Idiopathic Pulmonary Fibrosis
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Abstract
Idiopathic pulmonary fibrosis (IPF) is being more frequently diagnosed in India, due to its increased awareness, better availability of computed tomography (CT) and fiberoptic bronchoscopy. IPF has the histological appearance of usual interstitial pneumonia (UIP) on surgical lung biopsy. Recent research has given a new insight into the etiology of the disease. Clinical criteria have been specified for presumptive diagnosis of IPF and distinguishing IPF from other conditions. The conventional therapy has been steroids and immunosuppressive agents. But only a minority of patients respond to such a therapy. Immunomodulators (interferon Y1b), antioxidants (Acetyl cysteine) and antifibrotic agents (like pirfenidone) are being studied as novel therapies in this, otherwise, fatal condition. Lung transplantation is the only hope for those patients who show progressive deterioration on medical treatment. Living-donor lobar lung transplantation has been developed as a procedure for patients considered too ill to await cadaveric lung transplantation.

INTRODUCTION
A variety of acute and chronic lung diseases with variable degrees of pulmonary inflammation and fibrosis are collectively called as interstitial lung diseases (ILDs). The term interstitial is a misnomer, since the disease not only affects the interstitium, but also involves all the cellular and interstitial components of the alveolar wall extending into the alveolar space. Hence it is more appropriately called Diffuse Parenchymal Lung Disease (DPLD). Idiopathic pulmonary fibrosis (IPF) also known as cryptogenic fibrosing alveolitis (CFA) is a distinct entity amongst them. IPF is defined as a specific form of chronic fibrosing interstitial pneumonia limited to the lung, with the histopathology of usual interstitial pneumonia (UIP) on surgical (thoracoscopic or open) lung biopsy.

Epidemiology
Although the exact incidence and prevalence of the disease is not known, patients with this disease comprise about 15% of a pulmonary physician’s practice. Mortality from cryptogenic fibrosing alveolitis continues to increase in many countries. In India, this was earlier considered to be a rare disease. In 1979, Jindal et al published their data on 61 cases of DPLD seen over a period of five years. However, the scenario is different now and the disease is no longer rare or uncommon. Recently the same center published data on 76 patients with IPF diagnosed over a 16-month period showing a definite increase in the frequency of diagnosis.

A number of other publications from India have described various aspects of the disease.

Pathology
Hamman and Rich followed by Liebow described the pathologic features of interstitial pneumonias. The present accepted pathological classification has four histologically distinct forms, usual interstitial pneumonia (UIP), desquamative interstitial pneumonia (DIP)/ respiratory bronchiolitis interstitial lung disease (RBILD), acute interstitial pneumonia (AIP, Hamman-Rich disease) and nonspecific interstitial pneumonia (NSIP).

The pathologic changes that characterize UIP are distinguished by variation in the location and age of the lesions, with a predilection for the peripheral subpleural parenchyma. Fibrotic zones with associated honeycombing alternate with areas of relatively unaffected lung tissue. Fibrotic areas characteristically vary in age and activity. The major clinical and pathologic features of the four idiopathic interstitial pneumonias are contrasted in Table 1.

Pathogenesis
The hallmark lesion of idiopathic pulmonary fibrosis is the fibroblast foci. These foci reveal vigorous replication of mesenchymal cells and exuberant deposition of...
fresh extracellular matrix. Such foci are typical of alveolar epithelial-cell injury, with endoluminal plasma exudation and collapse of the alveolar space. Newer insights suggest that idiopathic pulmonary fibrosis results from sequential acute lung injury. The resultant wound-healing response to this injury culminates in pulmonary fibrosis (Table 2). Several interacting factors that modify the fibrotic response include the genetic background of the patient, the predominant inflammatory phenotype (Th1 or Th2), and a variety of environmental toxins.¹⁴

**Diagnosis**

Patients with idiopathic pulmonary fibrosis typically present with gradual onset of exertional dyspnoea and nonproductive cough. It presents in the fifth and sixth decades and is slightly more common in men than women.¹⁵ Some Indian studies suggest that the mean age of Indian patients is 50.6 years; almost one decade earlier as compared to Western series.⁶ Systemic symptoms, like low-grade fever and myalgia, may be present but are uncommon.

