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It takes a village to care for the patient with idiopathic pulmonary fibrosis

Idiopathic pulmonary fibrosis (IPF) is a devastating progressive fibrosing interstitial lung disease associated with a high burden of morbidity and death.[1] A clinical diagnosis of IPF is made only after careful interpretation of integrated clinical, radiologic, and often histopathologic data.

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Interstitial lung disease encompasses a broad spectrum of parenchymal lung diseases, and a classification of IPF is restricted to a lung injury pattern of usual interstitial pneumonia (UIP) based on high-resolution computed tomography or surgical lung biopsy, after all known causes of UIP have been excluded.[1]

However, a lung injury pattern of UIP is not synonymous with IPF, as UIP can be seen with connective tissue disease, chronic hypersensitivity pneumonitis, drug toxicity, and sarcoidosis.[1] As such, rendering a diagnosis of IPF requires a thorough evaluation to exclude such diverse potential etiologies.

In this issue of the *Cleveland Clinic Journal of Medicine*, Tolle and colleagues[2] provide an up-to-date, broad overview of IPF focused on what the primary care provider needs to know about the disease. Their review is timely and serves as a useful primer for the practicing clinician.

The field of IPF is actively evolving, as this era has been witness to a recent paradigm shift in pharmacologic management. Immunosuppression is no longer recommended[3] and may even be harmful.[4] And the US Food and Drug Administration has approved 2 antifibrotic drugs—pirfenidone and nintedanib—that have been shown to delay progression of IPF.[5,6]

Primary care providers have a unique opportunity to play an integral role in the evaluation and care of patients with IPF, in particular with earlier disease recognition, initial disease assessment, and timely specialty consultative referral—as well as implementing a comprehensive longitudinal care plan.

EARLIER DISEASE RECOGNITION

Routine health evaluations offer opportunities for early recognition of IPF

IPF is a rare disease primarily affecting men over the age of 65.[1] It is reasonable to presume that many or most of these individuals ultimately diagnosed with IPF are already seeking routine care for existing common medical conditions such as hypertension or dyslipidemia—or at least having periodic routine health maintenance assessments. Such evaluations may offer an opportunity for earlier recognition of an underlying fibrotic lung disease that may be subclinical in nature.

IPF has a lower-lung zone predominance. The importance of chest auscultation, particularly listening carefully to the lung bases, is poignantly highlighted in a recent editorial: "It is time that the stethoscope draped around the neck of physicians, which tends to be used for identification purposes rather than for medical diagnosis, be also the (presently only) genuine tool for an earlier diagnosis of IPF."[7]

Advances in imaging also provide an opportunity for earlier diagnosis. Many patients undergo screening computed tomography for coronary calcium scoring or lung cancer sur-

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veillance, and these studies may incidentally identify subtle interstitial lung abnormalities. These incidental findings should lead to further investigation, as they have been shown to be functionally important and carry risk of progression to clinical interstitial lung disease.[8]

INITIAL ASSESSMENT, TIMELY REFERRAL

But whether evidence of interstitial lung disease is detected incidentally or during testing for respiratory symptoms, further evaluation is necessary. Primary care providers are uniquely positioned to initiate the assessment and to expedite and guide further evaluation and specialty referral consultation to ensure an accurate diagnosis. They can also help grade the severity of the disease with pulmonary function testing, oxygen assessments at rest and with ambulation, and ordering thoracic high-resolution computed tomography to provide valuable information about disease extent and interstitial lung disease pattern.

General practitioners may assess for features suggesting connective tissue disease that would warrant specific serologic testing and dedicated rheumatologic consultation.

Refer patients to specialized centers rather than general pulmonary providers

Finally, given the rarity, complexity, and challenges of interstitial lung disease, an effective multidisciplinary team consisting of clinicians, radiologists, and pathologists enhances diagnostic accuracy.[9] This may also help general practitioners deviate from normal patterns of referral to general pulmonary providers, and instead refer patients to specialized centers with dedicated clinical and research expertise in interstitial lung disease.

IMPLEMENTING A COMPREHENSIVE, LONGITUDINAL CARE PLAN

The primary care practitioner often has developed long-term relationships with patients ultimately diagnosed with IPF, and because of this is particularly well positioned to help implement a collaborative and comprehensive care plan. Logistical realities such as distance to a specialty center, limited insurance coverage for specialty visits, and limited specialty availability all reinforce the central role that primary care practitioners play in ensuring that patients adhere to a comprehensive treatment program.

Primary providers may be very experienced and more inclined to manage a number of the common and often important comorbid conditions seen in patients with IPF, such as gastroesophageal reflux disease, obstructive sleep apnea, and depression. Reinforcing to the patient the need to adhere to adjunctive therapies such as supplemental oxygen and pulmonary rehabilitation is another key opportunity to actively engage in the management of patients with IPF.

Primary providers may also play a central role in IPF care through prevention strategies such as smoking cessation and ensuring appropriate immunization against seasonal influenza, pneumococcal pneumonia, and pertussis, among other age-appropriate vaccinations.

With the introduction and expansion of use of nintedanib and pirfenidone for IPF over the past few years, general practitioners may be called on to help manage common gastrointestinal side effects associated with pirfenidone (primarily nausea) and nintedanib (primarily diarrhea), and to be aware of potential drug-drug interactions and other medication-related toxicities.

Finally, as IPF remains a progressive disease, primary care practitioners are often well positioned to help implement palliative care, hospice care, and end-of-life care.

Despite recent advances in treatment, IPF remains a devastating lung disease with a high degree of morbidity and mortality. It takes a village to help care for the IPF patient. And as key members of the healthcare team, primary care providers have unique and important opportunities to help in the early recognition, thorough assessment, and comprehensive management of patients with IPF.

Dr. Fischer has disclosed consulting, membership on advisory committees or review panels, other activities from which remuneration is received or expected, and membership on clinical trial steering committees for Boehringer Ingelheim and Hoffman-La Roche.

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