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## How to Identify & Manage Patients with Progressive-Fibrosing ILD

Dr. Johnson: This is CME on ReachMD. I'm Dr. Shira Johnson and joining me to discuss Progressive Fibrosing ILD are our faculty from National Jewish Health in Denver, Colorado, Dr. Amy Olson, who is a Professor in the Division of Pulmonary, Critical Care & Sleep Medicine; Dr. Jeffrey Swigris, who is a Professor in the Division of Pulmonary, Critical Care & Sleep Medicine; and Katherine Rosen, a nurse practitioner in the same Division of Pulmonary, Critical Care and Sleep Medicine. Thank you for being here today.

Ms. Rosen/Dr. Olson: Thank you for having us.

Dr. Swigris: Thank you.

Dr. Johnson: Well, why don't we start with Katie. Can you tell us about an interesting patient, perhaps seen recently in your clinic by your team at National Jewish, maybe something about their symptoms, history, and workup?

Ms. Rosen: Of course. So, we've been following a 60-year-old woman with scleroderma-associated interstitial lung disease for several years. She was initially treated with cyclophosphamide for her first year of therapy, at which time her lung function stabilized. Unfortunately, use of the medication was associated with cytopenias and recurrent infections. We then transitioned her to mycophenolate, which she tolerated quite well. At present, she has been losing approximately 200 cc of her vital capacity per year for the last two years, despite mycophenolate, and with this, she is also more symptomatic, more breathless with an increased oxygen requirement, and she is concerned that her disease is progressing.

Dr. Johnson: What is the current treatment plan for this patient?

Ms. Rosen: Well, the current plan is to, of course, keep her on immunosuppressive therapy with mycophenolate, but we've also been discussing recent studies looking at both pirfenidone and nintedanib as a potential option, should either agent become FDA-approved.

Dr. Johnson: What are some of the limitations and challenges and quality of life issues that this patient is dealing with on a day-to-day basis?

Ms. Rosen: Well, first of all, regarding dyspnea, with her increased oxygen requirement, this is really difficult. With needing more oxygen, she is not able to get out of the house as much. Her oxygen will run out. She can't be gone from home for more than a few hours at a time. And, as well, despite her young age, she has severe esophageal dysmotility from her scleroderma and with this, is not deemed to be a transplant candidate. So, just coming to terms with, you know, her own mortality had a huge impact on her quality of life.

Dr. Johnson: Well, of course. Dr. Olson, can you tell us some more about what is PF-ILD?

Dr. Olson: Progressive fibrosing interstitial lung diseases are those interstitial lung diseases that we follow, and despite immunosuppressive therapy, tend to progress in terms of loss of lung function and increased oxygen requirement.

At this point in time, there's not a lot of information in literature about the incidence or the prevalence of progressive fibrosing interstitial lung disease. And I think at this time we need more epidemiologic studies to further define the prevalence, the incidence, and what this means in terms of morbidity and mortality in this disease entity.

Any disease pulmonary fibrosis is not good and has the potential to progress. A very thoughtful and attentive follow-up is necessary for all patients with diffuse fibrosing pulmonary fibrosis because we're still not great at predicting which patients with pulmonary fibrosis will have a progressive phenotype. We don't know which patients will progress rapidly, and we don't know which ones are destined to remain stable over a long period of time.

Dr. Johnson: Why is it especially important to identify these patients early?

Dr. Olson: Well, at this point in time, we do not know the optimal therapies to help fibrosis in those patients with a progressive fibrosing interstitial lung disease that don't have idiopathic pulmonary fibrosis. Thus, it's important to identify these people, should they be a candidate for lung transplantation. We do go through multidisciplinary discussions because we want to make sure we have them on the appropriate immunosuppressive therapy, and we want to make sure we identify patients that may be candidates for ongoing studies regarding antifibrotic therapy and those patients that do have a progressive fibrosing interstitial lung disease.

Dr. Johnson: For those just tuning in, you're listening to CME on Reach MD. I'm Dr. Shira Johnson, and today I'm speaking with Dr. Amy Olson, Dr. Jeffrey Swigris, and Katie Rosen about Progressive Fibrosing ILD.

