GB Syndrome ER and ICU management issues -

Moderator - Dr. Mihir Sarkar Panelist – Dr. Agnisekhar Saha Dr. Prabhas Prasun Giri Dr. Rohit Bhowmik Dr. Kaushik Maulik

History

- A 6 year old male, 20 kg
- Sudden onset weakness of lower limbs, inability to stand
- Progressed to UL over next 2 days
- Decreased volume of voice
- Sensorium was normal.
- No H/O fever, trauma, immunization, diarrhea or seizure.

What is your working diagnosis ?

CNS – Alert, conscious.

Quadriparesis – power LL -1/5, UL- 2/5

Global areflexia.

No sensory loss.

Resp – RR-30/min, CVS – Hr- 110/min,

BP – 98/58 mmHg



Kaushik

• What are the Emergency issues a patient with GB Syndrome might have ?

Rohit

Spectrum of Emergency Care



Dysautonomia – Labile BP, Arrythmia, ileus, bladder control, Cardiomyopathy

What are the indications of ICU admission in a patient f GB Syndrome ?

Prabhas

Indication to admission in PICU

- Rapid progression of motor weakness involving respiratory muscles
- Ventilatory insufficiency
- Pneumonia
- Severe bulbar weakness
- Autonomic instability: arrhythmia, or bradycardia
 - Autonomic dysfunction is an important cause of death due to hemodynamic instability and arrhythmias.

• How will you assess and monitor a patient of GB Syndrome in ER or PICU ?

Agnida

Assessment of Adequate Respiratory Function

Clinical

- Respiratory rate—Good index of response to hypoventilation caused by muscular weakness; tachypnea is the earliest response
- Swallowing and handling of secretions
- Quality of cough
- Volume of speech
- Single-breath count
- Chest expansion
- Presence of tachycardia/diaphoresis (nonspecific)
- Use of accessory muscles
- Orthopnea
- Inward movement of abdomen during inspiration
- Breathing pattern alternates between accessory and major respiratory muscles, signifying weakness of major respiratory muscles
- Change in status when sleeping—accessory muscle tone decreases
- Rate of progression of generalized weakness

Paradoxical vs Sea Saw Breathing



Laboratory Vital capacity Maximum inspiratory pressure Maximum expiratory pressure Sao₂, Pao₂, Paco₂, pH Chest radiograph

> Count up to 10 in one breath, the FVC is at least 15–20 mL/kg. Can count up to 25, the vital capacity is ~30–40 mL/kg.

Monitoring the trend in single-breath counting may help to determine disease progression

Ability to FLEX the neck

Increasing trend of PaCo2

Are there any predictors of need of Mechanical Ventilation ?

Kaushik

Anticipating Mechanical Ventilation in Guillain-Barré Syndrome

Nicholas D. Lawn, FRACP; Dade D. Fletcher, MD; Robert D. Henderson, FRACP; Troy D. Wolter, MS; Eelco F. M. Wijdicks, MD

- 1. Rapid disease progression,
- 2. Bulbar dysfunction,
- 3. Bilateral facial weakness, or dysautonomia
- 4. Vital capacity of less than 20 mL/kg,
- 5. Maximal inspiratory pressure less than 30 cm H_2O_2 ,
- 6. Maximal expiratory pressure less than 40 cm H_2O .
- 7. Reduction of more than 30% in vital capacity

"20/30/40 rule" – In Adults

Clinical predictors of mechanical ventilation in Guillain-Barré syndrome

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- •Simultaneous weakness of upper (UL) and lower (LL) limbs as the initial symptom (*P*<0.001);
- •UL power less than Grade 3/5 at nadir (P<0.001);
- Presence of neck and bulbar weakness (P<0.001);
- •Shorter duration from onset to bulbar weakness and confinement to bed (*P*=0.001)
- Bilateral facial involvement (P<0.01)
- An elective endotracheal intubation may prevent sudden respiratory arrest and its consequences

NIV vs Mechanical Ventilation – What and Whom ?

