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Interstitial Lung Diseases (ILDS)

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Abstract: *"Interstitial lung diseases'' (ILDs) refer to heterogeneous and complex group of conditions characterized by inflammation, fibrosis or both, in the interstitium of the lungs. This results in impaired gas exchange, leading to a worsening of respiratory symptoms and a decline in lung function. While the etiology of some ILDs is unclear, most cases can be traced back to factors such as genetic predispositions, environmental exposures (including allergens, toxins and air pollution), underlying autoimmune diseases, or the use of certain medications. There has been an increase in research and evidence aimed at identifying etiology understanding epidemiology improving clinical diagnosis and developing both pharmacological and nonpharmacological treatments. Aim of this review is to summarize the available data and recent advances about therapeutic strategies for ILD in the context of various CTD, such as systemic sclerosis idiopathic inflammatory myopathy and Sjogren syndrome, systemic lupus erythematosus, mixed connective tissue disease and undifferentiated connective tissue disease, and interstitial pneumonia with autoimmune features, focusing also on ongoing clinical trials*

Keywords: Interstitial lung disease, idiopathic pulmonary fibrosis, sarcoidosis, hypersensitivity, pneumonitis, nonspecific interstitial pneumonia

I. INTRODUCTION

Interstitial lung disease is an umbrella term for a number of different chronic lung conditions which involve inflammation and fibrosis of the lungs. Interstitial lung disease (ILD) is an umbrella term for ~200 different diseases that may result in inflammation and scarring of the lung tissue (Figure 1). ILD is characterized by progressive dyspnoea, cough, hypoxia, impaired lung function, diffuse bilateral infiltrates on imaging, inflammation, fibrosis, limited patient mobility and reduced quality-of-life (QOL). Most ILD cases result from an etiological factor, such as exposure to allergens, hazardous material, asbestos, drugs or an underlying autoimmune disease [(2–3)]. The development of these cases is a complex process that is influenced by a variety of factors, including the individual's genetic traits, and exposure to environmental pollutants. Idiopathic Pulmonary Fibrosis is the most aggressive form of ILD, causing progressive and permanent lung scarring. It causes a chronic and irreversible lung disease with a poor prognostic outcome with a median survival rate of 3–5 years postdiagnosis if left untreated [(7)]. Although two antifibrotic medications demonstrated a significant reduction in the rate of disease progression, it remains difficult to predict disease behavior for individual patients. The purpose of this review is to provide up-to-date information on interstitial lung disease (ILD), with a particular emphasis on definition, classifications, etiology, epidemiology, diagnosis, pharmacological, and non- pharmacological management. ILD has been classified into the following categories based on a recent publication by the American Thoracic Society Consensus Statements. The interstitial lung diseases (ILDs), also called diffuse parenchymal lung diseases, are a diverse group of pulmonary disorders classified together because of similar clinical, physiologic, or pathologic features. By the beginning of the 20th century, the gross and microscopic pathology of chronic ILD was well described. The focus turned to identifying occupational or environmental causes of ILD. Efforts from around 1950 to 1970 were aimed at understanding the radiographic, physiologic, and pathologic features of these diseases. By the 1960s, progress in categorizing ILDs was made: connective tissue diseases, drugs, occupational and environmental exposures, sarcoidosis, and in herited conditions were recognized as distinct entities. Those conditions that either remained unassociated with another process or without a

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clear cause were often lumped together under a single heading (e.g., "Hamman-Rich syndrome," "fibrosing alveolitis," or "idiopathic pulmonary fibrosis" [IPF]). Because the topic of ILD is vast, the themes chosen for discussion represent some editorial bias toward those that we have found most interesting or consider important to current practice or future studies in this field. Because of space limitations, we have not provided a complete list of references or discussed some entities where important advances have been made. Topics not discussed
include the following: familial lung fibrosis , connective tissue diseases , pulmonary vasculitis, pulmonary alveolar proteinosis, eosinophilic pneumonias, diffuse alveolar haemorrhage , pulmonary Langerhans' cell histiocytosis [8], lymphocytic interstitial pneumonia , and druginduced lung disease. Symptoms:- The most common symptoms of ILD are shortness of breath with exercise and a nonproductive cough, These symptoms are generally slowly progressive, although rapid worsening can also occur. Some people also may have a variety of other symptoms. They may include: fever, weight loss, fatigue, muscle & joint pain and abnormal chest sounds, depending upon the cause. [2]

