Inflammatory disorders of CNS:

Małgorzata Bilska

Encephalitis:

* Acute CNS dysfunction due to the presence of inflammatory cells in brain tissue
  Incidence: 1.5 to 7.4: 100.000 USA, 1.49 - 2.28 100.000 Poland
  Symptoms: seizures, focal neurological symptoms, impaired consciousness, psychiatric symptoms

Etiology:

Infectious:
bacterial 3- 16%
(N.meningitidis, Str.pneumoniae, H. influenzae, L.monocytogenes, M.pneumoniae)
Viral 9-26% :(HSV, VZV, enteroviruses, influenza virus, EBV, CMV, HHV-6)
Other <1%: fungal, protozoal

Non-infectious, immunological 20%:
(post vaccination, paraneoplastic syndromes)
Unknown 37-62%

Diagnostics:

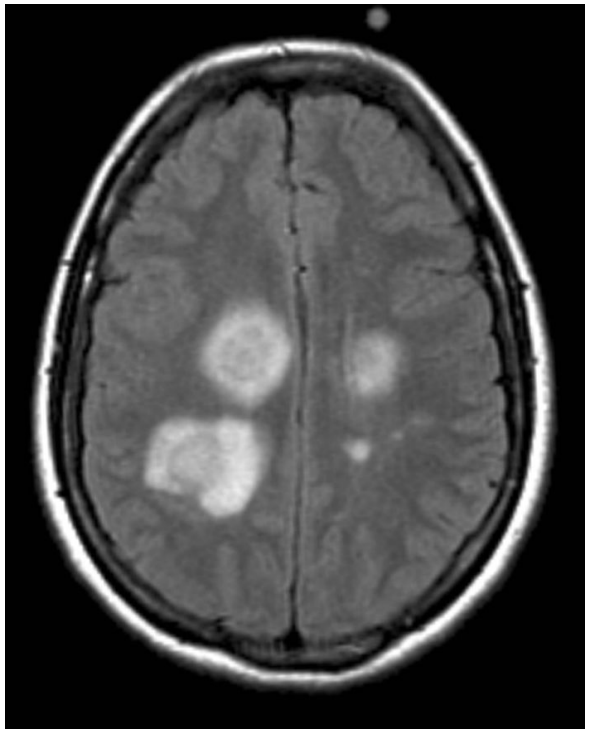
* MRI,CT, PET
* EEG
* Blood tests: serological,PCR ,
* urine, inflammatory markers, toxicological tests, antyneuronal antibodies,
* CSF examianation: latex tests, serology, PCR, oligoclonal bands, IgG index,
* Brain biopsy,
* Ophthalmological examination,
* Evoked potentials,

Pathomechanism:

* Molecular mimicry theory (the similarity of viral, bacterial proteins and neuronal antigens),
* Superantigen theory (bacterial antigens triggers the inflammatory cascade),
* Neuronal damage as a factor initiating antigen presentation

Acute Disseminated Encephalomyelitis (ADEM):

* inflammatory and demyelinating disease of the CNS
* mainly children (mean age 6 - 8 years), rare in adults
* neurological symptoms precedes infection (days - weeks), rarely after vaccination (eg. rabies, pertussis) and other immunological processes (eg. paraneoplastic syndromes, insect bites)



ADAM - diagnostic criteria:

Require the presence of all symptoms:

- multifocal neurological symptoms preceded by an inflammatory
- presence of symptoms of **encephalopathy**
- lesions in MRI undergoing fluctuations during the acute phase of illness lasting three months, then their gradual involution

-typically in MRI: diffused, poorly demarcated changes (> 1-2 cm)in white matter, seen in T2 and FLAIR sequences

- very rare lesions in T1 - possible, often symmetrical change involving deep gray matter (basal ganglia)

ADEM vs. SM:

