

Management of Respiratory Symptoms

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00:04

Good afternoon and welcome to this session. My name is Rebecca Moher and today I will be moderating, and we'll be talking about the management of respiratory symptoms from our special guest, Dr. Hollis cheney. To tell you a little bit about her Hollis or Holly cheney MD is an assistant professor in pediatrics at the George Washington University School of Medicine and Health Sciences, and the vice chair of the Division of pulmonary medicine at the Children's National Hospital. She is a board she is board certified in pediatric pulmonology. Dr. Cheney has been practicing in the children's division of pulmonary medicine for over 25 years. She is particularly interested in cystic fibrosis and rare diseases. We're also fortunate to have Dr. Cheney as a member of the Moebius syndrome Foundation's scientific advisory board. So welcome Dr. Cheney. Thank you. And would you like to say anything really quick before we get started? I just



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that, um, why do I like Mobius? Why am I working with it? Because I like working with rare diseases, because it's hard to do studies on and since they are rare, and I found that you can apply a lot of the knowledge from working with other diseases to Mobius. And it works quite well.



01:28

Excellent. And just a quick note, before we get into the content, if you do have a question for Dr. Chaney, please enter it into the q&a chat box. And we'll certainly try to answer it at

the end of the session. And also this session is being recorded and we will be at it will be available on the Moebius syndrome Foundation's website. And that'll be the on the resources page shortly after we finish. Okay. So I'm going to go ahead and get started. And I've got a handful of questions that were submitted ahead of time as well. So Dr. Chaney, first question for you is, you know, what area Do you specialize in?



02:04

pediatric pulmonary, so kids lungs, although, as I said, you can apply information about working with one disease to another, but that also works with adults. So if it involves the respiratory system in an adult, it still works. So I can I it's a children's hospital, but sometimes I see adults also. Excellent.



02:27

And specifically, Where do you work? You know, we're all in this virtual space right now. And everybody's curious to know exactly. Where do you practice? Where do you work?



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So I work at a children's National Hospital, which is in Washington, DC, and and we have satellite clinics in Maryland.



02:45

Excellent. And how do you be how did you become involved with Moebius syndrome?



02:52

So to be honest, um, as I say, I like to work with rare diseases. And I contacted the Nord, which is National Organization for rare diseases and asked them if there were if they knew of organizations that needed help from a pediatric pulmonologist and they said, Moebius.



03:17

Excellent. And so I think, you know, one of the questions that we get a lot is, you know, why are the lungs involved in Moebius syndrome.



03:26

So there's a lot of variety in Moebius because it hits it hits different areas, but the what it hits the biggest is the cranial nerves of face in the throat. And so it makes it hard to swallow to chew to suck, and in some cases, even cough. And so because of the difficulties with swallowing, a lot of the people end up aspirating they get food into their lungs, and from intermittently or long term getting queued into lungs, they end up with chronic lung disease.



04:05

And so how would someone avoid aspiration then?



04:10

So So the issue is partly a matter of coordination and if you drink anything too fast, it's gonna slip right down into the lungs. If you can slow down what's your game taking the end like with a parade food or a second food, you're you're back of your throat can handle it better and can make try and avoid it getting into the lungs. So the so like for infants, they should use a nipple with a very small hole so they can't drink fast. for toddlers, they can use a sippy cup with a small hole for the school age and the older it's a matter of when using an Open Cup not to cope never ever go up and never drink too fast. If it goes too fast. If you're coughing when you drink, it's going in your lungs. So Try not to drink too fast. The other thing that can be done is the second the liquid, which sounds pretty bizarre. I mean, some, some, some kids, they don't have any choice, even even if they try and drink water because they want water equals in their lungs, but you can put a thickener in it that doesn't have any taste. And then they can they can manage to get it into their, into their stomach instead of into their lungs.



05:25

Interesting. Okay, so then, you know, another one of the questions that was submitted ahead of time is, you know, how can they cough so much and especially with colds.



