APPROACH TO GBS

GBS - an acute, frequently severe, and fulminant polyradiculoneuropathy that is autoimmune in nature. It generally follow monophasic course, typically does not recur.

Common Subtypes

- Acute inflammatory demyelinating polyneuropathy (80-95%)
- Acute motor axonal neuropathy
- Acute motor and sensory axonal neuropathy
- Miller Fisher syndrome (can present upto 20%)
- Bickerstaff encephalitis
- Pharyngeal-cervical-brachial weakness

Uncommon subtypes

- Pure sensory GBS
- Facial diplegia and distal limb paresthesia
- Acute pandysautonomia,
- Bilateral lumbar radiculopathy
- Sixth nerve palsy and distal paresthesia

DIFFERENTIAL DIAGNOSIS

Cerebral

- Bilateral strokes
- Psychogenic symptoms

Cerebellar

- Acute cerebellar ataxia syndrome
- Posterior fossa structural lesion

Spinal

- Compressive myelopathy
- Transverse myelitis
- Anterior spinal artery syndrome

- Poliomyelitis
- Other infectious causes of acute myelitis (eg, West Nile virus, coxsackieviruses, echoviruses)

Peripheral nervous system

- Toxic neuropathy – Drugs/ Toxins
- Critical care neuropathy
- Diphtheria
- Tick paralysis
- Porphyria
- Lyme disease
- Vasculitis

Neuromuscular junction

- MG
- Botulism
- NM blocking agents

Muscle disease

- Acute viral myositis
- Acute inflammatory myopathies
- Metabolic – hypokalaemia/
- Periodic paralysis

HOW WOULD YOU DIAGNOSE?

Diagnostic Criteria for Typical Guillain-Barré Syndrome

Features required for diagnosis

- Progressive weakness of more than one limb
- Areflexia

Supportive features include:

- Progression of symptoms over days to four weeks

- Relative symmetry
- Mild sensory symptoms or signs
- Cranial nerve involvement, especially bilateral facial nerve weakness
- Recovery starting two to four weeks after progression halts
- Autonomic dysfunction
- No fever at the onset
- Elevated protein in CSF with a cell count <10/mm 3
- Electrodiagnostic abnormalities consistent with GBS

Features make the diagnosis of GBS doubtful:

- Sensory level (decrement or loss of sensation below a spinal cord root level as determined by neurologic examination)
- Marked, persistent asymmetry of weakness
- Severe and persistent bowel and bladder dysfunction
- More than 50 white cells in the CSF

CLINICAL FEATURES

- cardinal features are progressive, fairly symmetric muscle weakness accompanied by absent or depressed deep tendon reflexes
- Patients usually present a few days to a week after onset of symptoms.
- The weakness can vary from mild difficulty with walking to nearly complete paralysis of all extremity, facial, respiratory, and bulbar muscles.
- GBS usually progresses over a period of about two weeks.
- By four weeks after the initial symptoms, 90% of GBS patients have reached the nadir of the disease.
- Disease progression for more than eight weeks is consistent with the diagnosis of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)
- Approximately two-thirds of patients give a history of an antecedent respiratory tract or gastrointestinal infection Campylobacter jejuni infection is the most commonly identified precipitant of GBS
- In the absence of disease-modifying treatment, most patients with GBS show continued progression for up to two weeks, followed by a plateau phase of about two weeks, and then recovery of function over several weeks to months.

INVESTIGATION FOR DIAGNOSIS

Cerebrospinal fluid analysis

- albuminocytologic dissociation, is present in

  - 50 to 66% of patients with GBS in the first week after the onset of symptoms, therefore may be normal in earlier than one week

  - ≥75% of patients in the third week

- The elevated protein may be due to increased permeability of the blood-nerve-barrier at the level of the proximal nerve roots.

