus dorsalis l/m.abductor pollicis brevis/m.extensor digitorum communis/m.flexor carpii radialis
- m.vastus lateralis/m.vastus medialis/m.tensor fascia latae
- m.tibialis anterior

**Neurography**
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus, n.tibialis, n.ulnaris and n.medianus on one side

**Effect of benzodiazepines**
monitor the effect of IV injection of diazepam with EMG, in stiff man syndrome the EMG activity should be considerably reduced following the injection

**Note**
- abnormal long loop reflexes have been described in stiff man syndrome
- abnormal flexor reflexes have been described in stiff man syndrome

**References**

**Modified**
- 6.11.1996 BF, 26.3.1997 BF

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**TETANUS**

**Etiology**
- toxin produced by Clostridium tetani under anaerobic conditions
- toxin transported after endocytosis by retrograde transport to the spinal chord
- inhibits release of glycine and GABA
- both α and γ motoneurones are disinhibited
- in high concentrations the effects of tetanus toxin are similar to botulinus toxin

**Clinical features**
- occurs very rarely because of effective immunization
- may occur in patients with proper immunization
- presents 3 to 20 days following exposure to bacteria
- localized type: painful spasms near injury, limited spasms last for two weeks
- generalized type is more common: trismus, opisthotonus, reflex spasms, stiffness, neck rigidity, dysphagia: tetanic spasms
- autonomic over activity: hypertension, dysrythmias
- in long standing cases there is denervation of muscles
- diagnosis is clinical there are no specific tests

**Strategy**
- demonstrate abundant EMG activity in axial muscles and limb muscles
- differentiate from stiff man syndrome and epilepsy

**Expected abnormal findings**
- abundant EMG activity, especially in axial muscles
- after one week signs of acute denervation on EMG

**Neurography**
- initially normal findings
- after one week reduced MCS amplitude

**Expected normal findings**

**Neurography**
- SCS

**Procedure**

**EMG**
- m.deltoideus/m.biceps brachii
- m.interosseus dorsalis l/m.abductor digit minimi/m.abductor pollicis
- m.vastus lateralis/m.vastus medialis
- m.tibialis anterior

**Neurography**
- SCS: n.suralis and n.radialis on one side
- MCS: n.peroneus, n.tibialis, n.ulnaris and n.medianus on one side

**References**
- Kaeiser HE, Muller Müller HR, Friedrich B: The nature of tetraplegia in infectious tetanus. Eur Neurol 1968;1:17
- Crone NE, Reder AT. Severe tetanus in immunized patients with high anti-tetanus titer. Neurology. 1992;42:761-764

**Modified**
- 16.4.1997 BF