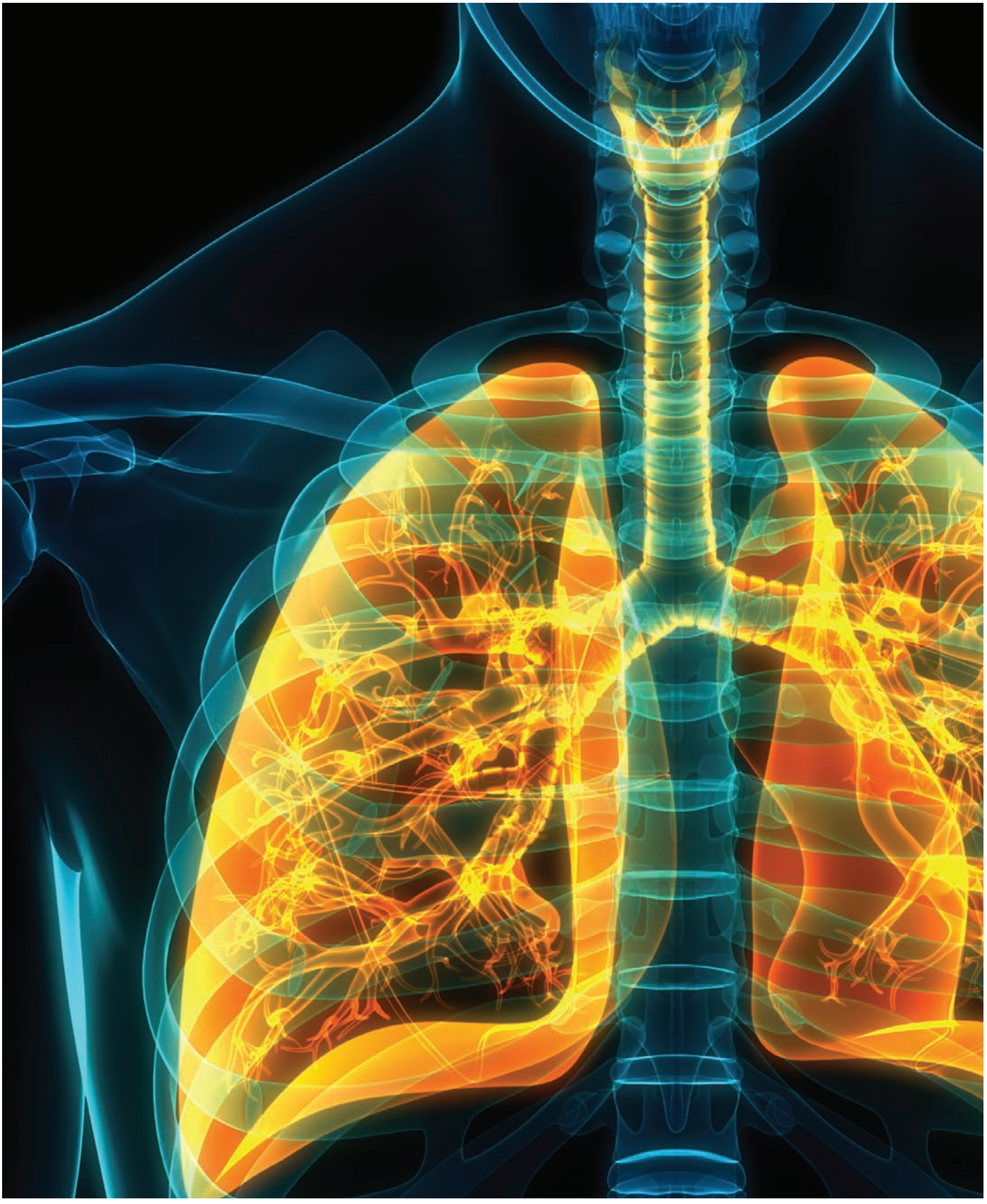

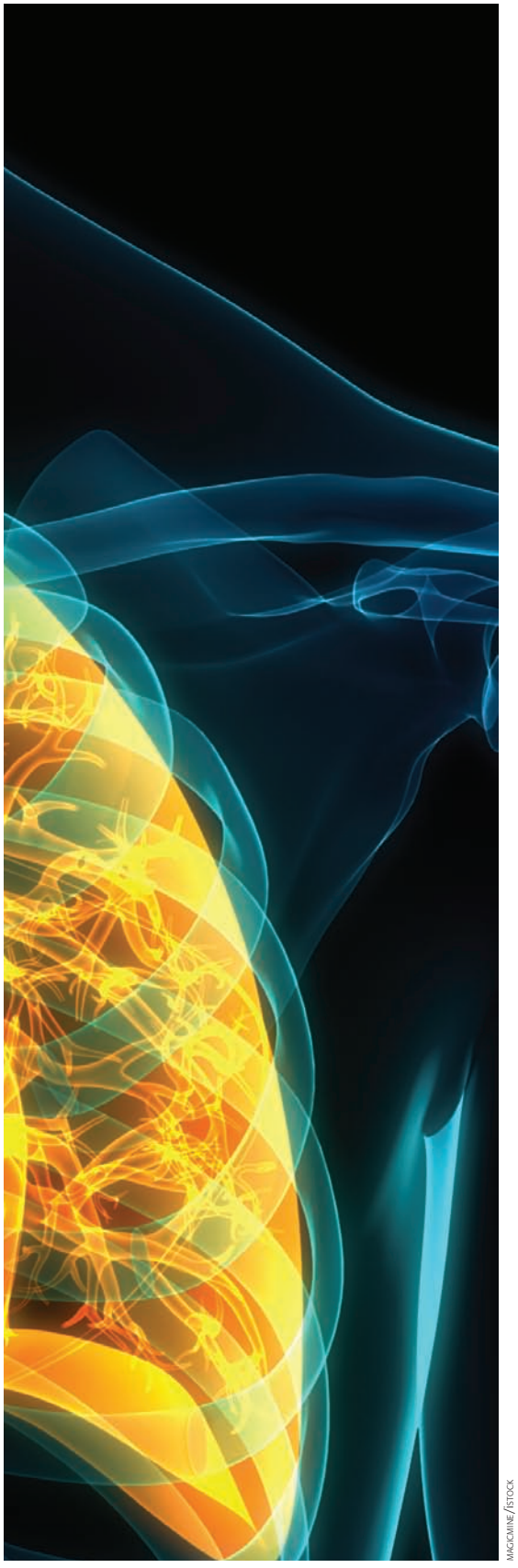

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Idiopathic pulmonary fibrosis: What nurses need to know

BY BILL PRUITT, MBA, RRT, CPFT, AE-C, FAARC

Abstract: Idiopathic pulmonary fibrosis (IPF) is a restrictive lung disease in which the cause cannot be determined. This article discusses restrictive lung diseases that fall under the general category of interstitial lung disease with a focus on IPF—a fatal disease characterized by progressive fibrosis and interstitial pneumonia, dyspnea, and decreasing pulmonary function.

Keywords: gastroesophageal reflux disease, GERD, high-resolution computed tomography, HRCT, idiopathic pulmonary fibrosis, interstitial lung disease, ILD, IPF, PFT, pulmonary function testing

CHRONIC LUNG DISEASES are generally classified as either obstructive or restrictive, terms that relate to the changes seen in pulmonary function tests (PFTs). There are many different restrictive lung diseases with a wide range of causes, including such issues as pneumothorax, pleural effusion, pneumonia, and acute respiratory distress syndrome to name a few (see *Obstructive versus restrictive lung disease*).[1] Idiopathic pulmonary fibrosis (IPF) is a restrictive lung disease in which the cause cannot be determined. This article discusses restrictive lung diseases that fall under the general category of interstitial lung disease (ILD) with a focus on IPF—a fatal disease characterized by progressive fibrosis and interstitial pneumonia, dyspnea, and decreasing pulmonary function.[2]

