Editorial

IPF in Saudi Arabia: Lessons for all

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In this issue of the Annals of Thoracic Medicine, Alhamad *et al*. reported the characteristics and outcomes of idiopathic pulmonary fibrosis (IPF) patients seen and evaluated at a large tertiary care interstitial lung disease (ILD) center in Riyadh.[1] There are a number of "take-home messages," or perhaps more apt in the current COVID-era, "stay at home messages" within this report. A notable attribute of this cohort was the relatively advanced nature of their disease with an average forced vital capacity of only 57% and over half already oxygen dependent at the time of their initial evaluation. Indeed, the average time from the onset of symptoms to diagnosis was about one year, but the average disease duration was just over 2½years, inferring an undue delay in referral to the ILD specialty center of about a year and a half.

It is interesting to note that two parameters from the 6-min walk test (final oxygen saturation <85% and walk distance <300 m), but no pulmonary function parameter were predictive of outcomes after multivariate analysis. This underscores the importance of obtaining this functional test during routine evaluation and follow-up of IPF patients, in addition to standard PFTs, since the two can provide complimentary prognostic information.[2]

The authors of this paper are to be commended for applying the latest definition of Group 3 pulmonary hypertension (PH), which enables one of the first reports of the prevalence of PH among a cohort of IPF patients. Interestingly, about half the patients had right heart catheterization (RHC) proven PH; this high prevalence is somewhat surprising even given the lowering of the threshold for PH to include patients with a mean pulmonary artery pressure between 21 and 24 mmHg accompanied by a pulmonary vascular resistance of ≥3.[3] A common retort from those who question the utility of RHC in patients with IPF and other ILDs, is *"why do it, if we not going to act on it?"* However, RHC does enable more accurate prognostic information as well as identifying any component of heart failure. In addition to this, the positive results of the recently reported INCREASE study of inhaled treprostinil might change this narrative, as hopefully this agent will be approved for use in IPF patients with complicating PH.[4]

It is reassuring to know that even after a multivariate analysis, the use of antifibrotic therapy was associated with prolonged survival. This report joins many others that have demonstrated the same association.[5-7] While there might be some bias as to who is prescribed antifibrotics, this consistent and unequivocal association from multiple worldwide cohorts raises the question of when can we move from calling this an "association" to an appreciation that antifibrotics clearly save lives. This important messaging will emphasize the role of antifibrotics and underscore that the sooner they are started, the more likely it is patients will derive long-term benefit. This message is to pharmacy and therapeutic committees of all hospitals in Saudi Arabia, many of whom apparently do not include antifibrotic therapies as formulary medications. In terms of cost as a perceived issue, there are data to suggest that preservation of lung function in IPF is associated with fewer admissions and less healthcare resource utilization.[8] This

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appeal for broader antifibrotic availability extends beyond the Kingdom of Saudi Arabia and applies to all countries where antifibrotics are approved. This is even more important in the current COVID-19 era, where travel to distant hospitals might not be as feasible as during the pre-COVID-19 era. While referral to ILD specialty centers has been shown to result in improved outcomes, in this current stay at home and do not travel too far era, it is incumbent to make sure that all therapies are available to every IPF patient and not those that are lucky to live within close proximity to an ILD Center.[9] This applies even more so in countries such as Saudi Arabia with a wide geographic expanse and few ILD specialty centers.

Another notable feature of this paper is the reported incidence of acute exacerbations (AEs), which at 21% is right in line with the literature. This, however, does further underscore the need for early referral to a center adept at monitoring these patients. There are also data demonstrating that antifibrotics can help avert IPF-AEs, which represents another reason for the early implementation of therapy. Physicians should not be lulled into a false sense of security by IPF patients who are asymptomatic, since these potentially morbid events tend to be unpredictable and can occur in patients who are "early" with well-maintained lung function.

It is difficult in the current polarized political environment and isolationism to not think how those of us on the forefront of medical care can set an example for society and our political leaders, through our cooperation and willingness to learn from each other. This report on IPF in Saudi Arabia has lessons for all and underscores that unity accelerates progress; indeed, a unified path forward is how we will impact relatively rare entities such as IPF, as well as more urgent medical threats such as COVID-19.

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