An Introduction to Guillain-Barré Syndrome

(Acute inflammatory Demyelinating Polyneuropathy AIDP)

Neuro ICU
Bitesize

Learning Outcomes

To Discuss the pathology of GBS Progression of illness and the clinical features

To develop an awareness of potential complications with a GBS Patient

To improve knowledge to improve quality of care for GBS patients from a holistic perpesctive

Introduction

Guillain-Barré Syndrome (GBS),

- A disorder in which the bodies immune system attacks parts of the peripheral nervous system
- Acute generalised flaccid paralysis

The bodies immune system the goes wrong and it attacks and damages the nerves

It is not fully understood why this happens, but it can be the result of

An Infection

- Such as food poisoning Flu Or CMV
- HIV or AIDS

A Vaccination

- Ie The flu Vaccination

Surgery or a medical procedure

Demographics

- UK Incidence 1 - 2 per 100,000 population(Layon et al 2004)
- It Can affect anyone at any Age
- Not Contagious
- It is not known why it strikes some people and not others but men are 1.5 times more likley to have GBS

GBS

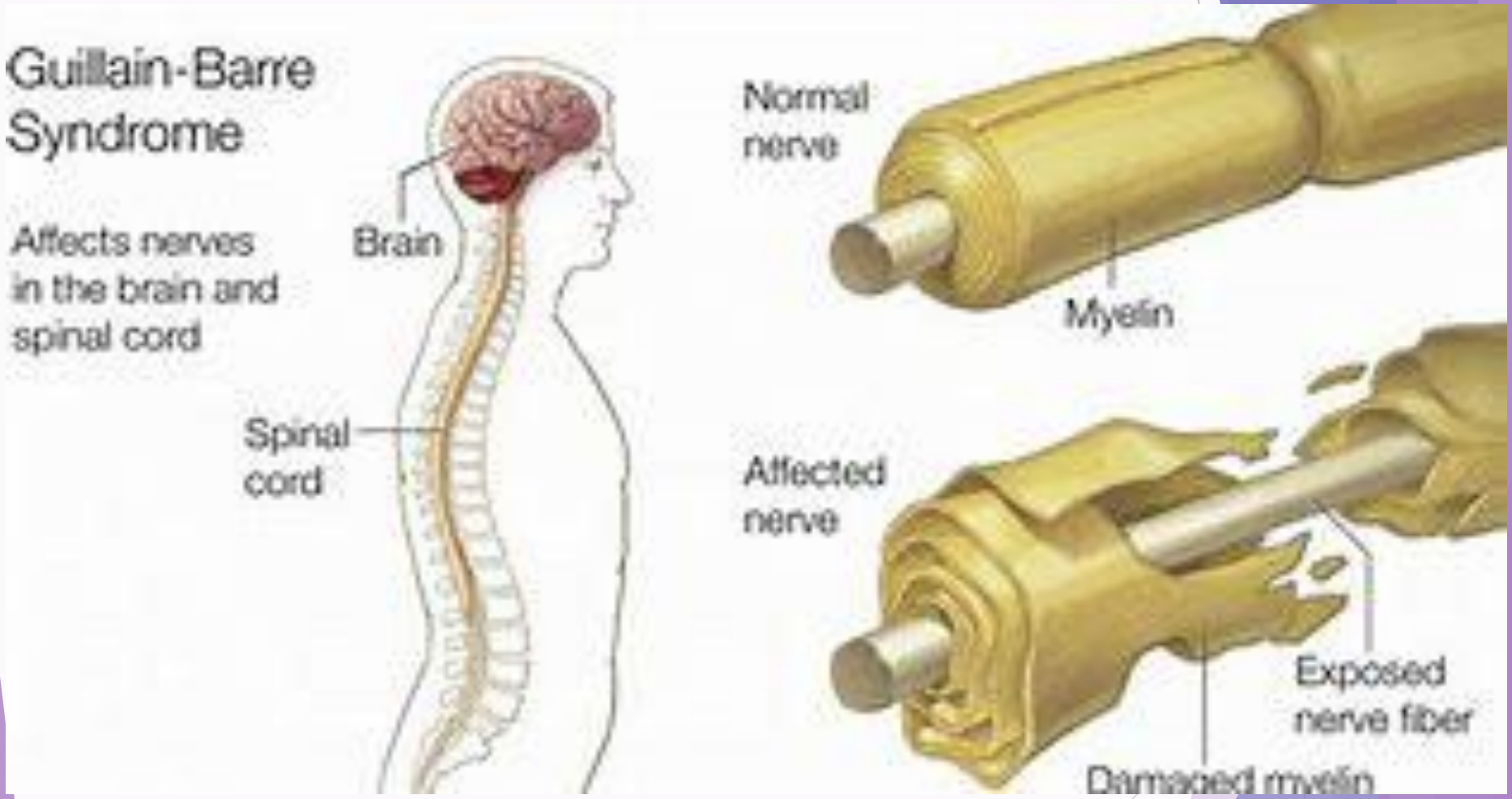
Acute onset with rapid progression.

