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Navigating Neuromuscular Diseases: CHEST Guidelines for Respiratory Care

Announcer:
You're listening to *NeuroFrontiers* on ReachMD, and this episode is supported by UCB. Here's your host, Dr. Charles Turck.

Dr. Turck:
Welcome to *NeuroFrontiers* on ReachMD. I'm Dr. Charles Turck, and here with me today is Dr. Bimaje Akpa, who's an Assistant Professor of Medicine in the Division of Pulmonary, Allergy, Critical Care, and Sleep Medicine at the University of Minnesota. Together, we'll be reviewing key recommendations from the American College of Chest Physicians clinical practice guidelines for managing respiratory complications in patients with neuromuscular diseases. Dr. Akpa, thanks for joining me today.

Dr. Akpa:
Yeah, the pleasure is mine.

Dr. Turck:
Well, to start us off, the guidelines suggest that pulmonary function testing is key in assisting with treatment decisions. For instance, performing a polysomnography can help determine if noninvasive ventilation is the right option for symptomatic patients with normal overnight oximetry and function test results. So with that recommendation in mind, Dr. Akpa, how important is testing in mitigating the respiratory complications that patients with neuromuscular diseases experience?

Dr. Akpa:
Evaluations and testing are very important. The respiratory complications related to neuromuscular disease is usually due to hypoventilation. So the screening and the detection in hypoventilation early on is one of the core principles of managing these patients.

Now, one of the most vulnerable periods where hypoventilation can be very evident is usually during a period of supine REM sleep. And so polysomnography can help detect the onset of hypoventilation. Having said that, in clinical practice, the availabilities of sleep labs and the cumbersomeness of getting sleep labs on all our patients is not really applicable. So that's where overnight oximetry tests and pulmonary function tests are very comparable alternatives for detecting hypoventilation in these patients.

Dr. Turck:
So now that we've discussed testing, let's move on to treatment. The guidelines recommend noninvasive ventilation for patients with neuromuscular disease and chronic respiratory failure or sleep-related disorders. But how can patient factors such as rate of disease progression impact putting that recommendation into practice?

Dr. Akpa:
So the use of noninvasive ventilation is key in managing neuromuscular disease patients. However, you have to tailor the use of noninvasive ventilation to the needs of the patients. So for example, the guidelines would suggest that noninvasive ventilation should be started on neuromuscular disease patients when setting criteria are unmet based on their respiratory parameters. However, if I have a rapidly progressive disease such as ALS, then I'm more likely to start noninvasive ventilation early on. And in fact, the pulmonary function test will actually be done even more frequently, although the guidelines suggest that they should be done every 6 months. On the flip side, if I have a slowly progressive neuromuscular disease such as Duchenne's dystrophy, then those patients are more likely to start a noninvasive ventilation later, and the frequency of the testings will be less frequent. Sometimes we can even go as far as just doing it annually.

Dr. Turck:
And then once a patient is receiving noninvasive ventilation, the guidelines recommend adjusting parameters like mode of ventilation, inspiratory and expiratory pressures, and inspiratory time to help optimize a patient's outcome. So what role does ongoing monitoring and assessment of these parameters play in ensuring that the treatment is effective?

Dr. Akpa:
You can imagine that patients have their own intricate neural breathing system, and then when you apply an external system, whether it's a noninvasive ventilator or mechanical ventilation, it's key that the ventilator and the patient are in synchrony. And so when we start noninvasive ventilation, we need to ensure that the patients are tolerating the settings. And in fact, the prescribed settings are doing exactly what the clinician wants it to do. So, for example, when we start noninvasive ventilation on patients, we're trying to mitigate the work of breathing. So a very good parameter you can get on the downloads is a rapid shallow breathing index. And if we find out that the number is a little bit elevated, we can always change the inspiratory time or increase the pressure support. We can always get an overnight oximetry test while those patients are using the noninvasive ventilation to make sure that there is no evidence of nocturnal hypoventilation. So it's very important that we make those adjustments: one, to ensure that the ventilatory goal is being met, and two, to ensure that there's a good synchrony between the ventilator and the patient to increase their compliance and tolerance.

Dr. Turck:
For those just tuning in, you're listening to *NeuroFrontiers* on ReachMD. I'm Dr. Charles Turck, and I'm speaking with Dr. Bimaje Akpa about the most recent CHEST guidelines for respiratory management in neuromuscular diseases.

So now that we've discussed noninvasive ventilation as a first-line treatment, Dr. Akpa, let's turn to what the guidelines have to say about mechanical ventilation. This alternative is recommended for patients who are intolerant to or unresponsive to noninvasive ventilation, or for patients who have worsening bulbar function, frequent aspiration, insufficient cough, episodes of chest infection despite adequate secretion management, or declining lung function. Would you talk a bit about your experience using mechanical ventilation in patients like these?