Clubbing is noted in up to 50% of patients.¹⁵ Crackles are present in majority of patients. These are typically ‘dry’, end-inspiratory, and ‘Velcro’ in quality, mostly in the lung bases. Cyanosis, cor pulmonale, accentuated pulmonary second sound, right ventricular heave, and peripheral edema are seen in terminal stages of IPF.

Routine laboratory evaluation of a patient with suspected IPF is for excluding other causes of parenchymal lung disease. An elevated erythrocyte sedimentation rate and hypergammaglobulinemia may be found. Positive anti-nuclear antibodies or rheumatoid factor occur in 10-20% of patients and are usually in low titers.¹⁶

Pulmonary-function tests typically reveal a parenchymal restrictive ventilatory defect, with reduction in total lung capacity, functional residual capacity, and residual volume.¹⁷ However, patients who smoke may also have a concurrent obstructive ventilatory defect. Impairments in gas exchange may be demonstrated by a decrease in the carbon monoxide diffusing capacity or by hypoxemia with graded exercise testing.

Bilateral basal symmetrical peripheral reticular opacities with decreased lung volumes are characteristic findings on the chest radiograph.¹⁵ Volume loss is characterized by diaphragmatic elevation and depression of the fissures. Progressive fibrosis ultimately leads to cystic dilatation of the distal air spaces, which is visible as peripheral “honeycombing.” An alternative diagnosis or superimposed complicating illness should be suspected if chest radiograph shows additional features like pleural effusions, air bronchograms, confluent shadows, or hilar adenopathy. Most of the patients with IPF have abnormalities on chest radiograph. Rarely patients can have a normal chest radiograph but have evidence of IPF either in High Resolution Computerised Tomography (HRCT) or surgical lung biopsy.

HRCT has greatly enhanced the evaluation of interstitial lung diseases by increasing spatial resolution, facilitating visualization of parenchymal detail at the level of the pulmonary lobe. The protocol for HRCT scanning of patients with suspected IPF includes a section thickness of 1-1.5 mm collimation and interval size between sections of 1-2 cm. The HRCT pattern of

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**Table 1 : Contrasting clinical and pathological features of idiopathic interstitial pneumonias**²¹⁻²³,¹⁵

<table>
<thead>
<tr>
<th>Clinical features</th>
<th>UIP</th>
<th>DIP/RBILD</th>
<th>AIP</th>
<th>NSIP</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age, years</td>
<td>50-60</td>
<td>36-42</td>
<td>49</td>
<td>49</td>
</tr>
<tr>
<td>Onset</td>
<td>Insidious</td>
<td>Insidious</td>
<td>Acute</td>
<td>Subacute/insidious</td>
</tr>
<tr>
<td>Mean survival</td>
<td>3-5 yr</td>
<td>12 yr</td>
<td>1-2 months</td>
<td>17 months</td>
</tr>
<tr>
<td>Response to steroids</td>
<td>Poor</td>
<td>Good</td>
<td>Poor</td>
<td>Good</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Pathological features</th>
<th>Temporal appearance</th>
<th>Interstitial inflammation</th>
<th>Collagen fibrosis</th>
<th>Fibroblast proliferation</th>
<th>Honey-combing</th>
<th>Intraalveolar macrophage accumulation</th>
<th>Hyaline membranes</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Variegated</td>
<td>Uniform</td>
<td>Minimal</td>
<td>Variable</td>
<td>No</td>
<td>Yes, diffuse or peribronchiolar</td>
<td>No</td>
</tr>
<tr>
<td></td>
<td>Uniform</td>
<td>Minimal</td>
<td>Minimal</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Occasional</td>
</tr>
<tr>
<td></td>
<td>Uniform</td>
<td>Uniform</td>
<td>Prominent</td>
<td>Variable, diffuse</td>
<td>No</td>
<td>Occasional</td>
<td>No</td>
</tr>
</tbody>
</table>

**Table 2 : Pathogenesis of IPF**

![Diagram of Pathogenesis of IPF]

