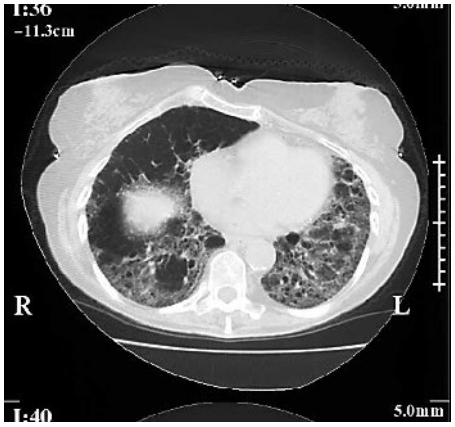
CE 2.5 HOURS
Continuing Education

Idiopathic



Idiopathic pulmonary fibrosis in an adult. Chest computed tomographic scanning (axial view) shows a pattern of diffuse interstitial lung disease; fibrosis with honeycombing is seen. The four images are in sequence (from left to right) at 10-mm intervals.

Rose Dohany, a 60-year-old executive secretary, visits her primary care physician for evaluation of shortness of breath and nonproductive cough. She has no other symptoms, but a chest X-ray shows reticular opacities in the lower lungs, indicating pneumonia, and she's prescribed a seven-day course of antibiotics. Two weeks later, though, her symptoms persist, and tests reveal restrictive abnormalities in pulmonary function: forced vital capacity (FVC) is 1.6 L, or 56% of predicted value; forced expiratory volume in one second (FEV_1) is 1.35 L, or 67% of predicted value. Her physician refers her to a pulmonologist for further evaluation.

The pulmonologist orders additional testing, which shows decreases in FVC and FEV_1 on spirometry (1.47 L, or 52% of predicted value, and 1.28 L, or 64% of predicted value, respectively), decreased lung volumes, and decreased carbon monoxide– diffusing capacity (D_{LCO}) (which is 13.7 gm/dL, or 38% of predicted value). A repeat chest X-ray shows no changes, and a high-resolution computed tomo-

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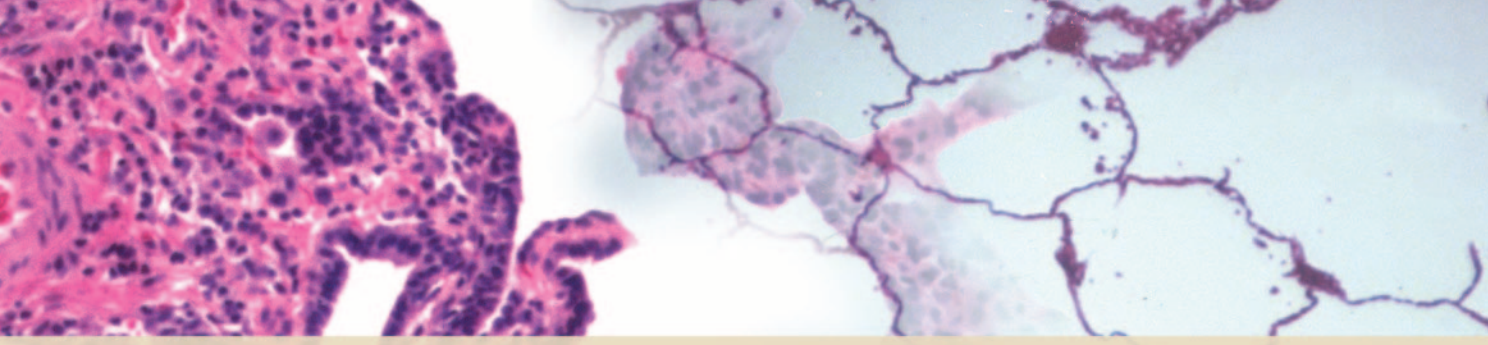
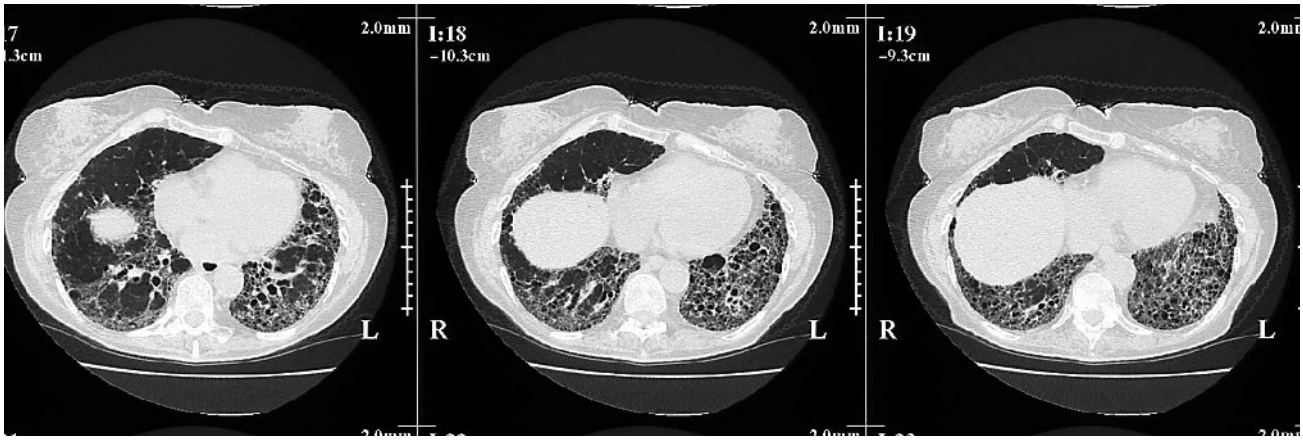