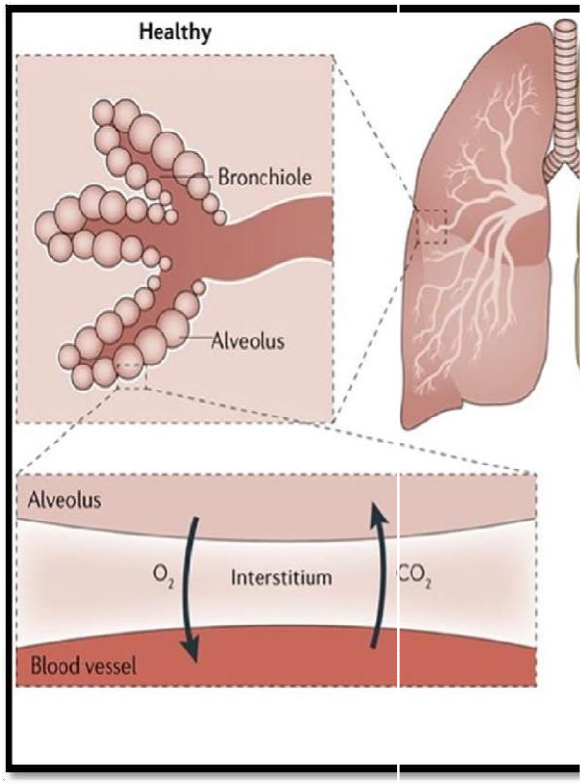

Figure 1. Illustrates normal lungs and lungs with a type of ILD (Interstitial Lung Disease)

II. LITERATURE REVIEW

Peter M George et al 2020, Within the spectrum of fibrosing interstitial lung diseases (ILDs) is a subset of patients who have inexorable progression of pulmonary fibrosis despite treatment, which is known as the progressive fibrotic phenotype. Although the concept of progressive fibrosing ILD has been applied largely to patients with idiopathic pulmonary fibrosis (IPF), there is now an increasing focus on irreversible progressive fibrosis in a proportion of patients with a range of underlying ILD diagnoses.
Evidence has emerged to support a possible role for antifibrotic therapy in these patients.

Marsha H. Antoine et al 2023, Interstitial lung disease (diffused parenchymal diseases) are a heterogeneous group of disorders characterized by fibrosis (scarring) of the lungs. These are classified on the basis of histopathological, radiologic and clinical parameters. This activity describes the evaluation and management of interstitial lung disease and reviews the role of the interprofessional team in improving care for patients with this condition.

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AIM

"A REVIEW ON INTERSTITIAL LUNG DISEASE (ILT)"

TREATMENT OBJECTIVES IN ILD

1. Provide symptom-relief
2. Slow down disease progression
3. Prevent complications
4. Improve quality of life
5. Prolong survival
6. Prevent treatment-complications
7. End-of-Life care and palliative treatment

PLAN OF WORK

I. Literature Review (Weeks 1-4)

- Conduct a comprehensive review of existing literature on intestinal lung disease
- Identify key findings, gaps in knowledge, and areas for further research

II. Study Design (Weeks 5-8)

- Develop a study design to investigate the gut-lung axis in intestinal lung disease
- Choose appropriate methods (e.g., experimental, observational, or computational

III. Data Collection (Weeks 9-16)

- Collect data on gut microbiome, cytokines, tight junctions, and intestinal permeability
- Use techniques such as 16S rRNA sequencing, ELISA, and immunohistochemistry

IV. Future Directions (Weeks 29-30)

- Identify areas for future research based on study findings and literature gaps
- Develop a plan for future studies to investigate intestinal lung disease
- This plan of work provides a general outline for investigating intestinal lung disease.

