Guillain-Barré syndrome in children

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EPNS teaching course , Budapest march 2015

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GBS

Guillain-Barré syndrome (GBS) is the most common cause of acute flaccid paralysis in children since the successful vaccination programs against poliomyelitis.

Estimated incidence of GBS

in children (0-15 yrs) is 0.34 to 1.34 /100.000 per year

*(*in adults: 1:100.000 persons/ year)

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Symptoms of periferal versus central nervous system

Periferal nervous system:

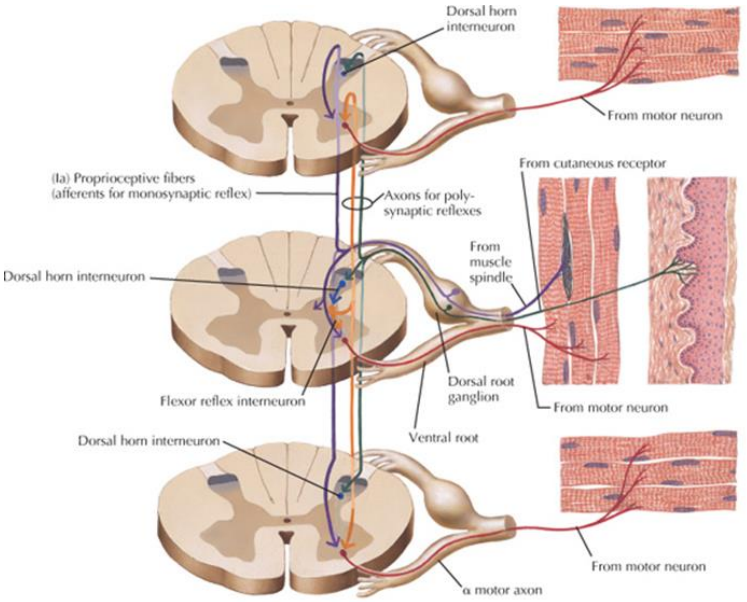
Anterior horn motor neuron

Afferent and efferent roots

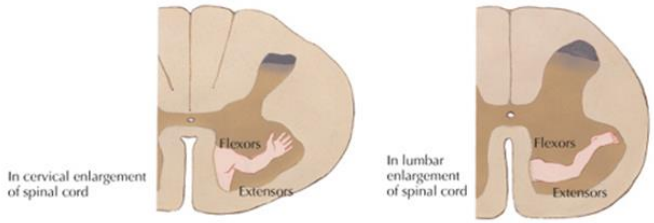
Sensory-motor nerve

Neuromuscular junction

Muscle



Schematic representation of motor neurons



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Symptoms of periferal versus central nervous system

Periferal nervous system:

Anterior horn motor neuron

Afferent and efferent roots

Sensory-motor nerve

Neuromuscular junction

Muscle

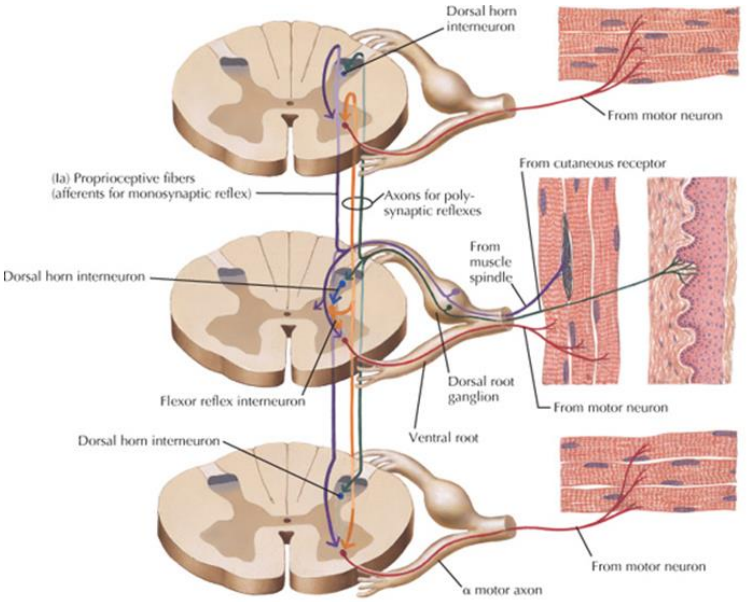
Symptoms:

