DR. PRASHANT PRAKASH

M.D. (Medicine), M.D. (Chest)
M.A.M.S., F.I.C.P., F.I.A.C.M.
Professor & Head,
Super Specialty Department of Pulmonary Medicine
S.N. Medical College, Agra

Past Affiliation	:	**Former Sr. Resident, Dr. R.M.L. Hospital, New Delhi** **Former Visiting Fellow, Dept. of Pulmonary Medicine, PGIMER, Chandhigarh**
Membership	:	**Indian Chest Society, Indian Association of Bronchology**
Fellowship	:	**Fellow of Indian College of Physicians (FICP),** **Fellow Indian Association of Clinical Medicine (FIACM)**
Organising Chairman/ Secretary of conference	:	**Joint Organizing Secretary- National Conference of Indian Association of Clinical Medicine (2014),** **Treasurer – National Conference of Association of Physicians of India (2020).**
Conference Presentations	:	**25**
Publications	:	**32**
Book Chapters	:	**22**
Editor/Reviewer	:	**Asstt. Editor in "Review in Geriatrics – 2015"**

Awards :

- **Received Short-term Visiting Fellowship by ICMR in 2010.**
- **Received Short-term Visiting Fellowship by ICMR in 2013.**
- **Received Pawan Kumari Jain Oration Award in Nov. 2017 Awarded by Delhi Chapter of Association of Physicians of India.**
- **Received GB Jain Oration Award in Nov. 2019 Awarded by the Indian Association of Clinical Medicine.**
- **Received Prof. Rathnavelu Subramaniam Oration Award in 2021 Awarded by Association of Physicians of India.**
- **Received 8th IACMCON 2010 Oration Awarded by the Indian Association of Clinical Medicine, West Bengal Chapter in Sept. 2022.**

Areas of Interest :

- **Interventional Pulmonology**
- **Lung Physiology & Lung function testing.**

RECENT TRENDS IN THE MANAGEMENT OF ILD

DR. PRASHANT PRAKASH

M.D. (Medicine), M.D. (Chest)

M.A.M.S., F.I.C.P., F.I.A.C.M.

Professor & Head,

Super Specialty Department of Pulmonary Medicine

S.N. Medical College, Agra

INTRODUCTION

- Interstitial Lung Diseases (ILDs) are a heterogenous group of more than 150 disease entities that differ significantly with respect to prevention, therapy and prognosis.
- The current classification scheme of ILDs is shown in Fig 1.

