Oxygen is NOT for Hypoventilation in Neuromuscular Disease

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Editor's Note: The IVUN office continually hears from people with post-polio or other neuromuscular diseases and conditions being inappropriately prescribed O_2 therapy. This anecdotal evidence (confirmed by the Mayo Clinic case series) supports the need for accurate information from the physicians most expert in the pulmonary aspects of neuromuscular disease to be disseminated more widely to alert people to the reasons why they should be wary of O_2 therapy.

If progressive respiratory failure occurs in people with neuromuscular disease, an abnormal nocturnal oximetry study is often an early indication that hypoventilation is occurring. There are significant periods of decreased oxygen levels in the blood or hypoxemia during sleep when lying flat, in addition to decreases in vital capacity (VC), maximum inspiratory force (MIF), and maximum expiratory force (MEF). Decreased oxygen saturation (SaO_2) combined with increasing carbon dioxide (CO_2) retention or hypercapnia are the hallmarks of hypoventilation. This is sometimes called ventilatory pump failure, due to the weakened respiratory muscles.

Patients with neuromuscular diseases who are developing progressive respiratory failure due to respiratory muscle weakness will die unless mechanical ventilation is used. The rate of progression is often hard to predict. Some patients seem suddenly to experience life-threatening hypercapnic respiratory failure. They may not have been aware of gradually increasing symptoms and signs, particularly since they are often not physically active and are often not being regularly monitored with simple pulmonary function tests.

Administering oxygen does not provide assistance to the weakening respiratory muscles, but gives both the patient and the doctor the false impression that appropriate treatment is being provided. While in fact hypoventilation is mistaken for an oxygen transfer problem. Indeed, administering oxygen can mask the problem. Also there is a danger of causing respiratory depression by giving oxygen. Oxygen is NOT the treatment for hypoventilation. It will improve the SaO_2, but not the hypoventilation and may increase the danger of dying of sudden respiratory failure.

In hypercapnic respiratory failure due to hypoventilation, the SaO_2 falls due to the rise of the CO_2. The alveoli in the lungs (tiny gas exchange units) should clear most of the CO_2 out with each breath. Instead, with hypoventilation, CO_2 accumulates and thus there is decreased room in the alveoli for oxygen. When mechanical ventilation using room air is provided, it lowers the CO_2 in the alveoli, corrects the SaO_2, and rests the respiratory muscles. The ventilator should be adjusted to

achieve a normal SaO_2, on room air. If oxygen is being administered, one cannot use noninvasive oximetry to tell whether enough assisted ventilation is being provided; repeated arterial blood gas specimens (ABGs) would be needed.

When there is respiratory failure in neuromuscular patients (ALS, post-polio, SMA, muscular dystrophy, etc.) who have no additional pulmonary disease that impairs oxygen transfer, the ventilator set-up is adjusted to:

- be comfortable for the patient;
- achieve SaO_2 of 95% or higher on room air (this can be measured with a finger-sensor oximeter);
- assist the patient to effectively cough and clear secretions;
- provide improved oral communication (if vocal communication is possible).

It has been common for people using noninvasive nasal ventilation (NPPV) with a bi-level positive pressure unit to use inadequate settings; frequently, they are not monitored with clinical evaluation and oximetry. The EPAP is often set too high – usually it should not be higher than 3-4 cm H_2O; the IPAP is set too low – usually it needs to be 12-16 cm H_2O and adjusted to achieve an oxygen saturation of 95% or higher.

Some situations may require administering oxygen, such as pneumonia due to infection or aspiration. If this occurs in patients with respiratory muscle weakness and hypoventilation, then it is important to provide both assisted ventilation and supple-mental oxygen, and use ABGs to monitor them.

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Additional Observations about Oxygen in Neuromuscular Disease

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I agree completely with Dr. Oppenheimer that assisted ventilation is the appropriate therapy for alveolar hypoventilation. Apart from a limited number of situations such as pneumonia or lung fibrosis, oxygen therapy is usually inappropriate and may prove hazardous. Clearly, in an acute pneumonia O_2 therapy can be entrained into the ventilator system. Fortunately, in the United Kingdom, this message is getting across to healthcare workers and patients. There is still some inequity in providing noninvasive ventilation, but the situation is improving.

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The analogy that I often use in regard to patients with respiratory failure from neuromuscular disease is that their lungs are like a deflated balloon that they are not strong enough to inflate. To inflate the balloon, mechanical assistance to force air into the balloon is needed. Blowing oxygen across the mouth of the balloon (the equivalent of using supplementary oxygen delivered by nasal cannula) will do nothing to inflate the balloon.

The case series published by the Mayo Clinic (see reference to Gay & Edmonds, 1995) demonstrates the dangers of administering as little as 1 to 2 L/min of nasal cannula oxygen. Patients with a variety of neuromuscular disorders experienced marked CO_2 retention; several became obtunded and required intubation or died when they were placed on 0.5 to 2 L of nasal cannula oxygen.