Paresis/ paralysis

Hypotonia

Low or absent reflexes

fasciculations



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Diagnostic criteria

Essential criteria

- Progressive, more or less symmetrical weakness of 2 or more limbs
- Areflexia / hyporeflexia
- Most severe paresis < 4 weeks
- No other cause

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Findings supporting a diagnosis of GBS

Essential criteria

- Progressive, more or less symmetrical weakness of 2 or more limbs
- Areflexia / hyporeflexia
- Most severe paresis < 4 weeks
- No other cause

Findings supporting GBS diagnosis (not always present)

- Sensory deficits, pain, facial weakness
- Difficult in swallowing, respiratory insufficiency
- Spinal fluid abnormalities
- EMG abnormalities

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Spinal fluid in GBS

- in the first week after onset in 50 % of GBS patients normal

- after two weeks 80% has a raised spinal fluid protein content

- cell number is normal or slightly raised

(< 50 cells x 106 mononuclear cells

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EMG

It is typically prudent to wait at least 7-10 days for electrical studies to be informative.

If electrical studies are performed too early, normal results can be falsely reassuring.

EMG

First week after onset of symptoms

- dispersed, impersistent, prolonged, or absent F response (88%)
- increased distal latencies (75%)
- conduction block (58%) or temporal dispersion of compound muscle action potential (CMAP)
- reduced conduction velocity (50%) of motor and sensory nerves

Second week of illness

- reduced compound muscle action potential (CMAP, 100%)
- prolonged distal latencies (92%)
- reduced motor conduction velocities (84%) are prominent.

.

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EMG

Criteria for axonal forms of GBS include

- lack of neurophysiologic evidence of demyelination

- loss of amplitude of CMAP or sensory nerve action potentials to at least less than 80% of lower limit of normal values for age

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Symptoms that may point to another diagnosis than GBS

- Relatively mild paresis of arms and legs and:
    - severe pulmonary dysfunction
    - severe sensory deficits
    - clear deficits of bladder and/ or bowel
- Fever
- Sensory level
- Slow progression (> 4 weeks)
- Clear and persisting asymmetry of muscle weakness
- Persisting bladder and bowel dysfunction

- Spinal fluid > 50 x 106/ mononuclear cells

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Clinical GBS variants

- acute inflammatory demyelinating polyradiculopathy

- acute motor axonal neuropathy

- acute motor and sensory axonal neuropathy

- Miller Fisher syndrome (ophthalmoplegia, absent tendon reflexes, ataxia) GQ1b ganglioside often positive!