- CSF cell count is typically normal, ie, less than 5 cells/mm 3

- CSF pleocytosis is common in patients who have GBS and concurrent HIV infection

- concurrent HIV infection or an alternative diagnosis should be considered in patients with a CSF cell count >10/mm 3

Neurophysiology study

Demyelinating features at EMG

- earliest abnormalities - ***prolonged or absent F waves and absent H reflexes*** reflecting demyelination at the level of the nerve roots

- ***Increased distal latencies and conduction blocks with temporal dispersion*** of motor responses follow

- Sensory nerve conduction studies (NCS) reveal absent responses or slowed conduction velocities. ***Typically, the sural sensory response is normal***, while median and ulnar sensory responses are affected (sural sparing)

Antibodies

- In clinical practice, commercially available testing for serum IgG antibodies to GQ1b is useful for the diagnosis of MFS, having a sensitivity of 85 to 90 percent

- Antibodies to GQ1b may also be present in GBS with ophthalmoparesis, Bickerstaff encephalitis, and the pharyngeal-cervical brachial GBS variant, but not in disorders other than GBS

- Currently, laboratory testing for antibodies to glycolipids other than GQ1b is not performed routinely because of limited clinical utility

INVESTIGATION FOR MANAGEMENT

- FBC/ESR/
- Urea, Creatinine/Electrolytes
- LFT/ALT/AST/CKMB
- Viral screening esp HIV
- Stool culture and serology for *C jejuni*
- Stool culture for poliovirus in pure motor syndromes
- Acute and convalescent serology for cytomegalovirus, Epstein-Barr virus, Mycoplasma

Treatment

IV Immunoglobulin

- precise mechanism of action in GBS is unknown
- as effective as plasma exchange
- is recommended for nonambulatory adult patients with GBS who start treatment within two or possibly four weeks of the onset of neuropathic symptoms
- Intravenous immune globulin is given for five days at 0.4 gram/kg per day
- Side effects include
    1. aseptic meningitis,
    2. rash,
    3. acute renal failure (mostly related to sucrose containing products), and
    4. (rarely) hyperviscosity leading to stroke.
    5. IgA deficiency can lead to anaphylaxis

Plasma exchange

- is recommended for nonambulatory adult patients with GBS who start treatment within four weeks of the onset of neuropathic symptoms.
- is also recommended for ambulatory patients who start treatment within two weeks of the onset of neuropathic symptoms
- is usually given for four to six treatments over eight to 10 days
- main complications are
    1. hypotension,
    2. sepsis, and
    3. problems with intravenous access

Significance

- Treatment with plasma exchange or IVIG hastens recovery from GBS
- The beneficial effects of plasma exchange and IVIG are equivalent
- Combining the two treatments is not beneficial
- Glucocorticoid treatment alone is not beneficial

SUPPORATIVE CARE

Respiratory failure

- Vigilance is essential when caring for a patient with GBS, since deterioration due to progression of muscle weakness can occur rapidly.
- is common, and 15 to 30 percent of patients need ventilatory support. Close respiratory monitoring with frequent measurement of vital capacity and negative inspiratory force (NIF) is needed.

- **predictors of respiratory failure**

  - Time of onset to admission less than seven days

  - Inability to cough

  - Inability to stand

  - Inability to lift the elbows

  - Inability to lift the head

  - Liver enzyme increases

  At least four of these six predictors, mechanical ventilation was required in >85 percent

- **Indication for intubation**

  1. Forced vital capacity <20 mL/kg
  2. Maximum inspiratory pressure <30 cmH2O

3. Maximum expiratory pressure <40 cmH2O

Autonomic dysfunction

- Dysautonomia occurs in 70 percent of patients and manifests as symptoms that include tachycardia (the most common), urinary retention, hypertension alternating with hypotension, orthostatic hypotension, bradycardia, other arrhythmias, ileus, and loss of sweating
- close monitoring of blood pressure, fluid status, and cardiac rhythm is essential
- Sustained sinus tachycardia occurs in 37 percent of patients and requires no treatment

Pain control

- Neuropathic pain occurs in about 40 to 50 percent of patients during the course of GBS and often requires treatment.
- Gabapentin or carbamazepine may be used for intensive care unit pain control during the acute phase of GBS

Others

- Prophylaxis for deep vein thrombosis, bladder and bowel care, physical and occupational therapy, and psychological support are essential.