Rohit

Assessing Ventilation and the Need for Urgent Intubation and Ventilatory Support

Oropharyngeal weakness –

Airway collapse Poor cough Aspiration



Cranial nerves Facial weakness or asymmetry, Poor cough reflex

Swallowing dysfunction

Intubation Ventilation

Diaphragmatic and intercostal muscle weakness



What precautions will you take during intubation ?

Prabhas

Special considerations for intubation

 Dysautonomia may exaggerate the hemodynamic responses to the drugs

Emergency intubation (with full stomachs or ileus)

- Modified RSI technique Apnea Oxygenation, cricoid pressure
- Prepare atropine/glycopyrrolate, fluids, and vasopressors (risk of associated autonomic instability
- Avoid use of succinylcholine. Consider 1.0–1.4 mg/kg rocuronium. Reduce the dose in MG and myopathis
- Volatile aneasthetic agents are preferable
- A titrated or reduced dose of a sedative and analgesics

What strategy will you take while ventilating this patient ?

Agnida

Ventilator Settings

- Volume-controlled / SIMV + PS ventilation
- Normal V T (8 ml/kg) and Respiratory rate.
- Although high V T have been recommended by some authorities, not necessary for most patients.
- Set V T and respiratory rate that the patient considers comfortable.
- Low pressures and a low FiO2
- PEEP is set (eg, 5 cm H₂O) to prevent atelectasis.

Weaning

Train the diaphragm and respiratory muscle

Gradual weaning

Initially Day time PS mode and Night time controlled mode

Gradually decrease the PS , even can reduce to ZERO PS and......gradually night PS also

Can give T-Piece trail

Patient is not moving at all - Sedation and analgesia does it require ? Kaushik

Pain is often overlooked feature of GBS, occurring in 55% to 89% of patients. Anxiety occurs in 82% of patients.

Cause - Inflammation of proximal nerve roots, Sensory nerve fiber dysfunction Musculoskeletal pain from immobility

Analgesics options – Opiates - ileus may limit their use Gabapentine, Carbamapazemines. Sedation and Anxiolytic - Benzodiazepines, SSRI

Ruts L, Rico R, van Koningsveld R, et al. Pain accompanies pure motor Guillain-Barré syndrome. J Peripheral Nervous System. 2008;13(4):305–306

D - 2

 Patient had sudden onset of tachycardia (HR – 180), hypotension (BP- 70/40 mmHg) and profuse perspiration.

Rohit

- What are the possibilities ?
- How will manage the situation ?
- Autonomic dysfunction occurs in 2%–10% of patients
- Volume repletion, and if refractory to fluids, a pure α-agonist such as phenylephrine.
- Fluctuating BP Monitor ABP to guide fluid.

What are the Lab tests will you do ?

Acute weakness checklist for the first hour

- \Box Assess airway, breathing, and circulation
- \Box Characterize the weakness by detailed exam
- \Box Build an initial differential diagnosis of the causes of weakness
- \Box Consider emergency causes in the differential diagnosis
- □ Initial labs: Glucose, electrolytes, Ca, Mg, PO4, BUN/Cr, LFTs, PT, PTT, CBC, and ABG
- □ Special Labs: TFTs, CPK or CK, ESR, parathyroid hormone, GGT
- \Box Relevant MRI and CT imaging

Nerve Conduction Study, CSF Analysis





Course

• Nerve conduction study - low CMAP amplitudes in all 4 limbs suggestive of Acute Motor Axonal Neuropathy (AMAN) variety

 Cerebrospinal fluid (CSF) analysis done on day 10 of weakness showed albumin-cytological dissociation

Child received intravenous immunoglobulin (IVIG) 2 g/kg over 5 days on Day 2 and 3 of admission - no clinical improvement

Plasma Exchange done after 10 days

D-7

- Child worsened
- Fall in saturation to 85%, heart rate increased to 180/min, although BP was normal. High rade fever with profuse secretion in ET.
- EtCO2-56, expiratory tidal volume corresponding to inspiratory tidal volume, ventilator working well.
- Apparently chest movements normal on both sides, Crepitation bilateral.