Pulmonary Fibrosis

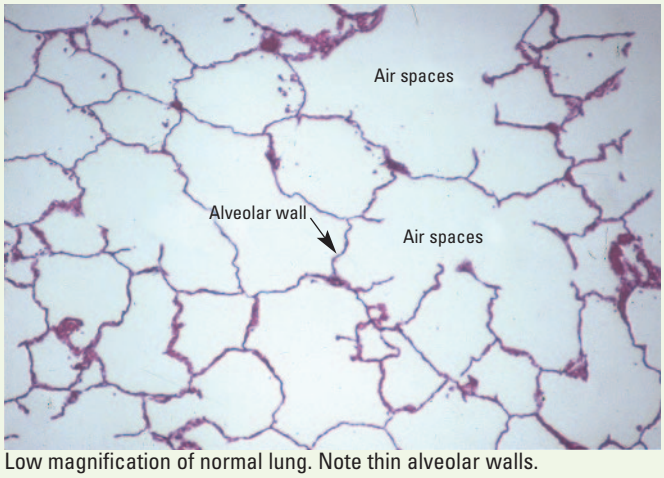
New guidelines for diagnosing and managing the disease demand a fresh approach to nursing care.

By Kathleen Oare Lindell, MSN, RN, and Susan S. Jacobs, MS, RN

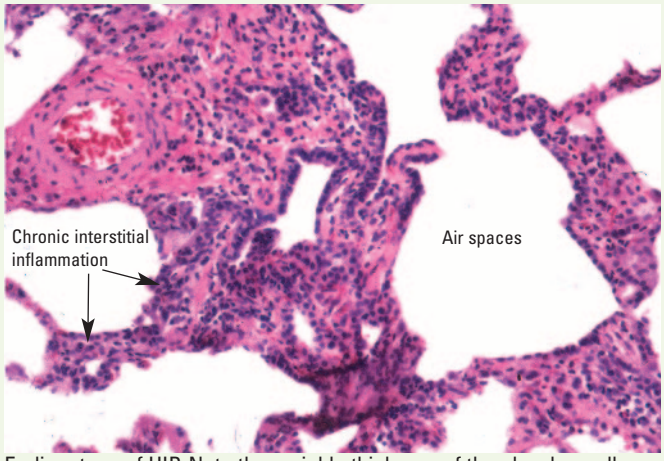


graphic (CT) scan reveals "honeycombing" (a pattern of dense, slightly irregular circular shadows) and irregular bronchial dilation ("traction" bronchiectasis) at the bases of the lungs. An exercise desaturation study reveals decreased oxygen saturation (SaO_2) upon exertion, such that Ms. Dohany requires 3 L/min of oxygen to maintain a saturation of greater than 90%. Results of an echocardiogram to evaluate for pulmonary hypertension, which is often associated with chronic hypoxia, are normal. Six weeks after Ms. Dohany's initial symptoms, the pulmonologist advises an open-lung biopsy. The results are consistent with usual interstitial pneumonia (UIP), a hallmark of idiopathic pulmonary fibrosis (IPF), a rapidly progressive, debilitating disease that afflicts an unknown number, though one source estimates about 50,000 people in the United States.[1]

IPF is more common than cystic fibrosis and amyotrophic lateral sclerosis (Lou Gehrig disease). Yet many people have never heard of it, and clinicians have struggled to understand it. Recent advances in research and new diagnostic guidelines are giving IPF increased visibility among providers and the public. The following is an overview of diagnosis, disease progression, treatment, and ongoing research.



Low magnification of normal lung. Note thin alveolar walls.



Earlier stage of UIP. Note the variable thickness of the alveolar walls and the interstitial chronic inflammation.

End-stage fibrotic remodeling in the lung ("honeycomb" lung) as a result of UIP.

TERMS AND CHARACTERISTICS

IPF is one of the approximately 200 diseases known as interstitial lung diseases, characterized by inflammation, scarring, and stiffening of the alveolar walls. "Honeycombing fibrosis," a process of cyst formation, is a hallmark of IPF and causes loss of lung compliance or elasticity. IPF results in irreversible loss of the lung tissues' ability to transport oxygen, causing dyspnea, and is often accompanied by a dry, hacking cough. There is no cure.

