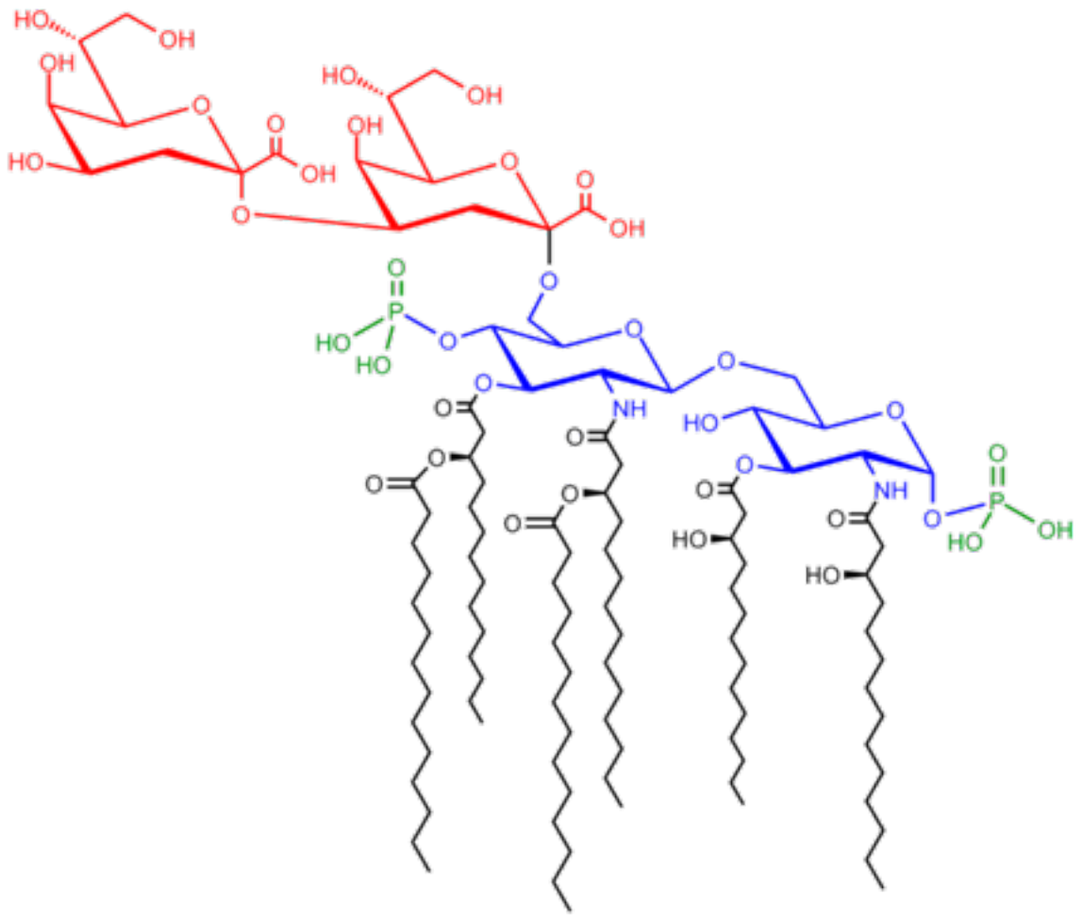
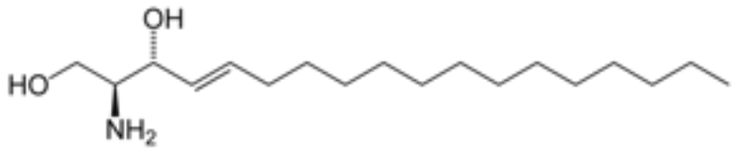
Guillain-Barré Syndrome

