Guillain-Barre Syndrome

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GBS

- Eponym that encompasses acute immune-mediated polyneuropathies
- Peripheral nerve myelin is target of an immune attack
- Starts at level of nerve root=conduction blocks & muscle weakness
- Eventually get widespread patchy demyel= increased paralysis

Pathophysiology

- Usually postinfectious
- Immune-mediated: infectious agents thought to induce Ab production against specific gangliosides/glycolipids
- Lymphocytic infiltration of spinal roots/peripheral nerves & then macrophage-mediated, multifocal stripping of myelin
- Result: defects in the propagation of electrical nerve impulses, with eventual conduction block and flaccid paralysis

Clinical Features:

- Progressive, fairly symmetric muscle weakness

  -typically starts in proximal legs

  -10% will 1st develop weakness in face or arms

  -severe resp muscle weakness in 10-30% pts

  -oropharyngeal weakness in ~ 50%

Clinical Features:

- Absent or depressed DTR
- Often prominent severe pain in lower back
- Common to have paresthesias in hands and feet
- Dysautonomia is very common: tachycardia, urinary retention, hypertenison alternating w/ hypotension, ileus

Diagnosis:

- Albuminocytologic dissociation: elevated CSF protein w/ normal WBC (80-90% pts)
- Electromyography (EMG) helps confirm diagnosis = prolonged or absent F waves

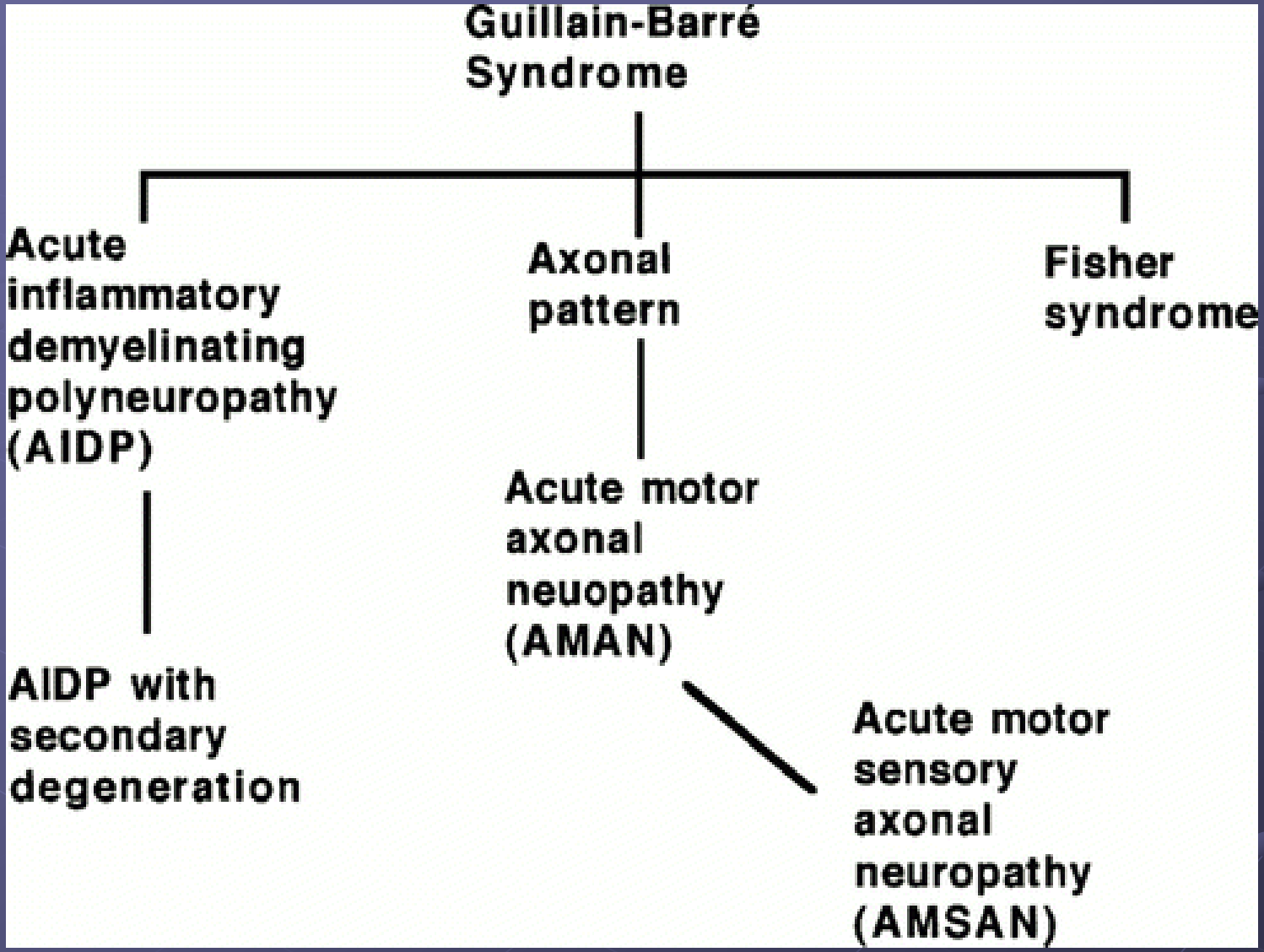
NINDS Expert Consensus:

- Req'd Features for dx:

1. Progressive weakness of > than 1 limb
2. Areflexia

- Supportive Features:

~progression of Sx over days to 4 weeks
~relative symmetry
~CN involv esp b/l facial n weakness
~autonomic dysfunction ~EMG features
~elev CSF protein w/ cell count ,10 mm3

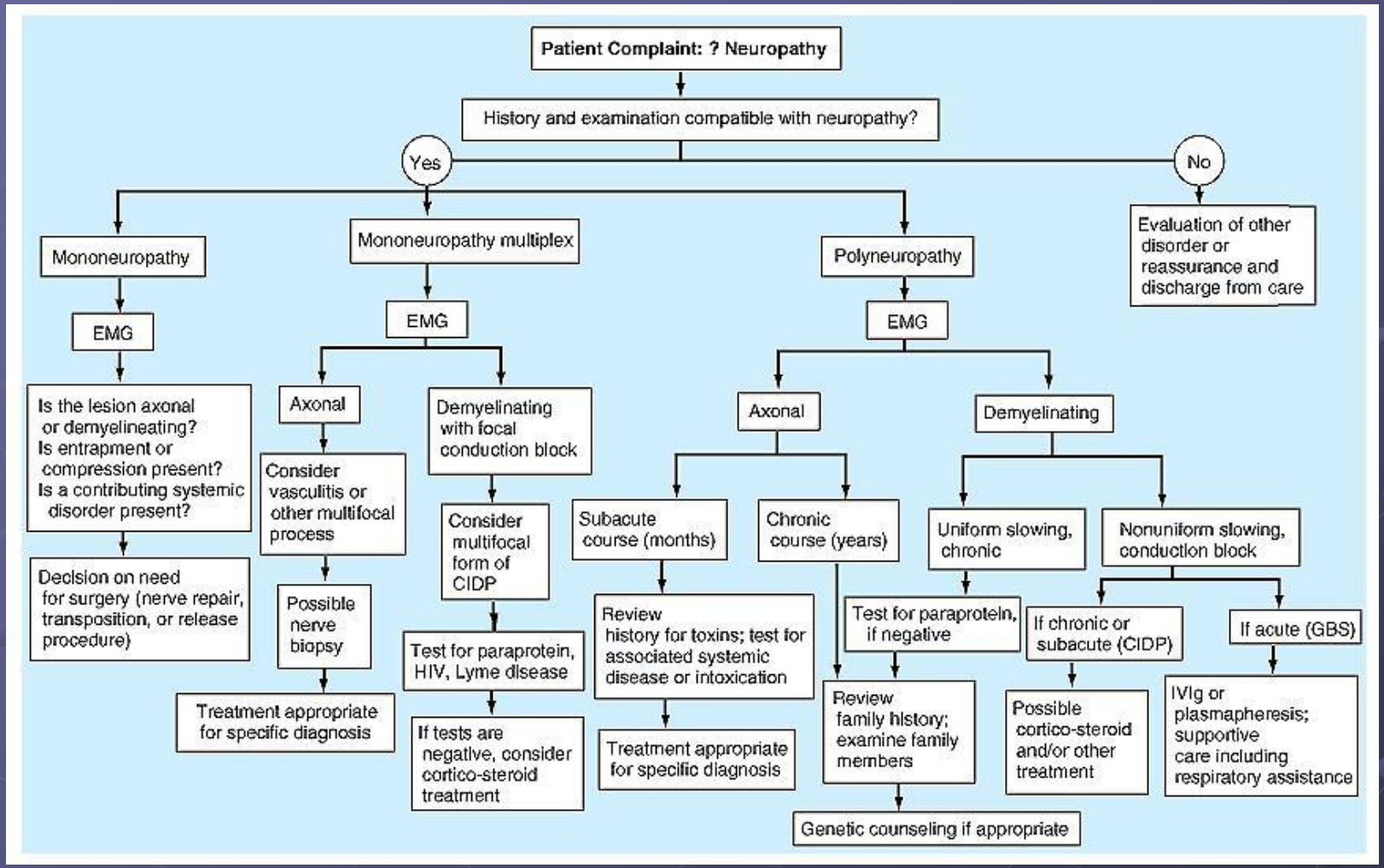

Guillain-Barré Syndrome
Acute inflammatory demyelinating polyneuropathy (AIDP)
Axonal pattern
Fisher syndrome
AIDP with secondary degeneration
Acute motor axonal neuopathy (AMAN)
Acute motor sensory axonal neuropathy (AMSAN)

GBS=heterogenous syndrome w/ variant forms

- Think of AIDP as the traditional form as described previously, accts for 85-90%
- **Miller Fisher Syndrome**: opthalmoplegia, ataxia, and areflexia (5%). GQ1b antibody. Only 1/4th w/ extremity weakness
- AMAN: selective involv of motor nerves, DTRs are preserved, more common in Japan/China, almost all preceded by Campylobacter infxn
- AMSAN: more severe form of AMAN +sensory

DDx of Polyneuropathy:

- Arsenic poisoning
- N-Hexane (glue sniffing)
- Vasculitis
- Lyme Disease
- Tick paralysis
- Sarcoidosis
- Leptomeningeal Dz
- Paraneoplastic Dz
- Critical Illness


Patient Complaint: ? Neuropathy
History and examination compatible with neuropathy?
Yes
No
Evaluation of other disorder or reassurance and discharge from care
Mononeuropathy
EMG
Is the lesion axonal or demyelineating? Is entrapment or compression present? Is a contributing systemic disorder present?
Decision on need for surgery (nerve repair, transposition, or release procedure)
Mononeuropathy multiplex