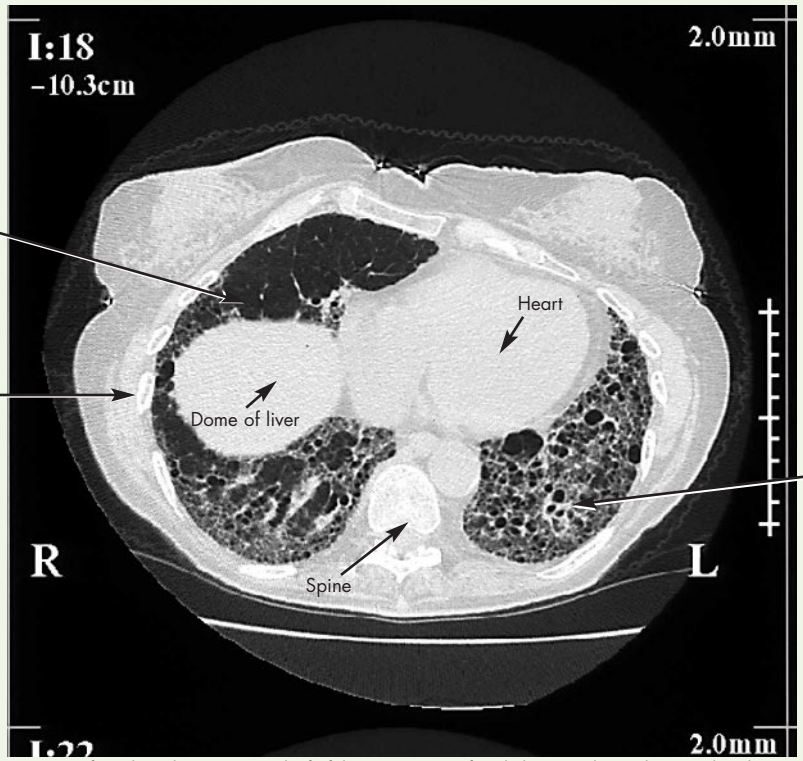
The insidious nature of IPF and its similarities to other interstitial lung diseases make it one of the most difficult pulmonary conditions to diagnose and manage. Pulmonary fibrosis can sometimes be linked to a particular cause (such as exposure to occupational or environmental contaminants or to certain medications) or associated with a connective tissue disease (such as rheumatoid arthritis or scleroderma). But because most cases have no known cause, they're termed idiopathic.

That the disease is known by various names has led to further confusion about IPF. For example, IPF is known in Europe as "cryptogenic fibrosing alveolitis." And until recently, in the United States "idiopathic pulmonary fibrosis" referred to a group of diseases now known as the idiopathic interstitial pneumonias, a subcategory of interstitial lung disease that includes UIP, desquamative interstitial pneumonia, and respiratory bronchiolitis–associated interstitial lung disease. IPF is now recognized as a distinct clinical entity, a specific form of chronic fibrosing interstitial pneumonia of unknown etiology not associated with disease in other organs.[2]

In 2000 the American Thoracic Society (ATS) and the European Respiratory Society, in collaboration with the American College of Chest Physicians, published an international consensus statement on the diagnosis and treatment of IPF.[3] The statement provided a revised, more specific definition of the disease and named UIP as "the histopathological pattern that identifies patients with idiopathic pulmonary fibrosis."[3] Found on lung biopsy, UIP is marked by mixed areas of normal tissue, interstitial inflammation, fibrosis, and honeycombing. Because the presence of UIP is considered conclusive evidence of IPF, the consensus statement recommends surgical lung biopsy for most patients suspected of having the disease.

EPIDEMIOLOGY AND RISK FACTORS

Until recently, varying terminology and lack of standard diagnostic criteria have complicated the gathering of accurate statistics about people with IPF.



Courtesy of Mark L. Shapiro, MD, Chief of the Department of Radiology, Englewood Hospital and Medical Center, Englewood, NJ

Virtually all patients have abnormal high-resolution computed tomographic scans.

The disease appears to be relatively uncommon, with about 15,000 new cases diagnosed annually in the United States.[1] Incidence is estimated at 20 to 30 per 100,000.[1] Patients typically present with the disease between ages 50 and 70; about two-thirds of people are older than 60 when it arises. IPF occurs more often in men than in women.[1] Survival is estimated at three to five years postdiagnosis, although individual case outcomes vary.[2]

Several risk factors have been identified: among them are tobacco use, use of antidepressants, presence of gastroesophageal reflux disease, occupational or environmental exposure to irritants such as asbestos or metal dust, history of viral infection, and genetic predisposition. When the disease appears in families, it's known as familial pulmonary fibrosis, and its manifestation, diagnosis, treatment, and prognosis are similar to those of the idiopathic form, except that the disease arises at an earlier age.

PATHOGENESIS

Although etiology remains a mystery, research indicates that IPF results from the lungs' normal healing processes going awry.[2] Inflammation is the body's normal response to injury, followed by rapid restoration of healthy tissue. But in patients with IPF (or other interstitial lung diseases), the lungs become chronically inflamed, leading to injury and uncontrolled fibrosis. Eventually, the lung parenchyma becomes irreversibly scarred, thereby compromising gas exchange.