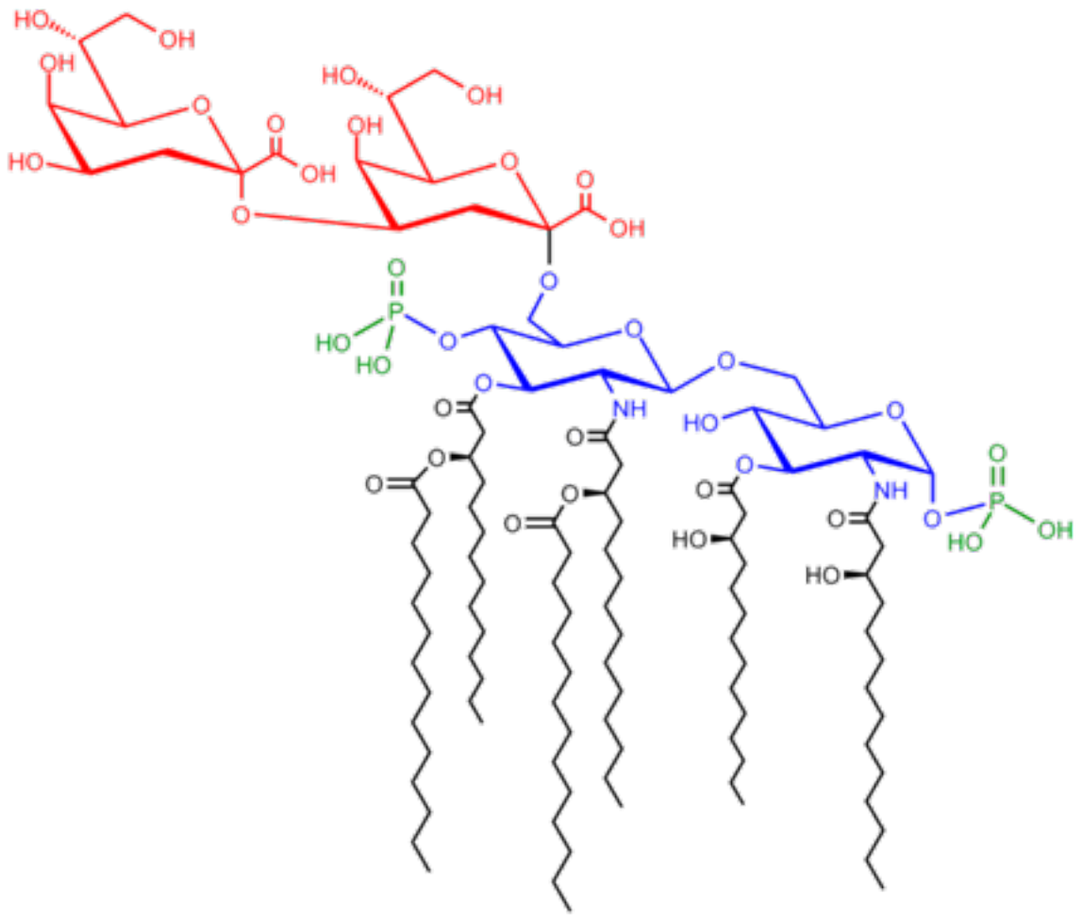
Ouch!

Guillain-Barré Syndrome

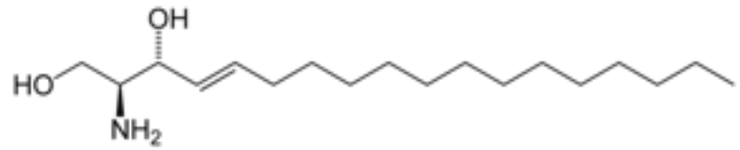
- Acute post-infective polyneuropathy
- Heterogeneous condition with several variant forms