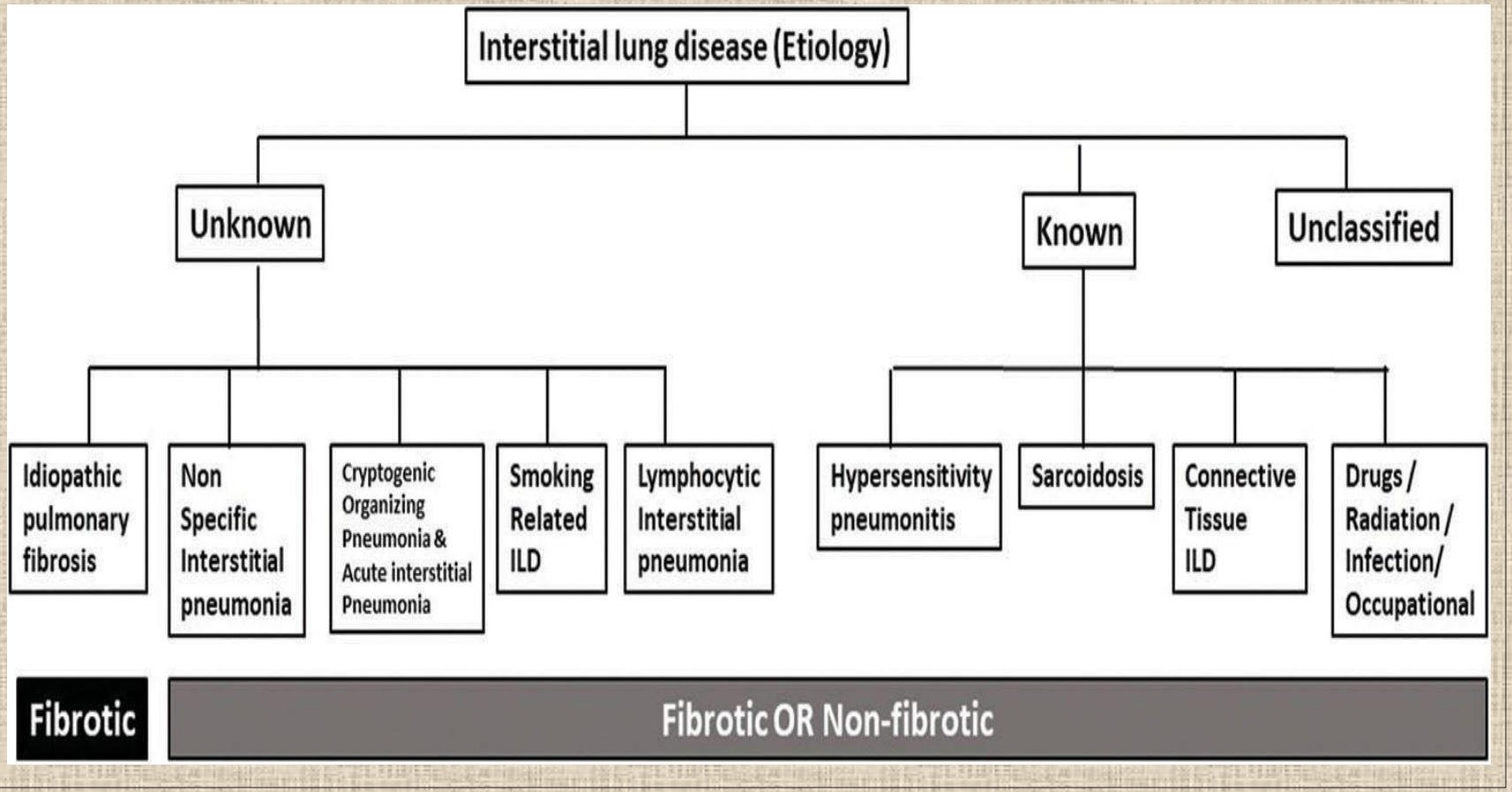

Figure 1: Classification of interstitial lung disease on the basis of known or unknown etiology

CLASSIFICATION OF ILDs

- The Multi-disciplinary classification of the Idiopathic Interstitital Pneumonias (IIPs) has been updated by an official American Thoracic Society (ATS)/European Respiratory Society (ERS) Statement in 2013 by which **IIPs have been classified into major IIPs, rare IIPs and unclassifiable cases.**
- **The major IIPs are grouped into chronic fibrosing, smoking- related and acute/subacute IIPs (Table-1).**

TABLE-1 CATEGORIZATION OF MAJOR IDIOPATHIC INTERSTITIAL PNEUMONIAS

Category	Clinical-Radiological-Pathologic Diagnosis	Associated Radiologic and/or Pathologic-Morphologic Patterns
Chronic fibrosing IP	Idiopathic pulmonary fibrosis Idiopathic nonspecific interstitial pneumonia	Usual interstitial pneumonia Nonspecific interstitial pneumonia
Smoking related IP*	Respiratory bronchiolitis-interstitial lung disease Desquamative interstitial pneumonia	Respiratory bronchiolitis Desquamative interstitial pneumonia
Acute/subacute IP	Cryptogenic organizing pneumonia Acute interstitial pneumonia	Organizing pneumonia Diffuse alveolar damage

Definition of abbreviation : IP = intersititial pneumonia.

*** Desquamative interstitial pneumonia can occasionally occur in nonsmokers.**

- The diagnostic strategy in a patient with ILD is based on considerations regarding the
  - dynamic time course (**acute, subacute, chronic**),
  - cause (**known or unknown**),
  - context of the disease at presentation (**presence of extrapulmonary/systemic disease manifestations**).

- The management of ILDs can be broadly divided into
  - General management of common comorbidities,
  - Management of symptoms such as cough, dyspnea
  - Pulmonary hypertension
  - Specific therapies.