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RARE GBS variants

- pharyngeo-cervical brachial variant

- pure sensory axonal neuropathy

- Bickerstaff encephalitis

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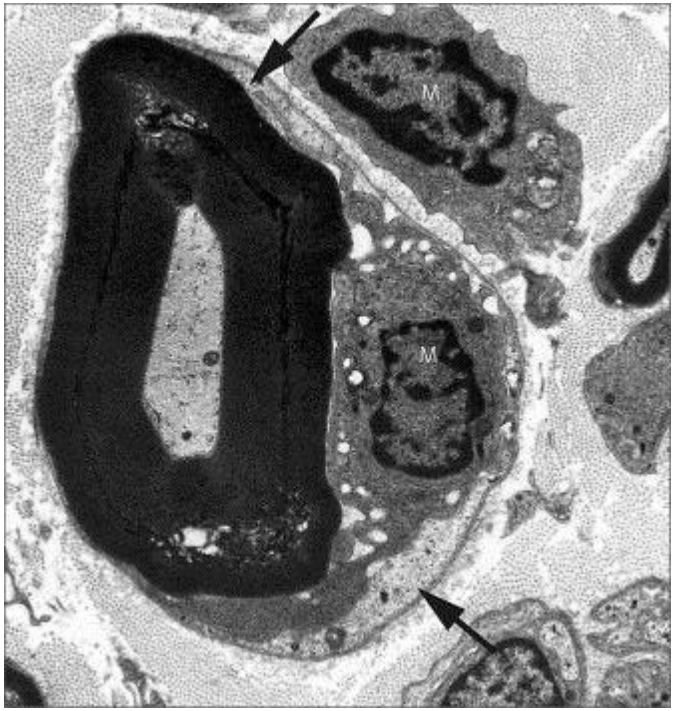
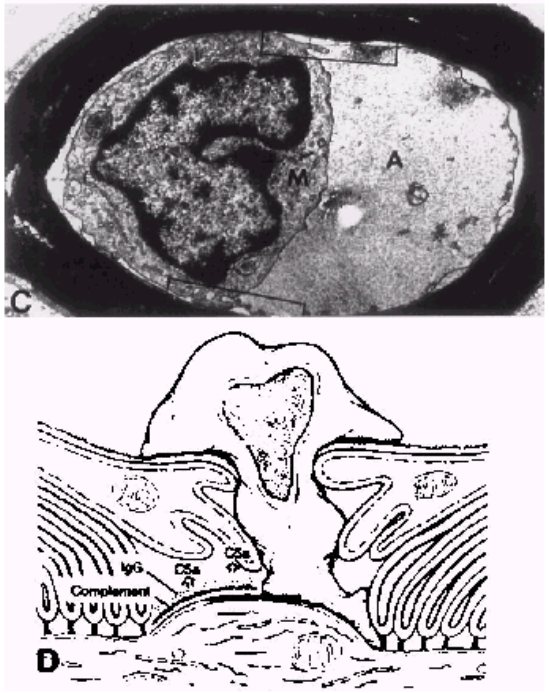
Histological classification of GBS

Demyelination



Acute inflammatory demyelinstin neuropathy (AIDP)

Axonal degeneration



Acute motor axonal neuropathy (AMAN)

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Differential diagnosis

(Sub)acute flaccid paresis in children

- Poliomyelitis
- Lyme disease (Borrelia)
- Botulism
- Spinal Muscular Atrophy (SMA)
- Myelopathie
- Myasthenia gravis
- Toxic: arsenic acid, thallium, organic phosphates
- Wernicke disease
- Critical illness polyneuropathie
- Vasculitis
- Toxins of weird animals