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

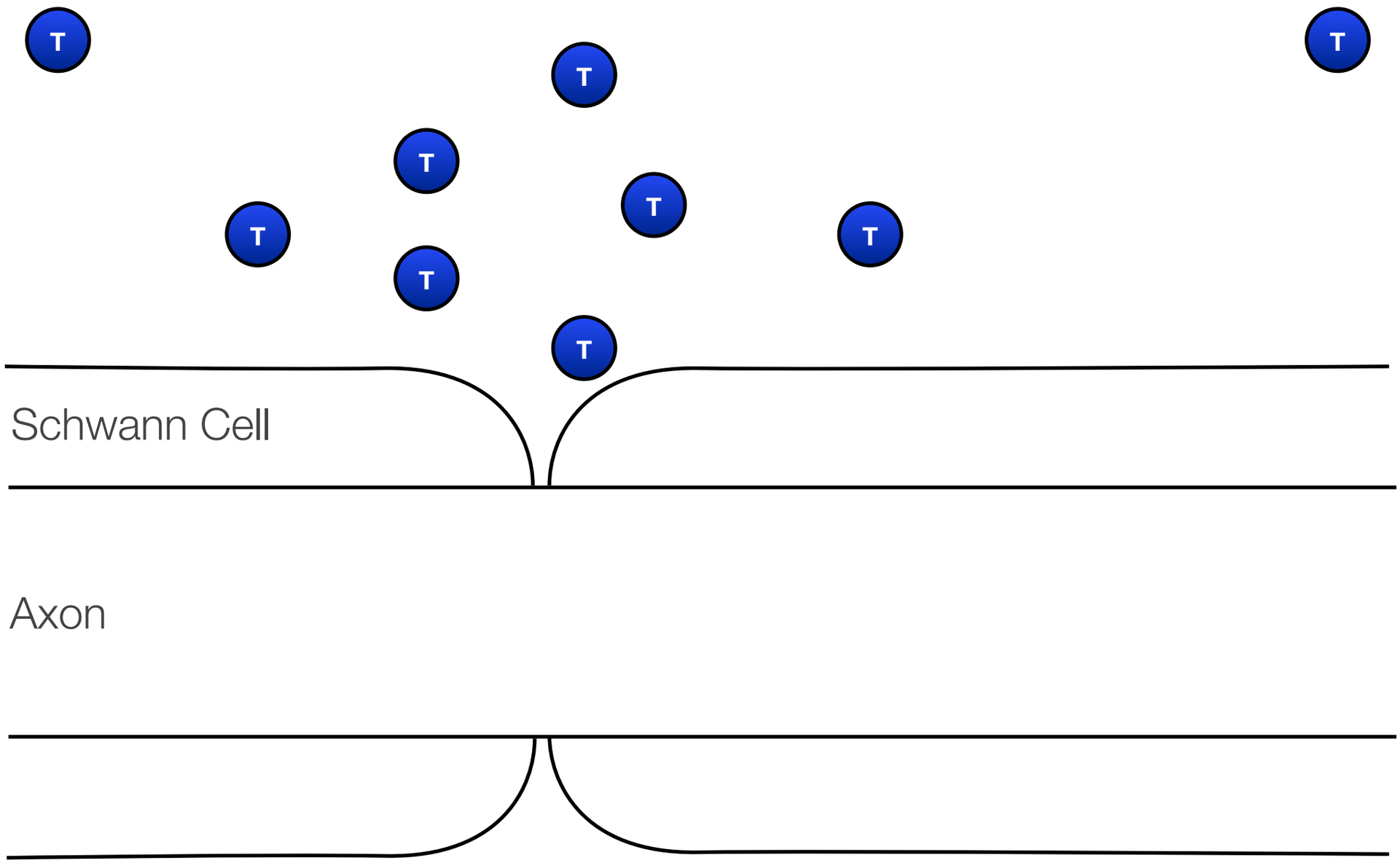
UK study of 103 GBS patients
26% had evidence of recent c. jejuni
1% of age matched controls
2% of household controls

