

Thus, many clinicians consider NMDs to be terminal because they know of no way to prevent morbidity and mortality from respiratory failure and feel justified by the inevitably fatal results of nihilistic conventional approaches.<sup>63,64</sup> Instead of helping to keep patients alive in anticipation of possible medical treatments and cures, they more often transmit hopelessness, provide "palliative care" that only expedites death, and make no valid effort to prevent respiratory morbidity. Thus, patients either die from acute cardiopulmonary failure or undergo tracheotomy during a usually preventable episode of acute respiratory failure. Tracheostomies can prolong survival, but many if not most patients with tracheostomies eventually die from related complications, and tracheostomies adversely affect quality of life (see below).

### The Most Common Evaluation Errors

#### Misinterpretation of Symptoms

The symptoms of hypoxia due to lung disease can include shortness of breath, difficulty with thinking, and anxiety. There are many more symptoms of hypercapnia (Table 1), yet hypercapnia is often ignored. Even underventilating (hypercapnic) patients themselves often do not recognize early symptoms. Many people have few or no symptoms when sitting but require the use of 3 or 4 pillows to sleep because they cannot breathe for very long when lying down. This problem is often due to diaphragm dysfunction out of proportion to weakness of other inspiratory muscles. Sometimes it takes a trial of nocturnal use of inspiratory muscle aids before the user feels more energy and less fatigue and has more restful sleep.

Although people with either ventilatory impairment or lung disease with oxygenation impairment often complain of shortness of breath when walking, wheelchair users with ventilation impairment rarely complain of shortness of breath despite severe hypercapnia and impending respiratory failure. Instead, they complain of anxiety and inability to

fall asleep. Unfortunately, these symptoms are often treated as though they are due to oxygenation impairment until decompensation results in death or translaryngeal intubation. Ventilatory failure can also cause the syndrome of inappropriate secretion of antidiuretic hormone.<sup>645</sup>

For small children with NMD, the initial signs of nocturnal underventilation may be frequent arousals from sleep and episodic perspiration, flushing, and tachycardia. If recognized at all, such symptoms may be mistaken for the simple need to be turned. Certainly, any small child with NMD who was once sleeping most or all of the night without arousals and then requires increasingly more frequent turns, especially the child with paradoxical breathing who awakens frequently despite using a bed that continuously turns him or her, is quite likely to have nocturnal hypoventilation.

#### Inadequate Pulmonary Function Studies

People with neuromuscular conditions are routinely sent to pulmonary function laboratories, where they undergo a comprehensive and expensive battery of tests designed for people with lung disease. These tests include spirometry (i.e., the measurement of forced expiratory volumes and flows to evaluate for airway obstruction), studies to test the integrity of the respiratory exchange membrane, diffusion studies, and, often, painful and inaccurate arterial blood gas analyses (see below). However, because people with neuromuscular conditions do not usually have a long history of cigarette smoking or asthma, all of these tests except for spirometry are unnecessary. Furthermore, because lung underventilation begins during sleep, spirometry or simple determination of vital capacity (VC) is best done with the person lying down. For people with many diagnoses, the VC in the supine position is often much less than in the sitting position. There can also be a great difference in VC when the patient is lying on one side or the other and when the patient is wearing or not wearing a back brace or body jacket. A well-fitting body jacket can increase the VC, whereas a poorly fitting body jacket can decrease VC. Unfortunately, pulmonary function laboratories do not measure the VC in various positions or situations, and if the physician knew enough to ask the lab to do so, he or she would perform the tests with a simple hand-held spirometer.

Although they are the most important tests of all, pulmonary function laboratories do not measure assisted cough peak flows (see Manually Assisted Coughing) or maximum insufflation capacities and are rarely equipped for or asked to measure end-tidal carbon dioxide levels.

#### Failure to Monitor Sleep Appropriately

Either the ability to breathe supine or during sleep is entirely ignored, or expensive and unnecessary polysomnography is ordered to diagnose sleep-disordered breathing (Chapter 10). To perform polysomnography, patients are usually asked to sleep in the hospital and are observed for at least one night. Electroencephalography, plethysographic measurement of chest and abdomen movement, air flow at the nose, and SpO<sub>2</sub> are typically monitored. It can be difficult to sleep under these circumstances. The test attributes symptoms to central or obstructive apneas; it does not attribute them to underventilation secondary to inspiratory muscle weakness and, therefore, has little relevance to people with NMDs. Furthermore, once the polysomnographer has the interpretation

Table 1. Symptoms of Chronic Alveolar Hypoventilation and Sleep-disordered Breathing\*

Fatigue	Lower extremity edema
Dyspnea (particularly in patients capable of ambulation)	Irritability
Morning or continuous headaches	Anxiety
Multiple nocturnal awakenings with dyspnea/tachycardia	Nocturnal urinary frequency
Difficult arousals	Polycthemia
Hypersomnolence	Impaired intellectual function
Difficulty with concentration	Depression
Frequent nightmares	Decreased libido
Nightmares about breathing difficulties (e.g., suffocation, drowning)	Weight loss or gain
Congestive heart failure	Muscle aches
	Memory impairment
	Poor control of airway secretions

\* In order of appearance.<sup>567</sup>

oxygen therapy, CPAP, or bilevel PAP at insufficient levels to fully rest or adequately assist the inspiratory muscles during sleep or otherwise (see below). In reality, the only tests needed to monitor sleep are oximetry and end-tidal carbon dioxide. Polysomnography is warranted to evaluate symptoms only when the VC and maximum inspiratory pressures are essentially normal and nocturnal  $\text{ErCO}_2$  and  $\text{SpO}_2$  are unremarkable despite symptoms.