Unidentified repeated stimulus  
Repetitive lung injury  
Inflammation  
Abnormal wound healing  
Th1-Th2 balance  
LUNG FIBROSIS  
Genetic factors  
Abnormal lung fibrosis  
Abnormal wound healing  
Th1-Th2 balance  
LUNG FIBROSIS  
Genetic factors
IPF commonly shows patchy, predominantly peripheral, subpleural, bibasal reticular abnormalities, and areas of traction bronchiectasis with limited amount of ground glass opacity. When early fibrosing alveolitis is suspected HRCT scans need to be performed in the prone position to prevent any confusion with the increased opacification seen in the dependent lower lobe postero-basal segments of many normal individuals scanned in the usual supine position. The areas of severe involvement show subpleural honeycombing. The accuracy of a confident diagnosis of UIP made on HRCT by a trained observer is about 90%. Thus, experienced clinicians can make a confident diagnosis of IPF in many patients without the need for biopsy. However, over half the patients with proved IPF had an uncertain diagnosis on HRCT and clinical evaluation. When the diagnostic studies do not support a confident diagnosis of IPF or the clinician is less experienced, a lung biopsy is needed for diagnosis. HRCT has also been proposed as a technique for determining the ‘activity’ of IPF. Some studies have suggested that ground glass opacity on HRCT can be associated with alveolar inflammation and predicts physiologic improvement after steroid treatment. But ground glass opacity is predominantly associated with fibrotic thickening of alveolar septa and intra-alveolar granulation tissue. In patients with predominant ground-glass opacification in the absence of traction bronchiectasis, the pattern generally seems to reflect the NSIP or DIP end of the spectrum of the interstitial pneumonias which are more steroid-responsive.

Bronchoalveolar lavage (BAL) shows increase in polymorphonuclear leukocytes, neutrophil products, eosinophils, eosinophil products, activated alveolar macrophages, alveolar macrophage products, cytokines, and growth factors. BAL is a useful research tool, but its diagnostic usefulness is limited. BAL may substantiate a variety of alternative specific diagnoses provided appropriate laboratory studies are performed (e.g. malignancy, eosinophilic pneumonias, infections). Increase in percentage of neutrophils or eosinophils in BAL fluid have been associated with worse prognosis while BAL lymphocytosis has been associated with a more cellular biopsy, less honeycombing and a greater responsiveness to treatment.

Because of the limited tissue obtained, tranbronchial lung biopsy (TBLB) is not helpful in making the diagnosis of UIP. But in a given patient TBLB may exclude UIP by identifying an alternative specific diagnosis in the right clinical setting (e.g. sarcoidosis, malignancy, infections).

Surgical lung biopsy by open or video-assisted thoracoscopic methods is needed to make a diagnosis of UIP. But it is performed only in a minority of patients with atypical clinical or radiological features. The potential risks and cost associated with surgical lung biopsy need to be balanced against the accuracy of a clinical diagnosis. The usual clinical practice is to make diagnosis of IPF relying largely on clinical and radiological features.

According to American Thoracic Society, in the immunocompetent adult, the presence of all of the following major diagnostic criteria as well as at least three of the four minor criteria increases the likelihood of correct clinical diagnosis of IPF. The major criteria include 1) Exclusion of other known causes of ILD, such as certain drug toxicities, environmental exposures, and connective tissue diseases, 2) Abnormal pulmonary function studies that include evidence of restriction (reduced VC often with an increased FEV1/FVC ratio) and impaired gas exchange (increased alveolar arterial oxygen gradient with rest or exercise or decreased DLco), 3) Bibasilar reticular abnormalities with minimal ground glass opacities on HRCT scans and 4) Tranbronchial lung biopsy or bronchoalveolar lavage (BAL) showing no features to support an alternative diagnosis. The minor criteria include 1) Age > 50 yr, 2) Insidious onset of otherwise unexplained dyspnea on exertion, 3) Duration of illness ≥ 3 months and 4) Bibasilar, inspiratory crackles (dry or ‘Velcro’ type in quality).

Risk factors for IPF

Cigarette smoking has been identified as a potential risk factor with the odds ratio from various regions of the world ranging from 1.6 to 2.9. Environmental exposures to metal dust, wood dust and solvents have been linked with increasing risk of developing pulmonary fibrosis. Viruses (Epstein Barr virus, influenza, cytomegalovirus, hepatitis C) chronic aspiration secondary to gastroesophageal reflux and exposure to antidepressants have been implicated in the pathogenesis of IPF. Hereditary factors may contribute to the risk of developing IPF but no specific genetic marker has been identified. Familial IPF is probably inherited as an autosomal dominant trait with variable penetrance.