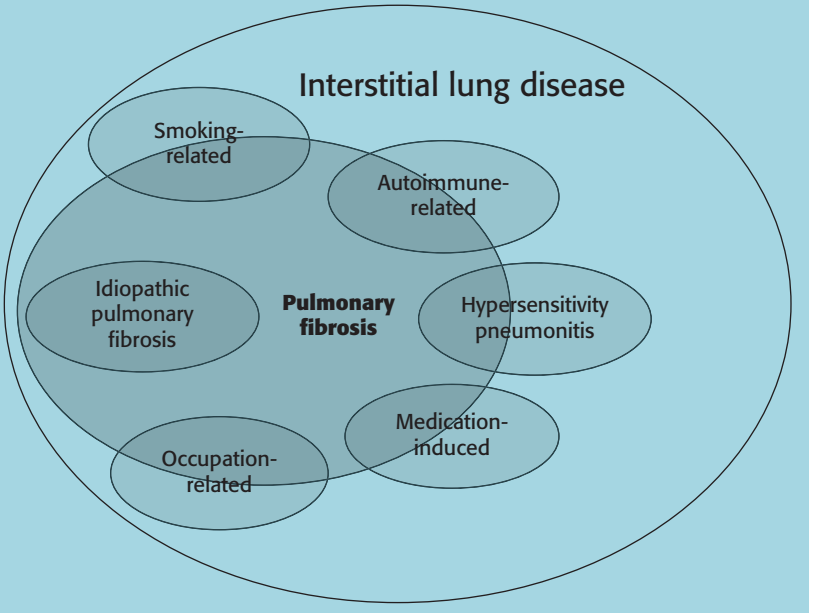
The terms *pulmonary fibrosis* and *ILD* are often used interchangeably, and the disorder may also be called diffuse parenchymal lung disease. Depending on the reference, ILD has between 180 and 200 subtypes and a multitude of abbreviations and acronyms to describe them. This proliferation of subtypes and their associated abbreviations and acronyms has created some confusion among healthcare professionals caring for patients with suspected or diagnosed ILD.[3] The intent of this article is to provide nurses with a better understanding of the overall disorder and care needed for people who have been diagnosed with a restrictive lung disease, particularly IPF.

Epidemiology

ILD has six main causes: smoking, systemic autoimmune or connective tissue issues, hypersensitivity pneumonitis, medications, occupational (and avocational) exposures, and idiopathic (see *Six causes of ILD*).[3] Between 20% and 50% of all cases of ILD are classified as IPF. Using conservative estimates, the incidence of IPF is 3 to 9 cases a year per 100,000 persons in Europe and North America, with a lower incidence in Asia and South America.[4-6]

IPF tends to affect more males than females, typically males ages 50 to 70. After diagnosis, survival usually ranges from 3 to 5 years.[5,7] Worldwide, IPF appears to be increasing in both incidence and mortality.[4,8]

Six causes of ILD[3]



Schematic overview of interstitial lung disease (ILD) and pulmonary fibrosis, which includes six common ILDs. Other less common ILDs would occupy space within the ILD and fibrosis ovals and would likewise manifest varying degrees of inflammation and/or fibrosis depending on the particular disorder.

Risk factors

Older age, male gender, cigarette smoking, and certain environmental exposures are considered risk factors for development of ILD and IPF (see *Common causes of ILD and IPF*). Environmental exposures include dust from certain metals and from wood and vegetable/animal dust; this has been defined in research that distinguished exposure between farmers working with crops versus those working with animals.[9] Infectious agents such as Epstein-Barr virus, hepatitis C virus, adenoviruses, and herpes viruses may also increase the risk but research on these topics has not been definitive so far.[10]

Gastroesophageal reflux disease (GERD) and its association with aspiration or microaspiration is a possible risk factor and is undergoing further study. GERD has been observed in up to 90% of patients with IPF. Aspiration can lead to pneumonitis, which is suspected of causing or worsening IPF.[11]