Conventional treatment with corticosteroids and cytotoxic agents has sought to eliminate or suppress the inflammatory process. But this approach generally doesn't improve patients' chances of survival or the quality of their lives.[4]

Recent research indicates that fibrosis is more prevalent than inflammation in IPF and that fibrotic areas vary from case to case. In addition to the honeycombing fibrosis that results from chronic inflammation, some areas show "exuberant" fibroblastic activity attributed to sequential, acute alveolar injury.[2] Current research is focusing on how an imbalance of cytokines (protein growth factors that activate inflammation) contributes to pulmonary fibrosis.[5, 6]

SYMPTOMS

IPF affects each person differently and at varying rates, which often causes delayed or incorrect diagnoses. Symptoms are nonspecific and insidious, appearing like those of other lung diseases.

Typical early symptoms include dyspnea during physical activity and a nonproductive cough. Patients may also complain of fatigue. As the disease progresses, dyspnea in some patients may occur even at rest and with mild exertion such as talking and eating. Weight loss may occur as well. Enlargement

or clubbing of the fingertips may develop in as many as half of patients.[2] Virtually all patients will have abnormal high-resolution CT scans, although in mild cases a chest X-ray can appear normal.

A majority of patients with IPF present with dry rales on chest auscultation (the sound has been described as that of hook-and-loop fasteners being opened, or "Velcro-like")[2] heard early in inspiration and often at the lung bases. It's not unusual for patients presenting with dry crackles in the lung bases to be misdiagnosed with pulmonary edema and treated with diuretics.

DIAGNOSIS

The differential diagnosis of IPF spans a broad spectrum: patients are often assessed for asthma, acute bronchitis, or heart disease before being correctly diagnosed with IPF.[7] Researchers have described the process of diagnosing IPF as one of exclusion.[8] A multidisciplinary approach is necessary, and nurses should be familiar with the steps in a typical assessment for IPF. The initial evaluation should include

- a thorough physical examination.
- a complete patient history, including possible exposure to environmental irritants.
- an assessment of pulmonary function by spirometry (including lung volumes, carbon monoxide–diffusing capacity, and pulse oximetry readings at rest and after exercise).
- chest X-rays and a high-resolution CT scan.

According to the international consensus statement, surgical lung biopsy is also recommended for most patients, especially those with suspected IPF who have clinical, physiologic, or radiologic features atypical of the disease.[3] Video-assisted thoracoscopic surgery is the preferred technique. The biopsy allows pathologists to distinguish UIP—the histopathologic hallmark of IPF—from other idiopathic interstitial pneumonias. Bronchoalveolar lavage alone may rule out alternative diagnoses but is not diagnostic of UIP.

Although not conclusive, expiratory flow patterns can provide valuable clues. For example, in patients with chronic obstructive pulmonary disease (COPD), expiratory flow rates are decreased because of the loss of elastic recoil and "floppy airways," but in patients with restrictive lung disease elastic recoil and flow rates increase. Total lung volumes increase with COPD as a result of hyperinflation and air trapping, whereas with IPF total lung volumes decrease as a result of tissue scarring and shrinkage. Pulmonary function tests will reveal these differences. In patients who smoke or previously smoked, these tests may also show a combination of obstructive and restrictive disease. For patients with IPF, the diffusion capacity test is the most important to follow, as it reflects the amount of function at the alveolocapillary membrane.

DISEASE PROGRESSION

The prognosis for patients with IPF remains poor. The median survival time after diagnosis is less than three years.[9] The survival time may be shorter in older patients but is often longer in younger patients with fewer risk factors. Other factors associated with a poor prognosis include

- histologic features (evident on lung biopsy) that indicate production of scar tissue.
- male sex.
- tobacco use.
- a more rapid onset of symptoms.
- a more rapid disease progression.
- a greater degree of fibrosis (evident on high-resolution CT scanning).
- poor lung function (evident on pulmonary function tests).
- the presence of neutrophils and eosinophils in bronchoalveolar lavage fluid.
- no response to drug therapy.

Conventional approaches to treatment have included antiinflammatory drugs such as corticosteroids, immunosuppressive and cytotoxic agents such as azathioprine (Imuran) and cyclophosphamide (Cytoxan), and antifibrotic agents such as colchicine, either individually or in combination. Treatment may result in temporary relief of shortness of breath and enhanced results on pulmonary function tests. But there's no evidence that treatment improves patient outcomes, and the drugs' adverse effects are well known and may diminish the quality of patient's lives. Clinicians should therefore focus on managing symptoms and optimizing functional status.

SYMPTOM MANAGEMENT

Dry, hacking cough and dyspnea are the predominant symptoms of IPF. Nurses can help by teaching patients and their families strategies for symptom management.

Cough associated with IPF may be the result of interstitial lung receptors sensing inflammation and alveolar scarring. When coughing is prolonged and severe, it can worsen dyspnea and cause oxygen desaturation, but it rarely results in presyncopal episodes. Patients may describe profound breathless-

Practicing Optimism

A patient's view of his pulmonary rehabilitation program.