MANAGEMENT OF COMORBIDITIES IN ILD

- Most common comorbidities encountered in ILD are
  - **Gastroesophageal Reflux Disease (GERD)**
  - **Pulmonary Hypertension (PH)**
  - **Lung Cancer**
  - **Obstructive Sleep Apnea (OSA)**
  - **Venous Thromboembolism (VTE)**

MANAGEMENT OF COUGH IN ILD

- Evolving therapies for cough in ILD include:
  - **Prednisolone:** A short trial of oral prednisolone in distressing cough associated with IPF is an appropriate consideration.
  - **Gabapentin:** Gabapentin may be tried for intractable cough.
  - **Thalidomide:** Thalidomide may be tried for intractable/ distressing cough associated with IPF.

MANAGEMENT OF DYSPNEA IN ILD

- Evolving therapies for dyspnea in ILD include:
  - **Pulmonary rehabilitation:** Pulmonary rehabilitation is advocated in dyspneic patients with ILD. **Pulmonary rehabilitation has no beneficial effects on survival. The effects last as long as program is continued.**
  - **Supplemental oxygen:** Supplemental oxygen **is recommended in patients with documented resting hypoxemia and/or exercise-induced hypoxemia, and desaturation while sleeping. Long-term oxygen therapy (LTOT) is indicated for patients with ILD who have persistent resting hypoxemia**
  - **Nebulized opioid therapy:** Nebulized opioid therapy **is not beneficial to relieve dyspnea in all ILD patients** and may be used only for patients receiving comfort and palliative care.

VACCINATION IN INTERSTITIAL LUNG DISEASES

- According to a consensus statement of Indian Chest Society (ICS) & National College of Chest Physicians (NCCP) on management of ILDs, **vaccination (influenza and pneumococci) are advocated for all patients with ILD.**

MANAGEMENT OF PULMONARY HYPTERTENSION IN ILD

- The consensus statement of ICS & NCCP on management of ILDs endorses the guidelines for management of chronic Pulmonary hypertension (PH) **specific therapy for patients with PH and the treatment of underlying lung disease as the mainstay of therapy and supplemental oxygen in cases of hypoxemia.**
- **Ambrisentan is contraindicated in patients with PH related to IPF. The therapeutic benefits of other PH specific therapy in ILD related PH remains unknown.**

ROLE OF NON-INVASIVE VENTILATION (NIV) AND MECHANICAL VENTILATION (MV) IN ILD

- Consideration of NIV is advocated as early as possible **in patients who require high flow supplemental oxygen at rest, especially in patients manifesting acute exacerbation (AE) ILD with respiratory failure** as it has been associated with better short-term outcomes.
- The consideration of MV in patients with AE ILD with respiratory failure should be made only after proper counseling.

ROLE OF LUNG TRANSPLANTATION IN ILD

- Lung transplantation is **the only treatment with clearly proven survival benefit in advanced ILD, especially IPF,** and should be considered in carefully selected patients.
- Posttransplant survival is variable in lung transplant programs. **While the 5-year survival in most experienced lung transplant programs is about 70%, less experienced programs have lesser survival rates.**

MANAGEMENT OF IDIOPATHIC INTERSTITIAL PNEUMONIAS (IIPs)

- **The diagnosis of IIPs requires exclusion of known causes of interstitial lung disease such as drug or inhalational exposure and CTD-ILD.**
- The major IIPs are grouped into chronic fibrosing, smoking- related and acute/ subacute IIPs (Table-1).
- **The doses, side effect and management of side effects of commonly used drugs in ILDs is outlined in Table-2.**

TABLE-2:
DOSES, SIDE EFFECTS AND MANAGEMENT OF SIDE EFFECTS OF COMMONLY USED DRUGS IN THE TREATMENT OF INTERSTITIAL LUNG DISEASE

