

### Transcript Details

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### Experiencing COPD Cases: A Clinician's Perspective

#### Announcer:

You're listening to *Clinician's Roundtable* on ReachMD, and this episode is brought to you by Grifols. Here's your host, Dr. Charles Turck.

#### Dr. Turck:

This is *Clinician's Roundtable* on ReachMD. I'm Dr. Charles Turck, and joining me to discuss his experiences in managing patients with chronic obstructive pulmonary disease and alpha-1 antitrypsin deficiency, or AATD-associated COPD for short, is Dr. David Mares, who specializes in Adult Pulmonology and Critical Care in Anderson, Indiana. Dr. Mares, welcome to the program.

#### Dr. Mares:

Thank you.

#### Dr. Turck:

Let's begin with the patient case. Dr. Mares, would you share a memorable experience you had managing a patient with AATD-associated COPD?

#### Dr. Mares:

I'd be glad to. I have a couple of ladies that are called—their sisters—and they're called, in our office, the alpha twins. They're not twin sisters, but they're sisters. I worked with one of these ladies who worked in our building, and frequently would be seen, we'd see one another in the hallway and just say, "Hello." I didn't even know her name. But she one day stopped me in the hallway and asked if we could talk for a few minutes because her brother had been diagnosed with, as she described, a rare form of COPD with emphysema. And I asked if she thought that that was alpha-1 or if that was the term she remembered. And she says, "Yes." So we then walk to the office, and I screened her for alpha-1. On the way to the office, I had the opportunity to ask her questions about her symptoms. And she felt that she wasn't having significant shortness of breath or cough, sputum production, or anything else that would suggest a diagnosis of COPD.

In the meantime, she also told me that her sister would need to be screened, and we made arrangements for that to happen. And her sister basically, had the same discussion, really not symptomatic, just here to be screened.

After talking with their families and discussing this situation and hearing the questions I had to ask them, both of them basically, realized that they had been attributing their symptoms to other things, but they had actually been in a little bit of denial about the disease process; both of them had been short of breath, stories similar to, 'Well, my husband and I walk frequently, but he's having to slow down so that I can keep up,' or 'I used to carry my lab specimens in a backpack, but now I carry them in a wagon. I do get antibiotics a couple times a year for bronchitis episodes.' So really both of these ladies were already developing significant symptoms of COPD.

And both of these ladies have been in our practice now and on augmentation therapy for alpha-1 antitrypsin deficiency for now roughly 10 to 12 years. And it has been a great pleasure evolving our knowledge of the disease through caring for these two very delightful ladies. So we've spent some time together going and talking to a local college genetics class where they had the opportunity to communicate with the genetic students. And then the genetic students got a real experience with somebody who actually has a genetic disease so that they can expand their knowledge of genetics through that. And I presented the details, the clinical details of alpha-1 to the genetics class. We've had just a great time together. And the outcomes for both of these ladies have been very good. And particularly important is that neither of these two ladies would have felt that they were individually sick enough to seek out the care of a doctor. They were both in that denial phase of COPD that's so common. So we were able—based upon screening of family members—

we were able to get an early diagnosis in these ladies.

**Dr. Turck:**

Now why is it so important to identify this deficiency early in our patients with COPD?

**Dr. Mares:**

So our patients are experiencing oxidative damage to their lungs because they don't have alpha-1 antitrypsin to defend their lungs from neutrophil elastase. And so the length of time that they spend without augmentation therapy, it's an amount of time where additional damage is occurring to their lungs. So we'd like to catch them as early as possible before that additional damage has taken too much of their lung function. And we would like to be able to initiate therapy for them. And that therapy, not only augmentation therapy but modification of exposures, particularly getting off of cigarettes and changing one's life exposures so that we can minimize the oxidative damage and minimize the amount of lung tissue that they lose before we catch them and can minimize that loss.

**Dr. Turck:**

And, Dr. Mares, based on your experience, how should we approach detecting AATD to help identify early signs in our patients with COPD?

**Dr. Mares:**

So our problem is that we cannot, nobody, not me that has been doing this for nearly 30 years and has many alpha-1 patients, nor anyone else, we can't recognize what an alpha patient looks like because they share the same symptoms with all the rest of our COPD patients. And frankly, with many of our asthma patients as well. So we have to start the concept of screening these people and screening everybody that has COPD, screening the people that have asthma that's difficult to control screening, the family members, screening bronchiectasis patients. It's a matter of recognizing in our own practices that screening for alpha-1 is just as important as screening for glucose elevations, cholesterol, colon cancer, and breast cancer. We need to incorporate screening into our everyday practice with COPD patients and those others that I already mentioned.

**Dr. Turck:**

For those just tuning in, you're listening to *Clinician's Roundtable* on ReachMD. I'm Dr. Charles Turck, and I'm speaking with Dr. David Mares, who's sharing some of his experiences managing patients with COPD and alpha-1 antitrypsin deficiency, or AATD-associated COPD.

Now, Dr. Mares, shifting gears for just a moment and looking at potential treatments you touched on a little bit before, but once you've detected AATD in your COPD patient, how do you initiate therapy? And is every patient a candidate for augmentation therapy?