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CLASSIFICATION : (3) , (7-1O)

Idiopathic :-

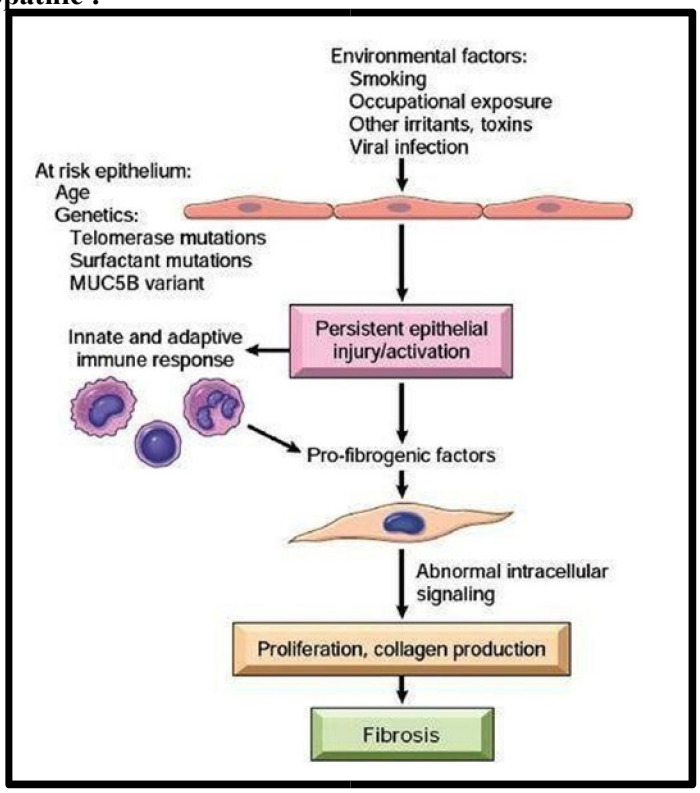

Figure 3. (5)

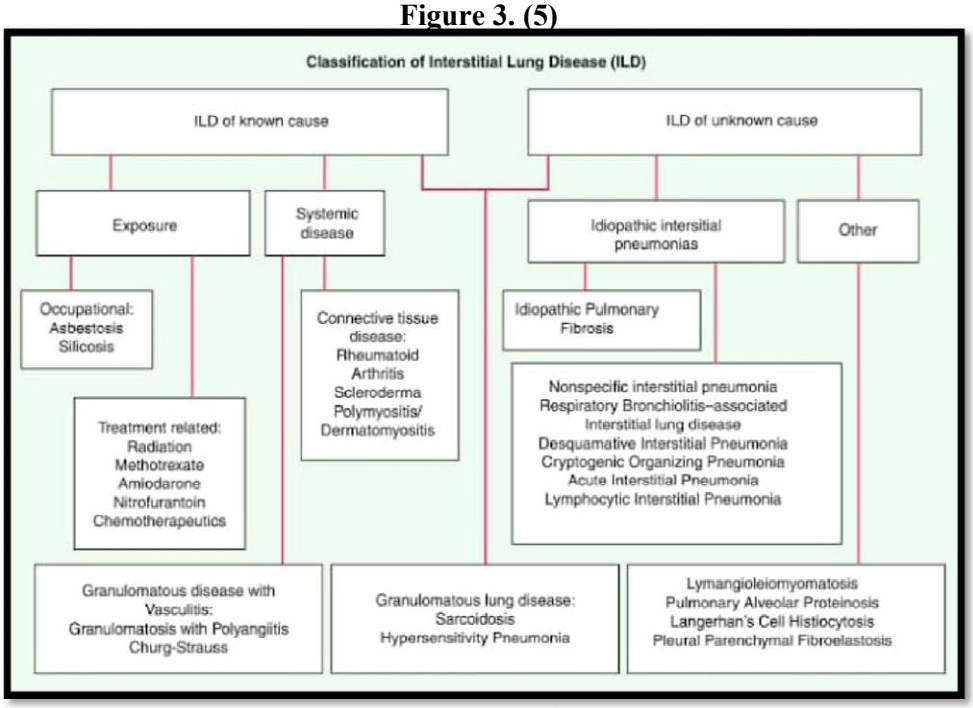

Figure 2. (4)

