

Best Practices for Incorporating Non-invasive Ventilation in ALS Care

Non-invasive ventilation supports a patient's ability to take deeper or more frequent breaths than they can on their own. Many patients with amyotrophic lateral sclerosis (ALS) use a bilevel positive airway pressure (BPAP) machine or other ventilator at some point in the disease course, but these devices may be warranted earlier than you think.

In 2023, the [American College of Chest Physicians](#) (CHEST) released guidelines for the respiratory care of patients with neuromuscular diseases, including those with ALS. "One of the big things that emerged from that statement is that we should probably be initiating non-invasive ventilation sooner in all patients with neuromuscular diseases, but especially in ALS," says Finbar Foley, MD, Assistant Professor of Pulmonary, Critical Care, and Sleep Medicine at Yale School of Medicine.

The 2023 CHEST guidelines recommend starting non-invasive ventilation in patients with ALS if their forced vital capacity (FVC) is less than 80% and they have symptoms suggestive of developing respiratory failure, including fatigue, shortness of breath, and sleep or cognitive difficulties. However, Center for Medicare Services coverage guidelines state that non-invasive ventilation is indicated for patients with neuromuscular disease if their arterial carbon dioxide level is 45 or higher or their FVC is less than 50%.

"The difficulty is the payment guidelines don't align with early initiation, so pulmonologists are trying to find ways to get patients devices sooner," Dr. Foley says.

Potential solutions include testing patients sitting up and lying down. "Sometimes, if we check lung function in the supine position, we find that the vital capacity drops below that threshold for which the device can now be approved by insurance," he says. Dr. Foley also recommends a sleep study if there are any signs of a patient experiencing nighttime breathing disturbances.

“We know that of all the things that we can do for ALS, ventilatory support currently provides the largest survival benefit,” he says. “It has a larger survival benefit than riluzole, for example. There’s such a focus on the medical therapies that are available, but in fact, ventilatory support is currently the thing that most guarantees survival benefit.”

Resources

- 2023 CHEST Guidelines: [Respiratory Management of Patients With Neuromuscular Weakness](#)
- [Practical Guide to Management of Long-Term Noninvasive Ventilation for Adults With Chronic Neuromuscular Disease](#)
- Mouthpiece ventilation resources:
 - [Mouthpiece ventilation in neuromuscular disorders: Narrative review of technical issues important for clinical success](#)
 - [Daytime Mouthpiece for Continuous Noninvasive Ventilation in Individuals With Amyotrophic Lateral Sclerosis](#)
- [ALS Respiratory Care Course for Clinicians](#) (free, 19 lectures, CME/MOC credit available)