Practice guidelines for interstitial lung diseases: Widening the reach

Clinical practice guidelines serve as the cornerstone of management recommendations for a myriad of medical conditions. They are based on the rigorous and systematic assessment of evidence-based literature. These guidelines form "best practice" patterns, solidifying standard of care approaches to diagnosis and treatment. Clinical practice guidelines are among the most widely read and cited medical articles. They are particularly useful in diseases where uncertainty or practice variation has existed in management approaches. This issue of *Lung India* features the first interstitial lung disease (ILD) management guidelines from the Indian Chest Society and the National College of Chest Physicians. This is a welcomed and important addition to the field of ILD.

Until now, ILD clinical practice guidelines have primarily been published by the American Thoracic Society (ATS), with the idiopathic pulmonary fibrosis (IPF) guidelines also including the European Respiratory Society (ERS), the Latin American Thoracic Association, and the Japanese Respiratory Society.[1-5] Additional key statements that have had a major impact on the approach to ILD diagnosis have also originated from the ATS and the ERS and, more recently, the Fleischner Society.[6,7] While the goal of these guidelines and statements is to provide a framework of broad scope, there are several limitations in regard to their widespread applicability to patients in different regions of the world. First, the evidence on which they are based is primarily informed by data from ILD patients residing in North America, Europe, Australia, and certain parts of Asia. However, the epidemiology of ILD differs throughout the world, influenced in part by differences in occupational and environmental exposures,[8] and limited information exists as to the natural history of ILD in large portions of Asia and in Africa.[9] Second, enacting current medical society-endorsed ILD diagnostic and treatment recommendations is not feasible on a global scale due to patient cost constraints or differences in available diagnostics or treatments among countries. While surgical lung biopsy is part of the algorithm in diagnosing IPF in the setting of certain high-resolution computed tomography patterns,[3] a large number of patients worldwide lack access to surgical services.[10] There exists increasing recognition that diagnostic criteria for certain respiratory diseases, acute respiratory distress syndrome, for example, need to be modified to enable applicability to all patients, especially those lacking access to the medical care available in other countries.[11] In addition, current ILD guidelines and statements have primarily centered on idiopathic interstitial pneumonias (IIPs), mainly IPF. As IIPs comprise only a proportion of ILDs, additional guidelines that offer management recommendations specific to non-IPF ILDs are needed.

The guidelines published herein build on currently available ILD clinical practice guidelines in several important and commendable ways. First, the scope of the guidelines extends past management of IPF to include treatment recommendations for other common ILDs, including rheumatoid arthritis-associated ILD (RA-ILD), systemic sclerosis-associated ILD (SSc-ILD), hypersensitivity pneumonitis, sarcoidosis, and certain occupational lung diseases. Within the past 5 years, there have been advances in the treatment of non-IPF subtypes of ILD, such as SSc-ILD,[12] which are included here. Second, given that these guidelines address the use of immunosuppression for certain non-IPF ILDs, the need for assessing for infection with mycobacterial tuberculosis (TB) in patients from high prevalence countries is essential and is importantly addressed. The diagnosis of an IIP hinges on the exclusion of alternative etiologies. While prior guidelines and statements have recognized the need to evaluate for other ILD entities, such as hypersensitivity pneumonitis and connective tissue disease-associated ILD, additional alternative etiologies, such as current or prior TB infection, become relevant considerations as potential contributors to abnormal chest imaging findings in parts of the world with a high TB burden. Thus, recommendations incorporating the need to evaluate for TB at the discretion of the clinician as a usual practice point within the context of ILD evaluation reflect an important tailoring of society guidelines based on the needs of the patients they represent. Third, the use of the Medical Research Council (MRC) dyspnea scale is included as a method for ILD monitoring in patients unable to perform spirometry. While the spirometry measurement of forced vital capacity has been the primary outcome measure of several recent ILD clinical trials,[13-16] for many patients, especially those with advanced disease, performing spirometry is a challenge. The MRC dyspnea scale is an easy to perform and cost-effective means to assess functional limitations of dyspnea and has, along with its modified form, been demonstrated to have value in predicting outcomes in ILD.[17,18] Lastly and notably, these guidelines include recommendations aimed at improving quality of life and palliation of symptoms of cough and dyspnea. Palliative care is an under underutilized resource

in advanced lung disease and often not employed until the very end of life for ILD patients.[19] The authors should be applauded for incorporating a recommendation for offering palliative care to all patients with advanced ILD. Whether or not palliative care may have a survival benefit in the setting of ILD, as has been demonstrated in other conditions, has yet to be determined.[20]

Several issues remain and represent areas of uncertainty in the ILD literature. While gastro gastroesophageal reflux disease (GERD) is a common comorbidity in patients with ILD, notably IPF, there remains equipoise in terms of whether patients with ILD should be systematically initiated on GERD-targeting medications. In addition, as is acknowledged in these guidelines, more data are needed, especially in terms of randomized controlled trials, to define the most optimal treatment approach to non-IPF ILDs, such as RA-ILD, especially in terms of first-line immunosuppressive alternatives to prednisone. This is an area of marked variability in clinical practice and merits further study. Given that these guidelines include recommendations for the use of immunosuppression for certain non-IPF ILDs, additional guidance in regards to indications for bone health prophylaxis in the setting of prolonged corticosteroid use and for prophylaxis against pneumocystis jirovechi pneumonia depending on the degree of immunosuppression would be helpful additions.

In conclusion, the guidelines published here expand on available ILD clinical practice statements by including recommendations for the management of certain non-IPF ILDs. The Indian Chest Society and the National College of Chest Physicians should be commended for their efforts. Ongoing ILD statements from societies around the world would be a welcomed addition to the literature to make certain that ILD guidelines reach and are applicable to patients across the globe.

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