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DIAGNOSIS AND RESPIRATORY MONITORING OF THE NEUROMUSCULAR PATIENT

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INTRODUCTION

Patients with neuromuscular disease may develop respiratory failure on account of weakness of respiratory muscles:

- diaphragm and accessory muscles of inspiration
- Abdominal muscles involved in forceful expiration (cough impairment)

Respiratory failure also due to hypotonia of bulbar muscles (facial, swallowing), and decreased central respiratory drive

NEUROMUSCULAR DISEASES

Although there are numerous neurodegenerative diseases we will focus on those that commonly result in respiratory emergencies

NEUROMUSCULAR DISEASES...

- **ALS** - Amyotrophic Lateral sclerosis – *Lou Gehrig's Disease*
  - Irreversible degenerative and progressive muscle dysfunction ultimately leading to respiratory failure and death
- Spinal Muscular Atrophy
  - Causes generalized respiratory and bulbar weakness, severely impacting a patient's ability to clear secretions, leading to acute respiratory failure.

NEUROMUSCULAR DISEASES…

- Myopathies: Oculorpharyngodistal, Mitochondrial, Inflammatory, Proximal, etc.…
- Myotonic and Muscular Dystrophy
- Pompe Disease
- Guillain-Barre
- Myasthenia Gravis

} *Most common cause of neuromuscular respiratory emergencies*

RESPIRATORY COMPLICATIONS IN NEUROMUSCULAR DISEASE

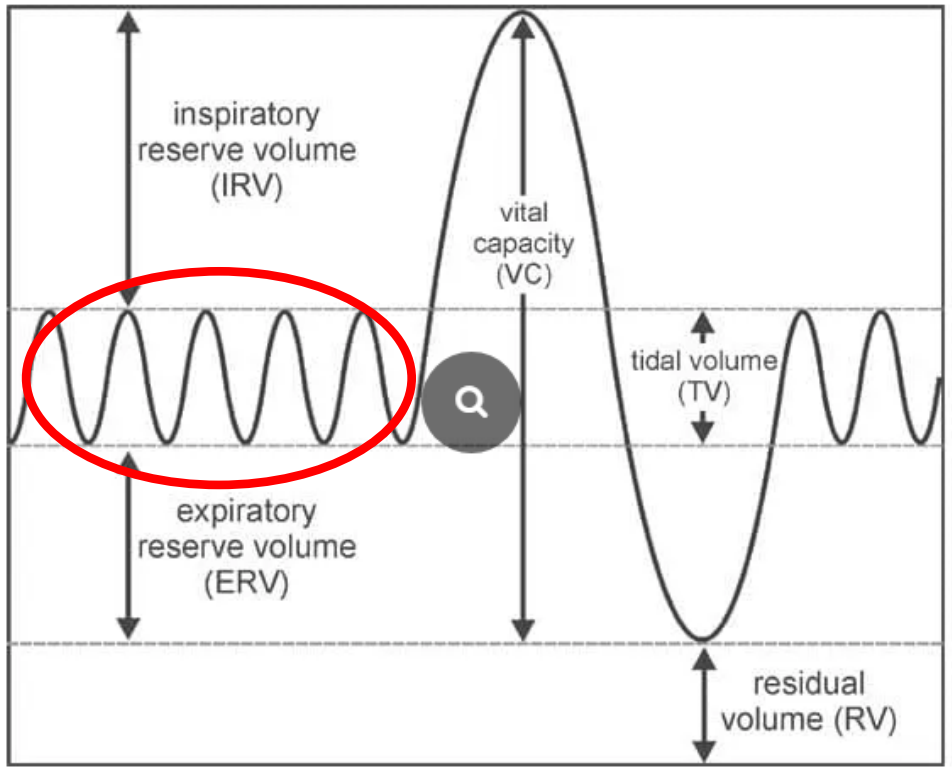
Table 2 Assessment for respiratory muscle weakness