Certain comorbidities such as diabetes mellitus and obstructive sleep apnea (OSA) are also considered possible risk factors. Finally, genetic factors are considered to increase the risk for both familial and sporadic cases of IPF.[2,4]

Pathophysiology

Both inflammation and fibrosis are involved in the pathology of ILD. The pathology of ILD is not clear, but continuing research into the diseases associated with this umbrella term is bringing more understanding.[12]

After an initial injury to the alveolar wall by infection or another noxious agent, alveolitis and vasculitis occur, triggering an immune response. The lung tissue may then move through the repair process and return to normal—or it can take an alternate abnormal pathway with persistent inflammation and infiltration by lymphocytes, macrophages, and plasma cells. Interstitial and alveolar wall thickening occurs, along with excess production of collagen and other connective tissue components. Fibroblasts, myofibroblasts, and extracellular matrix accumulate and fibrosis (also called scarring) becomes more apparent as tissue remodeling occurs.[2,3]

Radiographic studies performed in the early stages of IDL and IPF will have a ground-glass appearance, de-

Obstructive versus restrictive lung disease

With *obstructive lung disease*, expiratory airflow is reduced, which results in a prolonged exhalation (airflow obstruction) and often causes an abnormal amount of air to remain in the lungs over time (a condition known as air trapping). The two most common obstructive disorders are chronic obstructive pulmonary disease and asthma.[30] With *restrictive lung disease,* the lungs reflect reduced volumes and often have decreased lung compliance (becoming more rigid or stiff).[1,15] While obstructive lung disease is associated with expiration, restrictive lung disease is associated with inspiration.

scribed as an often-diffuse region of hazy lung radiopacity.[13] When cysts form, the resulting radiographic term "honeycombing" is used to describe the appearance of lung tissue. Honeycombing refers to clustered cysts with thick, well-defined walls.[14] The cysts have little to no gas exchange capability and reduce the number of functional alveoli as normal tissue is destroyed. As a result, hypoxia develops.

Fibrosis results in decreased compliance (lungs become "stiffer") and the patient's tidal volume and other lung volumes decrease while respiratory rate increases. The thickening of the interstitium combined with the decrease in functional alveoli leads to decreased diffusion and contributes to worsening hypoxia.[15,16]

Common causes of ILD and IPF[19]

Note that the occupational exposures are all inorganic entities while the causes of hypersensitivity pneumonitis are all organic entities. In addition, there is a link between hypersensitivity pneumonitis with occupational exposures because most of these relate to work environments.

Occupational exposures
- asbestos
- coal dust
- silica
- chlorine gas
- sulfuric acid
- hydrochloric acid
- ammonia

Hypersensitivity pneumonitis
- moldy hay
- silage
- moldy sugar cane
- mushroom compost
- wood pulp, bark, dust
- bird droppings

Medications
- antibiotics
- anti-inflammatory drugs
- cardiovascular drugs
- chemotherapy
- illicit drugs

Systemic autoimmune or connective tissue disorders
- scleroderma
- rheumatoid arthritis
- systemic lupus erythematosus
- sarcoidosis

Clinical presentation

A careful health history is essential for making an accurate diagnosis for ILD and IPF. The history needs to include details about exposure to inhaled substances such as cigarettes, cigars, marijuana, or illicit drugs such as inhaled cocaine. The practitioner should inquire about the patient's home, work, or hobby environment for the likely presence of substances such as mold, bird droppings, dust from wood or other organic sources, or dust/particles/fumes from substances such as asbestos, silica, heavy metals, or contaminated ventilation systems.[17]

Review the patient's medications, including the use of over-the-counter medications, herbs, and supplements, and assess for substances associated with ILD or IPF. Document any family history of lung disease to evaluate genetic risks.