Potentially Life Threatening

The immune system destroys the myelin sheath that surrounds the axons of many peripheral nerves

Nerves cannot transmit signals effectively:

- results in an inability of the brain to interpret textures ie heat and Pain
- Affects motor component of peripheral nerves (cranial & spinal).


Guillain-Barre Syndrome
Affects nerves in the brain and spinal cord
Brain
Spinal cord
Normal nerve
Myelin
Affected nerve
Exposed nerve fiber
Damaged myelin

Clinical Presentation

Sensory with distal paraesthesia.

- Tingling or prickly feeling in your fingers and toes
- Pain and Numbness

Weakness and paralysis follow usually this occurs in an ASCENDING pattern. Feet to Face – symmetrical.

- Patients are usually at their weakest by week 3

Clinical Features

Reflexes absence of normal reflexes
- hyporeflexia or areflexia.

bulbar weakness

Opthalmolegia,
- complete or partial Ptosis

respiratory muscle weakness

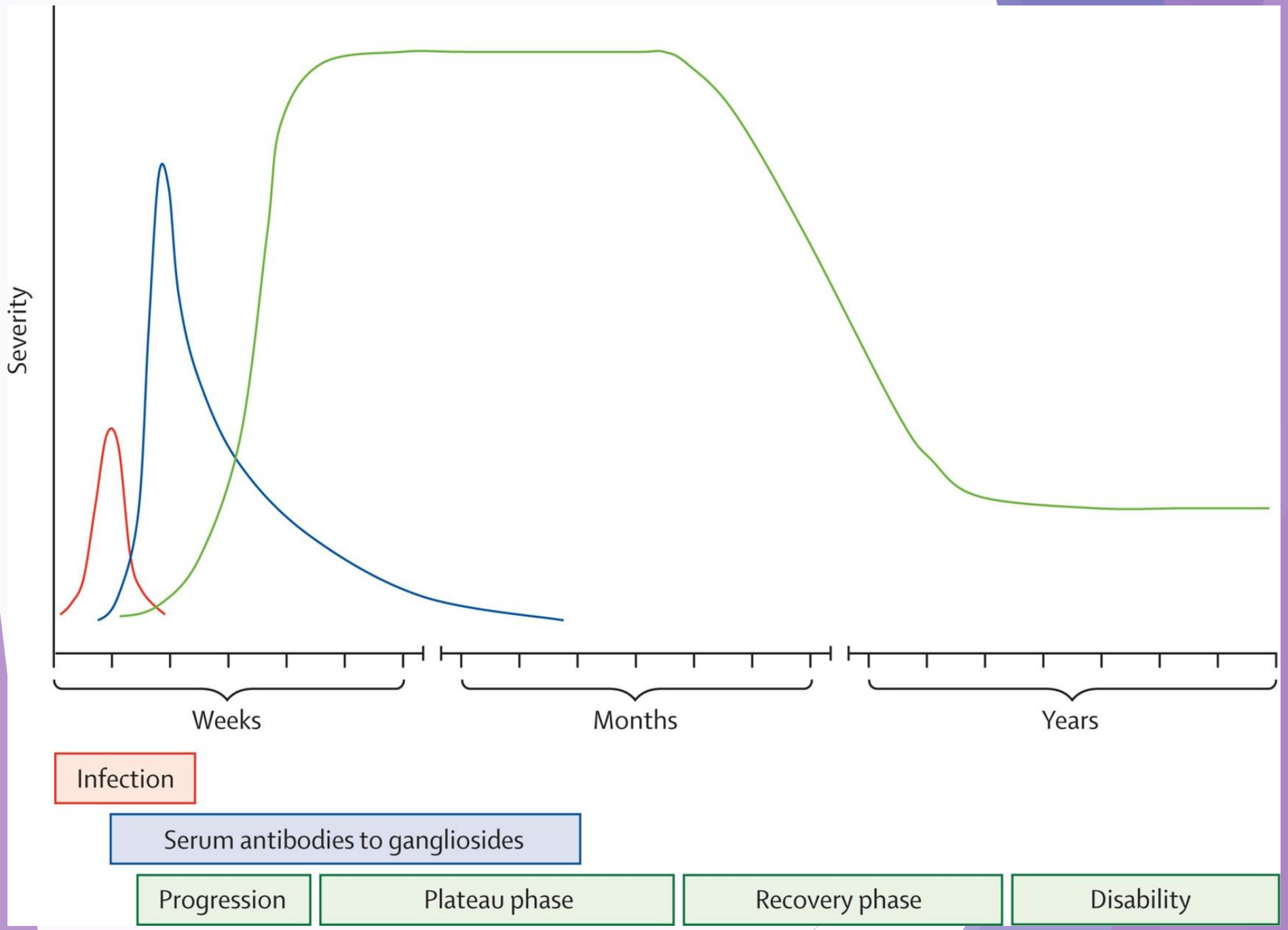
autonomic involvement (BP HR)

