Primary Acute Neuromuscular Respiratory Failure

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KEYWORDS

• Weakness • Acute • Neuromuscular respiratory failure • Recognition • Evaluation • Management • Guillain-Barré syndrome • Myasthenic crisis

KEY POINTS

- The diaphragm is the principal inspiratory muscle and normally carries two-thirds of the work of breathing. A paradoxic breathing pattern is a reliable marker of diaphragmatic failure.
- Concomitant disease of the airways, lungs, heart, or thoracic wall can precipitate ventilatory failure in patients with neuromuscular respiratory failure.
- The assessment of a patient with neuromuscular respiratory failure should include arterial blood gas analysis, chest radiograph, and bedside spirometry.
- Noninvasive ventilation can avert intubation and shorten the duration of hospitalization in patients with myasthenic crisis but should be initiated before the development of hypercapnia.
- Patients with Guillain-Barré syndrome who develop early signs of ventilatory insufficiency or have substantial bulbar weakness should be intubated early and mechanically ventilated.

INTRODUCTION

Patients with both defined and undiagnosed neuromuscular disorders are frequently admitted to hospital services and neurologists must be able to recognize and intervene appropriately when respiratory failure develops. Neurologists are also consulted to evaluate whether a neuromuscular disorder exists when there is difficulty liberating a patient from mechanical ventilation (invasive or noninvasive). Early recognition of acute neuromuscular respiratory failure (NRF) and determination of the cause is

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imperative, as there are survival implications. When the cause of neuromuscular weakness and resultant NRF cannot be identified, the outcome is usually poor.[1] This review focuses on the clinical recognition of primary NRF (eg, related to a neurologic disease), as well as its pathophysiology, diagnostic evaluation, and management. Secondary NRF resulting from systemic disease (eg, intensive care unit acquired weakness from multiorgan failure) is not covered here.

RECOGNITION OF NEUROMUSCULAR RESPIRATORY FAILURE

NRF may rarely be the presenting feature of a neurologic disorder, but more often results from disease progression or exacerbation by a superimposed respiratory illness, to the point that compensatory mechanisms are overwhelmed. The clinical presentation of dyspnea and rapid shallow breathing may be readily apparent in acute neurologic disorders, such as the Guillain-Barré syndrome (GBS), whereas patients with subacute neurologic disease, such as amyotrophic lateral sclerosis (ALS), who have developed the weakness over time may appear deceptively comfortable despite ventilatory failure.

The clinical signs of NRF are summarized in **Box 1**. Early in the development of NRF, the clinical signs are subtle. The patient may appear restless but otherwise relatively normal until questioning reveals that the patient pauses to take a breath in the middle of every sentence (so called staccato speech). Patients may describe a sensation of being unable to get a full breath. Close inspection may reveal sweat on the forehead or brow, rapid shallow breathing, and activation of the accessory muscles of respiration, signaling an increased work of breathing. Palpation of the accessory muscles will reveal contraction of the muscles before it is apparent by observation alone. Additional signs may include tachycardia, frequent coughing or throat clearing suggesting difficulty in handling oral and respiratory secretions, a weak cough, and abdominal paradox. The presence of a paradoxic breathing pattern is the most reliable sign of impending respiratory failure, as it reflects diaphragmatic failure[2] and may be missed if the patient is not examined in the supine position. In patients with frank failure, nasal flaring and sternal retraction also may be seen and the patient will appear anxious and fatigued.

ANATOMY AND PHYSIOLOGY OF NEUROMUSCULAR RESPIRATORY FAILURE

The clinical symptoms and signs of NRF result from failure of the breathing apparatus and therefore failure of ventilation. To review, the primary muscle of inspiration is the

Box 1
Clinical signs of neuromuscular respiratory failure

- Restlessness
- Diaphoresis
- Tachycardia
- Tachypnea
- Staccato speech
- Activation of accessory muscles during inspiration
- Weak cough
- Abdominal paradox