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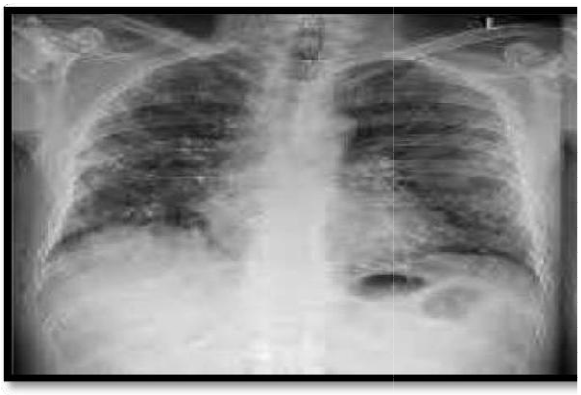
[A] Know cause - (11)
[a]Exposure – (24)
Occupational Disease – Asbestosis,
Silicosis
Treatment – Radiation Therapy
Drugs – Methotrexate, Nitrofurantoin, Chemotherapeutics, Amiodarone Bleomycin, Rituximab
[b]Systtemic disease -
Connective tissue disease: Rheumatoid Arthritis , Scleroderma
Polymyositis/
Dermatomyosins
Granulomatous disease with Vasculitis: GPA
(wegeners granulomatosinus)
RGPA [Churg Strauss syndrome]
Gmoulomatous lung disease :- Sarcoidosis Hypersensitivity Pneumonia (allergy)

[B] Unknow Causes – (11)
[a]Idiopathic interstitial pneimonias -
[1] Idiopathic Pulmonary Fibrosis :-
Chronic clinical presention
Most Commen in Male than Female



5th& 6th decade of life
Symptoms – Dry Cough, Dyspnea On Exertion (DOE)
On Examination shows – clubbing -late inspiratory Fine crackles (vectro crepitation) Dignosis :- Chest Xray (Bi Basale reticular infiltrate) (12)

HRCT [High Resalution Computed Tomography] – Use to investigation lung disease
Following find out :-
Posterior basal part involvement
Sabpleural
Honey combing
Septal thickening
Traction bronchiectasis
Radiologist changes in HRCT

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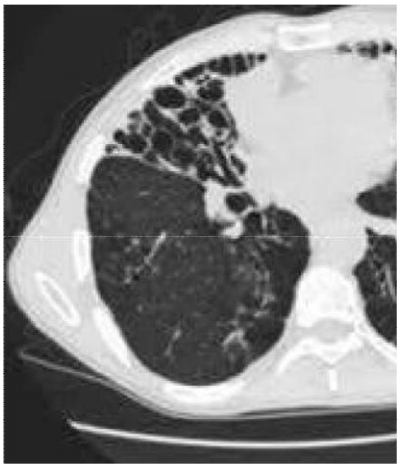
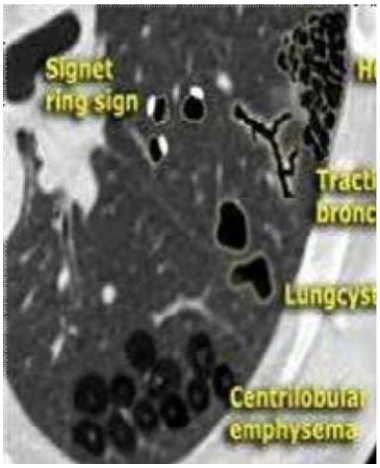
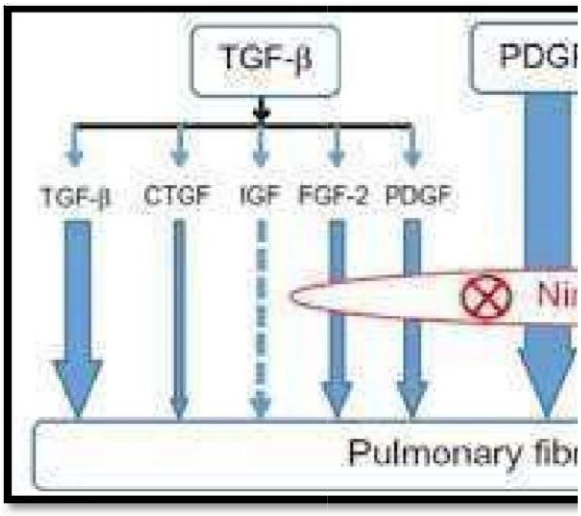




Pulmonary function tests (PFTs) :-decrease See that total lung capacity
FEV1/FUC Increase OR Decrease , decrease DLCO
These are example of Restriction Pattern all ILD.
TREATMENT :- 5 year mortality 50%
Nintedanib (tyrosine kinase inhibitor) Help to arrest to fibrosis IPF.
Lung Transplantion :- Late stage ,
Paranchyma tottaly destroyed then.
MOA :-



