APPROACH IS NONTRADITIONAL

Respiratory failure is an ominous sign in any patient, but it is particularly so in neuromuscular disease, where the chest wall is being compromised by paralysis. In patients with Guillain-Barré syndrome, it’s important to remember that at least 25% of patients with a relapse exhibit new neurological symptoms not present at the onset of the initial episode (Cochran et al. 1999; Medical Care 37: 1297–1304). The relapse rate with all types of Guillain-Barré syndrome, although they may be mild, is 50% (Barré 1904; Lancet 1: 1115–1116; Br. Med. J. 2: 1381). Approximately 10% of patients with Guillain-Barré syndrome die, and some who survive may develop chronic respiratory failure (Peters et al. 1999; Lancet 354: 1601–1608).

It is critical to intubate patients with neuromuscular disease in a timely fashion, especially those with early signs of respiratory failure. Physicians frequently ask the question, “When is the right time to intubate?” Dr. Ropper said, “We’re always a little too late.” The best reflection of the diaphragmatic function, he said, is forced vital capacity, which provides information about how much respiratory muscle is functioning and how much of a respiratory challenge is being accepted.

When the patient can no longer hold their breath, “That’s the danger point,” Dr. Ropper said. “They may have a 70% vital capacity and a 10-second forced expiratory volume, but they can’t hold their breath and are about to decompensate.”

Given the complicated nature of diagnosing respiratory failure, the best reflection of the diaphragmatic function, he explained, is forced vital capacity, which provides information about how much respiratory muscle is functioning and how much of a respiratory challenge is being accepted. Spirometry “doesn’t measure expiratory (or mid-inspiratory) flow rate. This is the best reflection of the diaphragmatic length–force relationship,” Dr. Ropper said, “and the muscle ‘begins to fall apart.’”

If neither patient can hold their breath, the medical team should be prepared to intubate. “If they can’t hold their breath, are they going to be able to cooperate if we say, ‘Let’s do this’? My ‘rule of thumb’ is to intubate patients with neuromuscular disease when they count to 10 in one breath with normal vital capacity, which is about two liters. If their condition continues to worsen, and the patient can’t hold their breath even when the rest of the neuromuscular illness is improving, it’s time to intubate,” Dr. Ropper explained.

Respiratory failure occurs when vital capacity falls to 25% or less of normal, he added. “Although the patient may appear normal, they’re flogging the few functioning motor units they have.”

FIRST, FACIAL WEAKNESS

When intubating patients with neuromuscular disease, the best reflection of the diaphragmatic function is normal vital capacity. “In neuromuscular respiratory failure, the work required for exhalation and ‘often hold their breath, thinking they can do it,’ he explained. “Another potential pitfall is the mismatch between inspiratory force and vital capacity. Dr. Ropper said that severe respiratory failure can develop in just hours depending on the patient’s condition. In neuromuscular respiratory failure, the number of motor units operating is small, lung volume is irreplaceably lost, and lung collapse, and lung volume is irreplaceably lost. They get a little tachycardic. That’s the way the body’s trying to compensate.”

Diaphragmatic function, he explained, is difficult to measure. “It’s very hard to do a manometric technique to measure the muscle function in the diaphragm.”

An alternate way to estimate diaphragmatic function, he said, is a method which was developed by Elizabeth’s Medical Center, Boston. In neuromuscular disease, the number of motor units operating is small, lung volume is irreplaceably lost, and lung collapse, and lung volume is irreplaceably lost. A patient’s capacity to perform a two-liter vital capacity is a useful method to determine diaphragmatic function. “Spirometry is an accepted standard. Spirometry doesn’t measure expiratory (or mid-inspiratory) flow rate. This is the best reflection of the diaphragmatic length–force relationship,” he explained.

If neither patient can hold their breath, the medical team should be prepared to intubate. “If they can’t hold their breath, are they going to be able to cooperate if we say, ‘Let’s do this’? My ‘rule of thumb’ is to intubate patients with neuromuscular disease when they count to 10 in one breath with normal vital capacity, which is about two liters. If their condition continues to worsen, and the patient can’t hold their breath even when the rest of the neuromuscular illness is improving, it’s time to intubate,” Dr. Ropper explained.

Neuromuscular respiratory failure is nontraditional because, in patients with Guillain-Barré syndrome, it’s unclear whether the patient could have held their breath if the motor units were functioning normally. “We are always a little too late,” Dr. Ropper ended.