Prognosis

IPF progresses in a relentless and often insidious manner with a mean survival ranging from 4 to 6 years after the time of diagnosis. (5-yr survival range 30 to 50%). Spontaneous remissions do not occur. Respiratory failure is the most frequent cause of death, accounting for approximately 40%. Other causes of death in patients with IPF being heart failure, ischemic heart disease, infection, and pulmonary emboli. Bronchogenic carcinoma has been identified with increased frequency (10-15%) in advanced idiopathic pulmonary fibrosis. Indicators of prolonged survival among patients with IPF include younger age (< 50 yr), female sex, shorter duration of symptoms (< 1 yr) with less dyspnea and relatively preserved lung function.
A clinical, radiographic, and physiologic (CRP) scoring system that includes seven variables (dyspnea, chest radiograph, spirometry, lung volume, diffusion capacity, resting alveolar-arterial oxygen difference, and exercise oxygen saturation) was shown to correlate with the degree of fibrosis and the cellular histopathological component of the open lung biopsy from patients with IPF. This scoring system is useful in staging the extent of IPF and monitoring the clinical course. The Composite Physiological Index (CPI) has been recently developed which correlates with the extent of pulmonary fibrosis on CT and is a better mortality predictor than individual pulmonary function indices. The CPI has advantages over the original CRP score in that it is easier to generate and takes into account the extent of emphysema.

In 1999, Akira described an accelerated variant of UIP in which patients progress to respiratory failure in few weeks after the onset of dyspnea. This condition is associated with evidence of peripheral ground-glass opacities and consolidation. Pathological findings were consistent with a diagnosis of UIP.

An acute exacerbation of IPF can occasionally occur similar to “Hamman–Rich syndrome”. It is characterized by acute progression of dyspnea over less than one month, with new, diffuse infiltrates and worsening hypoxemia clinically resembling the acute respiratory distress syndrome. Although the exact incidence of acute exacerbation is not known, 40% of patients who died of an IPF-related cause belong to this category. On pathological examination, some patients have acute alveolar injury without hyaline membrane. CT scans show bronchiectasis or honeycomb cysts or both and ground glass opacities or consolidation that is either peripheral, multifocal, or diffuse in distribution. The prognosis is very poor. In one series of 25 patients with acute exacerbation of IPF requiring mechanical ventilation, there was only one survivor. However, it is imperative that a diagnostic workup be performed to rule out an infection or other reversible causes of respiratory failure.

Treatment of IPF

Sufficient clinical evidence is lacking to show that any treatment definitely improves survival or the quality of life. The rarity and the heterogeneity in its clinical expression make therapeutic interventional prospective trials difficult.

Conventional Treatment Options

Treatment options include corticosteroids, immunosuppressive / cytotoxic agents and antifibrotic agents alone or in combination.

Corticosteroids

Despite their widespread use only 10-30% of patients with IPF improve on quantitative assessment when treated with corticosteroids. Forty percent respond on the basis of subjective or undefined assessment criteria (because of placebo effects or mood-enhancing effects of corticosteroids). Responses are usually partial and transient. If responses are to occur with corticosteroids, improvement is usually noted within 3 months. Prolonged treatment for a minimum of 1 to 2 years and sometimes indefinitely is reasonable for patients exhibiting unequivocal responses to therapy. High-dose intravenous pulse methylprednisolone has no proven advantage over oral corticosteroids. Treatment with corticosteroids alone is now considered inappropriate. Corticosteroids should be used in conjunction with cytotoxic agents.

Cytotoxic Agents

Since response to corticosteroids alone in treatment of IPF is poor, various cytotoxic agents have been studied in combination with steroids. Favorable responses have been noted in 15 to 50% of cases. Combination of immunosuppressive agents and prednisolone results in better survival when compared to prednisone alone especially in those with less severe disease i.e., FVC>70%.

Azathioprine

The combination of azathioprine and corticosteroids was associated with modest improvement and enhanced survival in some patients. In one prospective double blind randomized placebo controlled study with 27 patients, combination of azathioprine and prednisolone had a marginally significant survival advantage.

Cyclophosphamide

High dose intravenous cyclophosphamide administered every 2 to 4 weeks (dose range, 500 to 1,800 mg) has been tried in open trials of refractory IPF. Results are unimpressive. Toxicity associated with cyclophosphamide remains a major impediment to the routine use of this agent. A recent study suggested that combined corticosteroid and cyclophosphamide therapy has no impact on survival in patients with IPF.