[2] Other Disease –
Non specific interstitial pneumonia (NSIP)
5TH decade in life
Commen in female than male
Symptoms :- Dry cough , Dyspnea On Exertion (DOE)
On Examination shows – Not common clubbing
HRCT [High Resalution Computed Tomography] –
-lung architecture relatively preserved -ground-glass opacity (GGO)

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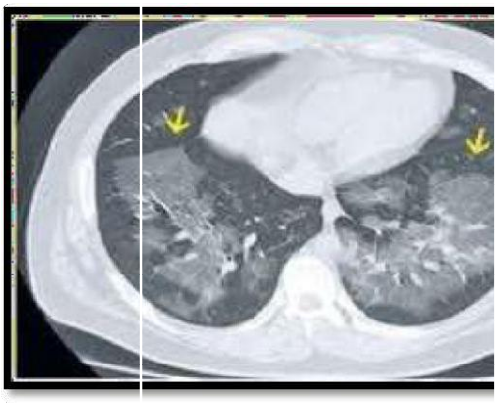
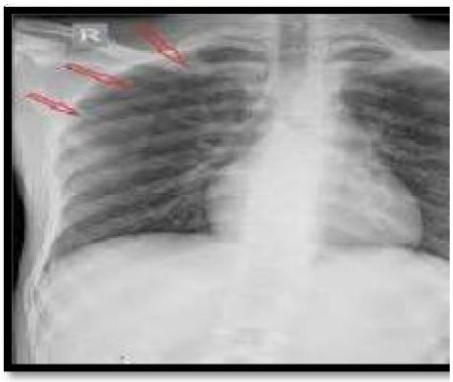


TREATMENT :- Steriods, Rituximab
Smoking associated ILD :-
- Respiratory Bronchiolitis-associated interstitial lung disease
- Desquamative Interstitial Pneumonia(DIP)

Cryptogenic Organizing Pneumonia (COP) :- Bronchiolitis obliteransorganizing pneumonia (BOOP)
TREATMENT :- Steriods
Acute Interstitial Pneumonia(AIP) :- Disorder like ARDS (Acute respiratory distress syndrome)
Lymphocytic Interstitial Pneumonia (LIP) :- Sjogrens syndrome & retro infection
(b) Other :-
Lymangioleiomyomatosis :- Seen in female age 20 – 40 years (13)
Spontaneous pneumothorax - abnormal collection of gas in the pleural space between the lungs and the chest wall.



Pulmonary Alveolar Proteinosis
Langerhan's Cell Histiocytosis
Pleural Parenchymal Fibroelastosis

Etiology:-While the pathophysiological mechanisms are not entirely understood, ILD is categorized as a restrictive lung disease that reduces lung expansion and total lung capacity (TLC) [14]. It causes scars and damage to alveoli leading to changes in lung function and decreased lung capacity and gas exchange [1]. Scarring and lung damage are linked to the creation of fibroblastic foci, where fibroblasts proliferate in response to alveolar cell injury. Triggered by transforming growth factor beta (TGF-β), this process transforms fibroblasts into myofibroblasts, which secrete collagen, leading to fibrosis [15]. Understanding the functionality of the lung and how ILD specifically changes lung structure and function helps to differentiate it from other lung diseases [2]. Although the impact of socioeconomic factors on clinical outcomes in patients with interstitial lung disease is not well characterized, they may also play a role [16]. A range of causes contribute to the development of ILD. These causes can be categorized into known and unknown

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Risk factors :- Factors that may make you more susceptible to interstitial lung diseaseinclude:[17]

Age:- Interstitial lung disease is much more likely to affect adults, although infants and children some times develop the disorder

Exposure to occupational and environmental toxins:- If you work in mining, farming or construction or for any reason are exposed to pollutants known to damage your lungs, your risk of interstitial lung disease is increased .

Gastroesophageal reflux disease:- If you have uncontrolled acid reflux or indigestion, you may be at increased risk of interstitial lung disease.

Smoking:- Some forms of interstitial lung disease are more likely to occur in people with a history of smoking, and active smoking may make the condition worse, especially if there is associated emphysema.

Radiation and chemotherapy:- Having radiation treatments to your chest or using some chemotherapy drugs makes it more likely that you'll develop lung disease.