Drug	Dose	Side effect	How to manage side effect
Pirfenidone	1800-2400 mg/day in divided doses, 200 mg 3 tablets thrice a day	**Nausea, vomiting** **Photosensitivity, rash** **Elevated liver enzymes**	Reduce or stop the drug, PPI Cover exposed skin, sunscreen Monitor LFT monthly for 6 months, thereafter 3 monthly
Nintedanib	150 mg twice a day	**Diarrhea** **Nausea, vomiting** **Elevated liver enzymes**	Reduce or stop drug, imodium Reduce or stop drug, PPI Monitor LFT monthly for 3 months, thereafter 3 monthly
N- acetyl cysteine	600 mg thrice a day	**Nausea, vomiting, diarrhea**	Self-limiting, reduce or stop the drug
Prednisolone	1 mg/kg BW tapered to 0.25 mg/kg BW*	**Hyperglycemia** **Hypertension** **Swelling face** **Osteoporosis** **Reduced immunity** **Weight gain**	Bring to lowest dose possible, sugar avoidance, oral hypoglycemics (if patient develops diabetes mellitus), exercise Salt avoidance, exercise Salt avoidance Calcium, bisphosphonates, exercise Bring to lowest dose possible, PCP prophylaxis, Influenza vaccine (once steroid dose is <7.5 mg/day), avoid crowded places Dietary modification, exercise, bring to lowest dose

TABLE-2:
DOSES, SIDE EFFECTS AND MANAGEMENT OF SIDE EFFECTS OF COMMONLY USED DRUGS IN THE TREATMENT OF INTERSTITIAL LUNG DISEASE contd…

Drug	Dose	Side effect	How to manage side effect
Azathioprine	50 mg twice a day	**Cytopenias**	Reduce or stop drug, Monitor CBC monthly till 6 months thereafter 3 monthly
		Infections	Reduce dose, avoid crowded places, PCP prophylaxis
		Nausea, vomiting	Reduce or stop the drug, PPI
Methotrexate**	10 mg/week may be increased to 20 mg/week and brought down to 5 mg/week	**Hemtological**	Reduce or stop the dose
		Neutropenia, thrombocytopenias	Folic acid is added once a week
		Gastrointestinal	Give with food
			PPI
			Split the dose
			Stop or reduce dose
		Hepatic	Monitor LFT
		Pulmonary toxicity	Stop drug
		Teratogenicity	Stop the drug
			Birth control till 6 months after stopping drug
MMF	1.5-3mg/day	**Leucopenia**	Reduce or stop drug
		Diarrhea	Reduce dose, hydration
Cyclophosphamide	500-1000 mg IV per 4 week or 1-2 mg/day orally	**Hemorrhagic cystitis**	Mesna
			Hydration
			Less with intermittent dosing
		Neutropenia	Monitoring CBC
		Infertility	Leuprorelin

TABLE-2:
DOSES, SIDE EFFECTS AND MANAGEMENT OF SIDE EFFECTS OF COMMONLY USED DRUGS IN THE TREATMENT OF INTERSTITIAL LUNG DISEASE contd…

Drug	Dose	Side effect	How to manage side effect
Infliximab	3 mg/kg at 0, 2, 6, 12, 18, 24 weeks	**Allergic reactions** **Infections**	Slow infusion, antiallergics, paracetamol corticosteroid loading Rule out pulmonary tuberculosis prior to initiation, PCP prophylaxis, monitoring
Rituximab	1000 mg IV repeat at 2 weeks	**Allergic reactions** **Cytopenias** **Infections**	Stop infusion Antiallergic medications, paracetamol corticosteroid loading CBC monitoring PCP prophylaxis, monitoring

CHRONIC FIBROSING IIPs

(I) IDIOPATHIC PULMONARY FIBROSIS (IPF)

- Idiopathic Pulmonary Fibrosis (IPF) is defined as a specific form of chronic, progressive fibrosing interstitial pneumonia of unknown cause,
  - **occurring primarily in older adults (6th to 7th decade),**
  - **limited to the lungs, and**
  - **associated with the histopathologic and/or radiologic pattern of Usual Interstitial Pneumonia (UIP) defined.**
- The definition of IPF requires the exclusion of other forms of interstitial pneumonia including other idiopathic interstitial pneumonias and ILD associated with environmental exposure, medication, or systemic disease.