INDICATION FOR ICU ADMISSION

- Respiratory failure
- Arrhythmia
- Dysautonomia
- Pharyngeal weakness
- Tachypnea
- Can't count upto 20
- Vital capacity – 1.5L

Causes of prolong stay in ICU

- Aspiration pneumonia
- Critical illness neuropathy
- Septic encephalopathy
- Malnutrition

Lethal complication of GBS

- Respiratory muscle weakness
- Arrhythmia
- Aspiration pneumonia &ARDS
- DVT & PE
- Pneumothorax (rare)

Prognosis

- 80% completely recover within 3-6 months
- 5% Mortality
- 10% Disablity
- 10% Recurrence
- Ptients who become ventilator dependent, about 20 percent will die

Poor prognosis factors for recovery of GBS

- Older age
- Rapid onset (less than seven days) prior to presentation
- Severe muscle weakness on admission
- Need for ventilatory support
- An average distal motor response amplitude reduction to <20 percent of normal
- Preceding diarrheal illness

Difference between neuropathy and myopathy

Neuropathy – decreased or absent reflex and sensory impairment

Myopathy - intact reflex and normal sensory

Difference between Cauda Equina $ and Peripheral neuropathy?

CE$ - sacral anaesthesia with or without sphincter involvement

PN - glove and stocking pattern with no sphincter

How to differentiate Conus medullaris and CE$?

Conus Medullaris

- UMNL
- bilateral saddle anaesthesia
- bladder and bowel dysfunction
- impotence
- absent anal reflex
- absence of lower extremity abnormality

CE$

- LMNL
- Low back and radicular pain
- asymmetrical leg weakness
- Variable areflexia
- Sparing of bowel and bladder function

Aetiological agents

Respiratory	- Mycoplasma
	- CMV
	- EB/VZ/HIV
GI	- C.jejuni
Others	- H influenza
	-Borrelia Burgcloferi
	- Parainfluenzae
	- Vaccine

FACTS

Causes of pure motor neuropathy

- CIDP
- Lead
- Porphyria
- Diptheria
- Drugs – Dapsone

Causes of palpable nerve

- Acromegaly
- Amyloidosis
- HMSN
- Leprosy
- Neurofibromatosis

Causes of Demyelinating polyneuropathy

- Chronic inflammatory demyelinating polyradiculoneuropathy
- Multifocal motor neuropathy
- Charcot–Marie–Tooth disease type I and type X
- Paraprotein-associated demyelinating neuropathy (multiple myeloma)
- HIV

Causes of Axonal polyneuropathy

- Diabetes mellitus
- Alcohol
- Uraemia
- Cirrhosis
- Amyloid
- Myxoedema
- Acromegaly
- Paraneoplastic

- Drugs and toxins
- -Vitamin deficiency (B1.6.12. E)
- Idiopathic
- Infection (HIV, Leprosy, Brucellosis)
- Hereditory

Causes of autonomic neuropathy

- DM
- GBS
- Botulism
- HIV
- Amyloidosis
- Chagas disease

Causes of Proximal Myopathy

Inflammtory

- Polymyositis • Dermatomyositis
- Inclusion body myositis (additional distal effects)

Endocrine and metabolic

• Hypothyroidism • Hyperthyroidism • Acromegaly
• Cushing's syndrome • Addison's disease
• Conn's syndrome • Osteomalacia
• Hypokalaemia • Hypercalcaemia

Toxic

- Alcohol (chronic and acute syndromes)
- Amphetamines/cocaine/ heroin
- Vitamin E
- Organophosphates
- Snake venoms

Drugs

• Corticosteroids • Statins
• Amiodarone • Beta-blockers
• Opiates • Chloroquine
• Ciclosporin • Vincristine
• Clofibrate • Zidovudine

Paraneoplastic

- Carcinomatous neuromyopathy
- Dermatomyositis