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

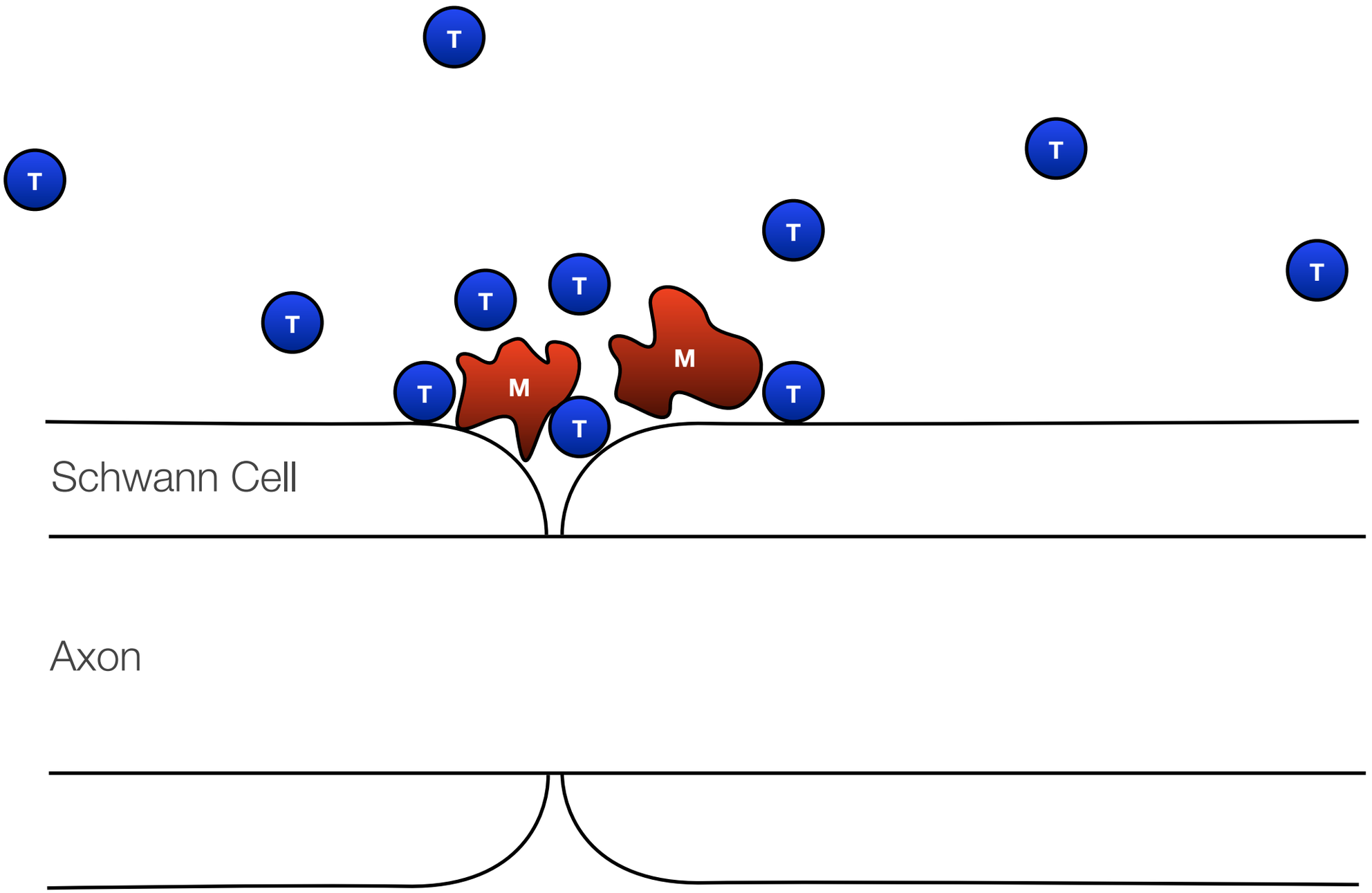
Only 70% of patients infected with c. jejuni report symptoms

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

Pathogenesis



Pathogenesis



Clinical Features at Time of Presentation

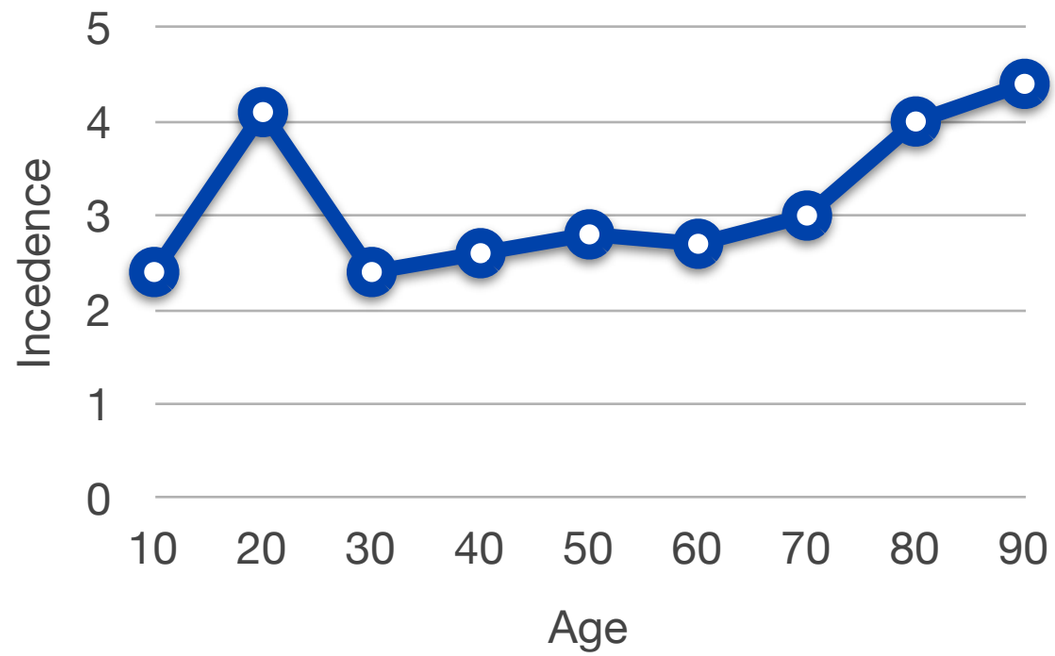
- Weakness
  - ascending in 90%
  - occulomotor in 15%
  - depressed deep tendon reflexes
- Parasthesia
  - hands and feet
- Pain
  - lower back
- Dysautonomia
  - arrhythmia
  - pulse rate ↓↑
  - blood pressure ↓↑
  - temperature ↓↑
  - urinary retention
- Rare
  - facial myokymia
  - hearing loss
  - meningeal irritation
  - vocal cord paralysis
  - altered mental status

