



SPRING 2019 PULMONARY UPDATES

New Research on Therapies for Patients with Pulmonary Fibrosis

Luca Richeldi, MD

So in regard to the treatment of patients with idiopathic pulmonary fibrosis we have additional recent data that show us that the existing treatments are safe and effective. And it's important to know also that there are emerging new data from basically from phase two data, which are additionally putting confidence on the potential for new drugs to become available in the future. In particular, recently, they've been published data on the Pentraxin drug that has been completing the phase two. And we have data now on the open label extension study, which is showing that after the six months of randomized placebo control treatment safety is still good, which is most important part and that is a signal or suggestion of a sustained treatment effect. So this is very promising in light of the phase three studies that will be upcoming with this drug. And we had additional data in IPF that also it's important to treat the palliation part of these patient because they are very sick. And we know that we don't have any standard of care for these patients. And we know that this is going to be supportive care, one of the big hits that we need to take care of in the next few years.

A. Whitney Brown, MD

So there's some an exciting new device and it is using pulsed nitric oxide that's bled into oxygen therapy. They're doing a series of studies that are embedded I believe they're calling it phase 2B onto phase 3. But what was presented in terms of initial data was their first cohort, which was a group of IPF patients, actually not just IPF it was enriched with other disease populations as well - IPF, chronic HP, NSIP. And they chose patients who the majority of which had echocardiographic suggestion of pulmonary hypertension, so more severe patients, who were already using oxygen therapy. And they did an eight week study randomized people to either stay on their normal oxygen therapy, or use this novel device that had pulsed nitric oxide bled at a 30 parts per million bled into oxygen. And then they followed the two groups out. And what they saw in this was a kind of a cool study, because a lot of the studies have been predicated on six minute walk. And we know that that's really important, but it is just a snapshot in time. So they used an activity monitor that was worn like a watch to, to track their level of physical activity over the eight week period. And the day, the 24 hours in the day, were divided up into sedentary which would include sleep and rest, light, physical activity, moderate, and then vigorous. And so what they saw over the eight week time period is for those who were wearing the nitric oxide device compared to regular oxygen that the percentage of time they spent during the day and the moderate level of physical activity increased. The vigorous would be defined by activities such as running, and heavy, heavy exercise, the majority of these patients who were pretty sick, I think the forced vital capacity went all the way down to 40% and higher, they were not spending a lot of time in the section but the moderate physical activity, those wearing the device, increased their percentage of time. And they also looked at six minute walk distance, Nadir oxygen saturation on the six minute walk and some other parameters. And there was not as clear of a signal in those parameters. But the activity monitor, which is kind of a novel concept, did show did show a change. And so I think that's kind of provocative, it may be showing us that the future of some of these interventions had a test them would be more of a continuous read on our activity rather than just performance on a six minute test. But the all of these

patients have now rolled into open label extension. And so we'll get more data going into the future. And they're enrolling their cohort two, and then there'll be a cohort three and I think we'll hear more in the future about this potential, novel device as well as the use of activity monitors.