Institute of Genomic Medicine and Rare Disorders
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Immun mediated neuropathies

Next supervion expected: 2017.06.01.

Definition and background

Immun mediated neuropathies represent a broad spectra of periferial nerve lesions that has common entities like good response to immun-suppressive agents and mostly demyelinating type of nerve lesions. They can be classified depending onset, time course, distribution, type of nerve fibers involved, etc. One of the most important aspect is the onset. Onset is acut in Guillaine-Barré syndrome (GBS) and subacut or chronic in CIDP. In GBS after a 4-6 weeks progression steady state and gradual improvement can be observed. While in CIDP the onset in most of the cases subacut and the time course is slowly progressive over mounths. Both (GBS and CIDP) have different subtype regarding the nerve fibers predominantly involved and the distribution of nerve lesions throughout the body (Table 1.). Different subtypes have different prognostic perspectives and in certain cases different treatment approach. In all immun mediated polyneuropathy the diagnose lies upon the triad of clinical presentation, electrophysiological studies and CSF analysis. Other causes of etiology have to be excluded.

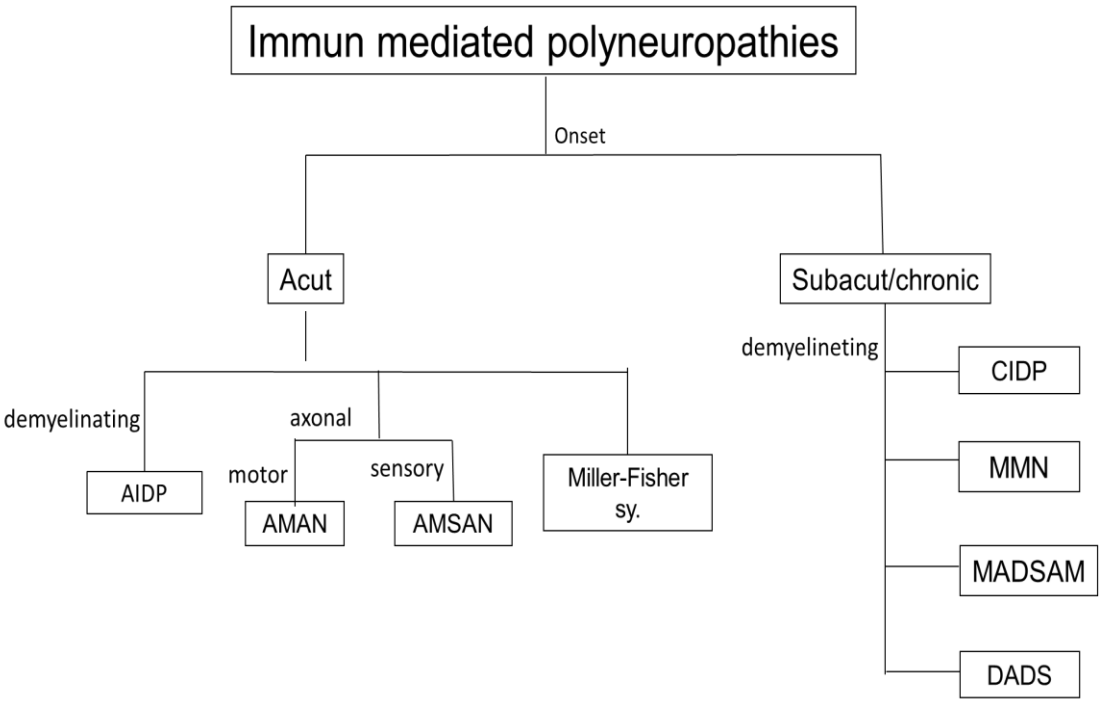

Table 1. Main subtypes of acut and chronic immun mediated polyneuropathies

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Acut immun mediated neuropathies

Guillain-Barré syndrome is an acut, immun mediated, inflammatory multifocal polyneuropathy. Typically it manifests with an ascending pattern beginning in the lower extremities. Main symptomes are muscle weakness, sensory deficits due to large myelinated fibers lesions, pain common but autonomic disturbances are minor and loss of deep tendon reflexes. Miller-Fisher subtype characterized by ophtalmoplegia, ataxia and loss of deep tendon reflexes. Electrophysiological studies reveal demyelinating (or in certain subgroups axonal) lesions. In the cerebrospinal fluid there is cytoalbuminaer dissociation (elevated protein level without cells).