Assessment	Clinical history	Physical exam	Pulmonary function testing	Sleep testing	Thoracic imaging
Bulbar weakness	Aspiration, drooling, voice change, post prandial cough	Change in appearance, drooling	Difficulty with testing due to poor mouth seal. False low values.		Aspiration
Diaphragm/ inspiratory muscles	Orthopnea, dyspnea on bending or immersion, sleepiness, morning headaches, decreased stamina speaking	Sleepy, increased respiratory rate, shallow breathing, orthopnea, accessory muscle use	Decreased forced vital capacity, decreased inspiratory pressure, decreased sniff nasal pressure, postural drop in forced vital capacity	REM or sleep hypoventilation	
Expiratory muscles	Recurrent infections, weak cough	Decrease in cough volume			
Not specific	Dyspnea	General respiratory exam	Decreased peak flow, decreased peak cough flow		Low lung volumes, pneumonia, pulmonary embolism

Abbreviation: REM, rapid eye movement.

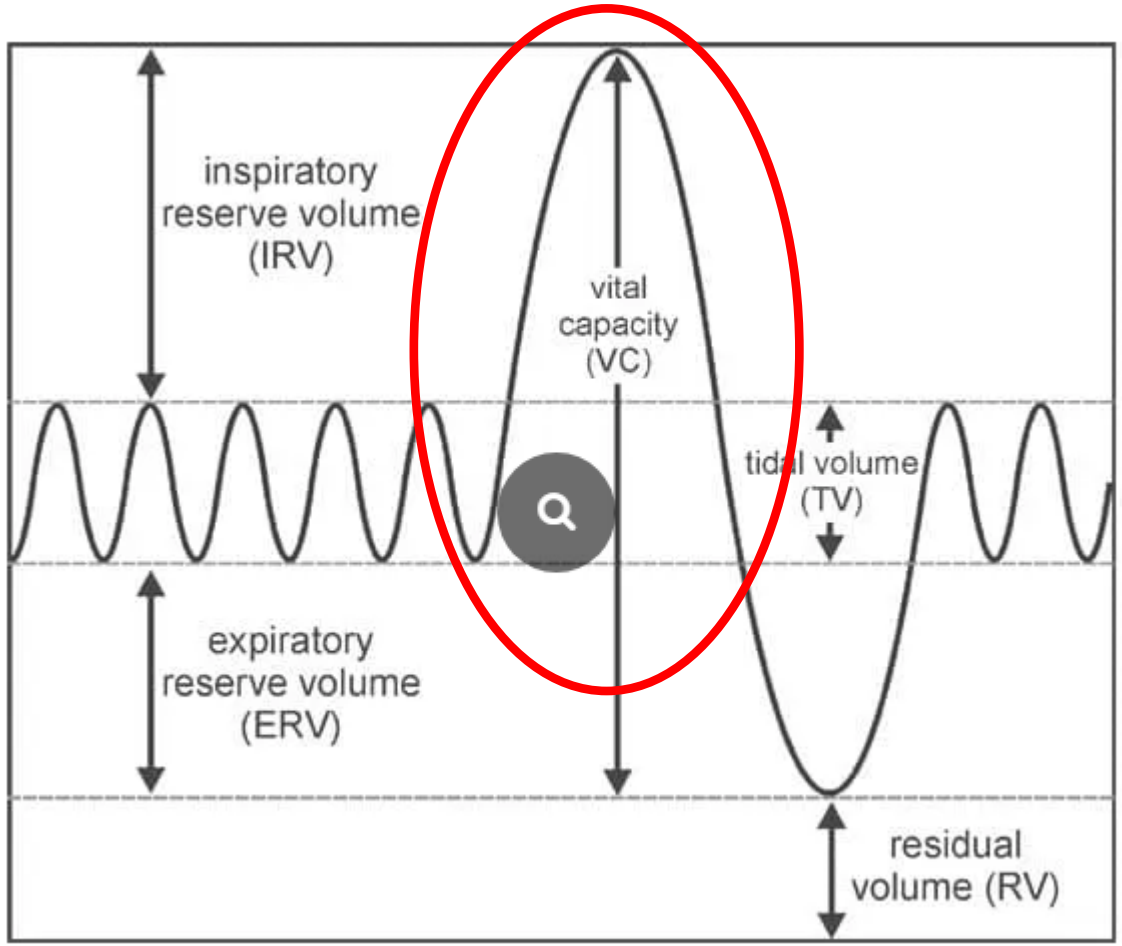
RESPIRATORY COMPLICATIONS- LOSS OF TIDAL VOLUMES

- Muscle weakness leads to impairment to creating effective volumes:
  - **Tidal Volume** is normally twice or three times anatomic deadspace of 1 ml/lb
    - For example a tidal volume of 500ml has Anatomic deadspace of 150ml & an Alveolar volume of 350ml.
    - When alveolar volume is less than anatomic deadspace, respiratory failure is imminent (for example, a tidal volume of 250ml would be 150 Vd and 100Av)
    - Decreased Vt causes increase in $PaCO_2$, patient attempts to compensate by increasing the RR leading to "rapid shallow breathing phenomenon"



RESPIRATORY COMPLICATIONS – VC & NIF

- **Vital Capacity** (the maximum useable volume of the lungs = Total lung capacity – residual volume)
  - A Vital capacity of < 15ml/kg is concerning
  - Bedside VC measurement is a useful tool in determining the degree of respiratory involvement
  - May be inaccurate in patients with facial paralysis that accompanies certain neuromuscular diseases (e.g. myasthenia gravis
- **NIF** – Negative Inspiratory Force indicates muscle strength



OTHER RESPIRATORY COMPLICATIONS...