Table 1. Interstitial lung disorders:- major categories (18)

Idiopathic interstitial pneumonia	**Sarcoidosis**
Idiopathic pulmonary fibrosis (IPF)	**Hypersensitivity pneumonitis**
Non-specific interstitial pneumonia (NSIP)	**latrogenic pneumonitis/fibrosis (druginduced ILD, radiation injury)**
Cryptogenic organizing pneumonia (COP)	**Eosinophilic ILD (e.g. eosinophilic**
Respiratory bronchiolitis interstitial lung disease (RBILD)	**Occupational lung disease**
Desquamative Interstitial pneumonia (DIP)	**Inherited disorders (e.g. familial pulmonary fibrosis, Hermansky-Pudlak syndrome)**
Acute interstitial pneumonia (DIP)	**Primary disorders pulmonary Connective tissue disease-associated (e.g. Langerhans cell histiocytosis)**

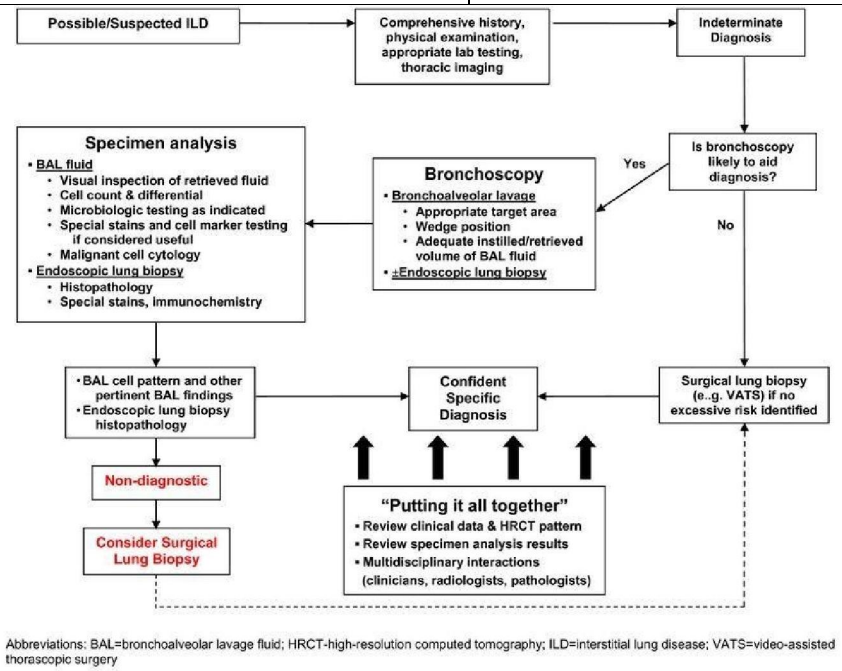

FIGURE 2. Suggested approach to the diagnosis of ILD (19) Making an accurate and confident diagnosis of a specific form of ILD (20-21)

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Patients with suspected ILD should have information from the history, physical examination, thoracic imaging and other testing (e.g. peripheral blood tests, pulmonary function testing) carefully and thoughtfully reviewed to determine whether or not additional procedures are needed and whether such procedures are likely to be helpful in reaching a confident diagnosis.

If one needs to obtain invasive testing (bronchoscopy with BAL and/or TBLBx , VATS biopsy) all findings should be reviewed (preferably in a multidisciplinary fashion) to identify the ultimate diagnosis that best fits with the combination of clinical information, imaging, and invasive testing results [22].

Risks and potential benefit of invasive testing should be carefully considered,

especially for frail, elderly patients. Some specific combinations of clinical data with imaging results and other findings can strongly support specific ILD diagnoses. iv.A younger patient with nodular changes along bronchovascular structures and bilateral hilar lymphadenopathy on HRCT imaging is highly likely to have a diagnosis of sarcoidosis and the presenceof a significant lymphocytosis on a BAL cell count determination would be highly supportive of this diagnosis.

A patient with clearly abnormal autoimmune serologies and either a NSIP or UIP pattern on HRCT imaging is likely to have CTD-ILD and may have a specific CTD diagnosis such as rheumatoid arthritis or scleroderma that can be detected and confirmed via peripheral blood serologictesting .

A patient with a significant exposure history to potential organic antigens (e.g. bird fancier or farmer) with a HRCT findings of upper lung field dominant centrilobular ground glass nodules, acute or sub acute onset of symptoms (e.g. dyspnea, myalgias) is quite likely to have acute HP, and this diagnosis is strongly supported by the findings of significant BAL lymphocytosis.