Black widow

ciguatera

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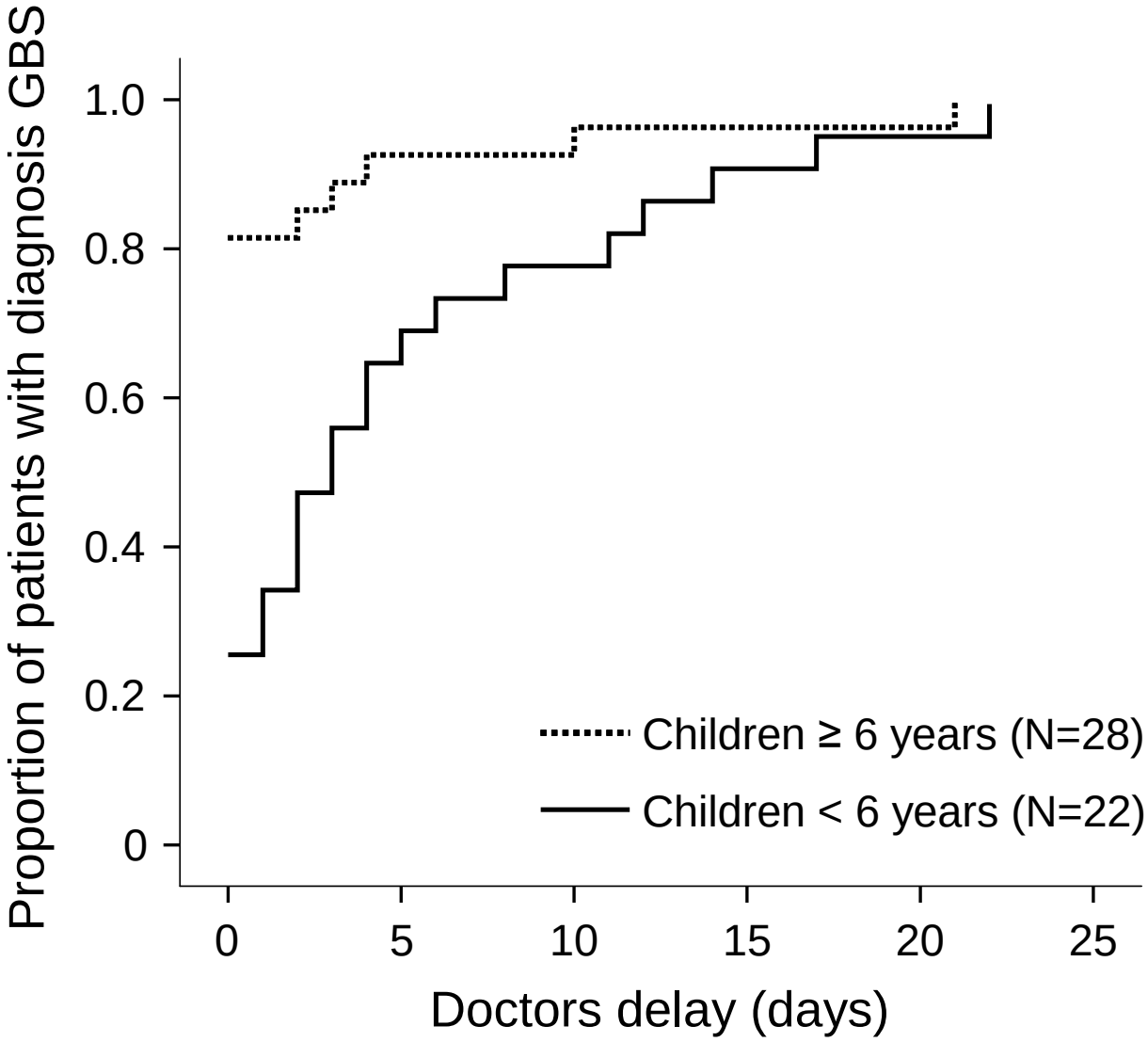
Diagnosis in children is often difficult….



Roodbol et al. Neurology 2011;76(9):807

First diagnosis in our cohort of 56 children

75% of the children < 6 years old: pain in neck and or legs

- First diagnosis in majority: ‘flue’, pneumonia, suspicion of a metabolic disease, myopathy, tonsillitis, acute rheumatic fever
- 3 malignancy, 2 meningitis, 4 coxitis
- 1 no idea
- 7 suspicion of Guillain-Barré syndrome