05:35

So what happens when you have chronic aspiration when you're getting your food intermittently into your lungs, or long term into your lungs, it irritates the lungs, it gives a lot of inflammation to the lungs, and often actually even bacteria because stuff going

from the mouth into the lungs has bacteria in it. So in essence gives a lot of people asthma, it's called reactive airway disease, meaning the lungs are irritated. But it's the same thing as asthma. It's just because of the aspiration. So it's easy for me to just call it asthma. But in other words, many of the people have asthma or reactive airways disease. Let me explain to you what that means. That That means that their lungs are very irritable and certain things are going to make are going to trigger coughing or trigger troubles breathing and the most common would be with a cold. When you get a cold, you're gonna cough a lot. When you go to cold, you may have trouble breathing, exercise can so particularly vigorous exercise, running very hard can trigger coughing or troubles breathing, and that probably relates to the weather. The weather is very irritating to the lungs, in particular the extremes. So like for instance, exercising in the really cold air, or exercising in a really hot air like in the summertime, it's going to just close things down that much faster, it'll be that much harder. Um, other things that can irritate the lungs. emotions can laughing crying can trigger coughing. odors can obviously smoke and people should stay away from smoke and never ever ever smoke. And by the way, or vape. But even perfumes are chemical cleaners like bleach, they can irritate the lungs and trigger coughing or troubles breathing. And then allergies. Now not everybody has allergies. But if you do have allergies, the allergies will aggravate the asthma so it causes more troubles with coughing and breathing. So what do you do about it? So there's there's two different medicines that you can take to help with the symptoms. When you have reactive airways disease, there's muscles around the outside of the airways. And when the lungs are irritated the muscles squeeze the airways and make it hard to breathe and make it cough. So there's an inhaler. And for little kids, they can use a nebulizer and the inhaler is a medication called albuterol. Which when you use it, it it relaxes the muscles around the airways and opens up the lungs, it helps to cough, it helps the breathing and you can actually think of it as a cough medicine when people are coughing like this medicine helps. The trick to using an inhaler is to always always always use a chamber with it, it can be any kind of a spacer, it doesn't have to be a particular brand. But to put the inhaler inside of the chamber and breathe in very slow. Now with little kids, they use a mask. I found over time you have to watch them do it because sometimes they breathe through their nose and not their mouth and they have to open their mouth or they're not getting any any medicine the noses make them walk out medicine. And anybody any age should use a spacer because you get a better dose with a spacer. So since it's your squirt man into a chamber, it's partly coordination. But it's partly that there's more medicine inside that chamber. And if you breathe back and forth a few times you get more into your lungs. The second medicine is is an inhaled steroids. Because of the chronic aspiration because of some people get recurrent pneumonia. There's a lot of inflammation in the lungs and and there's a second inhaler or through the nebulizer called us at night, which helps get rid of the inflammation. For people who have only symptoms with colds, they can just take the medicines with colds but a lot of people need to stay on the inhaled

steroids long term because they always have inflammation in their lungs. And if they take the inhaled steroids on a regular basis, it'll keep the inflammation down, keep them more stable and keep them from getting quite so sick when they do get a cold.



09:46

Excellent. No, we appreciate that information. And, you know, I guess a follow up question to that you touched on that. You know how the lungs are irritable or they get irritated with weather changes and things like that. So, specifically, why is it that somebody gets short of breath with exercise.



10:05

So that that is from the reactive airways disease when you exercise the muscles around the airway, squeezey airway, so it makes it hard to breathe. And it's going to be harder with vigorous exercise and harder with the weather and taking the albuterol before exercise will help the tolerance taking it after exercise can open them back up after the fact.



10:30

Excellent. I appreciate the points that you made also about the spacer and the arrow chambers, etc. Because that was a question that was submitted ahead of time as well. So thanks for proactively answering that for us.