Dr. Akpa:
I would say, over the last decade or even longer, there have been relatively less transitions from noninvasive ventilation to mechanical ventilation. And this is probably due to the fact that more recently, we've been able to use mouthpiece ventilation, also known as sip ventilation, in patients to support their ventilation. And so what we've seen is that a lot of patients are able to delay transition from noninvasive to mechanical ventilation by the use of a mouthpiece.

Having said that, transition to mechanical ventilation has to be an ongoing conversation in this subset of patients. If they're intolerant to noninvasive ventilation or have overwhelming secretions or bulbar functions, those discussions have to be had. And when those discussions are being made, they have to be in a patient-oriented basis and based on the patient's needs. We also have to make sure that patients and their caregivers understand that the demand for their care escalates when they go from a noninvasive ventilation to a mechanical ventilation.

A lot of those patients might actually even have to be institutionalized in LTACs and nursing homes, because they don't have caregivers that are able to take care of them. And as for palliative care, we need to understand that the placement of a tracheostomy will probably be required in all of the transition to a mechanical ventilation. So all of these conversations have to be made prior to this transition.

Mechanical ventilation can actually improve survival and prolong life in these patients, as well as increase sleep quality. And so it's not a hopeless situation transitioning from a noninvasive ventilation to mechanical ventilation.

Dr. Turck:
And now I'd like to shift gears and focus on select complications in patients with neuromuscular disorders. Starting with sialorrhea, anticholinergic medication is recommended as first-line therapy with either botulinum toxin therapy or radiation therapy to the salivary glands as an alternative if the patient is intolerant or unresponsive. So given that both treatments are viable options, how do you decide between botulinum toxin versus radiation?

Dr. Akpa:
Good question. The guidelines weren’t really clear as to how to choose one therapy over the other, and so those are therapeutic options. I would say that usually when you're choosing a therapeutic option as it pertains to the management of salivary secretions, you have to weigh the benefit over the harm. Now, some data suggest that the harm might outweigh the benefit in radiotherapy. And in fact, given that some of the adverse effects from radiotherapy could be debilitating and very permanent, I would say that we should be very selective in having patients perform these therapeutic options, particularly radiotherapy, in tertiary centers that have the volumes and the expertise. In my clinical practice and at the University of Minnesota, we tend to send our patients for the botulinum toxin injections

because we have the expertise and it's easily done.

Dr. Turck:
As we near the end of our program, I'd like to ask you about one more complication, which is reduced cough effectiveness. The guidelines recommend manually assisted cough techniques as initial treatment, which can be combined with lung volume recruitment or mechanical insufflation/exsufflation. What can you tell us about these options and the considerations that accompany them, like their associated costs and caregiver assistance and training?

Dr. Akpa:
So these are therapeutic options; there's no specific option that is superior to the other and they’re not mutually exclusive. Having said that, in clinical practice, you can actually combine some of these options to get superior effects and benefit. I would say, when choosing an option, one of the things you want to keep in mind is the independence of the patient and the presence of a trained caregiver. For example, glossopharyngeal breathing, also known as frog breathing, is one of the means of lung volume recruitment. And this can be done by the patient, and you probably don't even need a caregiver to be able to do that. And you can do this multiple times during the day, and it's easy to do. So I would usually choose this in my patients who are able to do that.

Having said that, a lot of the other choices will be determined on the dexterity of the patient. The presence of intellectual disability may make it difficult for some of these options to be applicable. The use of other lung volume recruitment, whether it's mouthpiece or resuscitation bag, will actually require the presence a trained caregiver. But again, it's available and it's not expensive. Now, when you begin to use the mechanical insufflation/exsufflation, that's where things can get really expensive. And not only is it expensive, you would also need the presence of a caregiver to be able to use it. And so those are the factors that play into whether you're choosing one or the other, dependent on the presence of a trained giver or your local resources.

And then one other last one that's usually used very frequently is the high frequency chest wall oscillation. That can be used with a manual cough assist, whether it's a lateral compression or an abdominal thrust. Combining those two has actually been shown to be more effective compared to using each of one separately.

Dr. Turck:
Now, we only have a few moments left, Dr. Akpa, so before we close, is there a call to action you'd like to give your colleagues concerning these guidelines?

Dr. Akpa:
I would say that the guidelines are key and are very timely. And just as I had previously said, these guidelines have to be tailored to the individual need of the patient and the type of neuromuscular disease. And so depending on your local resources, you may choose one or the other. Having said that, the quality of the evidence and the data were quite limited. And so clinicians should feel free to tailor those. Those guidelines actually serve as a guide, but clinicians should be able to tailor those to fit the individual needs of their patients.

Dr. Turck:
Well, as those final comments bring us to the end of today's program, I want to thank my guest, Dr. Bimaje Akpa, for joining me to review the American College of Chest Physicians clinical practice guidelines for managing respiratory complications in patients with neuromuscular diseases. Dr. Akpa, it was great speaking with you today.

Dr. Akpa:
Thank you.

Announcer:
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