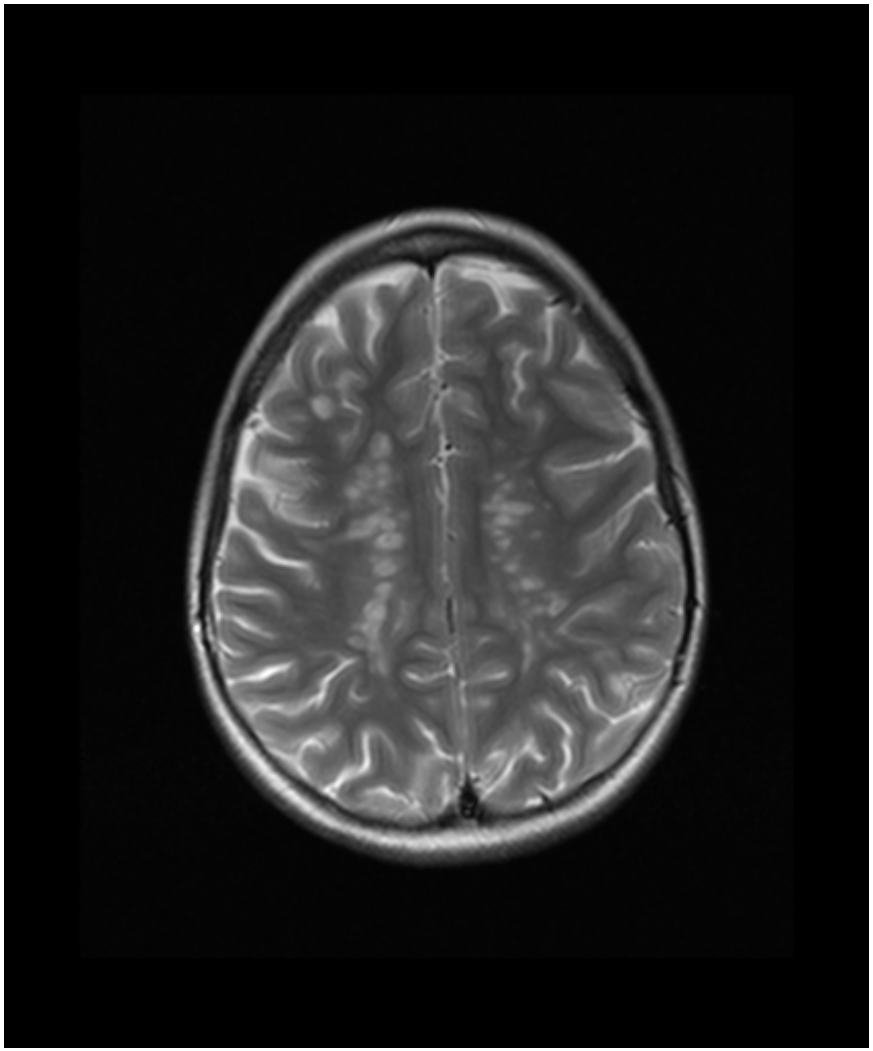
Control MRI examination performed after 6 months should show partial or total withdrawal of the changes in patients with ADEM

ADEM - treatment:

* Methylprednisolone 20 - 30 mg / kg /day (max. 1 g) for 5 - 7 days
* plasmapheresis or IVIG in a total dose of 1 to 2 g / kg for 2 - 5 days (in the case of lack of efficacy of steroids)
* cyclophosphamide

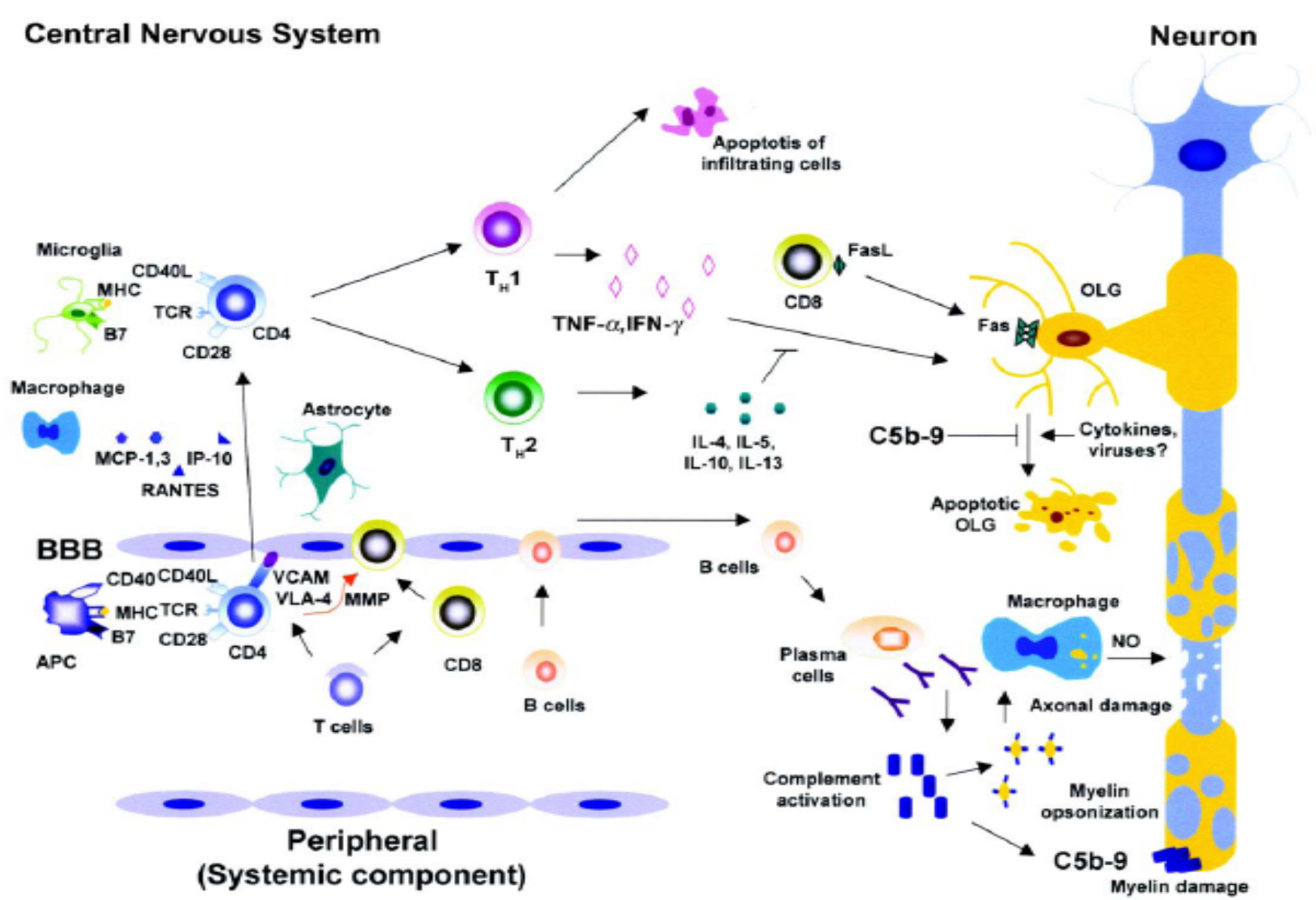
Multiple sclerosis(MS):