Specific Features

- Progresive
- 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

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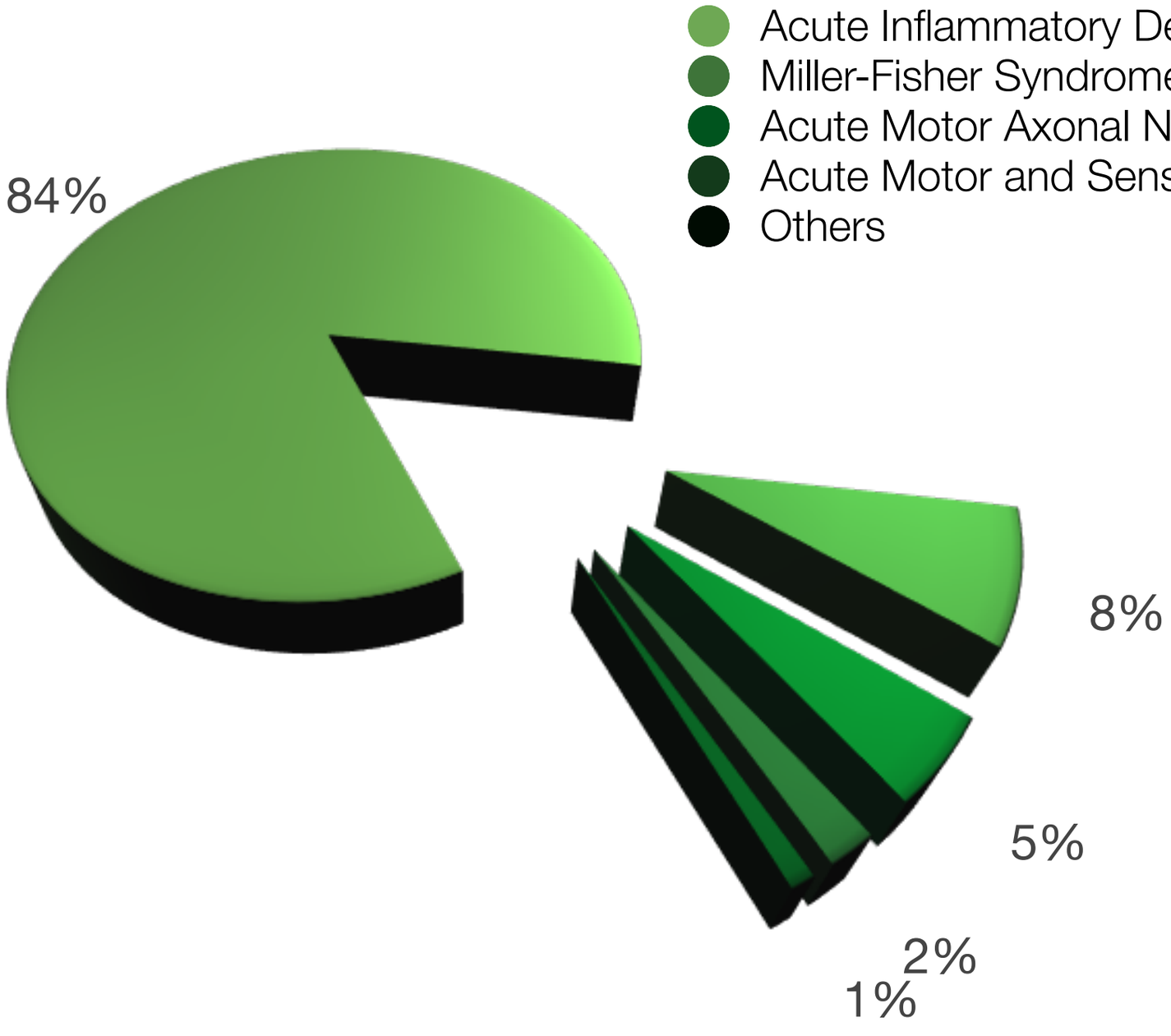