Aspiration – swallowing trouble in MG

Pulmonary embolus – from immobility

Pneumonia – impaired bronchial hygiene and retained secretions

Sleep disturbances

orthopnea

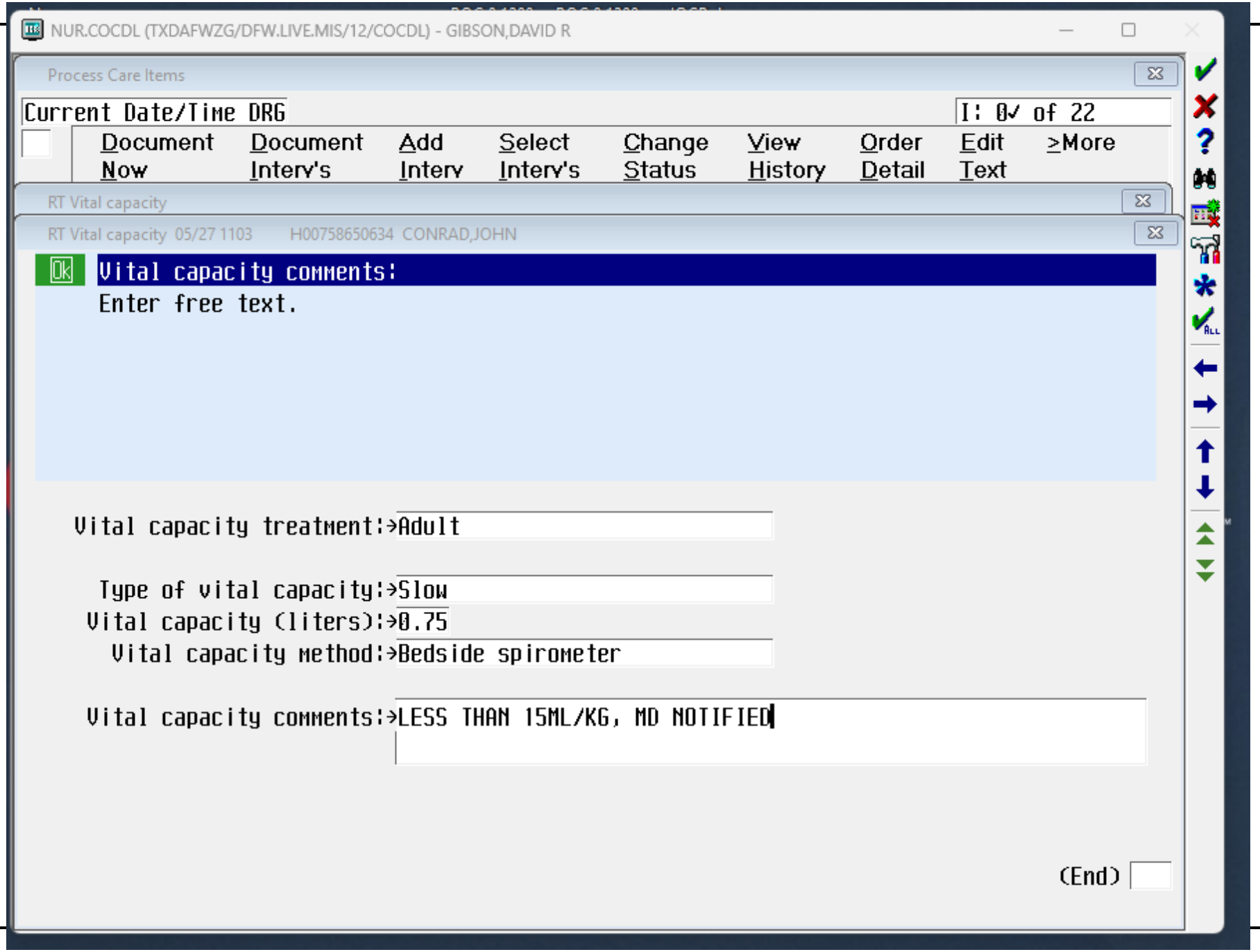
HOW TO MEASURE VITAL CAPACITY & NIF

“FVC” MEASUREMENT

- Patient must be able to maintain a seal around mouthpiece of results may be invalid.
- 3 attempts, which should be within 150ml of each other, take the best of the three.
- A full exhalation after the deepest breath, blow out for at least 6 seconds
- Not necessary to force the air, SVC is assessed even though the unit labels it as an FVC.

NIF

- < 30 cmH2O cause for concern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DISEASES IN WHICH RAPID RESPIRATORY DETERIORATION IS POSSIBLE

Myasthenia Gravis

Guillain-Barre (AIDP -Acute inflammatory demyelinating polyneuropathy)

MYASTHENIA GRAVIS

MYASTHENIA GRAVIS FACTS

Autoimmune disease of the neuromuscular junction causing muscular weakness

Bulbar (face) and respiratory muscles are commonly involved

14% of Myasthenia Gravis patients had respiratory failure as the initial presentation

Weakness, unlike Guillain-Barre which starts in the extremities, starts centrally and moves outward to extremities

No permanent cure although treatment provides manageable disease progression and significant symptom relief.

MYASTHENIA GRAVIS DIAGNOSIS AND TREATMENT

Diagnosis

- Diagnosing Myasthenia Gravis (MG) involves a combination of your clinical history, physical exams, blood tests, and electrodiagnostic tests.
- Tensilon test is no longer used to diagnose MG