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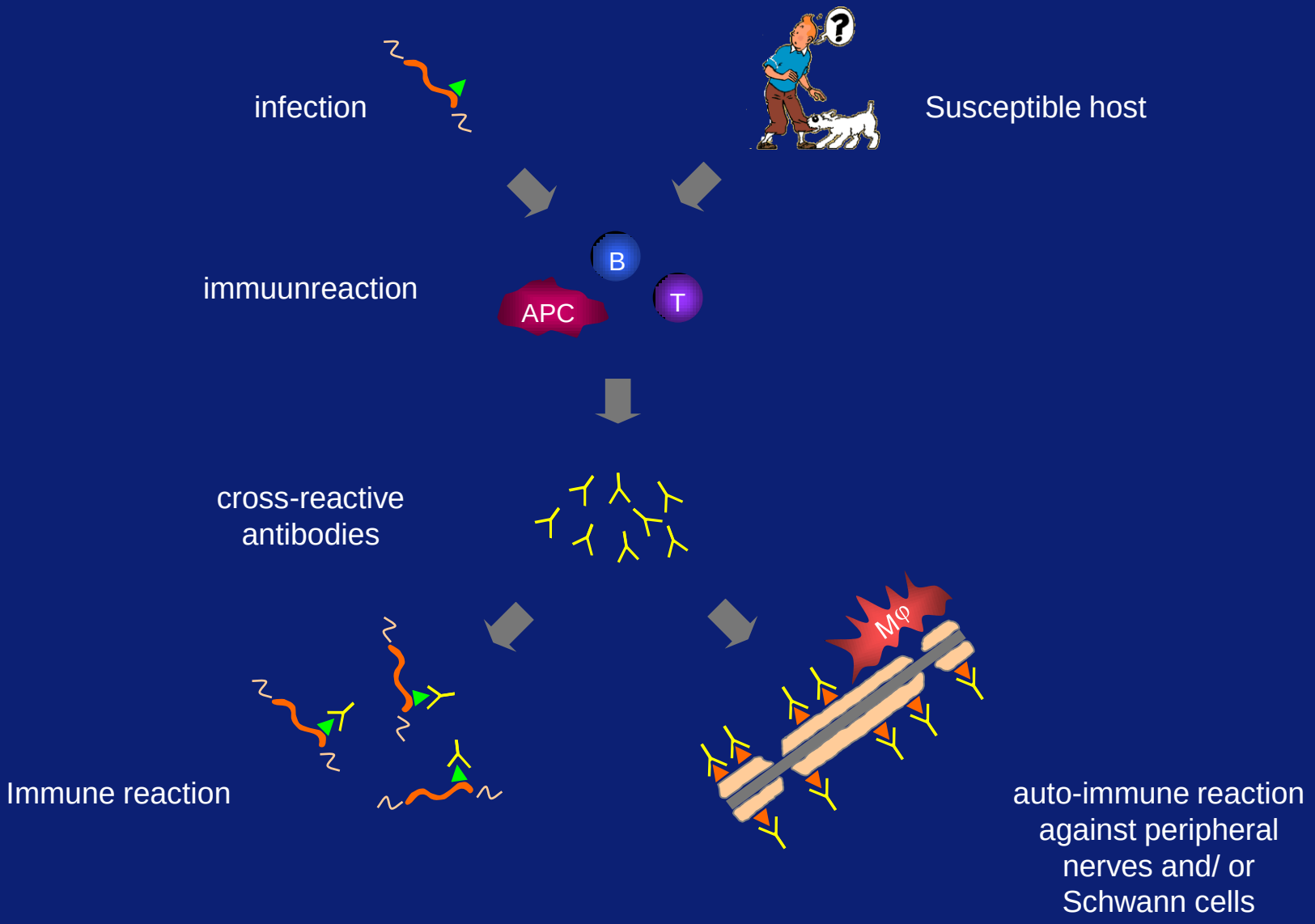