Acute Inflammatory Demyelinating Polyneuropathy (AIDP)

- 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

- Opthalmopegia, 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 inteferon 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 admision
- 35% need invasive ventilation

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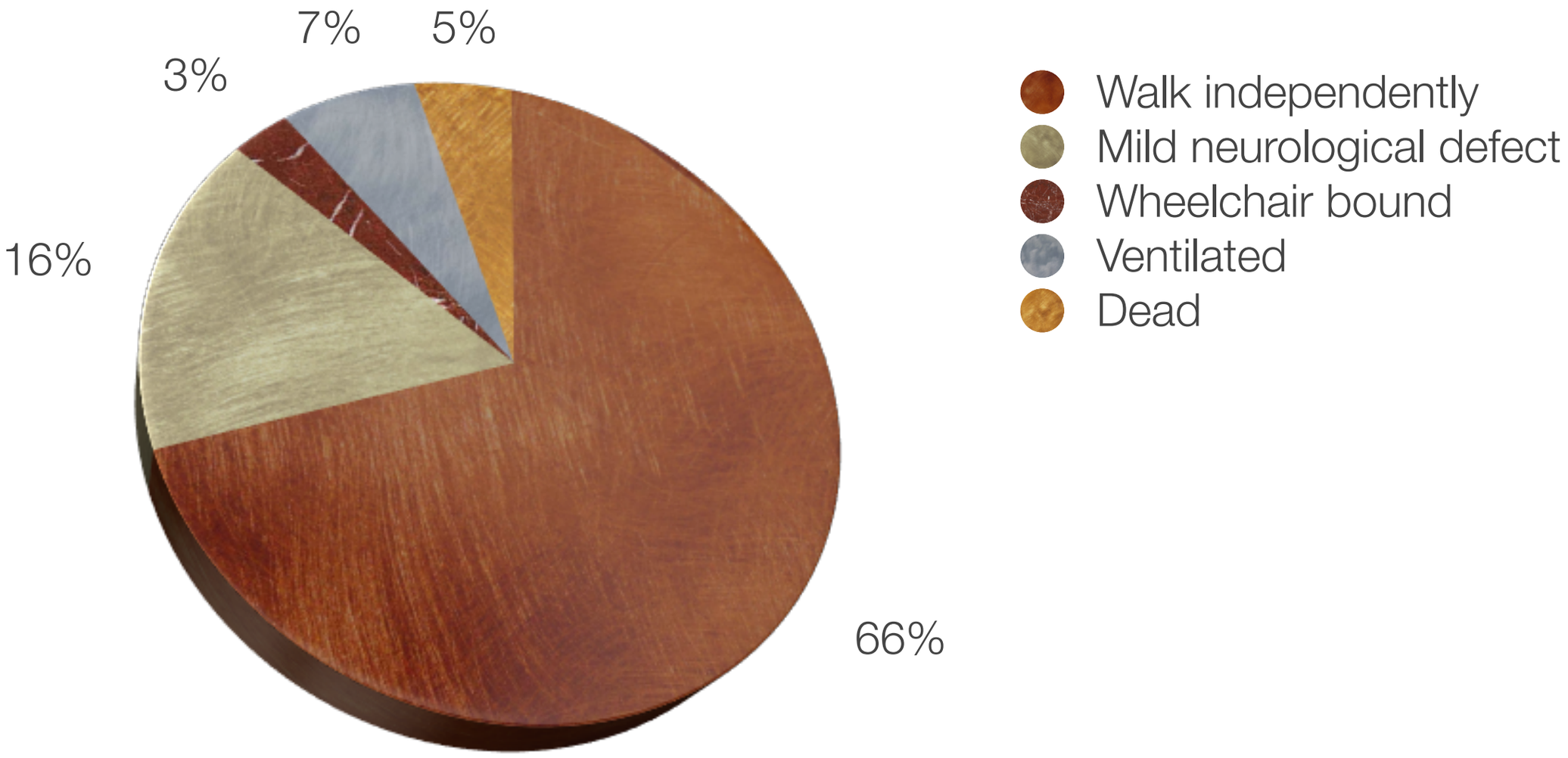
Poor Prognostic Features