**Dr. Mares:**

So first, I would mention that patients that continue to smoke are not going to be a candidate for alpha-1 augmentation therapy because tobacco abuse neutralizes the benefits of the augmentation therapy. So we have to work very aggressively on discontinuation of smoking before we consider augmentation therapy.

Secondarily, we want to work on other risk factor modifications, anything that would induce oxidative stress, things like potentially, occupations of welding or other occupational exposures, we'd like to have an impact on, although, we wouldn't necessarily delay augmentation therapy based upon somebody's occupational exposures.

What I would suggest in general is that when people have the severe deficiency states that are considered for augmentation therapy, which would be the ZZ phenotype, the SZ phenotype, the null phenotype, or many of the others that might fall under severe deficiency states, that we then discuss the situation with the patient, talk to them about what augmentation therapy is like and the fact that it's a weekly infusion therapy. And then we start by applying through one of the available programs so that we can get insurance coverage for augmentation therapy. And that is a process that has a great degree of assistance through some of the pharmaceutical companies.

**Dr. Turck:**

Now once you find the right treatment approach for your patient, what are some strategies you use to help them manage this deficiency?

**Dr. Mares:**

Well, we have a patient who is going to be cared for by the infusion company at great degrees through the assistance with an alpha-1 patient that is going to communicate through AlphaNet to that patient and share experiences so the patient understands what they're going to be going through and a registered nurse that's going to communicate with that patient. So in my practice, I don't have much to do to continue to promote the infusion therapy. Once I initiate that and get that ball rolling, it goes upon its own course and it's well managed, incredibly well managed in the background.

So what's left for me to do is to talk to that patient about minimizing exposures about any oxidative exposures—getting away from the campfires or sitting a little further back, maybe potentially occupational changes, or just small changes, getting away from welding in indoor areas or getting all of the necessary vaccines that should be accomplished in an alpha-1 patient—because of course, alpha-1 also induces some abnormalities of the liver. And we need to do hepatitis B vaccine and hepatitis A vaccine so that the patient doesn't have additional stressors on their liver that might lead to potential malignancies, pneumococcal vaccines, influenza vaccines. I like to be more diligent in making sure that patient gets the necessary vaccines because those sorts of infections are going to also induce a potential for respiratory complications. But even more importantly, in the alpha patient, they're going to induce that white blood cell-rich situation with any of those infections that will lead to more oxidative damage. And our alpha patients, when they call and have an acute exacerbation, they're going to get antibiotics and steroids because those exacerbations, our white blood cell-rich events that cause an increased risk of oxidative stress.

So in essence, we practice the same pulmonary medicine with that variety of COPD that we do with all of the other COPD patients, but we're more diligent to get that patient free of oxidative stress as soon as possible.

**Dr. Turck:**

And finally, Dr. Mares, are there any key takeaways you'd like to share with our audience today?

**Dr. Mares:**

Yes. I think when we should talk about diagnosis versus screening. When I see a patient and I look at that patient now and say, 'Gosh, you look like you have alpha-1,' most of those patients, I still screen them, most of those patients don't. When I say to myself, I'm screening all of my COPD patients, then I find people that I wouldn't have necessarily recognized as high likelihoods of alpha-1 by their clinical features, yet they have it. And so that's why it's so important to view this as a disease process that needs to be screened for in all of the population that's appropriate for screening. And we've gone through that list already. So think of this, again, as important as screening for any of the other diseases that we screened for. And recognize that in that COPD population when we screen, we find these people. And we all are unfortunately, involved in the care of very sick long-term chronic patients. And with COPD, unfortunately, we all feel like we're holding the hands of these unfortunate people sometimes improving upon their symptoms, maybe some of the medicines are going to change exacerbation rates, and that sort of thing. But when we screen for alpha-1 and we find an alpha-1 patient among that COPD population, we now have an opportunity to change that disease process for them. And that provides me a lot of personal satisfaction to take one of those patients out of that otherwise very sick and declining population and bring them into a new course of therapy that's going to help them in the long run.

So it's something we can do that's wonderful for our patients. But it also provides us a lot of personal gratification to be able to recognize that disease process by screening and even help a small part of that population with other therapies that would otherwise be unavailable to them.

**Dr. Turck:**

Well, with those final thoughts in mind, I want to thank my guest, Dr. David Mares, for joining me to share insights from his experience screening for and managing patients with AATD-associated COPD. Dr. Mares, it was great having you on the program.

**Dr. Mares:**

It's my pleasure to help you all as clinicians to understand and help the other potential alpha-1 patients out there that are going to be screened for and diagnosed as a result of your efforts. Thank you very much.

**Announcer:**

This episode of *Clinician's Roundtable* was brought to you by Grifols. For information about Alpha-1 screening or to order a free AlphaID™ screening kit to rule out Alpha-1 deficiency in your patients, visit this episode's landing page and click on the 'Click here' link to access the order form. To access this and other episodes in this series, visit [ReachMD.com/Clinicians Roundtable](https://ReachMD.com/CliniciansRoundtable), where you can Be Part of the Knowledge. Thanks for listening!