Similarly, a patient with such an exposure history plus a sub acute or chronic symptom onset and a HRCT that shows ground glass opacities or fibrotic changes with extensive mosaic attenuation due to airtrapping is likely to have chronic HP. viii.Lastly, an older patient who presents with sub acute or chronic disease onset and has bibasilar Velcro crackles on chest auscultation is highly likely to have IPF as their specific ILD diagnosis, and this diagnosis can be confidently confirmed if the HRCT shows a typical UIP pattern (peripheral and basilar predominance of fibrotic changes with reticulation and honeycomb change (Figure 1) and very little or no ground glass opacities) and alternative etiologies are lacking (e.g. presence of CTD, asbestosis, drug reaction with fibrosis)

Therapies for select types of ILD Treatment of IPF:-

The prognosis of IPF is generally poor, and the majority of patients have progressive loss of lung function and may suffer acute exacerbations with acceleration of lung function loss that often leads to death [23, 24].

Traditional therapies that were suggested to benefit patients with IPF included corticosteroids and cytotoxic drugs (e.g. azathioprine, cyclophosphamide) [25]. However, these agents have never been shown to have significant benefit in any adequately powered, prospective, randomized, placebo-controlled clinical trial. Further more, it was recently demonstrated that azathioprine, an agent that has been suggested to have efficacy for the treatment of IPF [25-27], was associated with significant harm compared to placebowhen administered to patients with IPF [28]. iii.This observation triggered the termination of the azathioprine/N-acetylcysteine (NAC)/prednisone arm of the NIH-sponsored IPF PANTHER clinical trial when it became obvious that excess mortality and other complications occurred in this cohort versus the other study arms of either NAC alone or placebo.

There are no treatment options for patients with IPF that have been approved by the U.S. Food and Drug Administration, and any pharmacologic treatment given in the US would be considered off-label.

Many new agents that target fibrogenesis have been evaluated in Phase 3 clinical trials (Table 8), but some of these agents (e.g. bosentan, macitentan, ambrisentan, interferons gamma and beta) have not shown benefit despite pre-clinical studies or phase 2 clinical trials that suggested potential efficacy.

Indeed, there can be considerable inter individual variability in genetic abnormalities

that have predisposed an individual to develop the disease, in pathophysiologic characteristics of the disease process, and in responses to specific drugs.

It should be recognized that a subset of patients that may benefit from a promising drug are very unlikely to be identified in a prospective, double blind, randomized phase 3 clinical trial in which these patients are combined with a

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much larger number of enrolled subjects for whom the drug has little or no effect, and the conclusion may be reached that the drug lacks benefit despite its potential to help a subset of patients.

Nonetheless, the results of some recently completed clinical trials suggest that pirfenidone [29,30] or nintedanib (BIBF 1120) [31]may have a significant impact on disease progression versus placebo, and pirfenidone has been licensed and is clinically available in Japan, Europe, and Canada.

Stem cell therapy, specifically the use of mesenchymal stem cells (MSC), has shown potential benefit in preclinical trials [32], and early results of a phase 1 clinical trial with adipose-derived MSC were recently reported [33].

Comorbidities can have a significant impact on disease course and quality of life for patients with IPF and other fibrotic lung diseases [34,35].

These include secondary pulmonary hypertension, coronary artery disease, venous thromboembolism, obstructive sleep apnea, coexistent emphysema, osteoporosis, diabetes mellitus, anxiety, and depression.

Coronary artery disease is highly prevalent in patients with IPF [36,37], and a significantly increased risk of developing primary lung cancer has been observed [38].

An increased risk of venous thromboembolism has also been observed [39], and sleepdisordered breathing is frequently present [40].

An IPF Net phase 3 clinical trial was performed to assess the effect of sildenafil in patients with idiopathic pulmonary fibrosis, but despite a trend toward improvement, a significant increase in 6MWT distance (the primary endpoint) was not attained [41], although a recent analysis of these data suggests that a subset of patients with right heart dysfunction may benefit from sildenafil therapy [42].

Similarly, anticoagulation, when given to disrupt the contribution of the coagulation cascade to the fibrotic process, provided no benefit and was associated with increased risk of significant adverse events [43].An abnormal degree of GER, which is present in a majority of IPF patients and has been linked to the presence of pepsin and/or bile acids in BAL fluid [44], has also been considered to be an IPFassociated comorbidity.