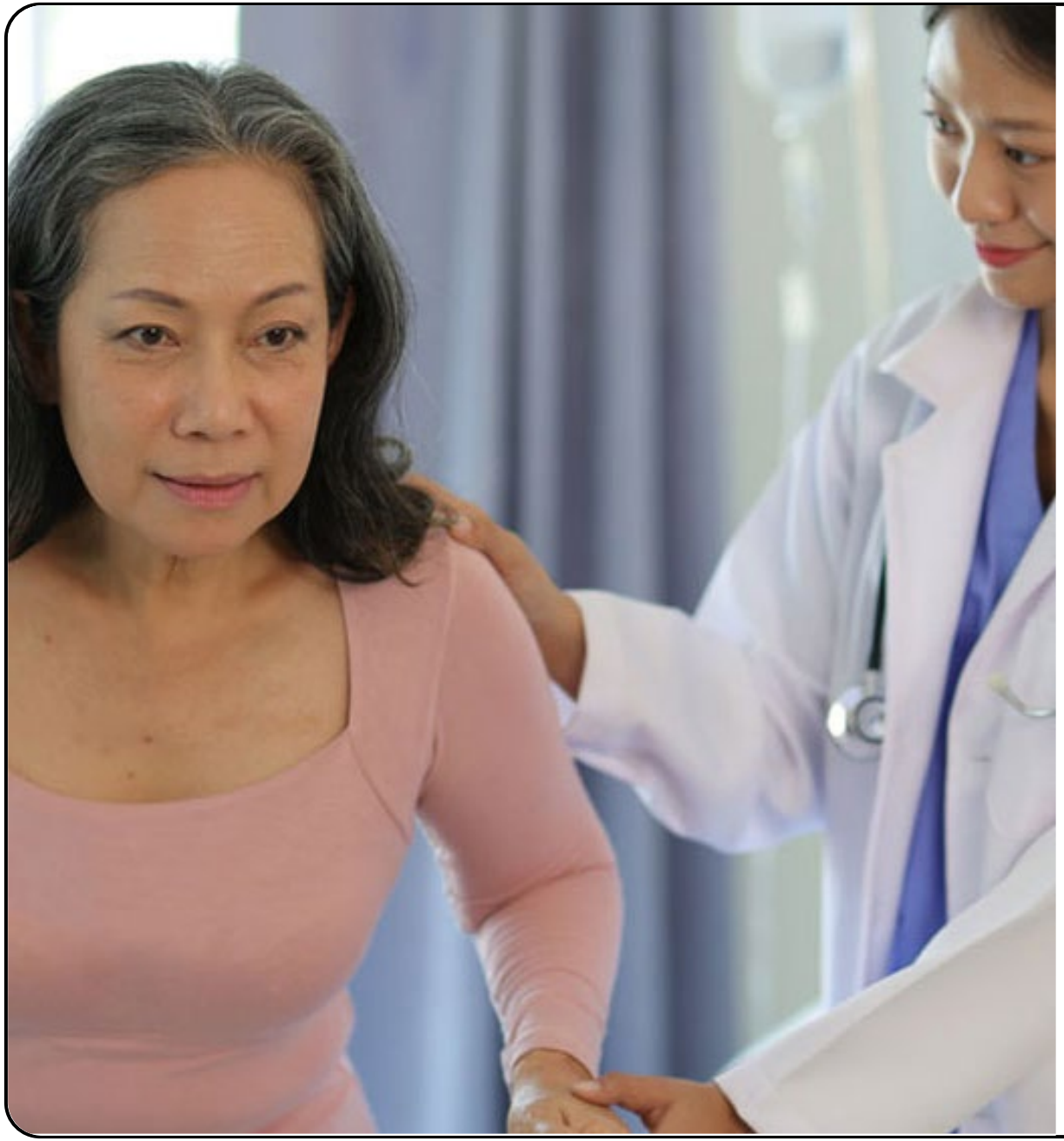
Treatment

- Drugs: Cholinesterase inhibitors (pyridostigmine), Corticosteroids, Immunosuppressants
- Plasmapheresis
- Lifestyle modifications

GUILLAIN-BARRE

GUILLAIN-BARRE FACTS

- Most common cause of paralysis
- More common in males
- Presents with weakness and sensory signs in the legs
- Progresses to arms then cranial muscles
- Disease progression can be rapid and about 20% of patients with GBS develop respiratory failure and require mechanical ventilation support
- May affect the Autonomic nervous system causing cardiac arrhythmias and BP instability