CHRONIC FIBROSING IIPs contd…

(I)IDIOPATHIC PULMONARY FIBROSIS (IPF) contd…

- **IPF is the most common entity in the spectrum of IIPs accounting for 50-60% of cases.**
- **The prognosis is usually worse compared with other IIPs, with a median survival time of 2-4 years.**
- **The diagnosis of IPF requires (1) exclusion of other known causes of ILD, (2) the presence of a UIP pattern on HRCT in patients not subjected to surgical lung biopsy (SLB), and (3) specific combinations of HRCT and SLB patterns in patients subjected to SLB.**

CHRONIC FIBROSING IIPs contd…

(I)IDIOPATHIC PULMONARY FIBROSIS (IPF) contd…

- The consensus statement of ICS & NCCP on management of interstitial lung diseases outline the following management protocols in IPF.
- **All symptomatic IPF patients with FVC of >50% predicted should be initiated on Pirfenidone.**
- **The patients on Pirfenidone developing ≥10% subsequent decline in FVC in any 6–12 months period should be given a choice of continuation of therapy or switch to an alternative therapy depending on case to case basis.**

CHRONIC FIBROSING IIPs contd…

(I)IDIOPATHIC PULMONARY FIBROSIS (IPF) contd…

- **All symptomatic IPF patients with FVC of >50% predicted should be initiated on Nintedanib.**
- **The patients on Nintedanib developing ≥10% subsequent decline in FVC in any 6–12 months period should be given a choice of continuation of therapy or switch to an alternative therapy on case-to-case basis.**
- **Either Pirfenidone or Nintedanib may be chosen for patients with IPF based on patient preference and tolerability.**

CHRONIC FIBROSING IIPs contd…

(I)IDIOPATHIC PULMONARY FIBROSIS (IPF) contd…

- **Duration of treatment is life-long.**
- **N-acetylcysteine (NAC) is currently not recommended for routine treatment of IPF and may be considered in certain subgroups on a case to case basis.**
- **More evidence is needed to recommend use of Pirfenidone in combination with Nintedanib or NAC and the dose of individual drugs to be used in such therapy in patients with IPF.**

CHRONIC FIBROSING IIPs contd…

(I)IDIOPATHIC PULMONARY FIBROSIS (IPF) contd…

- **Pirfenidone has been associated with slowing of the absolute decline in FVC, increases progression-free survival, and reduces mortality.**
- **Nintedanib has been associated with reduction in decline in predicted FVC, acute exacerbation and risk of all cause, and respiratory related and on treatment mortality.**

CHRONIC FIBROSING IIPs

(II) IDIOPATHIC NONSPECIFIC INTERSTITIAL PNEUMONIA (NSIP)

- The typical clinical presentation in NSIP was:
  - **breathlessness and cough of 6-7 months duration,**
  - **predominantly in women,**
  - **in never-smokers, and**
  - **in the 6th decade of life.**
- **Importantly, the NSIP pattern occurs not only as an idiopathic condition, but also in a variety of settings including CTD-ILD, HP, and drug toxicity, and in some patients with familial pulmonary fibrosis.**
- MDD is especially important to establish the diagnosis of idiopathic NSIP.

CHRONIC FIBROSING IIPs contd…

(II) IDIOPATHIC NONSPECIFIC INTERSTITIAL PNEUMONIA (NSIP) contd…

- The consensus statement of ICS & NCCP on management of interstitial lung diseases outline the following management protocols in NSIP[1].
- **Oral corticosteroids are suggested for the treatment of NSIP.**
- **Immunosuppressants such as cyclophosphamide may be used as add-on therapy in patients not responsive to steroids.**

CHRONIC FIBROSING IIPs contd…

(II)IDIOPATHIC NONSPECIFIC INTERSTITIAL PNEUMONIA (NSIP) contd…

- **Corticosteroids improves symptom score and lung function with response more pronounced in**
  - **cellular NSIP,**
  - **concomitant consolidation,**
  - **seronegative ANA**
  - **shorter disease duration.**
- In asymptomatic or mildly symptomatic cases close observation is often done, as the risk of treatment outweighs the benefits.
- **In symptomatic patients, oral corticosteroids are the mainstay of therapy.**