Dr. Johnson: Can you please describe CTD-ILD and how this may represent a progressive fibrosing interstitial lung disease?

Dr. Olson: The incidence and prevalence of connective tissue disease-associated interstitial lung diseases, like progressive fibrosing interstitial lung disease, again is based really on limited epidemiologic data. I think the strongest data we have occurs in rheumatoid arthritis, as this tends to occur in approximately 1% of the population worldwide. And we know that clinically significant interstitial lung disease occurs in 10-20% of cases. We did a study here on rheumatoid arthritis-associated mortality and found that approximately 6.6% of all deaths were actually due to the underlying interstitial lung disease itself; thus, this phenotype for progressive fibrosing interstitial lung disease.

Dr. Olson: Connective tissue disease-associated interstitial ILD's well-described and, unfortunately, at this point in time, we don't know which patients will go on and have stability with the therapies we offer at this time, including immunosuppressive therapies, or which patients will have a progressive fibrosing phenotype.

Dr. Johnson: Dr. Swigris, what is the treatment or management approach for patients with PF-ILD?

Dr. Swigris: In terms of therapy for non-IPF progressive fibrosing ILD, we generally take a patient-by-patient approach and we're typically more aggressive with our immune suppression for those patients who have more severe disease, for those patients who have more rapidly progressive disease, and for those patients without significant comorbidities that would make such therapy harmful or overwhelmingly risky. So, there's no real cookie-cutter approach to therapy. It really does need to be individualized. Typically, we are talking about immune suppression in the form of glucocorticoids, either intravenous or oral, often in combination. We view it almost like vasculitis therapy where we typically use an induction phase, particularly in patients who are rapidly

progressive with severe disease, and we would use intravenous glucocorticoids in transition to oral glucocorticoids, and then we choose from a limited number of immune-suppressing or steroid-sparing drugs, like mycophenolate mofetil or azathioprine. We're tending to use cyclophosphamide a bit less frequently these days, although that remains in our armamentarium. We all think that antifibrotics could have a role here, and there are some really compelling data emerging on the use of antifibrotics in a number of forms of progressive fibrosing interstitial lung disease. In addition to immune suppression and drug therapy for these folks, we want to remember and not lose sight of nonpharmacological therapy. So, we want to look for comorbid conditions, like pulmonary hypertension, esophageal dysmotility, mood disturbance, sleep disorders, et cetera. We also want to get these people into pulmonary rehab, and we're constantly reminding our patients and community providers who refer patients to us that pulmonary rehabilitation is not just for COPD anymore, and there's plenty of data to suggest that patients with PF-ILD and other forms of interstitial lung disease benefit from pulmonary rehab. So, I can't think of a patient that should not go into pulmonary rehab. Staying up to date with vaccinations is also key, and I think regardless of the therapeutic regimen, both drug and non-pharmacological therapy, these patients need to be followed carefully over time.

Dr. Johnson: So, what do you think is particularly important for healthcare providers to know in order to better support patients with PF-ILD, and what really is the take-home message for the clinical practice of those taking care of these patients with progressive fibrosing interstitial lung disease?

Dr. Olson: I think that all patients with diffuse pulmonary fibrosis need careful follow up at 3-6 month intervals as you would with a patient with IPF. They also need ongoing management of their comorbidities and comprehensive non-pharmacologic management, including but not limited to pulmonary rehabilitation, oxygen therapy and support groups, and overall awareness of their disease process, very similar to what Dr. Swigris just said.

Dr. Johnson: And that's a great way to round out our discussion on how to identify and manage patients with progressive fibrosing ILD. I want to thank my guests for helping us better understand progressive fibrosing ILD and the therapeutic options for the management of PF-ILD. Drs. Olson, Swigris and Katie, it was great speaking with all of you today. Thank you for being here.

Ms. Rosen: Thank you for your time.

Dr. Olson: Thank you.

Dr. Johnson: I'm Shira Johnson with Reach MD. Thank you for listening.