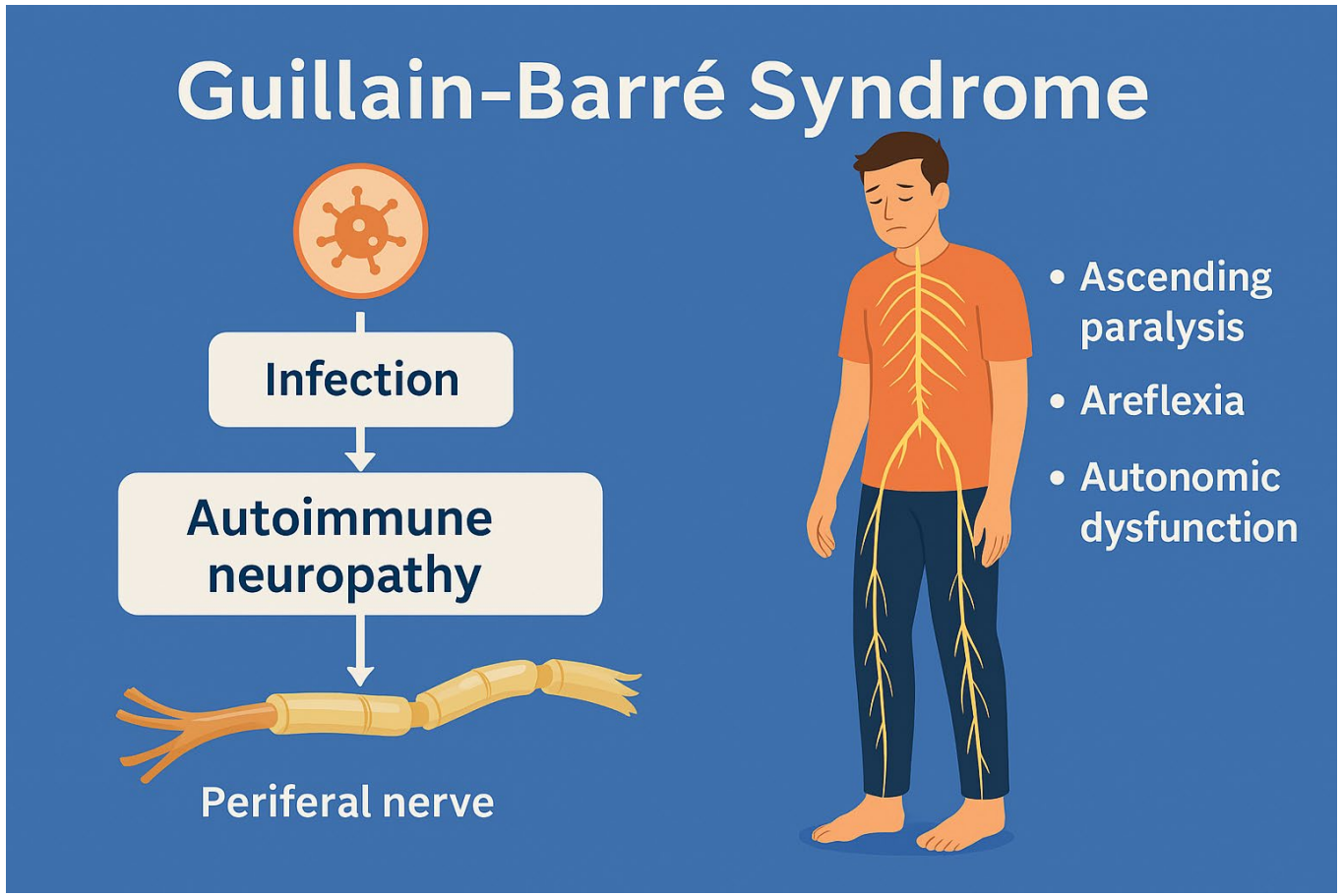
GUILLAIN-BARRE COURSE & PROGNOSIS

1. Paralysis may last for days, weeks, or months
2. Chronic GBS may occur is certain individuals with some degree of muscle weakness lasting for years
3. 80% regain the ability to walk at 6 months
4. Reoccurrence may occur years later

GUILLAIN-BARRE CAUSE

Thought to be caused by an aberrant immune response to infections (auto immune) resulting in damage to the peripheral nervous system.

