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Optimizing Neuromuscular Disease Care: How to Assess and Address Respiratory Risks

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

Dr. Turck:
This is *NeuroFrontiers* on ReachMD, and I'm Dr. Charles Turck. Joining me to share best practices for evaluating respiratory function in patients with neuromuscular diseases and mitochondrial myopathies are Drs. Jason Ackrivo and Bethany Lussier. Not only is Dr. Ackrivo the Associate Director of the Fishman Program for Home-Assisted Ventilation, but he's also an Assistant Professor of Medicine in Pulmonary, Allergy, and Critical Care at the Hospital of the University of Pennsylvania. Dr. Ackrivo, welcome to the program.

Dr. Ackrivo:
Thanks so much for inviting me.

Dr. Turck:
And Dr. Lussier is an Associate Professor of Internal Medicine at UT Southwestern Medical Center and a member of its Division of Pulmonary and Critical Care Medicine. She also serves as Director of the Neuromuscular Pulmonology and Home Ventilation Clinic at Parkland Health. Dr. Lussier, it's great to have you with us as well.

Dr. Lussier:
It's a pleasure to be here. Thank you.

Dr. Turck:
Well, to start us off, Dr. Ackrivo, why is it so important to proactively detect respiratory decline in patients with neuromuscular diseases and mitochondrial myopathies?

Dr. Ackrivo:
Well, in individuals with these diseases, the most common cause of morbidity and mortality is respiratory failure, and the earliest manifestations of respiratory impairment can be very subtle, occurring overnight before daytime symptoms develop. Patients may not be as forthcoming about respiratory symptoms because they feel that they might just be a part of the disease. But we do have treatments that can help improve quality of life and survival, and this includes non-invasive ventilation—often referred to as BiPAP—and airway clearance devices, sometimes called cough-assist device or mechanical insufflation/exsufflation. And early treatment can help prevent complications down the road such as pneumonia, hospital admissions, and respiratory failure.

Dr. Turck:
So given that importance, let's turn to you, Dr. Lussier. What are the key indicators of respiratory decline that we should be aware of?

Dr. Lussier:
Everybody who's experiencing a disease like mitochondrial myopathy experiences something a little different, and it depends how much of a burden the disease has presented over time. And so people adapt, right? Sometimes people will become short of breath with simple activity, and that's a flag for us. Sometimes it's not so obvious. Sometimes it's just that you're sleepier during the day or you're having trouble concentrating, indicating that you're getting into this pattern of shallow breathing. And these signs may represent problems with breathing—like shallow breathing—that are perpetuated and actually become exponentially worse overnight. And so those are the main things we look for.

Issues like bulbar weakness—which affects mostly the oral muscles—may affect somebody's ability to clear their airway if they have mucus or food in there. Issues with swallowing may also be signs. Other things we look for are if you're having difficulty projecting your voice and shouting over distances or if you're shortening your sentences. These are simple things that families may pick up on even before a patient does, like difficulty activating a remote sensor across the room and things like that. The subtle signs are the signs that we've adapted to for a while, and other people tend to pick up on it a little bit faster. It gets a little complicated with mitochondrial diseases, because sometimes people breathe more shallow and fast because of an underlying acidemia, which is different from most other myopathies. And so we take that into account as we address this in the clinic setting.

Dr. Turck:
For those just tuning in, you're listening to *NeuroFrontiers* on ReachMD. I'm Dr. Charles Turck, and I'm speaking with Drs. Jason Ackrivo and Bethany Lussier about the key indicators of respiratory decline in patients with neuromuscular diseases, and the critical importance of detecting those signs early.

Now, when it comes to evaluating respiratory function in patients with neuromuscular diseases, the CHEST guidelines recommend spirometry with forced or slow vital capacity, maximum inspiratory or expiratory pressure, sniff nasal inspiratory pressure, and peak cough flow. With that said, Dr. Ackrivo, would you tell us about your experience assessing patients?

Dr. Ackrivo:
I think that these tests are important. They're well described in the literature, but any physician caring for these patients has to make sure that they evaluate the entire patient and have a high index of suspicion that they could have respiratory impairment just due to the nature of having one of these diagnoses. As I mentioned earlier, respiratory impairment could be very subtle, and it could affect them overnight before it affects their symptoms during the daytime. The tests that you mentioned are very reproducible. Some of them have been shown to correlate with overall mortality, and so measuring them regularly is important, as trends can be used to evaluate changes in how much their respiratory function has been impaired by the disease.

