CNS Demyelination (GENERAL)

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1. **Demyelinating (s. myelinoclastic) diseases** - destruction of normal myelin - many ***acquired*** neurologic disorders.
2. **Dysmyelinating diseases** - inadequate myelin formation or maintenance - many ***congenital*** metabolic disorders.

N.B. **CNS myelin** (formed by oligodendroglia) differs chemically and immunologically from **PNS** **myelin** (formed by Schwann cells), but both types have same function - to promote transmission of neural impulse along axon.

**Idiopathic** (presumably **autoimmune**)

**Recurrent / chronically progressive demyelination** – most common CNS demyelination disorders:

1. Multiple sclerosis
2. Multiple sclerosis variants:
   1. neuromyelitis optica (s. Devic disease)
   2. concentric sclerosis (s. Baló disease, encephalitis periaxialis concentrica)
   3. Marburg variant of MS - clinically fulminant MS form
   4. Schilder disease (s. encephalitis periaxialis diffusa, diffuse sclerosis)

**Monophasic demyelination** (may be first clinical episode of multiple sclerosis!):

1. Optic neuritis
2. Acute transverse myelitis

**CNS complications of viral infections / vaccinations**:

1. Acute disseminated encephalomyelitis (ADEM)
2. Acute necrotizing hemorrhagic encephalomyelitis (ANHEM)

**Leukodystrophies** - inherited disorders that affect myelin synthesis / turnover:

**Primarily affecting CNS myelin**:

1. Adrenoleukodystrophy
2. Pelizaeus-Merzbacher disease
3. Spongy degeneration (s. Canavan's disease)
4. Alexander's disease

**CNS-PNS myelin**:

1. Metachromatic leukodystrophy
2. Globoid cell leukodystrophy (s. Krabbe's disease)
3. Cockayne’s syndrome

**Viral infections**

1. Progressive multifocal leukoencephalopathy (JC virus infection of oligodendrocytes)
2. Subacute sclerosing panencephalitis (measles virus infection of neurons and glia)
3. Human T-cell lymphotropic virus type I -associated myelopathy.

**Nutritional disorders**

1. Combined systems disease (s. vit. B12 deficiency)
2. Demyelination of corpus callosum (s. Marchiafava-Bignami disease)
3. Central pontine myelinolysis

**Anoxic-ischemic sequelae**

1. Delayed postanoxic cerebral demyelination
2. Progressive subcortical ischemic encephalopathy

Common features of CNS demyelination disorders

* 1. affect *young adults*
  2. **inflammation + selective destruction of CNS myelin** (with relative preservation of axons and PNS)
  3. clinical deficits are due to:
     1. ***effect of myelin loss*** on transmission of electrical impulses.
     2. limited capacity of CNS to regenerate normal myelin.
     3. secondary damage to axons.
  4. *no specific tests*; diagnosis is based on distinctive clinical patterns of CNS injury.

Demyelination may have either negative or positive effects:

***Negative conduction abnormalities*** - **slowed axonal conduction**, variable **conduction block** (in response to raised temperature or with metabolic changes in extracellular milieu) → fluctuations in function that vary from day to day, worsenings with body temperature elevation.

***Positive conduction abnormalities*** - **ectopic impulse generation** (spontaneously or following mechanical stress), **abnormal "crosstalk"** between demyelinated axons → Lhermitte's symptom, paroxysmal symptoms, paresthesia.

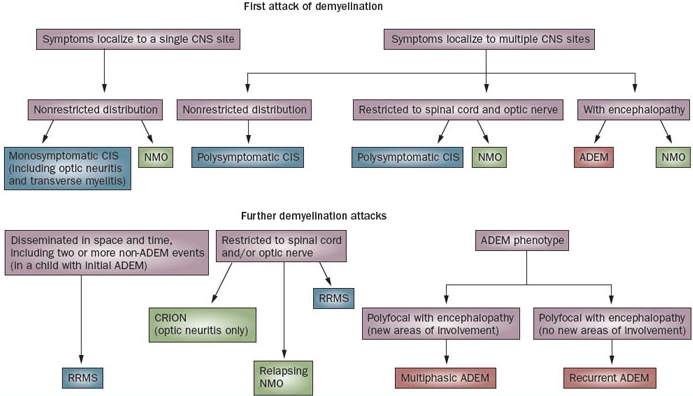
Incidental white-matter hyperintensities

- focal white-matter hyperintensities, often multiple.

* in deep parietal white matter.
* seen in up to 24% of men.
* *no clinical significance* - no associations with neurological abnormalities, CD4 count, alcohol or drug use, hypertension or smoking.

Diagnostic algorithm of pediatric onset demyelinating disorders

Abbreviations: ADEM = acute disseminating encephalomyelitis; CIS = clinically isolated syndrome; CRION = chronic relapsing inflammatory optic neuropathy; NMO = neuromyelitis optica; RRMS = relapsing-remitting MS.



Bibliography for ch. “Demyelinating Disorders” → follow this [link >>](http://www.neurosurgeryresident.net/Dem.%20Demyelinating%20disorders\Dem.%20Bibliography.pdf)

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