Diagnosis of Guillain-Barré Syndrome

DIAGNOSIS OF GBS :

1. Subacutely developing flaccid paralysis, with a nadir emerging after 2-4 weeks
2. Weakness from the onset involving both sides of the body with a strong tendency for symmetry
3. The deep tendon reflexes hypoactive or absent
4. Other causes for rapidly developing flaccid weakness are highly unlikely by history/additional tests

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SUPPORTED BY:

1. Albuminocytologic disassociation of the CSF
2. Electrophysiology supportive of a polyneuropathy

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GBS VARIANTS :

1. AIDP
2. AMAN/AMSAN (Axonal forms)
3. Miller Fisher Variant (Opthalmoplegia, Ataxia, Areflexia, Anti-GQ1b antibodies)
4. Pharyngocervicobrachial Subgroup
5. Pure Sensory/ Autonomic GBS

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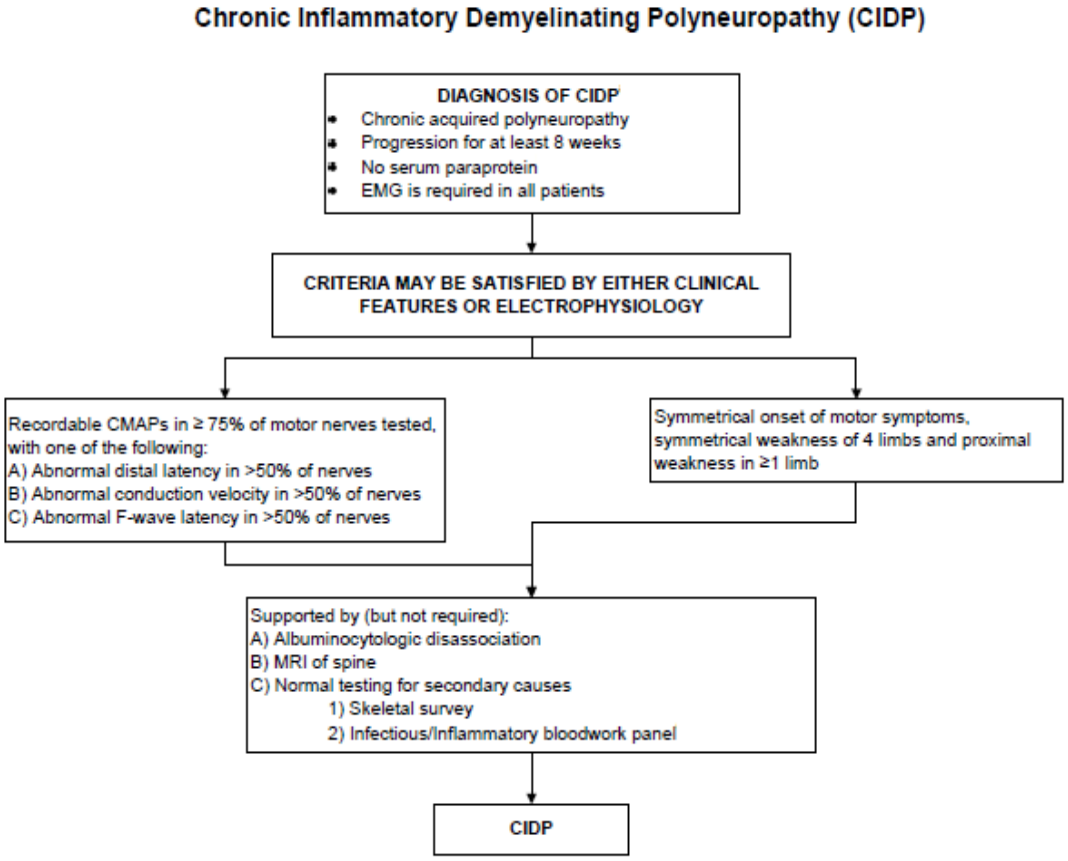
Chronic immun mediated neuropathies

Chronic inflammatory demyelinating polyneuropathy (CIDP) is a chronic, acquired, immune-mediated condition affecting the peripheral nervous system. The classic form of the disorder is characterized by: (1) progressive limb weakness, usually with a predilection for proximal muscles, sensory loss, and areflexia with a relapsing or progressive course; (2) electrophysiological features of demyelination, including prolonged distal motor and F-wave latencies, reduced conduction velocities, and conduction block and temporal dispersion; (3) laboratory features of albumino-cytological dissociation in the cerebrospinal fluid.



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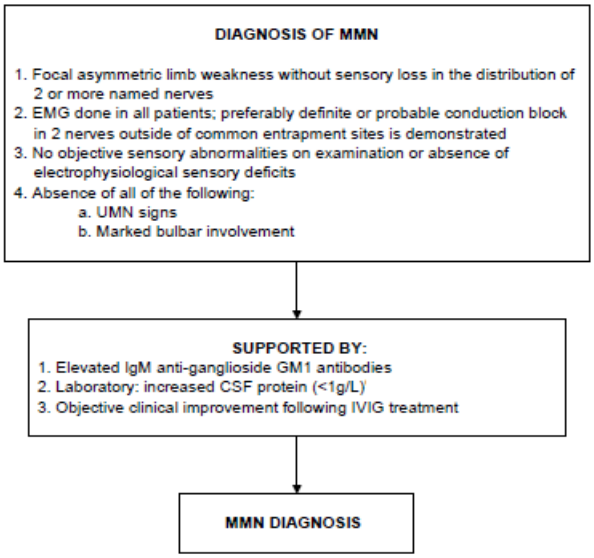
Mutiplex motor neuropathy (MMN) is a subroup of CIDP and it is of particular importance because of only reacting to IVIG treatment. In MMN there is an asymmetrical involvment of only motor nerve fibers.

Diagnosis and Treatment of Multifocal Motor Neuropathy (MMN)



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