Early Diagnosis and Detection of IPF

I try to highlight the fact that, by making an early diagnosis of IPF, we can allow early treatment. And we have interesting and quite solid data, mainly from the Impulses trials of Nintedanib, in idiopathic pulmonary fibrosis, showing that patient with preserved lung function, so patient with early disease, they benefit from treatment. And these patients, they do progress at the same rate as patient with more advanced disease.

So, in other words, the earlier the treatment, the better is the benefit for patient. And there are data coming from the UK, showing that tolerability of Nintedanib is better, when the drug is started with preserved lung function, when the drug is started earlier. So I think that we have initial data showing that probably we might improve patient outcome, by diagnosing these patients at early stages, and by starting treatment in these patients at early stages. Because they will have a reduced rate of progression. At the same time, they will tolerate the drug better.

So I think we need to move forward towards the concept that early diagnosis translate into early treatment, and translate into better outcome for our patients. The problem is that we see patients with IPF quite late in the course of disease, at the first time that we see them. And there are data coming from the US mainly, showing that the longer is the delay between symptom onset and diagnosis of disease, the worse is the outcome.

So I think we need to reiterate the message of early disease, and we need to reiterate that with both pulmonologists and probably chest radiologists. And probably also general medicine practitioner. Because they are going to see these patients for the first time when they come with a little bit of cough, a bit of shortness of breath. At that time, I think will be important to get IPF among the differential diagnosis, and to get the level of clinical suspicion for that disease higher than it is actually.

In this presentation, Dr. Hunninghake from Boston, he provided a very nice overview on where we are, and where we should move in the field of early pulmonary fibrosis detection. Because, clear, there is no doubt, this is a very challenging field. Very challenging. And, currently, we do not have a lot of tools to identify a patient with early disease. But we have some byproducts, like the program for lung cancer screening, or the program for detection of emphysema. So, in other words, Dr. Hunninghake can make very clear the point, that we need to use everything we have now to identify patient with early fibrosis.

Now, one specific point is related to genetics. We know that at least 10 to 20% of IPF forms are familiar. And we know that our genes, that they predispose to pulmonary fibrosis. And probably genetic screening, in family members of patients with IPF, is the way to go to identify these patients early. And Dr. Hunninghake has been publishing seminar papers in this field. And I know that he presented data on ongoing longitudinal prospective studies, that clearly will show how, by determining the genotypes of family member of patients with pulmonary fibrosis, we may actually improve the detection of early fibrosis, and therefore improving the management of these patients.

There's a very clear point, challenging, because it's not easy, is around the use of genetic medicine. But I think it's clearly the next step forward. Incorporating genetics into clinical practice, into routine clinical practice, is clearly still a challenge. Because we don't know which is the real predictive value of the genetic testing. And these genetics tests, they might be sophisticated to perform, and that means expensive. And, plus, they will need counseling. So what to do when you identify genetics.

So, clearly, IPF and pulmonary fibrosis is not a monogenic disease. We are not in the field of alpha one antitrypsin deficiency. We're not in the field of cystic fibrosis. It's a multigene disorder. So how to do a correct counseling of these patient, of these people? We don't know yet. But there are studies ongoing. So, in other words, probably large screening are not ready for prime time with genetics. But we are moving forward in that direction. And the studies which are ongoing, I'm sure we'll identify some genes that, in their polymorphism, will be able to improve the diagnostic accuracy in these patient. And I'm sure that these patients will want to know, in particular, the family members of patients with IPF.

How to diagnose patient with early pulmonary fibrosis? That was the topic that Dr. Lida Hariri, from Boston, discussed. It's a great challenge. It's very important. It's a combination of methods. They all go from wide use of spirometry, to testing symptoms with specific questionnaire, to the use of Chester's quotation. We should not forget the chest auscultation, in particular digital chest auscultation. It's a cheap, safe point of care tool, that can be used actually to screen patient with early pulmonary fibrosis. And that would be incorporated. We know that, every single pulmonology, we listen to the patient with interstitial lung disease.

And not to forget the genetics, of course. I think that the answer to the question of these talk by Dr. Hariri is, basically, it's going to be a combination of multiple tests, multiple technique. Most of them still in the experimental setting. And they will become ready for prime time, probably sometime from now. But it's an area where there is a lot of ongoing research. And I think that all pulmonologists need to stay tuned. Because these methodologies, probably not all of them, but some of them, will enter clinical practice.

I personally think that digital auscultation is the most promising tool for this. I mean, every pulmonologist is listening to the chest of their patients. And we all know the so-called Velcro crackle, to be present basically in any patient with IPF, and in most patients with pulmonary fibrosis actually. And they are very early signs. So it's a difficult area of research. But I think we have initial evidence that digital auscultation, in other words, the use of electronic stethoscopes recording the sounds from the chest of our patient, can be used, both for early detection, and for monitoring of disease progression in patients with pulmonary fibrosis.

So my personal opinion is that digital auscultation is going to be the next tool that we are going to use. And I think an important component is that it's cheap, it's safe, it's point of care, and every pulmonologist ... Every physician, actually, is very familiar with the scatter stethoscope. It's just an issue of translating these to a digital era.