* Chronic autoimmune disorder characterized by the formation in the CNS foci of inflammation, demyelination and gliosis **(multiple foci)**
* Changes are scattered in space and time



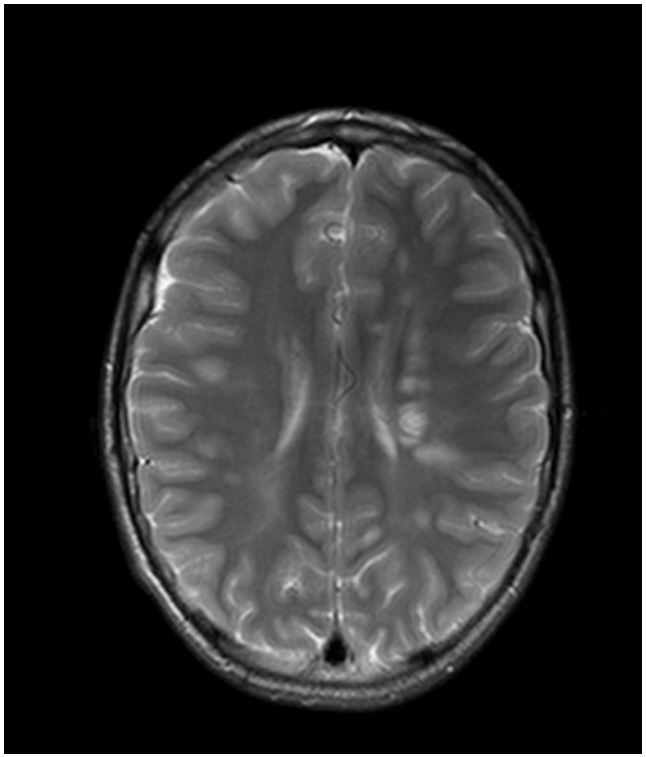
Pathogenesis:

* activation of the peripheral immune cells (dendritic cells, macrophages, lymphocytes),
* damage to blood-brain barrier penetration of inflammatory cells
* cross-reactive with myelin proteins
* influence of genetic and environmental factors on the development of the disease