10:41

Just one little word of wisdom, some of the chambers and make a noise when you breathe through them. And the kids like to make the noise, the noise is there to tell them they're breathing too hard. They have to slow down, it needs to be a very slow, deep breath. If you hear the noise, they need to slow down.



11:01

Good points. Definitely good points. Okay, so another question that came in was, how do I keep getting? or Why do I keep getting pneumonia? And how do I keep from getting recurrent bouts of pneumonia.



11:14

So the issue is that as you get older, and you get more inflammation in the lungs, and particularly depending on how much you're getting food into the lungs, that the little kids tend to get it into their lungs more than the as you get older, as you get older, you tend to learn how to handle it. But if you think about it, if you if food gets in the lungs, when you drink, if you ever get sick and vomit, you can't control that, and that gets into the lungs too. So it's over time you get more and more in the lungs. And it sets up these areas in the lungs that are they're kind of like pockets of mucus, it's full of mucus full of inflammation. And as long as that's in there, then the bacteria get in there and just set up camp. And also when you ask right, you got bacteria in your mouth that can get down into the lungs. So so when you get a cold you have you have more mucus up in your upper airway, you automatically get more in your lower airways. And then because there's bacteria in there, it turns into a lot of cough, a lot of congestion and even a pneumonia. So the trick is, when you start to get a cold start on the inhalers right away, start on the albuterol to open the airway start on the steroids to get really inflammation. There's something called chest PT, chest PT is just cupping the hand and clapping on the chest. If you can do it on yourself in the front, but you might be someone else to do it on your back. And it literally just kind of knocks that mucus out of there. So you can cough it out. Some of the people with Mobius don't have a real strong cloth. So it helps to get that extra pounding on their back to help mobilize the secretions. And if you're at all prone and ammonia and you're starting to cough up mucus, you can get one of the factors that gives you an antibiotic early in the course so it doesn't turn into an ammonia. If you get recurrent pneumonia, then it will happen again. So if you started to cough up mucus, then they should hopefully be willing to treat you.



13:22

Alright, very good. So another question that we tend to get a lot in the Moby's communities around sleep and sleep disorders. And, and and so a question that came in was, you know, why do I have trouble sleeping? And really, what can I do about it?



13:38

Okay, so that's actually a kind of a complex question. And I'm gonna break it down into four reasons. The most common, we're seeing a lot of these days with COVID, that people don't have a bedtime. And they go, I have very irregular hours, and they just upset their whole body sleep cycle, and they'll go to bed at 10 one night and to another night and get up all different hours, and then they'll take naps. And it just it just their body gets so

confused that they can't sleep. And that happens a lot in teenagers. Obviously, just getting into a normal cycle helps that one that one's not that hard. It's just behavior, having a bedtime and trying to keep regular hours. The second reason which pertains to Moebius is obstructive sleep apnea obstructions. obstructive sleep apnea means you're closing off the top of your windpipe while you're breathing when you're asleep. And so you have a hard time getting the air into the lungs. So like little kids will do that around 234 years old, because they have large tonsils and adenoids, and it blocks the airway. You can see when they're when they're snoring, they're laying down their snoring, they're pausing, they're gasping, they're having they're tossing and turning. They're all over the bed because they're trying to open their airway. In that case, taking out their tonsils and adenoids help With Mobius, some of them have abnormal shape to their jaw, or their pharynx. And sometimes EMT can find a way to open it up a little bit more. The next step is if there's nothing you can do that can actually correct the anatomy, there is a way to give positive pressure at night positive pressure would be through a machine that either goes over the the face like seat pepper, bipap, or through a trick, which is a hole in the neck. positive pressure into the airway through the trunk. That's sort of last case scenario. Um, the other one is obesity. And we got to be careful about obesity, because it just puts more tissue into the airway. And it makes more likely to have obstructive sleep apnea. So just trying to make sure there's not too much weight. The fourth reason for not sleeping well is central sleep apnea central sleep apnea is where the brain does not tell you to breathe properly. And some people with Mobius have this it tends to show up very young and it is much worse with sleep than it is awake. And these babies usually need actually positive pressure ventilation. And often a tray can have a ventilator to keep them breathing normally. And then the last reason for poor sleep is depending on how much lung disease there is, if there's quite a bit of lung disease where so we can we can compensate a lot when we're awake that we can't compensate when we're asleep. And if we don't have good lung function we name it may mean oxygen with sleep, but it won't show up when you're awake. And so if you need an oxygen, you may have very restless sleep and not feel rested in the morning, and just purely using oxygen at night can help that part of it. Excellent. Well, there's