Certain virus outbreaks have been associated with GBS such as the Zika virus



GUILLAIN-BARRE DIAGNOSIS

- Electrophysiology studies establish evidence of dysfunction and help distinguish between sub-types:
  - Acute Inflammatory demyelinating polyradiculoneuropathy (AIDP) Most common form
  - Acute motor axonal neuropathy (AMAN)
  - Acute motor sensory axonal neuropathy (AMSAN)
- CSF examination: Increased protein level with normal cell count (for example, not an increased number of WBCs)
- Diagnosis of exclusion: Rule out other causes of paralysis
- Watch for atypical presentation (e.g.. One sided weakness, upper instead of lower limbs)

GUILLAIN-BARRE TREATMENT

- Supportive care
  - DVT prophylaxis
  - Routine bronchial hygiene therapy to prevent respiratory infection
  - Ventilatory support
- IV immunoglobulin therapy
- Plasma exchange
- 40% of patients receiving treatment don’t seem to respond although GBS could have been worse without treatment

WHEN TO ICU?

- **Respiratory Compromise & The 20/30/40 Rule:** Imminent respiratory failure is the primary reason for ICU care. Admission is required if:
  - Forced Vital Capacity (FVC) drops below 20 ml/kg
  - Maximal Inspiratory Pressure (MIP) is not at least -30 cmH2O
  - Maximal Expiratory Pressure (MEP) falls below 40 cmH2O.
- **Rapid Progression:** A rapidly worsening condition or a precipitous drop in pulmonary function tests (e.g., a decrease of > 30% in 24 hours).
- **Bulbar Palsy:** Severe lower cranial nerve involvement leading to dysphagia, inability to clear secretions, or risk of aspiration.
- **Severe Dysautonomia:** Labile blood pressure, cardiac arrhythmias, or extreme fluctuations in heart rate.
- **Severe Motor Weakness:** Patients who are bedbound, cannot lift their arms, or have profound neck flexor weakness.

RT INTERVENTIONS FOR NEUROMUSCULAR DISEASE PATIENTS

1. Frequent monitoring:
    1. Oxygenation (PaO2 < 60 while on 60% FIO2 or greater)
    2. ABG – Acute respiratory acidosis (pH < 7.30 with CO2 > 55)
    3. Q4 FVC assessment
    4. Q4 NIF assessment

RT INTERVENTIONS FOR NEUROMUSCULAR DISEASE PATIENTS

1. Non-invasive ventilation with a pressure support (the difference between the IPAP and EPAP) to support adequate tidal volume
2. Intubation:
    1. To protect the airway
    2. When volumes drop below 5 ml/kg of ideal body weight
    3. FVC <15 ml/kg
    4. Rapid progression of respiratory weakness

INTERVENTIONS CONT....

- Bronchial hygiene therapy
  - Weakness of respiratory muscles may lead to:
    - Atelectasis-V/Q mismatch leading to hypoxemia, secondary pneumonia
    - Retained secretions
      - Assisted Cough
      - Volara
      - Nasotracheal suctioning

QUESTIONS & DISCUSSION

EVALUATE THIS PRESENTATION

SOURCES

- “Respiratory management of Patients with neuromuscular disease: current perspectives” Gerald Pfeffer, Marcus Povitz; Dovepress
- “Diagnosis and management of Guillain-Barre syndrome in ten steps” Sonja E. Leonhard, Melissa R. Mandarakas, et al. Neurology Vol 15, Nov. 2019
- “Respiratory Management of Patients With Neuromuscular Weakness, An American College of Chest Physicians Clinical Practice Guideline and Expert Panel” Akaram Khan, MD; Lindsy Frazer-Green, PhD; et al. Critical Care Guidelines and Consensus Statements. Chest Aug. 2023
- “Comparative Outcomes of Respiratory Failure Associated with Common Neuromuscular Emergencies: Myasthenia Gravis versus Guillain-Barre Syndrome” Anantha R. Vellipuram, Salvador Cruz-Flores, et al. Medicina 2019 55