Facial weakness

pain

Can appear comatose with absent brainstem reflexes

Sensory impairment – light touch, pin prick, vibratory sensation, proprioception


Severity
Weeks
Months
Years
Infection
Serum antibodies to gangliosides
Progression
Plateau phase
Recovery phase
Disability

Variants

Motor-sensory GBS

Pure motor GBS

Miller-Fisher Variant (MFS)

- Which affects the cranial nerves and not Limb weakness
- Another variant of MFS is Bickerstaffs brain stem encephalitis (BBE) Which causes an altered level of consciousness

Bulbar Variant

Primary Axonal GBS

- Axons are effected and not the Myelin Sheath
- 2 variants
  - Acute Motor Axonal Neuropathy (AMAN)
  - Acute Motor Sensory Axonal Neuropathy (AMSAN)

CIDP (Chronic Inflammatory Polyradiculoneuropathy)

Diagnosis & Investigations

History need to exclude other causes of progressive weakness e.g. hypokalaemia, tetanus, myasthenia

- Is it Symmetrical

EMG studies

- reduced nerve conduction

Lumbar puncture for CSF examination

- raised protein count (>0.5g/L). Normal cell count.

Reflexes such as knee jerks are lost

Prognosis

Disease can progress rapidly within 24-72 hours from onset to quadriparesis and respiratory paralysis.

65% recover by end of one year.

30% require mechanical ventilation.

Maximum neurological deficit occurs by 2 – 3 weeks from onset.

Recovery occurs over weeks to months and up to year.

Around 5-8% mortality due to complications associated with

- respiratory failure,
- sepsis,
- PE, respiratory/cardiac arrest
- co-morbidities

80% fully recover

- Therefore 20% will be left with a lasting weakness

Medical Treatment

Mainly supportive aiming to prevent complications.

Effective treatments to improve recovery;

Intravenous Immunoglobulin Ig (IVIg) usually for 5 days started as soon as diagnosis is made.

Plasmapheresis daily for 4 – 5 days and may need a second course if relapse.

Management

Respiratory support	ETT, Trache, Ventilation
Cardiovascular support	fluids, monitoring
Analgesia	Pain frequently underrated often worse at night
Sleep deprivation common.	
Nutrition	NG tube until bulbar function recovers
Maintain limb flexibility	

Respiratory Management

Vital Capacity measurements to detect deterioration of respiratory function

- VC's of <15ml/kg or less than 1L require intubation.

Elective intubation for signs and symptoms of respiratory distress

Do not wait for respiratory decompensation.

Respiratory Management

Tracheostomy early in patients with rapid onset.

Weaning as respiratory muscles recover usually 3 -4 weeks post onset. Diaphragm muscle recovery before limbs usually.

Cardiovascular Management

Autonomic dysfunction is a significant cause of morbidity and mortality.

- 3-10% of cases

Blood pressure fluctuates with hypotension and hypertension within minutes. Caused by impaired baroreceptor buffering.

Hypotension treated with fluid boluses and head down position.

Beta blockers should be avoided due to increased risk of cardiac arrest.

Hypertension (ongoing) can cause patients with pre-existing cardiac disease can develop congestive cardiac failure and ischaemia.

Cardiac arrythmias

Sinus Bradycardia or Tachycardia.

Sinus Bradycardia, Sinus arrest and atrioventricular block can occur during intubation and suctioning.

Complete heart block can occur and may require temporary pacing.

