Guillain-Barré Syndrome

Ouch!
Guillain-Barré Syndrome

- Acute post-infective polyneuropathy
- Heterogeneous condition with several variant forms
Neuronal Ganglioside

Lipid A

Neuronal Ganglioside
Pathogenesis

- **Antecedent infection**
  - Campylobacter jejuni
  - Cytomegalovirus
  - Epstein-Barr virus
  - HIV
  - Influenza virus
  - Escherichia coli
  - Haemophilus influenzae

- **Molecular Mimicry**
  - shared cross-reactive immunogenic epitopes

UK study of 103 GBS patients
26% had evidence of recent c. jejuni
1% of age matched controls
2% of household controls

Other studies suggest up to 60-70% in AMSAN and AMAN

Only 70% of patients infected with c. jejuni report symptoms

It is possible to induce flaccid paresis by inoculating mice with c. jejuni lipooligosacharide
Pathogenesis

Schwann Cell

Axon
Pathogenesis

Schwann Cell

Axon
Clinical Features at Time of Presentation

- **Weakness**
  - ascending in 90%
  - oculomotor in 15%
  - depressed deep tendon reflexes

- **Parasthesia**
  - hands and feet

- **Pain**
  - lower back

- **Dysautonomia**
  - arrhythmia
  - pulse rate ↓↑
  - blood pressure ↓↑
  - temperature ↓↑
  - urinary retention

- **Rare**
  - facial myokymia
  - hearing loss
  - meningeal irritation
  - vocal cord paralysis
  - altered mental status

**Specific Features**

- Progressive
- Monophasic
- Symmetrical
- Apyrexial
Epidemiology

- 3 cases / 100 000 / year
- Most common acute polyneuropathy
- Overall incidence similar across the western world
- Geographical variation in variants
Guillain-Barré Syndrome Variants

- Acute Inflammatory Demyelinating Polyneuropathy (AIDP)
- Miller-Fisher Syndrome (MFS)
- Acute Motor Axonal Neuropathy (AMAN)
- Acute Motor and Sensory Axonal Neuropathy (AMSAN)
- Pharyngeal-Cervical-Brachial (PCB)
- Pure Sensory
- Bickerstaff encephalitis
- Acute Pandysautonomia
Guillain-Barré Syndrome Variants

- Acute Inflammatory Demyelinating Polyneuropathy (AIDP)
- Miller-Fisher Syndrome (MFS)
- Acute Motor Axonal Neuropathy (AMAN)
- Acute Motor and Sensory Axonal Neuropathy (AMSAN)
- Pharyngeal-Cervical-Brachial (PCB)
- Pure Sensory
- Bickerstaff encephalitis
- Acute Pandysautonomia
Guillain-Barré Syndrome Variants

- 84% Acute Inflammatory Demyelinating Polyneuropathy (AIDP)
- 8% Miller-Fisher Syndrome (MFS)
- 5% Acute Motor Axonal Neuropathy (AMAN)
- 2% Acute Motor and Sensory Axonal Neuropathy (AMSAN)
- 1% Others
Acute Inflammatory Demyelinating Polyneuropathy (AIDP)

- 85% of presentations in Australia
- 30% have demonstrable recent c. jejuni infection
- Demyelination followed by inflammatory axonal damage
- Progressive, symmetric ascending muscle weakness accompanied by diminished deep tendon reflexes
Miller Fisher Syndrome

- Ophthalmoplegia, ataxia and areflexia (descending weakness)
- 90% have anti GQ1b
Acute Motor Axonal Neuropathy (AMAN)

- 30% of GBS in Japan and China
- Young patients
- 70% have demonstrable recent c. jejuni infection
- Direct axonal damage
- GM1, GD1a, GD1b and GalNac-GD1a
- No sensory involvement
Acute Motor and Sensory Axonal Neuropathy (AMSAN)

- AMAN + sensory involvement
Investigations

- **CSF**
  - raised protein, normal WCC (albuminocytological dissociation), sensitivity 70%

- **Nerve conduction studies**
  - useful for diagnosis and prognosis

- **Antibodies**
  - Anti GQ1b: sensitivity = 90%, specificity ~100% for Miller-Fisher / Bickerstaff encephalitis
Management

- Admit to hospital
- Early discussion with ICU
- 4 hourly vital capacity - 20mL/Kg
- Cardiac monitoring
- Attention to autonomic dysfunction
- Neuropathic analgesia - gabapentin
- DVT prophylaxis
Immunotherapy

- IV immunoglobulin and plasma exchange both hasten recovery
- IV immunoglobulin is equivalent to plasma exchange
- Onset of recovery is reduced by approximately 40 days
- Median time to walking unaided = 53 vs 83
- No benefit of combination
- No benefit of steroids
- No benefit of interferon beta

American Accadamy of Nerology (AMN) Practicle Paramater on Immunotherapy for Guillain-Barré Syndrome. 2003
Clinical Course

- Most patients (74%) get worse for 2 weeks, plateaux for 2-4 weeks and improved thereafter
- 67% have begun recovery at 4 weeks
- 50% need an ICU/HDU admission
- 35% need invasive ventilation
Clinical Course

- Most patients (74%) get worse for 2 weeks, plateaux for 2-4 weeks and improved thereafter
- 67% have begun recovery at 4 weeks
- 50% need an ICU/HDU admission
- 35% need invasive ventilation

Poor Prognostic Features

- Older age
- Rapid onset
- Need for Ventilation
- Distal motor response < 20%
- Preceding diarrheal illness
Long Term Outcome

- Walk independently: 66%
- Mild neurological defect: 16%
- Wheelchair bound: 3%
- Ventilated: 7%
- Dead: 5%
Long Term Outcome

- **Walk independently**: 66%
- **Mild neurological defect**: 16%
- **Wheelchair bound**: 3%
- **Ventilated**: 7%
- **Dead**: 5%

**Cause of Death**
- ARDS
- Sepsis
- PE
- Dysautonomia
Things to Remember

- Acute post-infective polyneuropathy
- Weakness, pain and dysautonimia
- Will present with symmetrical progressive weakness and/or parasthesia
- 5-10% will be limited to cranial nerves at time of presentation
- Admit to hospital, talk to ICU, start DVT prophylaxis and gabapentin early
- IV immunoglobulin or plasma exchange