

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. 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:
 - 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

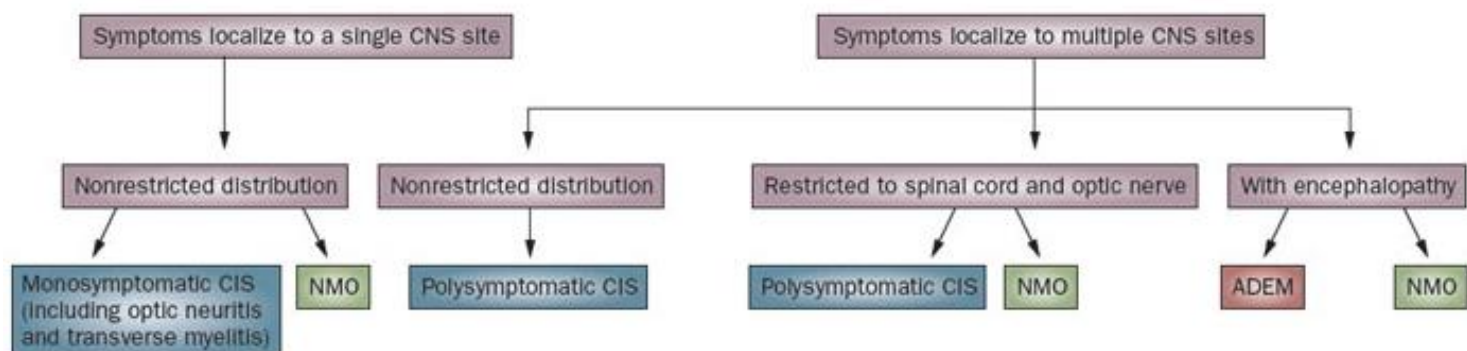
- focal white-matter hyperintensities, often multiple.

- 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.

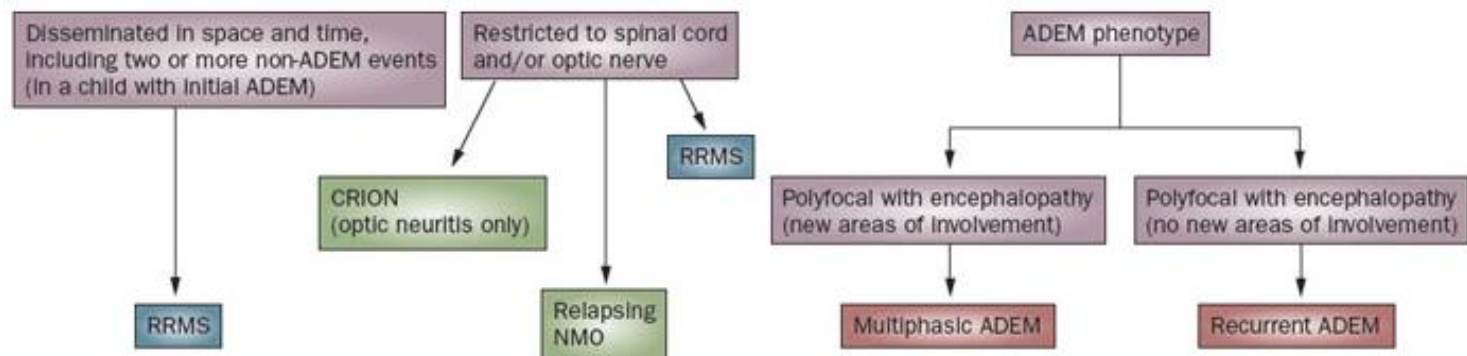
Diagnostic algorithm of pediatric onset demyelinating disorders

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



BIBLIOGRAPHY for ch. "Demyelinating Disorders" → follow this [LINK >>](#)