

# CNS Demyelination (GENERAL)

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- A. **DEMYELINATING (s. MYELINOCLASTIC) diseases** - destruction of normal myelin - many *acquired* neurologic disorders.
- B. **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. B<sub>12</sub> 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:
  - a) **effect of myelin loss** on transmission of electrical impulses.
  - b) limited capacity of CNS to regenerate normal myelin.
  - c) 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

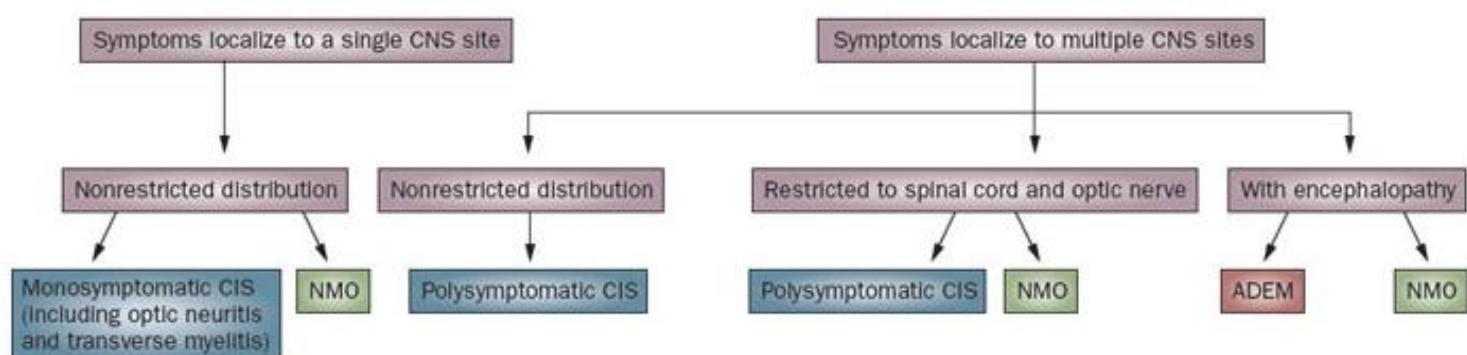
- focal white-matter hyperintensities, often multiple.

- 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.

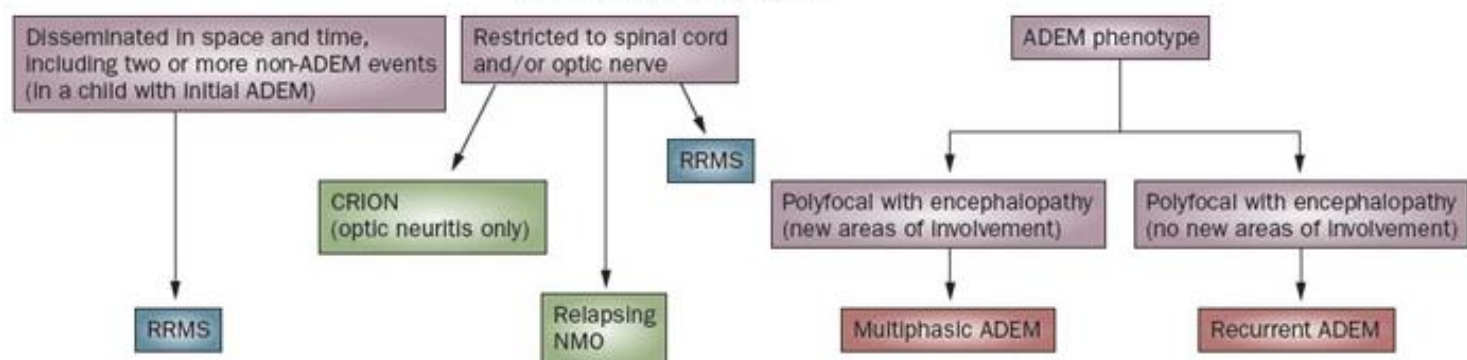
## Diagnostic algorithm of pediatric onset demyelinating disorders

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

First attack of demyelination



Further demyelination attacks



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