At the pulmonary rehabilitation session, when no one is looking, John Jennette inches the treadmill setting up past 4.2 mph. He's got his supplemental oxygen and feels good today. He feels like pushing himself.

A nurse appears at his side within minutes. "You're breathing too hard. What are you up to?" She makes him return the machine to the prescribed setting. When he describes the incident later, Mr. Jennette, who suffers from IPF, laughs.

"I have to watch to make sure I don't get busted," he says. "Going to pulmonary rehabilitation every Tuesday and Thursday for three years has been one of the best things I've done. The nurses read me like a book."

One nurse with a background in cardiology knew that Mr. Jennette was receiving interferon the nights before his rehabilitation sessions and suggested he change the night of injection.

"Once, when I was on interferon, my heart started to race during the session; she noticed it right away," Mr. Jennette says. "I feel secure in the rehab environment, where the nurses and trained personnel can look at these little details."

John Jennette

Attending pulmonary rehabilitation sessions is part of how Mr. Jennette has chosen to handle IPF since he was diagnosed with the disease three years ago at age 43. To stem the progress of the disease, he also prays, tries to stay optimistic, exercises, participates in clinical and National Institutes of Health (NIH) studies, and attends a support group.

Not every patient with IPF is as active as Mr. Jennette. Still, he says, even for people who don't push themselves as he does, pulmonary rehabilitation has numerous benefits.

"Some people who attend are elderly. If they weren't at rehab, they'd be sitting at home," he says. "It gives them a chance to mix it up, to have nurses see them, and to set goals—'this week I'll do another half mile on the bike.' Many are frail, and they still have to get around, climb stairs, shop . . . it's important to keep in shape."

Mr. Jennette credits the nurses at the pulmonary rehabilitation sessions with helping patients understand IPF. "Often people will get on exercise machines and feel out of breath and say, 'I don't know what's happening to me.' The nurses play videotapes to show people what's happening to them physically."

At times, Mr. Jennette is frustrated by the relative obscurity of IPF. "There's so much focus on the 'biggies'—cystic fibrosis, emphysema, bronchiectasis, asthma. I challenge you: try to find a chart of the IPF lung. Nurses can't know everything, but I would tell them: try to keep your eyes and ears open, find out more about IPF. A lot of cases go undetected or misdiagnosed until it's too late.

"The nurses can make or break the experience," he added. "When you have only five to 10 minutes with doctors, especially when you're first diagnosed, you're confused. Nurses are capable of sorting that kind of mayhem out. They can tell just by looking at you that you're not right today. Sometimes during rehab sessions they'll catch little things—a doctor said you should be on two liters of oxygen, but you're out of breath, and you really need three."

To Mr. Jennette, modest acts of caring have made the difference, especially when he hasn't felt well. Once, when he had a 104°F fever after a bronchoscopy, a nurse warmed a blanket for him in the microwave oven.

Nurses can serve as a significant source of support to families as well as to patients, he says. "If you take a little time to talk to people, you find out a lot more than you do with a stethoscope. If someone is balking at exercise or a lung transplant, sometimes a nurse will draw the husband or wife aside to talk. That really helps. When I get sick, my whole family gets sick. Everyone's life goes downhill.

"The other day I had a heart-to-heart talk with an NIH nurse. She told me that 113 people have died in her arms. She loves nursing, it's the only career she ever wanted to pursue . . . and she said that those times that she may be the last person in someone's life, you can't put a dollar amount on that."—*Emmeline Chang*

ness and panic. Although usually nonproductive, in some cases the cough may produce mucoid sputum, suggestive of concurrent chronic bronchitis.

The cough, frequently the symptom most troubling to the patient, is difficult to treat. Oral or inhaled prednisone (Deltasone), a corticosteroid, doesn't consistently relieve the cough but may reduce its severity. Benzonatate (Tessalon), an oral cough suppressant, acts directly on the brain's "cough center," the medulla. If these measures fail, the use of opioids may suppress the cough as well as provide some relief of dyspnea.

Alternative remedies such as inhaled steroids, throat lozenges or honey and lemon in hot water have been reported to provide relief. Patients should be instructed to avoid known irritants that may trigger cough, such as aerosol sprays and smoke from fireplaces or barbecue grills. If a patient is known to experience oxygen desaturation during coughing episodes, increasing the supplemental oxygen flow rate may help prevent other adverse effects such as dizziness and breathlessness.

Dyspnea. The shortness of breath that accompanies interstitial lung disease results from increased lung stiffness and the additional effort necessary to move air in and out of the lungs. Even at normal arterial oxygen levels, the brain senses that the body is expending a great deal of effort for very little return. The rapid, shallow breathing pattern seen in patients with IPF is unlike the slow rate with prolonged expiration seen in patients with COPD.

Managing dyspnea requires multiple approaches. It's important for patients with IPF to know that even when their oxygen saturation levels are normal,

they may still feel uncomfortably short of breath. Explaining the underlying disease mechanisms may alleviate anxiety and help them to understand how various interventions work. Dyspnea doesn't necessarily indicate a need for supplemental oxygen. However, if the patient's oxygen levels are low (partial pressure of oxygen below 60 mmHg or SaO_2 below 90%), supplemental oxygen should be given.