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

Long Term Outcome



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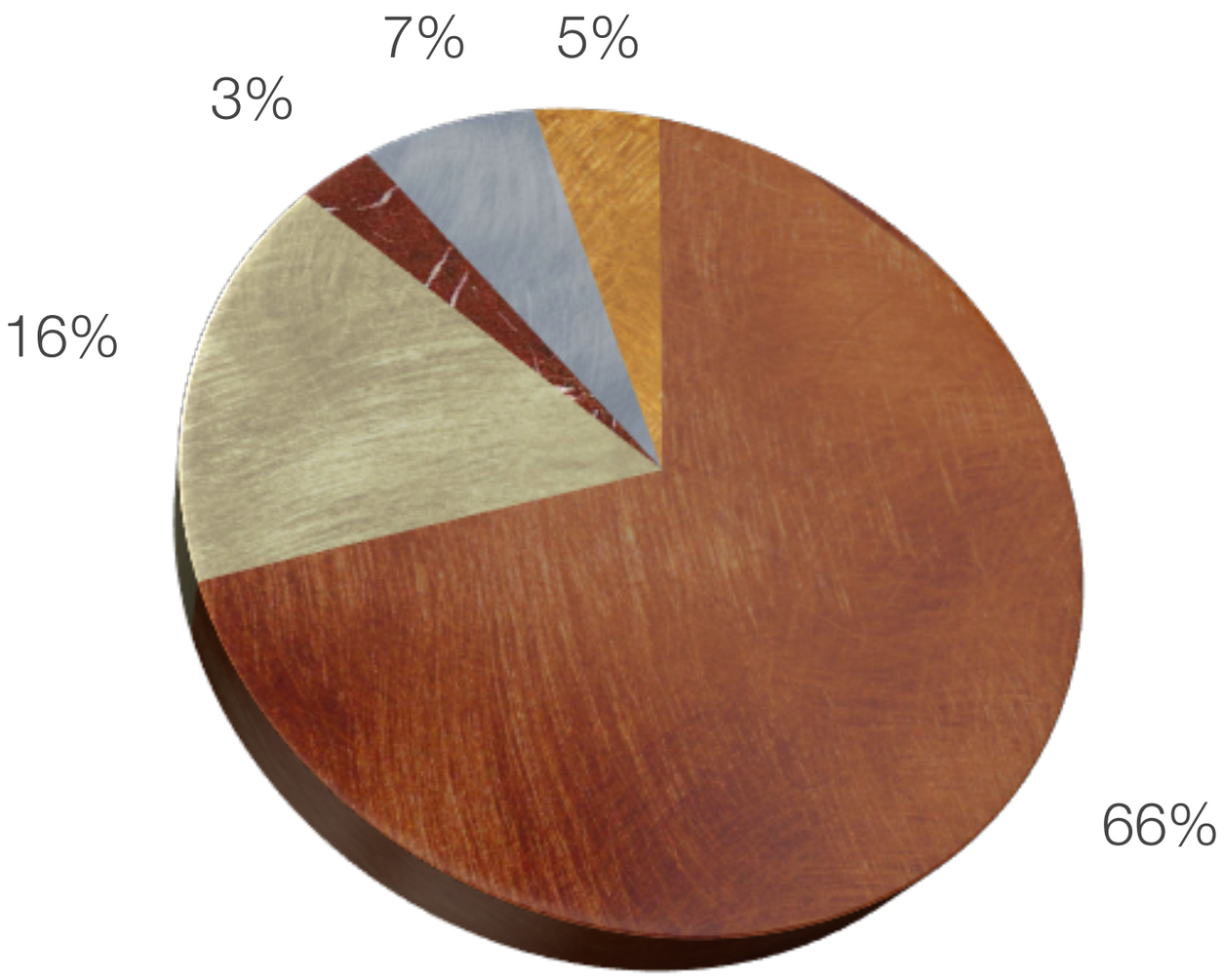

- Walk independently
- Mild neurological defect
- Wheelchair bound
- Ventilated
- Dead

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