Agents that alter collagen synthesis or fibrosis

Colchicine

Colchicine inhibits collagen formation and modulates the extracellular milieu and suppresses the release of alveolar macrophage-derived growth factor and fibronectin. Efficacy appears similar to corticosteroids and side effects with colchicine are rarely severe. Initial studies were encouraging with trend towards improved outcome. However subsequent studies have failed to demonstrate any benefit of treatment with colchicine over no treatment at all. With the present limited data, there is no evidence to suggest a beneficial role for colchicine in the treatment of IPF.

D-Pencillamine
Anecdotal evidence of responses to D-penicillamine have been noted but controlled studies have not been done.\textsuperscript{2,35}\textsuperscript{2} Moreover, it is toxic and has significant adverse effects like loss of taste, nausea, vomiting, stomatitis, nephrotoxicity.

**Interferon γ**

Interferon( IF)-γ inhibits the proliferation of lung fibroblasts in a dose-dependent manner and reduces the synthesis of protein in fibroblasts.\textsuperscript{56} In an open randomized study with 18 patients who had not responded to glucocorticoids and other immunosuppressive agents 200μg of IF γ-1b 3 times/week for 12 months along with prednisolone resulted in improvement in total lung capacity and partial pressure of oxygen.\textsuperscript{57,58} This generated great interest in IF as the magic cure for IPF but subsequent studies dampened the hopes. Honore et al reported 4 patients with advanced IPF who developed irreversible acute respiratory failure following treatment with IF γ-1b.\textsuperscript{59} Raghus G et al conducted a double-blind trial with 330 patients with IPF who did not respond to corticosteroid therapy.\textsuperscript{60} They found that IF γ-1b did not affect progression-free survival, pulmonary function, or the quality of life over 58 weeks. However a trend toward enhanced survival was seen in those who received IF (an absolute reduction in the risk of death of 7% and a relative reduction in the risk of 41%). Increased levels of Interleukin-18 in the induced sputum of patients with IPF have been found to decrease after treatment with IF γ-1b.\textsuperscript{61} A recent meta-analysis showed that IF-γ1b therapy is associated with reduced mortality.\textsuperscript{62} More studies are currently underway to exactly define the safety and efficacy of this form of therapy.

**Pirfenidone**

This is a new antifibrotic agent. In one study 42 consecutive patients with IPF and deterioration despite conventional therapy or who were unable to tolerate or unwilling to try conventional therapy were treated with oral pirfenidone.\textsuperscript{63} In these patients there was decrease in mortality and helped in reduction of prednisolone dosage. Patients with deteriorated lung functions showed stabilization. Adverse effects were minimal. A recent double-blind, randomized, placebo-controlled trial with 107 patients found that pirfenidone improved VC and prevented acute exacerbation of IPF during 9 months follow up.\textsuperscript{64} Future long-term studies are needed to clarify the overall safety and efficacy of pirfenidone in IPF.

Relaxin (increases procollagenase), halfuginone (inhibits collagen synthesis), suramin (profibrotic cytokine inhibition), and prostaglandin E\textsubscript{2} (inhibits collagen production) are the other antifibrotic agents tried.\textsuperscript{65,66}

**Antioxidant agents**

Oxygen radicals mediate epithelial injury in IPF and hence antioxidant strategies were speculated to be beneficial.\textsuperscript{67} Glutathione (an effective scavenger of toxic oxidants that suppresses lung fibroblast proliferation in response to mitogens), taurine (a natural free aminoacid), and niacin inhibit the development of experimental fibrosis in animal models.\textsuperscript{68}

Acetyl cysteine is a precursor of glutathione and can replenish pulmonary glutathione levels. In a study with 18 patients with established diagnosis of IPF treatment with 600mg N-acetyl cysteine (a glutathione precursor) three times a day for 12 weeks in addition to their latest immunosuppressive therapy resulted in significant improvement of pulmonary function tests.\textsuperscript{69} A recent double-blind, randomized, placebo-controlled multicenter study assessed the effectiveness over one year of a high oral dose of acetylcysteine (600 mg three times daily) added to standard therapy with prednisone plus azathioprine.\textsuperscript{70} This landmark trial showed that acetylcysteine slowed the deterioration of vital capacity and DL\textsubscript{CO} at 12 months without significant differences in the type or severity of adverse events.