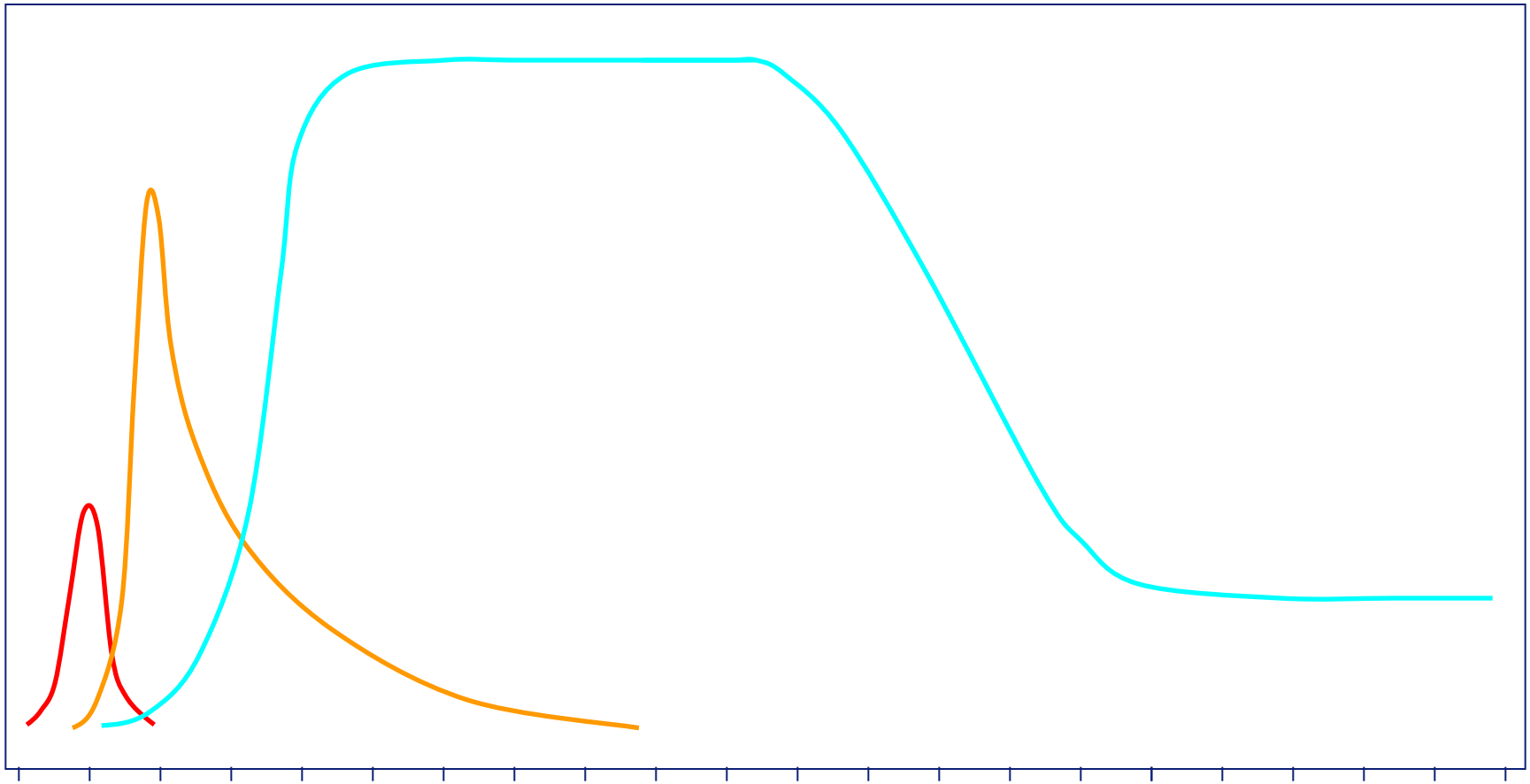
