Neuromuscular Disease Patient Management Cheat Sheet

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Types of Neuromuscular Diseases	
Myasthenia Gravis	Chronic Disorder, Acute events of muscle weakness and fatigue. Descending paralysis.
Guillain Barre	Ground up, Ascending paralysis. Usually triggered by an acute infectious process.
ALS	Amyotropic Lateral Sclerosis. Progressive neuro-degenerative disease
Muscular Dystrophy	Rare group of genetic diseases. Duchenne MD is the most common form among children and Myotonic is the most common in adults
Multiple Sclerosis	A slow progressive CNS disease, characterized by disseminated patches of demyelination in the brain and spinal cord. Patients usually experience exacerbations and remissions.
Spinal Muscular Atrophy Disorders	A group of hereditary disorders characterized by skeletal muscle wasting due to progressive degeneration of anterior horn cells in the spinal cord and of motor nuclei in the brain stem
Post-Polio Syndrome	A group of symptoms that develops years or decades after paralytic poliomyelitis and usually affects the same muscle groups as the initial infection
Diaphragmatic Paralysis	May be unilateral where only one side of the diaphragm is affected.

Clinical Presentation

Muscle weakness and fatigue
Exercise intolerance
Myopathic face (drooping face)

Pathophysiology of NM Disease

Inspiratory Muslces	Weakness of these muscles lead to decreased Vt, VC and FRC. May cause atelectasis and alveolar collapse
Expiratory Muscles	Weakness of these muscles lead to decreased ability to cough and move secretions. Increased risk of pneumonia.
Airway Muscles	Weakness of these muscles lead to decreased ability to protect the airway from aspiration and to speak/swallow. Risk of airway collapse and aspiration pneumonitis

Lab Findings

ABG	On arrival, these patient may present with acute ventilatory failure with hypoxemia.
PFT	These patients present with decreased lung volumes, decreased MIP and MEP. VC is the most commonly measured parameter for these patients.
Creatinine Kinase	Increased in many myopathies. Sign of muscle fiber necrosis.

Ventilation Strategy

Indications	20/30/40 rule: VC < 20 ml/kg, MIP <-30 cmH2O and MEP <40 cmH2O or pH <7.35/-PaCO2 > 45 mmHg
Main concerns	Typically these patient's lungs are healthy. Our main concern here is protecting the airway and preventing pneumonia.
Target Gas	Since the lungs are not the issue we would expect these patients to have a normal gas.
Type of Ventilation	These patient benefit from both positive pressure and negative pressure ventilation. Although PPV is seen most often in the hospital setting. PPV can be invasive or noninvasive
Mode	Typically VC-CMV will be used in these patients.
Parameters	Normal parameters would be used such as: VT 6-8ml/kg, RR 8-12, flow greater or equal to 60 LPM, Ti 1.0, PEEP 5 and FiO2 0.21. Pplat should be <30cmH2O.

Adjuncts and Other Treatments

Inline Suction	Because these patient's muscle are weak they often cannot generate a cough and bronchial hygiene may become difficult. To prevent disconnection with with ventilator, inline suction should be used.

Adjuncts and Other Treatments (cont)

Cough Assist	Helps the patient generate a strong cough and improve bronchial hygiene. Cough assist can be used in the long term tracheostomy patient.
Incentive Spirometry/Hyperinflation Techniques	Helps mobilize secretions and prevents infection. Breath stacking is often used in these patients.
Pharmacology	Steroids for MG, plasmapheresis, pain management, sedative and anxiolytic may be required.

Challenges With Patient Management

Most NM diseases have no treatment or have a long process leading to long term care. These patients would be ventilated for a long duration of time likely resulting in the use of a tracheostomy.
Be sure to check cuff pressures regularly and change trach as needed. Normal cuff pressure is 20-30cmH2O.

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