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# Intubate Early in Neuromuscular Respiratory Failure

**NEW YORK CITY**—"The diaphragm is the respiratory muscle that fails first in most neuromuscular diseases," said Allan H. Ropper, MD. This difference has implications for the evaluation and treatment (including timing of intubation) of patients with Guillain-Barré syndrome and other neuromuscular diseases, he said.

Dr. Ropper, who is Professor and Chairman of Neurology at Tufts University School of Medicine, Boston, presented the closing address at the Second Biennial New York Symposium on Neurologic Emergencies and Neurocritical Care. "Respiratory failure in neuromuscular disease is a very different entity from the parenchymal pulmonary failure seen in pneumonia, congestive heart failure, emphysema, adult respiratory distress syndrome, and other lung diseases so familiar to internists, critical care specialists, and emergency medicine specialists."

## Monitoring the Failing Diaphragm

The muscle fibers of the diaphragm are "tuned" to work efficiently only within a relatively narrow range along the length-tension relationship, said Dr. Ropper, who is Chairman of Neurology at St. Elizabeth's Medical Center, Boston. In neuromuscular disease, the number of motor units operating in the diaphragm may be sharply reduced, even before breathing problems or other signs of neuromuscular deficit become apparent. When the small number of remaining motor units are called upon "to produce the effort that was formerly required of the entire [diaphragm]," he noted, the ideal length-tension relationship is lost, and the muscle "begins to fall apart."

The best reflection of the diaphragmatic length-tension relationship, Dr. Ropper said, is the mid-expiratory (or mid-inspiratory) flow rate. This is unwieldy for clinical purposes, however, so indirect measurement by spirometer has come to be the accepted standard. Spirometry "doesn't measure anything in particular about the diaphragm," he admitted, but it provides consistent, reproducible readings from measurement to measurement and correlates well with body surface area.

Inspiratory force measurement provides a supplementary gauge to diaphragmatic performance, Dr. Ropper said. Flow-volume loops, which typically show a restrictive pattern in patients with neuromuscular respiratory disease, are not useful as indicators of diaphragmatic function.

## Evaluating Respiratory Failure

Respiratory failure may be a presenting sign in amyotrophic lateral sclerosis, myasthenia gravis, acid maltase deficiency, and critical illness myopathy. In contrast, in Guillain-Barré syndrome, porphyria, polymyositis, inclusion-body myositis, and muscular dystrophy, it is a later phenomenon—unlikely to be present initially but almost certain to develop eventually.

Dr. Ropper pointed out several pitfalls in bedside measurement of respiratory function that can be particularly important in older patients with neurologic disease, since they may have coexisting chronic obstructive pulmonary disease or pneumonia.

First, he said, facial weakness, a universal feature of Guillain-Barré syndrome, gives spuriously low readings of vital capacity. Because air leakage may not be apparent, Dr. Ropper recommended "putting nose clips on and manually sealing the mouth around the mouthpiece" to achieve an accurate measurement.

Another potential pitfall is the mismatch between inspiratory force and vital capacity. Dr. Ropper explained that this suggests either parenchymal or obstructive lung disease or an inconsistent respiratory effort.

In neuromuscular respiratory failure, Dr. Ropper said, diaphragmatic weakness typically occurs together with cervical muscle weakness. Neck weakness is a clue to neuromuscular respiratory failure, he explained, "because the myotome that enervates that diaphragm also enervates simultaneously the neck flexor muscles and the shoulder elevators." This observation is useful, Dr. Ropper pointed out, "when inconsistent [respiratory] effort, poor effort, or a pulmonary parenchymal problem [interferes with] the measurement of vital capacity."

Perhaps the simplest way to obtain a "ballpark estimate" of vital capacity, Dr. Ropper explained, is to have the patient count aloud during a single breath. A patient with normal vital capacity should be able to count to 20 in a single exhalation, he said, noting that this correlates approximately with a vital capacity of two liters.

At the high and low ends of lung volume, the method is inaccurate, but it can signal when a patient with Guillain-Barré syndrome or myasthenia gravis may need to be intubated.

When a patient with Guillain-Barré syndrome or myasthenia gravis can count only to 10 in one breath, vital capacity is approaching one liter, and it is time to consider intubation, Dr. Ropper said. The rub, he noted, is that patients frequently slow their exhalation and "often hold their breath, thinking that's what [the doctors and nurses] want them to do."

Given the complicated nature of diagnosing diaphragm weakness, Dr. Ropper's threshold for action is to intubate patients with neuromuscular respiratory failure when vital capacity falls to approximately 12 to 15 mL/kg (normal vital capacity is between 30 and 55 mL/kg).

## Approach Is Nontraditional

He acknowledged that this flies in the face of the traditional critical care approach to intubation. Patients may appear to be comfortable, semi-reclining, and not expending too much respiratory effort, but "they get a little antsy, and maybe raise a bead or two of sweat on their brow or on their cheek. They get a little tachycardic. That's the danger point," Dr. Ropper warned. Even when patients are not dyspneic and the chest film looks normal, he explained, "they're flogging the few remaining motor units, and the diaphragm is about to decompensate."

Dr. Ropper said that severe respiratory failure can develop quickly in patients with Guillain-Barré syndrome and other acute neuromuscular diseases, even when the rest of the neuromuscular illness is not advancing. Most problems that arise when these patients are in the critical care unit, he noted, result from "late intubations," performed after they already have secretions in the airway or have become autonomically unstable.

While mild hypoxia is the signature feature of early neuromuscular respiratory failure, the work required of the diaphragm is intensified by pulmonary arteriovenous shunting brought on by atelectasis. Unlike plate atelectasis, which "is evident on a chest film, [this] is a miliary phenomenon, widespread throughout the lung" and not radiographically visible, Dr. Ropper explained. Because the patient is "moving mainly central lobular air," surfactant in the peripheral alveoli is reabsorbed, those alveoli collapse, and lung volume is irreplaceably lost.

Giving the patient 100% oxygen will not correct the hypoxia, Dr. Ropper warned, because the problem is not a ventilation-perfusion mismatch, "it's a physiologic shunt"—unoxygenated blood is being directed to an unoxygenated part of the lung and returning to the heart unoxygenated.

## Managing Guillain-Barré Syndrome

The general treatment for Guillain-Barré syndrome involves plasma exchange or intravenous immunoglobulin (IVIg) as well as early intubation, Dr. Ropper said. He recommends IVIg, 0.4 g/kg/d for four to five days, or exchanging 250 mL of plasma in four or five sessions for seven to 10 days.

Repeat treatments are not usually useful in patients whose conditions continue to worsen, he noted. Similarly, corticosteroids alone are of questionable value, and are "now eschewed in the treatment of Guillain-Barré syndrome," although they may be beneficial after IVIg infusion. The relapse rate with all approaches is approximately 10%, he added.

Patients who have severe axial pain can receive epidural analgesia. As important as specific treatment measures, Dr. Ropper emphasized, are infection surveillance, prophylaxis against deep venous thrombosis and pulmonary embolism, skin and eye care, and nutritional support. The value of good physician-patient communication and psychological support during a potentially terrifying experience, he added, is not to be overlooked. It is "these little things that the patient remembers," he concluded, "not whether you were clever enough to choose gamma globulin or intubate early."

**—Elliot Richman, PhD**

**Reference**
1. Ropper AH. Neuromuscular respiratory failure. Presented at: The Second Biennial New York Symposium on Neurologic Emergencies and Neurocritical Care; May 19, 2001; New York City.

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