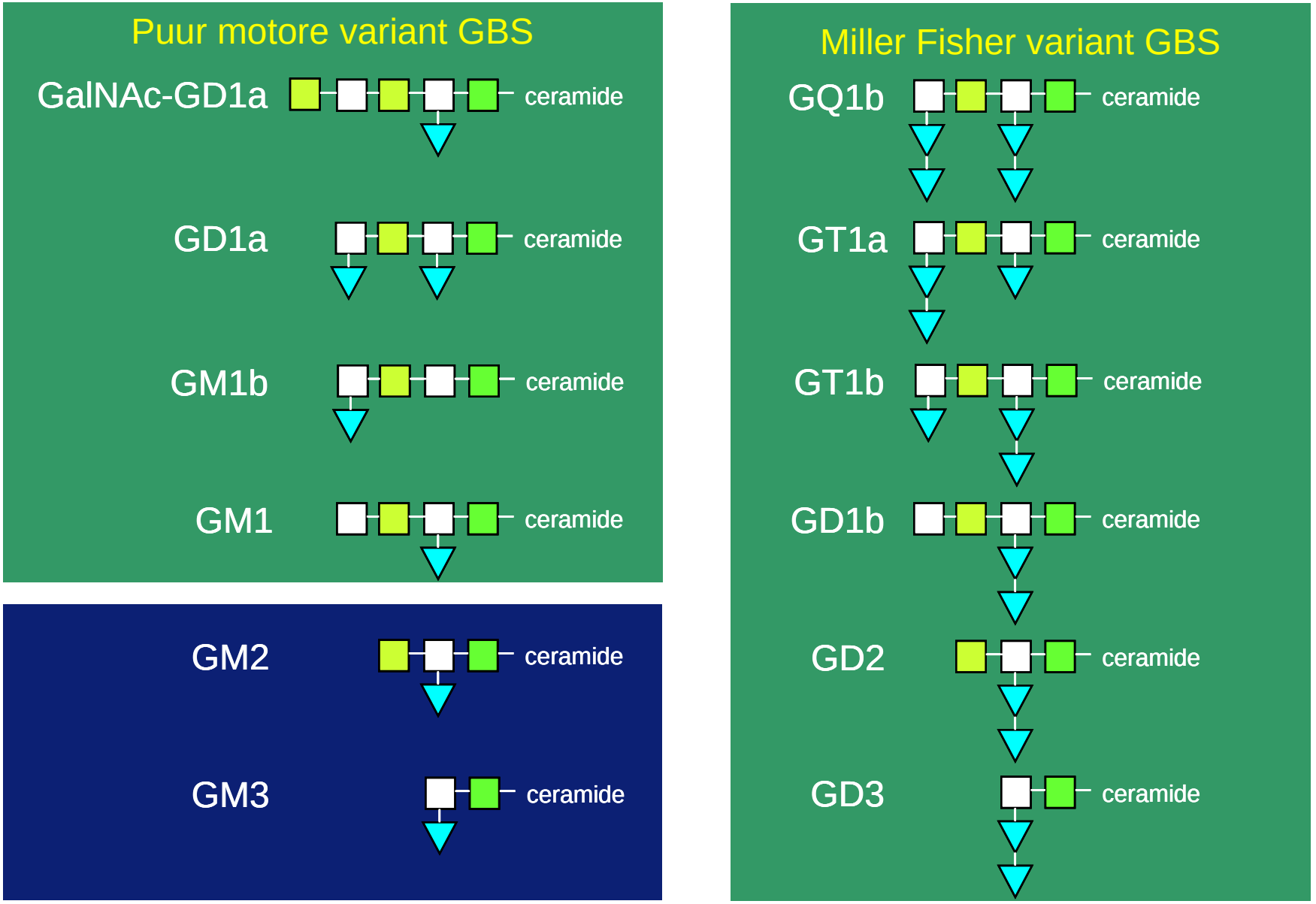
Hypothesis of pathogenesis GBS: *molecular mimicry*