16:50

a lot there. So I might need to come back to a couple of these points here. So regarding the regular bedtimes and COVID. And, you know, the body being confused, any tips that you can share there to help kind of normalize and make things regular? will power setting a schedule really important?



17:16

Yeah. So was the teenager the adult it's willpower with the child? It's the mother's willpower and being firm and making sure I know, you've taken away the the screens, the TVs, the games, the inputs, a teenager taken away the phones, they're on their phones all hours of the night, it's it's it's they call it a sleep hygiene issue, because they just let it happen and don't seem to care, but then it really does mess up the sleep.



17:46

Okay, well, that's that definitely helps. And then also, you know, appreciate that they extra information there about the obstructive sleep apnea. We've heard a little bit about that. So I think it's helpful to to hear more here. And you mentioned the central sleep apnea. Just a follow up question there. You know, any statistics or anything on how common that is? In the general population? I know it might be hard to boil that down to the Moebius population as well. Any additional comments there



18:17

were fortunately not fortunately not common and not common in Mobius. It's just a small sub part of it. And I say it say it usually shows up in infancy, because it's their brainstem, just not telling them to breathe effectively, and it's picked up in infancy.



18:35

Okay. Well, one of the questions that we often get also is when we do have physicians that present and, you know, share, a lot of different topics that the community has been involved or that have been interested for quite some time is, how do we contact you, if somebody wanted to meet with you make an appointment reach out to you for more information? How would they go about that?



18:59

So I do work at the Children's National Hospital in Washington, DC, and there's a they can go to the appointment line at that hospital, or I can even give our art our pulmonary number, which is 2476 to one to eight and they can make an appointment. The other thing is we because of COVID we all do everything virtually these days. I gotta say it's not all that easy sometimes to assess the lungs virtually. But a lot of what I do is based on history and actually been able to do quite a bit virtually.



19:36

Excellent. And let's see what else it looks like. We had another question that came in to the chat box. And that is should people with severe respiratory symptoms, be concerned about getting the COVID vaccine?



19:50

Yes, yes, yes. Yes. Yes. Can you maybe expand on it, they should get it they should get I would not I would be more concerned about Getting COVID and getting the vaccine, I have found the vaccine to be mostly side effect free and having seen COVID and the long term consequences is not about to get.



20:14

Okay. Yeah, I think we hear that a lot, a lot of a lot of encouragement to get the vaccine and keep everybody healthy. So it looks like I don't see any more questions that have come in. So maybe just one last call, see if there's anybody that had any additional questions out there. Let's see. Looks like no, I don't have any more. Any other tips that you'd like to share? Dr. Cheney?



20:46

Not really, but um, I am open to if people want to contact me just for more questions, you know, leave a message with our nurse or whatever. I'm happy to do that. I. I'm always I always want to be helpful if possible.



21:04

Yep. Absolutely. Well, we certainly appreciate that. So it sounds like if we don't have any additional questions, then it sounds like we're about at the end of our content and our session here. So we want to certainly extend a special thank you to all of our attendees today. And, of course, to Dr. Cheney for attending and sharing all this great information. We appreciate your time, and certainly your service to the Mobius community. And we know this session will be very helpful to the many folks that will view it in the future. And we certainly again, appreciate all your content and your time today, Dr. Cheney. So thank you very much, and have a great day, everyone. Thanks for joining