Patients sometimes limit their physical activities in an attempt to avoid shortness of breath. But physical inactivity can lead to significant deconditioning and can worsen dyspnea. Regular aerobic exercise such as walking or using a stationary bicycle may help decrease dyspnea and increase overall strength, flexibility, and endurance as well as improve mood and facilitate sleep.[10, 11] It's also vital that patients learn and practice relaxation techniques to manage the panic that often accompanies dyspnea. Such techniques are best learned when a patient isn't experiencing it. Other strategies include avoiding pulmonary infections, stopping tobacco use, maintaining optimal body weight, eating smaller and more frequent meals, and learning ways to conserve energy.

PHARMACOLOGIC TREATMENT

Drug treatment may be initiated in an attempt to suppress or reverse inflammation. The international consensus statement recommends that treatment begin at the first indication of clinical or physiologic impairment[3] and should include prednisone in combination with either cyclophosphamide or azathioprine. The statement also acknowledges that no drug has been proven to reverse the course of IPF.

Prednisone, a corticosteroid, has strong antiinflammatory effects. According to ATS dosage guidelines, patients should start with 0.5 mg/kg/day for four weeks, followed by 0.25 mg/kg/day for eight weeks, then taper to 0.125 mg/kg/day.[3] Adverse effects may include mood variability, sleeplessness, increased blood glucose levels, osteoporosis, hypertension, cataracts, ulcers, bruising, and infection resulting from immunosuppression. Patients must know that because adrenal suppression occurs with prolonged treatment, they must not suddenly stop taking prednisone.

Cyclophosphamide, an immunosuppressant, decreases inflammation and can be taken either with prednisone or alone. The ATS recommends starting at 25 to 50 mg daily and increasing it in 25-mg increments every one to two weeks until 150 mg daily is reached.[3] Leukopenia, quantified as a white blood cell count below 3,000/L, is the most common adverse effect and can be detected by performing a complete blood count (CBC) every two weeks; if it does occur, the cyclophosphamide dosage should be decreased. Bladder inflammation, although a relatively uncommon complication, can lead to gross hematuria and painful urination. Patients should be instructed to monitor their urine for blood and to drink 2 L of noncaffeinated fluids daily before late afternoon to prevent the drug's toxic effects.

Azathioprine, an immunosuppressive agent, can be used instead of cyclophosphamide and may be taken either with prednisone or alone. The dosage begins at 25 to 50 mg/day and is increased by 25-mg increments every one to two weeks until the maximal dose of 150 mg/day is achieved.[3] Adverse effects include hepatotoxicity, leukopenia, and, rarely, pancreatitis. Patients should be monitored with CBCs and pulmonary function tests every two to four weeks.

Colchicine, an antifibrotic agent occasionally used to treat IPF, appears to have no more beneficial effect than other agents do. However, its adverse effects, which include diarrhea and gastrointestinal upset, are less severe overall than those of cyclophosphamide and azathioprine.[4] A recent study revealed that colchicine also has fewer adverse effects than prednisone does, although neither drug resulted in improvement.[12]

Deciding whether to continue pharmacologic treatment despite disease progression can be difficult for all involved. Even when pulmonary function test results and symptoms show no significant improvement, stabilizing or slowing the disease process remains a reasonable goal. Two studies have identified some improvement when adding either azathioprine[13] or cyclophosphamide[14] to treatment with low-dose prednisone, and the ATS treatment recommendations are based on these studies.

TOBACCO USE AND IPF

Quitting the use of any tobacco product (cigarettes, cigars, pipe tobacco, and smokeless tobacco) is one of the most important steps people with IPF and their family members can take to preserve and improve patient health.

Nurses can help patients who use tobacco to develop a cessation plan, which should include a quitting date. One can quit either all at once ("cold turkey") or, with the aid of medications, in stages. Medications that suppress or ease withdrawal symptoms include nicotine replacement therapy products (which administer nicotine by gum, patch, or nasal spray) and nonnicotine drugs such as bupropion (Zyban and others). Nurses can also

teach patients to recognize cues that trigger or intensify the desire to smoke (drinking coffee or alcohol, or stress, for example) and to develop appropriate ways to cope (such as learning how to relax).[15] Keeping a diary may help. A professionally supervised behavior modification program increases the patient's chance of success.[16]

SUPPLEMENTAL OXYGEN

Because the tissue scarring caused by the disease makes it difficult for oxygen to cross the alveolocapillary membrane, patients often need supplemental oxygen, which increases oxygen pressure at the alveoli to maintain SaO_2 at greater than 90%. An arterial blood gas test, an exercise desaturation study, or a six-minute walk test will determine whether the patient needs oxygen at rest or on exertion. A nocturnal oxygen saturation study or a formal sleep study will determine if the patient needs oxygen during sleep.