It has been suggested that reflux of foregut contents into the proximal esophagus via a dysfunctional lower esophageal sphincter (e.g. presence of a hiatal hernia) can predispose to (micro) aspiration, which may initiate and/or drive lung inflammation that can progress to pulmonary fibrosis in a susceptible individual, and accumulating evidence has linked GER with aspiration to IPF pathogenesis [13].

Use of medical therapy that inhibits acid production or having undergone a Nissen fundoplication has been associated with significantly improved survival for IPF patients [45], and an analysis of combined, placebo-arm cohorts from three IPF Netsponsored studies has shown less FVC decline in subjects who were using acidsuppression therapy [16].

Additionally, high pepsin levels in BAL fluid have been linked to some cases of acute exacerbation of IPF [15], and a significantly reduced incidence of acute exacerbations of IPF was observed for subjects enrolled in combined placebo cohorts from the IPF Net phase 3 clinical trials if they were taking antireflux medication [46].

Lung transplantation

Lung transplantation remains the best option for patients with advanced IPF, with 66% of transplant recipients living for more than 3 years after the surgery and 53% surviving for over 5 years (47). The criteria for lung transplantation in ILD patients encompass UIP or NSIP evidence, a 10% FVC or 15% DLCO decrease over 6 months, an oxygen saturation <88%, a 6-min walk of fewer than 250 m, a 50-m decrease in 6MWD within 6 months, or pulmonary hypertension. The evaluation and waiting period for lung transplantation can last for years, making it a challenging and difficult time for patients and their families (49). While post-transplantation survival rates in IPF patients are around 66% beyond 3 years and 53% beyond 5 years (50), lung transplantation can be a life-saving procedure that significantly improves life expectancy and enhances quality of life for some patients with IPF (51). When lung transplantation is being considered, it is crucial to review the patient at a specialist center. Transplant medicine experts can thoroughly assess the patient's suitability for this major surgical procedure, help prepare the patient, and manage their post-transplant care. The shared care approach is continued in these cases to ensure the best possible patient outcome(51).

The field of lung transplantation has continually evolved, increasing both in application and success. In 1963, Dr James Hardy and colleagues at the University of Mississippi performed the first lung transplant. Despite the recipient having chronic obstructive pulmonary disease (COPD) and being a suboptimal candidate due to advanced lung cancer and

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renal insufficiency, this pioneering procedure paved the way for future developments.[4] The first successful combined heart and lung transplant followed in 1981, marking another significant milestone in the history of transplant surgery.
Over the past 6 decades, lung transplantation has seen remarkable advancements. This growth has been particularly notable over the last 10 years, driven by donor utilization and procurement innovations. The number of lung transplants has increased, and outcomes have improved due to advancements in medical and surgical management, as well as microbiological and immunological care.[5]

FUTURE SCOPE

Interstitial lung disease is a group of disease that causes progressive scarring of lung tissues. People with interstitial lung disease may experience chronic or dry cough, fatigue or inability to exercise, shortness of breath or weight loss, etc. And the treatment of patients with interstitial lung disease depends on severity and underlying cause but often includes steroids, oxygen therapy, and pulmonary rehabilitation **On This semester I just review on this disease further I study the drug and try to make effective formulation for this disease,** Identify areas for future research based on study findings and literature gaps, Develop a plan for future studies to investigate intestinal lung disease

SUMMARY

This article has reviewed the pathophysiology of ILD and its cardiorespiratory consequences at rest and during exercise. An understanding of the cardiorespiratory consequences of ILD at rest and during rigorous exercise will help to characterize the natural history of the disease, to differentiate cardiac and respiratory limitations to exercise, and to assess the functional consequences of the disease. In addition, a better understanding of the pathophysiology and cardiorespiratory consequences of ILD will enhance methods of clinical exercise testing, which are essential in assessing functional work capacity, dyspnea, disability, and occupational capability and in prescribing exercise programs. Descriptions of long-term exercise responses for patients with ILD are lacking in the literature. These descriptions are needed to characterize training responses for different categories of patients with ILD of various severities, to determine what intensity of training is apt to be beneficial, and to define the criteria for exercise prescription and there by optimize the effects of training

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