In the typical patient with ILD or IPF, progression of signs and symptoms and loss of lung function are usually gradual; however, in some cases the progression is rapid. Early in the course of disease, patients complain of dyspnea on exertion that slowly worsens and is accompanied by a chronic nonproductive cough. As inflammation, fibrosis, and diffusion capacity worsen, the patient develops hypoxemia and a decreased Spo_2 (with significant desaturation during exercise) along with tachypnea and increasing dyspnea. In 40% to 80% of cases, the patient will exhibit digital clubbing for reasons that remain unclear. Physical assessment findings may also include a nonproductive cough and bilateral basilar crackles.[3]

As the disease progresses, the patient may become cyanotic due to chronic hypoxemia and experience weight loss and malnutrition as increased dyspnea makes eating more difficult.[2,15,18]

Diagnosis

Diagnosis of ILD and IPF is greatly enhanced by collaboration between a pulmonary team consisting of pulmonologists, advanced practice providers, radiologists, and pathologists who have experience working with patients who have these diseases.

No lab tests are specific for a diagnosis of IPF, so the role of lab testing in patients with newly identified ILD is to identify or exclude processes in the differential diagnosis.[6] Complete pulmonary function testing is needed to gather baseline information and track disease progression. PFT results typically show a restrictive pattern: reduced forced vital capacity (FVC), but normal ratio of forced expiratory volume in one second (FEV1/FVC), reduced diffusing capacity for carbon monoxide (DLCO), and decreased lung volumes and capacities.[6] Reduced diffusion capacity is reflected in the reduced DLCO value.

Arterial blood gases (ABGs) in mild to moderate ILD will reflect an acute respiratory alkalosis (acute alveolar hyperventilation) with hypoxemia. As the disease worsens, ABG results will change to show a compensated respiratory acidosis (chronic ventilatory failure) with hypoxemia.[6,19]

High-resolution computed tomography (HRCT) is a key to the diagnosis of IPF and often can eliminate the need for invasive diagnostic procedures. HRCT will often show usual interstitial pneumonia (UIP), which refers to typical histopathologic

changes found in patients with IPF such as honeycombing (see *Honeycombing in IPF*).[20] If HRCT does not show honeycombing, results are considered indeterminate and a surgical lung biopsy (often performed by a video-assisted thoracoscopic approach) is needed to make a diagnosis.[2,18,21] For a diagnosis of IPF, the known causes of ILD (such as exposure to causative agents, connective tissue disorders, and drug toxicity) must also be excluded.[2]

Following a typical patient

AC, age 71, is a retired White male who presented to his primary care provider with complaints of progressive dyspnea on exertion and a nonproductive cough. AC had smoked cigarettes for 25 years, 1 pack-per-day, but quit smoking 4 years ago. His health history includes hypertension, type 2 diabetes, and GERD. He has no surgical history. His family history includes his mother who died of cardiac disease at age 79 and his father who died at age 76 of an unclassified lung disease. He denies any alcohol or illicit drug use.

AC worked for over 40 years as a general auto mechanic before retiring 8 years ago. He enjoys playing golf but has not been able to play due to increasing dyspnea. He denies any other occupational or environmental exposure. He has a pet dog and has not traveled recently. He lives alone after his wife died 2 years ago. They had no children.

AC's physical assessment reveals a normal examination of the head, eyes, ears, nose, and throat; "Velcro type" bibasilar crackles on auscultation; and normal first and second heart sounds. His abdomen is soft, nontender, and nondistended. No extremity edema or cyanosis is noted but mild digital clubbing is present.

Diagnostic testing

AC's lab work results include an antinuclear antibodies titer of 1:60 (normal, <1:40), an erythrocyte sedimentation rate of 10 mm/h (normal, 0 to 22 mm/h for men), and a rheumatoid factor of 11 U/mL (normal, <60 U/mL), which rules out an autoimmune cause for his signs and symptoms.

Because of AC's history of diabetes and hypertension, testing also includes an echocardiogram, which demonstrates a preserved left ventricular ejection fraction of 60% (normal, ≥55%) with mild diastolic dysfunction. Estimated pulmonary artery systolic pressure is 28 mm Hg (normal, 15 to 28 mm Hg). A chest X-ray shows small lung volumes with increased interstitial markings without cardiomegaly or pleural effusion.