EMG
Axonal
Consider vasculitis or other multifocal process
Possible nerve biopsy
Treatment appropriate for specific diagnosis
Demyelinating with focal conduction block
Consider multifocal form of CIDP
Test for paraprotein, HIV, Lyme disease
If tests are negative, consider cortico-steroid treatment
Polyneuropathy
EMG
Axonal
Subacute course (months)
Review history for toxins; test for associated systemic disease or intoxication
Treatment appropriate for specific diagnosis
Chronic course (years)
Review family history; examine family members
Genetic counseling if appropriate
Demyelinating
Uniform slowing, chronic
Test for paraprotein, if negative
Nonuniform slowing, conduction block
If chronic or subacute (CIDP)
Possible cortico-steroid and/or other treatment
If acute (GBS)
IVIg or plasmapheresis; supportive care including respiratory assistance

Supportive Care

- Monitor Resp status closely (follow NIFs), up to 30% may req ventilatory support
- In severe cases, intrarterial monitoring may be necessary given the gisngifcant blood pressure fluctuations
- Neuropathic pain plagues most, often managed w/ Gabapentin or Carbamazepine

Disease Modifying Treatment

- IVIG : typically given for 5 d at 0.4 gram/kg/d (may need to extend course depending on response)
- Plasmapheresis: usually 4-6 treatments over 8-10 days

The choice b/w plasma exchange and IVIG is dep on availability, pt contraindications, etc. Because of ease of administration, IVIG is frequently preferred. The cost and efficacy of the 2 treatments are comparable.
Glucocorticoids have NO ROLE!!