If supplemental oxygen is warranted, it's important to ensure that the patient be able to maintain the maximal possible mobility.[17] The prescription should specify oxygen flow rates for the patient at rest, during exertion, and during sleep, as well as the form and method of oxygen delivery, whose systems may be stationary (concentrator, large tank) or portable (small tank). The oxygen itself may be extracted from room air (concentrator) or supplied as a compressed gas or a liquid (large or small tank). Lightweight portable tanks filled with liquid oxygen allow patients the most mobility. (See *Home Supplemental Oxygen Delivery Systems,* page 40.)

Although most patients receive continuous-flow oxygen through a nasal cannula, some may do so through an oxygen-conserving device. Transtracheal catheters deliver oxygen directly into the trachea; patients reach the desired level of oxygen saturation after receiving only about half as much oxygen as they'd need to reach the same level with a nasal cannula. Reservoir cannulae have a reservoir near the nose that fills with oxygen during exhalation; the reserved oxygen is inhaled at the onset of the next inhalation. Intermittent-demand devices either pulse oxygen during early inhalation or they provide oxygen flow throughout inhalation but eliminate flow during exhalation.[17, 18] Oxygen conservation devices often aren't suitable for this patient population.

Patients with IPF should be tested on their abilities to use such supplemental oxygen equipment and to maintain the desired oxygen saturation, particularly during exercise. It's important to teach patients to use oxygen as prescribed; many don't out of fear they will become addicted. Patient education should emphasize the rationale for oxygen therapy (to help oxygen to cross a fibrosed alveolocapillary membrane), as well as the effects of inadequate oxygen on the heart and brain.

RESOURCES

American Lung Association
www.lungusa.org

National Jewish Medical and Research Center
www.njc.org

Pulmonary Fibrosis Association
www.pulmonaryfibrosisassn.com

Dorothy P. & Richard P. Simmons Center for Interstitial Lung Diseases
http://ipf.upmc.com

LUNG TRANSPLANTATION

Lung transplantation should be considered for patients who meet the established criteria and whose conditions are deteriorating despite medical management. Because IPF progresses rapidly and unpredictably, and the waiting period for a suitable donor organ may exceed two years, early placement on the transplantation lists is necessary. IPF is more common among the elderly, and advanced age along with other conditions may preclude transplantation.[19]

Single-lung transplantation is currently preferred to double-lung transplantation, although the latter is preferable in patients with pulmonary hypertension (transpulmonary gradient greater than 20 mmHg). After successful transplantation, the patient no longer needs supplemental oxygen. Lung volume and D_{LCO} values are typically 60% to 70% of those predicted,[20] and pulmonary hypertension and right ventricular dysfunction are improved. Lung transplantation remains the only treatment that improves both the survival rate and the quality of the lives of patients with IPF.[20, 21] National survival rates for patients with IPF who undergo lung transplantation are 65% after one year and 48% after five years.[22]

REHABILITATION AND SUPPORT

Pulmonary rehabilitation has become the standard of care for chronic lung disease because of its effectiveness in mitigating symptoms, decreasing hospital stays, increasing exercise tolerance, and maximizing functional ability.[23, 24] Such programs are typically offered by community hospitals and coordinated by an RN or a respiratory therapist. Participation

Home Supplemental Oxygen Delivery Systems

Delivery system	Advantages	Disadvantages	Comments
Liquid oxygen tank	Portable tank can be refilled from reservoir by the patient. A large portable tank holds a 6–8-hr supply at 2 L/min; reservoir will last approximately 7–10 days at 2 L/min continuously. Smaller portable tanks weighing less than10 lbs. also are available.	Liquid system can be slightly more expensive, depending on where the patient lives; availability generally limited to urban areas.	As liquid warms to gas, some gas is vented from the system. In hot weather, this evaporation accelerates and may decrease reservoir duration to less than 1 week.
Compressed gas tank	Good availability in most areas. Portability possible with cart. Aluminum E* and A cylinders that are markedly lighter than steel and easier to maneuver are available.	At 2 L/min flow, an H or J tank lasts about 50 hr; storage of 4–5 large cylinders in the home necessary to ensure 7–10 day supply; portable cylinder on cart is cumbersome and heavy. At 2 L/min, an E cylinder lasts about 4–5 hr and an A cylinder about 8–10 hr.	Some smaller tanks (D or M) may be used; these weigh about 10 lbs. and can be refilled from large cylinders. Tank can be carried on shoulder strap, backpack, or fanny pack, or placed on portable cart.
Concentrator	On wheels, movable from room to room; weekly delivery of supply unnecessary because unit extracts oxygen from room air and delivers it continuously; compact, excellent system for rural or homebound patient.	Older models can be noisy; electricity bill may increase by $20–$30/mo., not necessarily reimbursed by third-party payers; flow greater than 3L/min results in significant decrease in concentration. Patient will need backup oxygen tank in case electricity fails.	Concentrator should be kept in room other than the bedroom; extension tubing should be used if noise disturbs sleep.
Intermittent delivery	Simple to use; delivery rate changes with respiratory rate (that is, the faster the patient's respiratory rate, the higher the rate of delivery).	Mechanically complex; only safe to use with portable system when patient is awake, unless there's an alarm to detect disconnection of patient. Oxygenation possibly less efficient on exertion.	System may be a separate unit that can be used with liquid or compressed gas cylinder, or it can be built into portable liquid oxygen unit. Less desiccation than with other systems, rarely needs humidification. Higher flow rates (greater than 4 L/min) tend to dry out nose membranes.