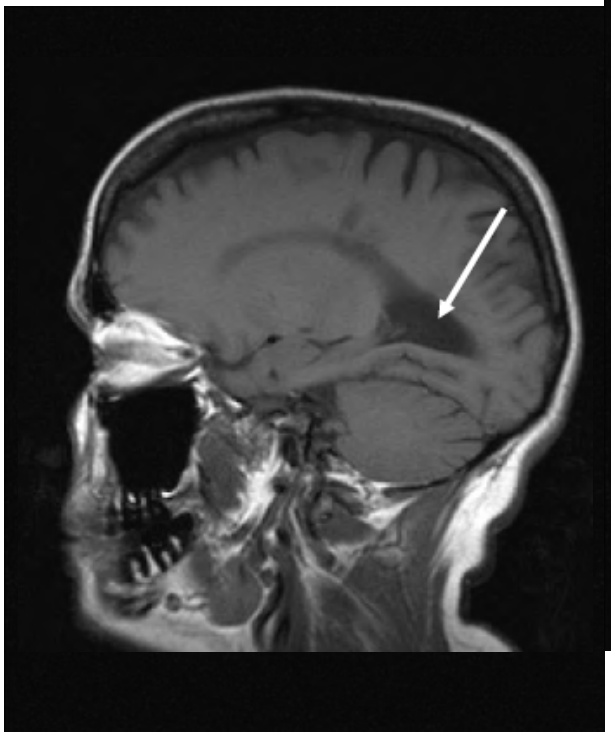


Cudrici C.,Niculescu T.,Niculescu F.J of Reh Research and Dev 2006,Vol 43(1)

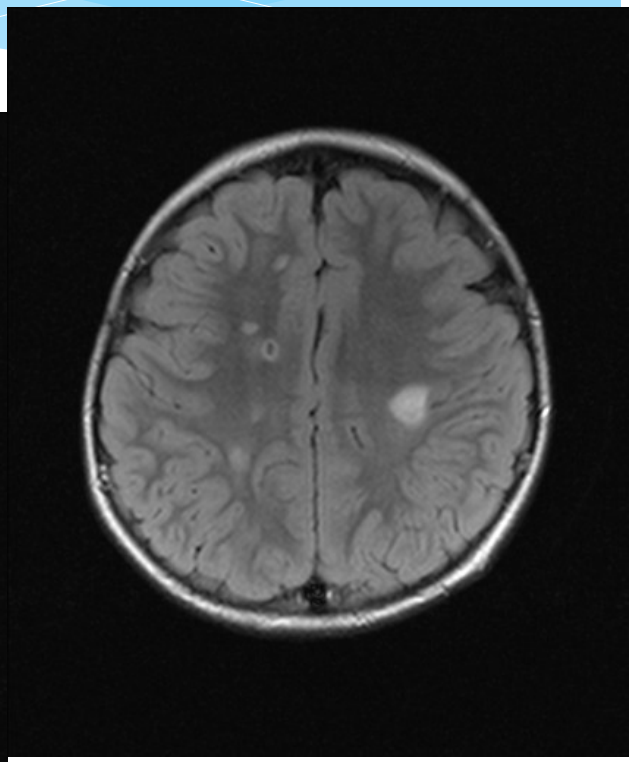
MRI in MS diagnostic:



T2



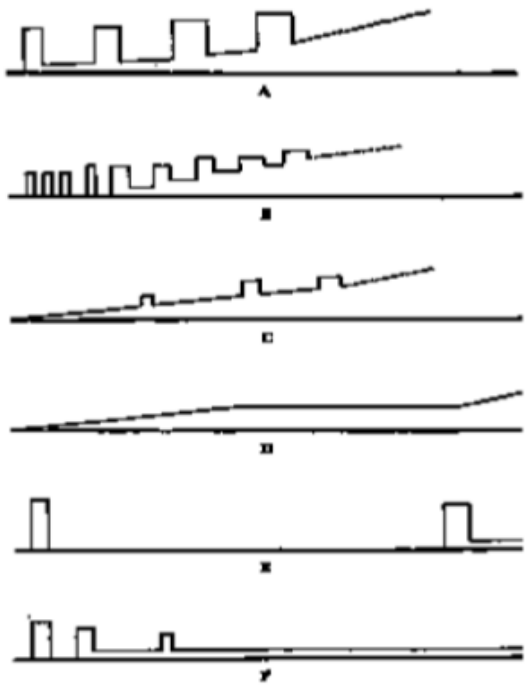
T1



FLAIR

Clinical course:

* relapsing-remitting (most common in children and adults)
* secondary progressive form
* primary progressive form
* progressive with relapses



McApline D, Compston ND, Lumsden CE: Multiple Sclerosis, 1955.

Multiple sclerosis -epidemiology :

* The risk in the general population - 1: 400, depending on the climate zone (Poland - high risk, 30 - 60 / 100.000
* The most common cause of neurological disability in young adults, the first symptoms usually occur about 20 - 40 years of age
* Symptoms before 15 (18) years of age,
  MS in children 3: 1 girls: boys
  (30% cerebellar symptoms, impaired balance, 20% of patients - retrobulbar inflammation of n.II , limb paresis, sensory disturbances, cognitive deficits, mood disturbances

MS treatment:

* **Treatment of relapses:**
  Methylprednisolone, IVIG, plasmapheresis

* **Immunomodulatory treatment:**
  First line: interferon beta, glatiramer acetate,
  Second line: natalizumab, fingolimod, dimethyl fumarate,
* Third line: alemtuzumab
  Daclizumab
  Cladribine
* **bone marrow transplantation**

* **neuroprotection**