### Overreliance on Arterial Blood Gases

Arterial blood gas analyses are not needed for medically stable people with NMDs (Chapter 11). Pain from the arterial stick causes hyperventilation and can cause falsely low carbon dioxide tensions 30% of the time.<sup>646</sup> Furthermore, unlike the use of end-tidal carbon dioxide and oximetry (which are practical and painless methods of blood oxygen and carbon dioxide analysis), an arterial blood gas analysis gives only a one-point rather than a continuous look at the body's gas exchange. Without hospitalization and the painful and hazardous placement of an arterial catheter, invasive blood gas analyses cannot be monitored during sleep, the most important time of all.

### The Most Common Conventional Treatment Approaches

1. "Palliation" with narcotics, sedatives, and supplemental oxygen to lighten and hasten death while obtaining "advanced directives" or "informed consent" indicating the use of "heroic measures," generally intubation or tracheotomy.
2. Treating ventilatory insufficiency and episodes of acute ventilatory failure as though they were respiratory insufficiency—with some combination of oxygen, bronchodilators, chest physical therapy, airway suctioning, and, ultimately, intubation and tracheotomy.
3. "Prophylactic" tracheotomy.
4. Using nocturnal bilevel PAP at low inspiratory and expiratory pressure spans that are ultimately inadequate to avert ventilatory failure, intubation, and tracheotomy.

### Easing and Hastening Death or "Not-so-benign Neglect"

Prescribing oxygen therapy with or without morphine is an especially popular approach with clinicians who do not offer noninvasive respiratory aids and who preach "therapeutic nihilism."<sup>644</sup> This approach, commonly used for amyotrophic lateral sclerosis (ALS) and motor neuron disease (MND), decreases dyspnea while hastening hypercapnic narcosis and death.<sup>647-648</sup> Eighty-two percent of patients with ALS die while receiving morphine, and 64% receive benzodiazepines; few if any are provided with noninvasive aids in Great Britain.<sup>649</sup> Often, without consulting the patient, the physician judges the patient's quality of life as unacceptable and the disease as terminal, ignores options that prevent respiratory complications, and biases the family against any ventilator use, which the physician usually associates with tracheostomy.<sup>643</sup> This approach emphasizes "palliation" but results in hopelessness and mental anguish. Patients die from carbon dioxide narcosis, aspiration of airway secretions, or an intercurrent chest cold that results in pneumonia and acute respiratory failure.<sup>14,650</sup> If morphine levels are not high enough to obtund the patients and they become short of breath, they

### Treating Ventilatory Insufficiency Like Respiratory Insufficiency

Conventional approaches to managing ventilatory insufficiency are identical to those for respiratory insufficiency and include supplemental oxygen administration, bronchodilators, and chest physical therapy, as is typical for chronic obstructive pulmonary disease (COPD). The goals of medical therapy can be to increase diaphragm contractility, to dilate the airways and facilitate airway secretion clearance, to improve ventilatory drive, and to improve oxygenation without exacerbating hypercapnia. Unfortunately, no medical therapy has been shown to be effective and safe for any of these goals for patients with NMD.<sup>651</sup> It has also been suggested that tricyclic antidepressants or monoamine oxidase inhibitors (e.g., safrazine hydrochloride) can be used to suppress rapid-eye-movement (REM) sleep to correct nocturnal hypoxemia in patients with DMD.<sup>652</sup> Nocturnal REM hypoxemia, however, is often the first sign of nocturnal hypoventilation and is rarely symptomatic until it progresses to cause long periods of nocturnal oxyhemoglobin desaturation ( $\text{dSpO}_2$ ) and hypercapnia. When patients become symptomatic, noninvasive ventilatory assistance is appropriate rather than further sleep disruption.

Bronchodilators, although often helpful for mobilizing peripheral airway secretions during intercurrent upper respiratory infections, are too often used on a routine long-term basis with no subjective or objective benefits. Bronchodilators often cause anxiety and, by increasing heart rate, can exacerbate cardiac dysfunction in patients with cardiomyopathies. Bronchodilators can even increase production of airway secretions.

Oxygen therapy became widely accepted for treating hypoxia in the mid-1960s. It has been demonstrated to improve survival significantly for patients with COPD.<sup>653</sup> This success and general familiarity with oxygen therapy have led to its use for treating hypoxia due to ventilatory impairment. However, considering the pathophysiology of ventilatory impairment (Chapter 9), such patients fare poorly because hypercapnia is exacerbated. Oxygen supplementation for such patients results in a significantly higher incidence of pneumonia, hospitalization, and hospitalization days than for untreated patients or when respiratory muscle aids are used.<sup>14</sup> In addition, because of its relatively high pressure gradient for absorption by comparison with nitrogen, supplemental oxygen can cause and help to maintain atelectasis.<sup>654</sup> Most patients with NMD who become comatose from carbon dioxide narcosis do so while receiving oxygen supplementation rather than assisted ventilation. Oxygen should not be used for people with NMDs other than for acute resuscitation, unless they are hospitalized in intensive care and monitored intently for intubation.

Although most conventionally managed patients avoid carbon dioxide narcosis, they inevitably undergo tracheotomy or die from retention of airway secretions during chest infections because of failure to teach and equip them to use mechanically assisted coughing (MAC).<sup>655</sup> Attempts to suction patients' airways via the nose or mouth are rarely effective and can further impair breathing. There is even less chance to enter the left main stem bronchus in this manner than with suctioning via a tube. Lung suctioning via the upper airway should rarely if ever be required or used as an alternative to MAC.

### Prophylactic Tracheotomy

Because a higher rate of complications is associated with emergency rather than elective tracheotomy, and because respiratory failure is inevitable for conventionally man-