

Synonym: acute inflammatory polyneuritis

Introduction

Peripheral demyelination and axonal degeneration, resulting in acute, ascending and progressive neuropathy. Characterised by weakness which may → respiratory failure, paraesthesiae, and hypo/areflexia. Sev autonomic dysfunction may also occur. Mort 2-5%. *Miller Fisher syndrome (MFS)* is a rare variant that presents with ataxia, ophthalmoplegia and areflexia with minimal weakness. Recovery usually <1-3 months. *Campylobacter* assoc.

Epidemiology

Commonest acute neuromuscular paralytic syndrome. Annual incidence ~1-2/100,000 pop. M>F. Biphasic age peaks: 15-35y and 50-75y

Risk Factors

~75% have a recent history of usually GI or respiratory infection, e.g:

- *Campylobacter jejuni*
- *Mycoplasma pneumoniae*
- EBV
- Hepatitis B
- CMV
- HIV

Other risk factors include:

- Vaccinations - live and dead vaccines have been implicated
- Malignancies: e.g. lymphomas, especially Hodgkin's disease
- Pregnancy - incidence ↓ in pregnancy but ↑ in postpartum months

Presentation

Features

- **Weakness:** Ascending, progressive & symmetric. Max sev @2wks & progression stops after 5wks. Facial weakness (Bell's palsy), dysphasia or dysarthria may develop.
- **Pain:** Neuropathic pain may develop, particularly in the legs. Also back pain.
- **Reflexes:** These may be reduced or absent.
- **Hypotonia**
- **Respiratory muscle paralysis**
- **Sensory:** Variable ascending symptoms & can include paraesthesiae and sensory loss.
- **Autonomic:** ↑HR, ↑BP, ↓sweating, ↓heat tolerance, paralytic ileus and urinary hesitancy.

Investigations

Diagnosis usually clinical.

Urine: Osm & electrolytes (SIADH)

Bloods: UEC, Osm (SIADH). Serology - *Campylobacter* & antiganglioside antibodies.

LP/CSF: ↑Protein (may be delayed). WCC<10 (mostly monocytes).

Spirometry: FVC sensitive to respiratory dysfunction & helps assess for ICU/intubation

Nerve conduction studies: Measurable slowing of nerve conduction may take 2-3 weeks.

MRI: shows enhancement of affected nerves

ECG: May be normal or 2° & 3° AV block, T-wave changes, ST depression, QRS widening, etc.

Management

- **Immunoglobulin** IV OR plasma exchange ↓recovery time by up to 50% - Cochrane r/v
- Supportive: 33% req intubation (VC<15ml/kg, PO2<70mmHg on RA, airway issues), Mx dysautonomia, DVT prophylaxis (TEDS/LMWH SC), analgesia for pain (**carbamazepine**, **gabapentin** may help). Steroids not beneficial.