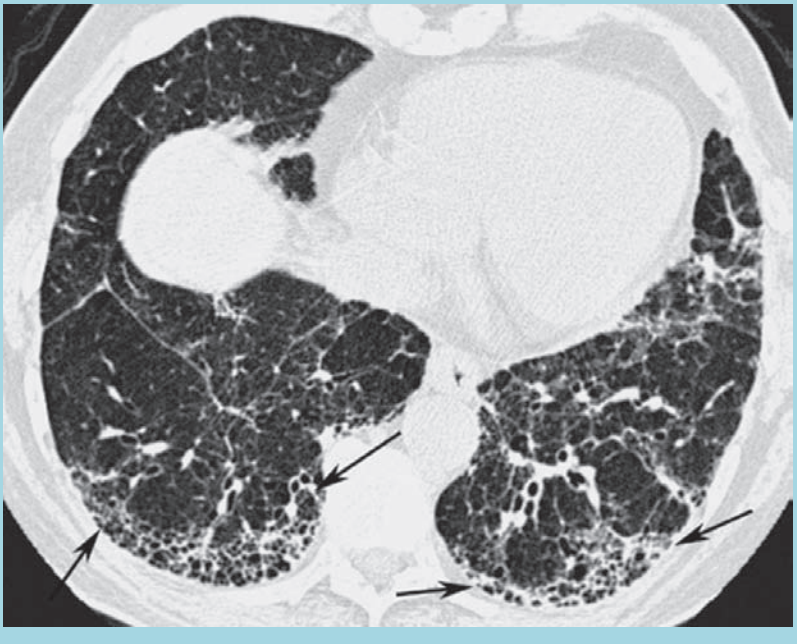
PFT results show a FEV1/FVC of 73% (normal, >70%), an FVC% predicted of 65% (normal, >80%), a total lung capacity of 61% predicted (normal, >80%) consistent with reduced lung volumes and lung capacities (reflecting a restrictive pattern), and a DLCO of 45% of predicted (normal, >80%) reflecting reduced diffusion capacity (see *Take a breath*).

AC is able to walk 420 meters during a 6-minute walk test (6MWT) with oxygen saturation dropping from 98% at rest to 91% during the test. The 6MWT measures the distance walked over 6 minutes and is commonly used to assess patients' functional capacity. During a 6MWT, healthy subjects can typically walk 400 to 700 meters.[22]

HRCT shows subpleural basal predominant reticulation with associated honeycombing and traction bronchiectasis (where the airways are distorted and pulled apart by the surrounding fibrosis) in both bases. Considered a key feature of IPF, traction bronchiectasis ranges from subtle irregularity of the bronchial/bronchiolar wall to marked airway distortion and varicosity.[23]

No evidence is found of ground-glass opacities, cysts, nodules, consolidation, air trapping on expiratory images, or lymphadenopathy. In addition, no evidence of pleural plaques or pleural effusion is found.

Honeycombing in IPF[31]



This thin-section CT image shows peripheral honeycombing (see arrows) in a patient with end-stage pulmonary fibrosis.

AC's work as a mechanic is significant because it may have involved prolonged exposure to asbestos from brake linings, leading to asbestosis. However, more detailed questioning reveals that he had almost never worked with brake repairs because this job was done by a brake specialist in the shop. In addition, the HRCT scan does not show calcified pleural plaques, which also supports ruling out asbestosis.

Given the typical UIP pattern on his scan with no clear evidence pointing to other diseases, AC is diagnosed with IPF. Initial referral is made for lung transplantation, but AC is not a candidate due to his age and comorbidities.