What is happening ?

Outcome depends on smooth sailing throughout the long course.

• What are the complications of prolonged Mechanical Ventilation? Agnida

What are the complications of prolonged Mechanical Ventilation?

- Nosocomial pneumonia
- Atelectasis
- Long-term, invasive ventilation may result in
- Tracheomalacia,
- Tracheoesophageal fistula formation, or
- Tracheal stenosis.
- Adhere to VAP Bundle .Practice sterile techniques during suctioning.

General Supportive Care – Key to Success ? Kaushik

- Regular physiotherapy,
- Provision of splints to prevent joint contractures,
- Prevention of deep vein thrombosis
- Careful positioning and repositioning.
- Early Enteral nutrition Inadequate calories and protein may result in ventilator dependence.
- Handwashing, strict policies regarding intravenous and urinary catheter asepsis and antimicrobial stewardship.

Nutrition – How will you go about it ?

Rohit

- GBS is a hypermetabolic and hypercatabolic state .
- Inadequate nutrition -

Fluid and electrolyte abnormalities, Decubitus ulcers, as well as Nosocomial infections

- Early enteral feeding.
- High-protein diet at 1.3 to 1.5 × their calculated basic energy expenditure (BEE) plus an additional 30% to 50% for weight.
- Monitor nitrogen balance, prealbumine, transferrin.
- Bowel care Prevent constipation

Duration of MV



The figure indicates the Duration of mechanical ventilation in 149 patients with GBS. Median duration of mechanical ventilation was 28 days, interquartile range of 12–60 days, absolute range 1 to >181 days (follow-up of the studies ended at 181 days)

Let's talk about tracheostomy

Why Tracheostomy ?

- To prevent laryngeal and upper airway damage due to prolonged translaryngeal intubation
- To allow easy or frequent access to the lower airway for suctioning and secretion removal
- To provide a stable airway in a patient who requires prolonged mechanical ventilation or oxygenation support

• When will you consider tracheostomy and how will you approach ? Prabhas



ORIGINAL ARTICLE



Tracheostomy or Not: Prediction of Prolonged Mechanical Ventilation in Guillain–Barré Syndrome

Christa Walgaard¹ · Hester F. Lingsma² · Pieter A. van Doorn¹ · Mathieu van der Jagt³ · Ewout W. Steyerberg² · Bart C. Jacobs^{1,4}

Conclusions -Ventilated GBS patients who are unable to lift the arms from the bed and patients who have axonal degeneration(AMAN) or unexcitable nerves at 1 week are at high risk of prolonged MV, and tracheostomy should be considered in these patients.

WHEN ?

- Published recommendations and experts' opinions mostly consider 10 to 15 days as the optimal delay for performing tracheotomy .
- Tracheotomy after 21 days might be associated with longer ICU stay and higher mortality.

• Tracheotomy: why, when, and how? *Respir Care* 2010, **55:** 1056-1068.

Benefits of Changing From a Translaryngeal Endotracheal Tube to a Tracheostomy Tube

Benefit	Type and Quality of Literature Support Showing Benefit
Improved patient comfort	Uncontrolled reports, clinical opinion
Less need for sedation	Several RCTs
Lower work of breathing	Theoretical analysis, one small study
Improved patient safety	Clinical belief but minimal data
Improved oral hygiene	Clinical observation
Oral intake more likely	Opinion only
Earlier ability to speak	Uncontrolled reports
Faster weaning from mechanical ventilation	One RCT
Lower risk of ventilator-associated pneumonia	Controversial, data support for both sides
Shorter intensive care unit and hospital stay	Several meta-analyses

Rehabilitation and permanent disability

- Early Mobilization:
- Physical Therapy:
- Occupational Therapy:
- Respiratory Therapy:
- •Speech and Swallowing Therapy:
- •Emotional and Mental Support:

Course



Key Points

Asses the early indications of ICU admission and Mechanical Ventilation

Stabilize the patient before transport and continue support during transport

Be aware of autonomic dysfunction

Prevent nosocomial infection

Good and meticulous supportive care is the key to success







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