But again, remember that the tests that you mentioned are all tests done in the clinic during the day, and you have to have a high index of suspicion that there could be impairment of their breathing while they're sleeping. So nocturnal testing will be important as well. Most commonly, that's done using nocturnal pulse oximetry, because that's what the guidelines describe. But also recognize that nocturnal pulse oximetry does not measure carbon dioxide levels, and a person can certainly have nocturnal hypercapnia without having nocturnal hypoxemia.

Dr. Turck:
And as a follow-up to that, Dr. Lussier, how can these assessments help inform treatment decisions?

Dr. Lussier:
Knowing a patient's baseline is actually really important, because, as we said, not everybody's going to present the same way. And so as soon as somebody has a diagnosis that puts them at risk for shallow breathing or any sort of hypoventilation syndrome, we start screening with any one of those tests or multiple of them. Depending on the interpretation, it may guide you to next steps. If there is a trend towards an abnormality, if you are repeating them yearly and you find that there is a downtrend that exceeds what is normal or expected, or if somebody is going through a stressful life event, you may see a sudden decline in one of those numbers, which may prompt us to do an earlier screening or intervention. For example, if I see somebody's forced vital capacity has dropped by 15 to 20 percent or is nearing 60 percent, I may push that patient to either get an overnight oximetry or even pursue a sleep study, depending on my resources, and say, 'I'm seeing early signs of hypoventilation that are most present at night, and I think we can make a move here and get you acclimated to non-invasive ventilation or BiPAP at night so that we can make an impact in long-term care.'

And so those early interventions and discovery is really important based on these tests. Increasing the frequency of testing may be necessary when somebody's going through an acute illness, hospitalization, or perioperative planning.

Dr. Turck:
Now, we're almost at the end of our program, so before we close, I'd like to hear from each of you one last time. Starting with you, Dr. Ackrivo, what are some common challenges in evaluating the patient's respiratory function? And how do you overcome them?

Dr. Ackrivo:
I think one of the biggest challenges is knowing when to start therapy. We have some general guidelines, and the United States has their own guidelines, which can differ from what you see in other international ones. And you also have to couple what the guidelines say with what the patient wants to do, because providing one of these therapies to a patient requires an extensive amount of buy-in from the patients and their caregivers.

There are some limitations to the testing that we mentioned earlier. The daytime testing, such as forced vital capacity or inspiratory pressures, are a static measurement that represent a snapshot in time and don't necessarily correlate with how well the patient's breathing at other parts of the day or even overnight. As Dr. Lussier mentioned earlier, the bulbar function can be affected, so there are limitations to the accuracy of the testing if they have significant bulbar impairment. And also remember that these tests are really a surrogate for ventilation, which refers to carbon dioxide levels, and that's not always so easy to measure, especially in the outpatient setting. You can compensate for bulbar impairment using things like an anesthesia mask rather than a cylindrical mouthpiece. And you should also be aware of some of the respiratory tests that are not affected by bulbar dysfunction, such as sniff nasal inspiratory testing, pulse oximetry, and if available, transcutaneous carbon dioxide monitoring. You can also do diaphragm ultrasound as well.

Dr. Turck:
And how about you, Dr. Lussier, do you have any best practices for overcoming challenges and optimizing respiratory function assessments?

Dr. Lussier:
There's always a problem with overcoming some limited resources in certain patients. But once they're initiated, I would say optimizing therapy is considered best practice—and easier said than done—as well as recognizing where the pitfalls may be in your approach to practice, and then escalating and adapting over time, recognizing that not one size fits all. So the reactive component to it and finding the right healthcare team to address the problems that are inevitably going to creep up over the long term is what's necessary.

Dr. Turck:
Well, as those final comments bring us to the end of today's program, I want to thank my guests, Drs. Jason Ackrivo and Bethany Lussier, for joining me to discuss the latest evidence-based strategies for assessing respiratory function in patients with neuromuscular diseases and mitochondrial myopathies. Dr. Ackrivo, Dr. Lussier, it was great having you both on the program.

Dr. Ackrivo:
Thank you.

Dr. Lussier:
Thanks so much for having us.

Announcer:
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