*Letters refer to tank size: the earlier the letter's position in the alphabet, the smaller the tank.

Source: Lindell KO, Van Sciver T. Obstructive pulmonary diseases. In: Lewis SM, et al., editors. *Medical-surgical nursing: assessment and management of clinical problems.* 5th ed. St. Louis: Mosby-Year Book; 2000. p. 660-715.

requires a physician referral and is usually covered by health insurers.

Usually, a group of six to 10 patients meets for two to three sessions weekly for six weeks. Each two-to-four-hour session incorporates education and exercise components provided by an interdisciplinary health care team. Topics include an overview of chronic lung diseases, medications, nutrition, energy conservation techniques, relaxation techniques, and supplemental oxygen use. Individualized exercise sessions involve stretching, weight training, and aerobic exercise using a treadmill or stationary bicycle. Oxygen saturation monitoring and telemetry are provided. The sessions also provide social and emotional support. For example, patients who become anxious and short of breath when they exercise alone often feel safer exercising with others in a monitored environment. (See *Practicing Optimism*, page 37.)

Graduates of such programs may choose to participate in a maintenance exercise program. Those who can't attend a hospital-based maintenance program should follow a home-based program tailored to their needs.

Patients newly diagnosed with IPF and those facing the daily challenges of living with the disease

may benefit from a peer support group. Many communities have American Lung Association–affiliated "Better Breather Clubs" for patients with lung disorders, and the Coalition for Pulmonary Fibrosis (www.coalitionforpf.org) offers detailed guidelines for establishing support groups.

EXPERIMENTAL TREATMENT AND RESEARCH

Survival of patients with IPF will improve only when more effective therapies are developed, which depends on a better understanding of the underlying disease. Recent research using molecular biologic approaches has provided new insights into the pathogenesis of IPF, and as a result, a wide range of potential mediators of inflammation and fibrosis have been identified, and some are now the focus of clinical research.

Such molecular biologic approaches examine specific steps in the course of the disease. Therapies currently under investigation include

- antifibrotic agents to suppress scarring (such as interferon gamma and certain antihypertensive medications).
- antioxidants to prevent lung damage (such as acetylcysteine and glutathione).
- monoclonal antibodies to inhibit "bad" cytokines and antagonists of their receptors.

For example, a recently completed multicenter, phase-III study examined the safety and efficacy of interferon gamma-1b in patients with IPF in whom treatment with prednisone failed.[25] Preliminary data analysis indicates that although interferon gamma-1b didn't appear to have treatment benefit, it may have some favorable effect on survival in patients with mild or moderate disease (FVC greater than 55%). Other, ongoing studies focus on identifying possible gene markers for pulmonary fibrosis and factors that can influence prognosis and help to predict the length of survival in patients with interstitial lung diseases.

END-OF-LIFE CARE

The goals of treatment of IPF include managing symptoms and relieving discomfort. Palliative care should be offered from the time of diagnosis, even while the patient pursues treatments to slow or stabilize the disease. But a time may come when the patient and family acknowledge that treatment is ineffective, that pulmonary function tests and oxygen levels have deteriorated, and that dyspnea is increasing. As the disease progresses, the patient may wish to consider other care options, such as enrollment in clinical trials that offer experimental drugs, lung transplantation (as appropriate), or the cessation of current treatment of IPF in favor of focusing on a plan for end-of-life care. Nurses should provide reassurance that end-of-life care is available, that dyspnea is manageable, and that both the patient and family have the right to participate in decisions about care. Nurses should support the patient's access to palliative care.

Managing end-of-life care entails deciding whether and when to begin hospice care, which can profoundly change the patient's and family's experiences of death. Hospice nurses educate families in how to provide or help in providing care, obtain medical equipment for the home, have medications delivered, arrange respite care for family caregivers, help titrate medications for relief of dyspnea and discomfort or pain, and provide emotional and spiritual support. ▼

Editor's note: In May 2002, *AJN* began publishing a bimonthly series on palliative nursing, with an emphasis on end-of-life care. One of the premises of this series is that for people with progressive and debilitating illnesses not responsive to curative treatment, palliation of symptoms is a responsibility of all nurses and should not be deferred until the patient is at the end of life. Idiopathic pulmonary fibrosis (IPF) requires a palliative approach regardless of the patient's participation in treatments or clinical trials. Nurses are encouraged to review this continuing education series, offered free online at www.ajnonline.com. The series, which will continue through 2004, is funded in part by a grant from the Robert Wood Johnson Foundation.

Complete the CE test for this article by using the mail-in form available in this issue or by going to Online CE at www.ajnonline.com.

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