Lack of vagal stimulation can cause Ventricular Tachycardia.

Pain Management

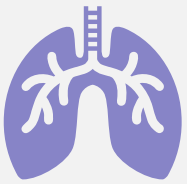
Neuropathic pain is often severe and may require opiates for relief.

Most patients will require regular analgesia, paracetamol, NSAID's

Other drugs used to control pain

- Tricyclic antidepressants used for pain given at night by altering the pain threshold.
- Anti-epileptic agents e.g. Gabapentin and carbamazepine - inhibit GABA uptake at receptor sites.

Neurological Observations



If ventilated may only be sedated 24-48 hours.

GCS baseline and then review.

No need for hourly observations on an on going basis when diagnosis established.

Assess motor strength and sensation.

Autonomic Dysfunction

- Cardiovascular – monitoring and support.
- Bladder most will require a catheter.
- Bowel – stool softeners, suppositories etc.
- Paralytic ileus – NG tube, free drainage

Psychosocial Management

Patients with GBS especially if quadriplegic and on a ventilator are unable to communicate normally, if at all therefore may experience;

Feelings of helplessness, fear, anxiety,

Isolation, depression, sadness.

Difficult to understand as facial paralysis

makes even grimacing to pain, frustration impossible.

Case Study
BOB

45 years old

Weight 85 kg
height 6ft

Has been unwell for the last 2 weeks with flu and was admitted to kent ward with limb weakness

He has now developed a respiratory weakness and has been admitted to the NICU for respiratory support his VC on the ward is 500mls

Respiratory rate is 35 b/min

Unable to speak a full sentence

Sats 89% on 15L non rebreathe mask

He is very anxious and distressed

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What do you think bob needs and what do you need to do?

ⓘ Start presenting to display the poll results on this slide.

Bob's Initial needs

Ventilation

Check the bedspace has emergency eq ie adjuncts, Green bag and mask, working suction

Prepare the ventilator

Prepare Intubation drugs and sedation drugs for after ventilation

Check that the difficult intubation trolly has been checked and it complete

Ensure that NIC and reg are aware of the severity of Bobs breathing

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What do you need to think about now Bob is ventilated?

ⓘ Start presenting to display the poll results on this slide.

Bobs Needs After Intubation

Airway Check cuff pressure	Breathing , Suctioning, Sat up	Cirulation , Bp Parameters? M HR Teperature Electroyte management , Teperature	, Neurological Assessment how often?
Is he on enough sedation?, What pain relief is he on? Is he comfortable	Gut Protection	DVT Prolaxsis	Mouth care and eye care
Limb position, needs OT and Physio	Communication he will need SLT	Nutrition Dietition referral needs an NGT	Intravenous Immunoglobulin or plasma Exchange

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Bob 5 days Later.....
What do you consider to be Bobs needs and Priorities for his care?

ⓘ Start presenting to display the poll results on this slide.

Bobs Care Priorities after 5 days in ICU

Chest management
- Assisted cough
- sputum clearance,
- suctioning,
- consider if he needs 5% saline
- Early Trache

Cardiac Stability
- HR
- BP
- Temperature
- Is he showing any signs of sepsis

Pain, what analgesia is he prescribed

Sedation, is he comfortable

GCS? How often would you do this and would you cause central pain

Mouthcare and Eye care

Reassurance and communication

? Music

Family Visits

Day and night rhythm, Sleeping tablets

Antidepressants

Bowel care

Monitor for signs of emergence
- Shoulder shrug when coughing

2 Weeks Later

Bob Now has a trache and he is off sedation

You notice that bob has started to intermittently trigger the ventilator

What are bobs care needs are?

Bob 2 Weeks Later

Trache site and position	Respiratory rate and sputum clearance, ? Start slow ventilatory weane	Cardiovascular , monitor for infection and dehydration, check electrolytes and temperature	Neurological, Daily GCS do not cause pain
Stimulation, TV Radio what does he like to watch/ listen to ,	If managing to sit him out on a chair take him off the ICU (careful not to weane and mobilise together)	Consider Side room	Mouthcare and eye care
Repositioning and Limb stretches , splints	Reassurance	Communication aids	Day and night Routines ? He may have a personalised care plan

Bob Months Later

Bob comes back to the ICU he is well on way to making a full recovery

Resources

- Guillian-Barré Support Group www.gbs.org.uk
- This is an excellent resource for family, friends and the patient. Booklets for download and helpline to speak to someone directly.

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