Clinical disease pattern in GBS



Antibodies in GBS patients are against ganglioside



Treatment

Admit a child to ICU

- rapid progression of motor deficits

- threat of airway obstruction or respiratory insufficiency

- impairment of autonomic functions

- severe swallowing problems

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Treatment with IVIg of children with GBS

Indicated when:

- Unable to walk independently
- (threat of) respiratory insufficiency
- Bulbar weakness
- Autonomic dysfunction
- Fast progression of symptoms

- **Repeated infusions of IVIg are indicated when:**
- Deterioration after initial stabilisation or amelioration
- No or insufficient effect of first IVIg treatment ?

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Prognosis

More favourable than in adults!

- at the moment of maximum neurological symptoms
  - walking with aid : 45%
  - Bed/wheelchair : 40-60%
  - Mechanical ventilation: 15-20%
  - Death: : 1-2 %
- Treatment related fluctuation : 10%
  relapse < 4weken
- Transition tor CIDP : 2%
  relapse > 4 weken

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Prognosis

Mortality

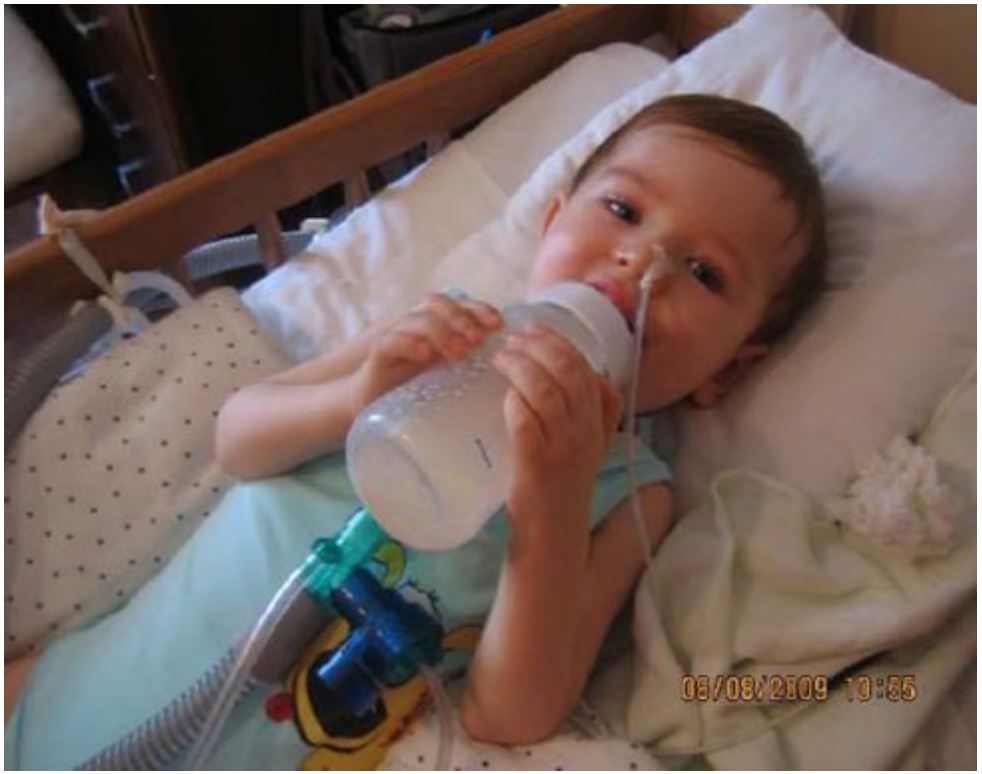
- Adults: 2,8%
- Children 2%

Needs ventilation

- Both 25%

Outcome 6 months

- Non-ambulant
  - 6% children
  - 20% adults



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Long term sequelae in 37 patients with childhood GBS

23/ 37 were now adults (age range 4-39 y, median 20 years)

Follow up: 1-22 years (median 11 years)

26% had problems at school after GBS

- 13% had to repeat a class
- 11% had to continue at a lower level at school
- 3% (one child) could not go to school because of evere fatigue

11% could not work because of sequalae

Roodbol et al. J Peripher Nerv Syst. 2014

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Long term sequelae

Risk of recurrence of GBS is low (1 à 2%)

Prognosis for regaining ambulation is excellent

In our cohort 10% had a persisting handicap

BUT

50% still suffered from fatigue, which was very disabling in some

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Conclusions GBS in children

- In children < 6 years diagnosis can be difficult
- Often atypical presentation with a lot of pain
- In case of rapid neurological deterioration ICU monitoring
- Prognosis of GBS in children is in general better than in adults, maybe because of better nerve regeneration
- At long term persisting neurological impairments, handicaps and fatigue may occur

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Thanks to: GBS- kids study group

Paediatric Neurology

Joyce Roodbol

Marie-Claire de Wit

Coriene Catsman

Paediatrics

Annemarie van Rossum

Emiel Spuesens

Matthijs de Hoog

Neuroimmunology

Bart Jacobs

Christa Walgaard

Neurphysiology

Judith Drenthen

Joleen Blok