Leucocyte adhesion molecules play an important role in the process of leukocyte retention in the lung. Antibodies to adhesion molecules have been shown to prevent collagen deposition in an animal model of lung injury.\textsuperscript{71} Inhibitors of specific fibrogenic cytokines or growth factors may help to retard the fibrotic process.\textsuperscript{72} Endothelin receptor 1 antagonist (Bosetan), anti-TNF alpha (Etanercept), PDGF receptor antagonist are in various phases of trial for IPF.\textsuperscript{73}

**Anticoagulation therapy**

Kubo H et al prospectively evaluated 56 patients with IPF who received treatment with prednisolone or prednisolone with oral anticoagulation therapy in outpatient setting and low molecular therapy when admitted in hospital.\textsuperscript{74} They found that plasma d-dimer levels were associated with mortality in patients with an acute exacerbation of IPF. The mortality associated with acute exacerbations of IPF in the anticoagulant group was significantly reduced compared to that in the non-anticoagulant group (18% vs 71%, respectively; p = 0.008). These results suggest the presence of an activated coagulation system in patients with IPF and a relationship between intravascular coagulation and mortality in acute exacerbations of IPF. Heparin may directly down-regulate the expression of various factors implicated in the progression of interstitial fibrosis such as transforming growth factor-β1, endothelin-1, and fibroblast growth factor-2.

**Lung transplantation**

The evolution of cadaveric lung transplantation has been the greatest hope for improving the quality of life in patients with IPF. Currently, it is recommended that patients who have TLC or VC values less than 60 to 65% of predicted values, resting or exercise hypoxemia PaO\textsubscript{2} values less than 55 mm Hg, and/or failed to respond...
to corticosteroids or immunosuppressive therapy be referred for lung transplantation. This procedure can prolong life and more importantly, may improve the quality of life of patients with end-stage lung disease. But factors like shortage of donor lungs, chronic graft dysfunction, and recurrent disease in the graft prevent transplantation from becoming a feasible solution in most of the patients. Chronic lung rejection that is characterized by bronchiolitis obliterans, a rapidly progressive inflammatory disorder of the small airways that cause severe airflow limitation, has become a major impediment for the long-term survival of lung allograft recipients. Single lung transplantation is the preferred surgical option. Relative contraindications to lung transplantation include unstable psychosocial profile or extrapulmonary disorders involving liver, kidney or heart and advanced age that may decrease survival. Owing to limited donor availability early enrollment is important as the waiting time for procuring a suitable donor organ may exceed 2 years, keeping in mind the limited survival of 3-5 years in these patients. In India this treatment option is still not widely available.

Living-donor lobar lung transplantation (LDLLT) is a new and evolving option for patients with end-stage lung disease. Date H et al reported a series in which LDLLT was performed in 9 patients with end stage IPF using two lower lobes donated by two healthy relatives. Early follow-up data support the option of LDLLT in patients with advanced IPF who would die soon otherwise waiting for cadaveric transplantation. There has been no donor mortality, and morbidity has been relatively low.

Other supportive measures

The primary goal in managing a patient with IPF is to restore them to the highest possible functional state. Patients should be encouraged to enroll in a pulmonary physical rehabilitation program. Daily walks or use of a stationary bicycle are useful. Those with hypoxemia at rest or during exercise should be managed by supplemental oxygen. Higher flow rates than those needed in COPD may be frequently required. Dry cough may be controlled with antitussives. Low dose opioids are effective and safe in the palliative management of dyspnea in terminally ill IPF patients. Pulmonary hypertension, cor pulmonale and right heart failure, which complicate late stages of IPF, should be appropriately managed. In randomized controlled, open-label trial a phosphodiesterase type 5-inhibitor sildenafil causes preferential pulmonary vasodilatation and improves gas exchange in patients with severe lung fibrosis and secondary pulmonary hypertension.

Conclusion

With increased awareness and better diagnostic facilities there is increase in the diagnosis of IPF in India. IPF used to be an unrelenting progressive disease with poor response to immunosuppressive treatment. But recent trials on immunomodulators, antioxidants and antifibrotic agents are promising.

References

5. Jindal SK, Malik SK, Deodhar SD, Sharma BK. Fibrosing alveolitis; A report of 61 cases seen over the past five years. Ind J Chest Dis All Sc 1979;19:174-9.