Managing IPF

Management goals for patients with ILD and IPF include slowing the fibrotic process and relieving the patient's dyspnea. In cases where the cause is exposure to a noxious agent, stopping this exposure should be a first step. For example, any patient who is a smoker should be strongly encouraged to stop smoking. Anti-inflammatory therapy may help depending on the specific disease subtypes of ILD, but in IPF many drugs have been shown to be ineffective or even harmful.[4,11,18]

Corticosteroids in combination with other medications such as N-acetylcysteine and immunosuppressive drugs are often used.[24] Two antifibrotic medications, pirfenidone and nintedanib, have been available in the US since 2014. Both are oral medications that have been shown to slow the rate of decline in FVC and prolong survival. However, these medications are expensive, with an estimated cost of over $100,000 annually for each.[25]

Many other drugs and combinations have been studied for use in treating patients with ILD and IPF. Depending on the particular subtype, some drugs may be recommended for use in one case but not in another. The clinical practice guideline for IPF issued by an international panel of experts in 2011 and updated in 2015 recommends against the use of glucocorticoid monotherapy; combination therapy with azathioprine, prednisone, and N-acetylcysteine; and monotherapy with N-acetylcysteine for routine maintenance treatment of IPF.[11] The guideline states that the recommendations against using these treatment regimens are strong due to insufficient evidence to support routine use. Recommendations for some other treatment approaches are weak due to the need for more high-quality research to evaluate the risks and benefits of these agents.[2,10] For management of cough, several agents are being studied, including thalidomide, pirfenidone, gefapixant, and inhaled cromolyn, but none is supported with enough evidence to be included in published guidelines.[25]

The 2015 clinical practice guideline also suggests that patients with IPF be screened for GERD and treated with antacids to decrease the risk of aspiration.[11] A recently published article that reviewed the links and risks of IPF and GERD calls for more high-quality clinical studies to evaluate the efficacy and safety of both proton pump inhibitors (PPIs) and antireflux surgery for treating IPF, "also taking into account that all the new antifibrotic and immune-suppressive therapies have relevant limitations."[26]

High-flow nasal cannula (HFNC) oxygen therapy is helpful to relieve dyspnea on exertion and treat hypoxemia in patients with ILD and IPF. HFNC systems deliver between 20% and 100% oxygen at flow rates up to 60 L/minute. These systems are often better tolerated, allow for eating and improved communication, and give the patient more freedom to move about compared with other oxygen delivery systems such as non-rebreather or partial rebreather masks or noninvasive ventilation.[27] In late stages of disease, oral, parenteral, and/or aerosolized morphine may be prescribed to relieve the sensation of dyspnea.

Take a breath[22]

FVC is the total exhaled volume (from full inspiration to full expiration with maximum effort to exhale). FEV1 is the volume exhaled in the first second (measured during the FVC maneuver). FEV1/FVC represents the ratio of these values.

Some patients with IPF qualify for lung transplantation but the selection criteria, cost, and availability of organs make this option difficult to achieve.[6,13] Currently, indications for transplant include histologic or radiographic evidence of UIP and any of the following: a DLCO below 39% predicted, more than a 10% drop in FVC over 6 months, or an Spo_2 less than 88% during 6MWT.[27]

Research has shown that a drop in distance walked of more than 50 meters for patients with IPF over a 24-week period is associated with a fourfold increase in death within 1 year.[28] Management of comorbidities such as chronic obstructive pulmonary disease, OSA, secondary pulmonary hypertension, GERD, anemia, anxiety, or depression can improve the patient's quality of life.[16]

The progression of IPF can follow several pathways but ultimately results in death, usually within 3 to 5 years after diagnosis. Some patients who have a slow, steady decline in their lung condition and overall health will survive at the high end of the continuum, but others have a rapid decline and very short survival time after diagnosis, sometime within months.

A minority of patients experience a slow decline after diagnosis until an acute event such as pneumonia, acute exacerbation of unknown cause, or

heart failure occurs, triggering a substantial negative change in health. Then another slow decline may occur until an additional sudden change occurs and another substantial negative change follows. This pattern repeats until the patient's death.[2]

AC's case progression

For AC, treatment is started with pirfenidone. He experiences nausea and diarrhea but is able to tolerate these adverse reactions. Loperamide and hydration are prescribed to treat his diarrhea.

Liver function tests are measured at baseline and throughout treatment because pirfenidone can cause elevated liver enzymes and drug-induced liver injury. He is prescribed a PPI for GERD. Due to his desaturation during the 6MWT, supplemental oxygen is added at 2 L/min by nasal cannula. AC completes a 6-week program in pulmonary rehabilitation, which improves his exercise tolerance and quality of life.

Six months after starting treatment, AC's FVC drops to 60% predicted and his DLCO decreases to 33% predicted. Palliative care is started and he is treated for depression. His oxygen therapy is increased to 3 L/min for a few weeks, then 4 L/min to help reduce issues with desaturation.

Unfortunately, AC suffers an acute exacerbation of IPF at 13 months after diagnosis and requires hospitalization. End-of-life care is discussed and AC elects do-not-resuscitate status. Systemic steroids are initiated without success and AC dies 9 days after admission.

Nursing considerations

Nursing care for patients like AC includes educating the patient and family about the disease process and recommending community resources for support. Education should include strategies for avoiding respiratory irritants such as dust, mold, and animal dander; performing optimal hand hygiene practices; and preventing respiratory infections by avoiding crowds and people who are sick. Helping patients gain access to clinical trials can aid in research on the disease and on treatment options and provide an avenue for patients and families to connect with others who have the disease while contributing to a better understanding of ILD and IPF.[29]

Patients and their families should be encouraged to receive the pneumococcal vaccine and annual influenza vaccinations.[16] Smoking cessation should be included for all who smoke. Education on good nutrition is also indicated to prevent weight loss and support the immune system.

To manage issues related to fatigue, increased work of breathing, and dyspnea, patients need to learn about energy conservation measures and breathing techniques (such as diaphragmatic and pursed-lip breathing). These techniques help reduce drops in oxygenation and the work of breathing.[13] Educate patients and families about the patient's treatment regimen and prescribed medications, including possible adverse reactions. For example, pirfenidone has been associated with many adverse reactions including anorexia, nausea, vomiting, insomnia, and rash. Nurses need to assess for adverse reactions to medications and other treatment approaches. If problems are noted, the provider may prescribe additional treatments such as antiemetics or topical agents as indicated.[25]

Depression, anxiety, and fatigue can increase stress and can make symptoms worse. Strategies for managing these issues are important to maintain quality of life. If any local support groups for ILD and IPF are available, patients and families should be encouraged to join. Support groups can help provide disease education, improve communication and emotional well-being, and help shape advance care planning.

Palliative care should be considered early in the disease process. Discussing expectations and wishes for end-of-life care can help patients improve their mental outlook and decrease the stress on both patients and the families. Hospice care may come into play for some patients. Nurses can help them understand the benefits of palliative care versus hospice care, answer questions, clarify misconceptions, and facilitate access to these care options.[18,29]

Research is going forward on several fronts to discover improved diagnostic and treatment options for IPF. For example, examination of epithelial cell injury has led to research into use of antioxidants and antireflux agents such as PPIs or histamine$_2$-blocker receptor antagonists. In addition, the use of biopsy in making a firm diagnosis may be impacted as research goes forward in finding a reliable noninvasive biomarker that can help identify the various subtypes of ILD.[21]

Future directions

At present, ILD and IPF are difficult to diagnose and treatment options are very limited. These diseases are not curable and the short timeline from diagnosis to death brings tough challenges to the patients, families, and healthcare team. However, the increasing numbers of centers specializing in these diseases is contributing to progress in treatment options and the quality of care. Palliative and hospice care are being brought into the care model more often and are helping patients and families as the disease progresses. Education is essential for patients and families to help them understand and cope. Ongoing research on several fronts in the diagnosis and management of ILD and IPF may reduce the burden of these diseases and improve patient outcomes in the future. ■

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