SMOKING-RELATED IIPs

(I) RESPIRATORY BRONCHIOLITIS–INTERSTITIAL LUNG DISEASE (RB-ILD)

- Histologic RB is always present in current smokers and can be viewed as a physiological response to smoking, which in a few individuals becomes extensive enough to result in an interstitial lung disease (RB-ILD).
- **The only proven therapy for RB-ILD is smoking cessation.**
- **Only those who continue to be symptomatic in spite of smoking cessation should be considered for corticosteroid therapy.**

SMOKING-RELATED IIPs contd…

(II) DESQUAMATIVE INTERSTITIAL PNEUMONIA (DIP)

- DIP has been recognized in nonsmokers, perhaps reflecting extension of childhood DIP into adult life (with the latter often due to surfactant protein [SP] gene mutations).
- **Ten-year survival remains approximately 70%, with resistance to treatment in a significant minority.**
- **There is clinical response to smoking cessation alone or in combination with corticosteroid therapy, as well as some possible spontaneous resolution.**

ACUTE OR SUBACUTE IIPS contd…

(I) CRYPTOGENIC ORGANIZING PNEUMONIA (COP)

- Patients with COP typically present with a subacute illness of relatively short duration (median, less than 3 months) with variable degrees of cough and dyspnea.
- **The majority of patients recover completely with oral corticosteroids, but relapse is common.**
- **Cytotoxic drugs and steroid sparing agents are used in addition to corticosteroid where there is slow response to steroids alone.**

ACUTE OR SUBACUTE IIPS contd…

(I)CRYPTOGENIC ORGANIZING PNEUMONIA (COP) contd…

- A small group (10-15%) may progress to pulmonary fibrosis and one third of patient can relapse.
- **Factors that may predict poor outcome include**
  - **the lack of lymphocytosis on BAL**
  - **predominantly interstitial pattern of imaging**
  - **histological features of scarring and remodeling of lung parenchyma.**

ACUTE OR SUBACUTE IIPS contd…

(II) ACUTE INTERSTITIAL PNEUMONIA (AIP)

- **AIP is a distinct IIP characterized by rapidly progressive hypoxemia, mortality of 50% or more, and no proven treatment.**
- Survivors usually have a good long-term prognosis (similar to adult respiratory distress syndrome [ARDS] survivors) but some experience recurrences or chronic, progressive interstitial lung disease.
- **AIP is idiopathic and should be distinguished from ARDS with known cause.**

ACUTE OR SUBACUTE IIPS contd…

(II) ACUTE INTERSTITIAL PNEUMONIA (AIP) contd…

- **Since there is no proven treatment, supportive care with oxygen supplementation and mechanical ventilation is indicated.**
- Early corticosteroid therapy and combination therapy with intravenous cyclosphosphamide and vincristine have been reported.

MANAGEMENT OF HYPERSENSITIVITY PNEUMONITIS (HP)

- **The more common form of HP are farmers lung, budgerigar (parakeet) keeper's lung (keeping of domestic birds), and pigeon breeder's lung in Europe.**
- **HP appears to be more common in men and during middle age,** although it is not known whether this reflects host predisposition or merely frequency of exposure to relevant antigens.
- **Inhalation of organic particles is the commonest cause of HP, but it can occur following inhalation of inorganic chemicals and, occasionally, ingestion of drugs**.

MANAGEMENT OF HYPERSENSITIVITY PNEUMONITIS (HP) contd…

- **Traditionally, HP is categorised as acute, subacute or chronic.**
- Acute HP presents 4–8 hrs after often heavy exposure with fever, malaise, cough, dyspnoea and chest tightness.
- The symptoms remit over 24–48 hrs in the absence of further exposure.

MANAGEMENT OF HYPERSENSITIVITY PNEUMONITIS (HP) contd…

- **The subacute and chronic forms usually occur with ongoing lower-level exposure.**
- Dyspnoea, cough and fatigue develop insidiously and weight loss is a common feature.
- **Patients with chronic HP typically do not give a history of acute symptoms but present with diffuse pulmonary fibrosis which must be distinguished from other conditions including IPF and fibrotic NSIP.**

MANAGEMENT OF HYPERSENSITIVITY PNEUMONITIS (HP) contd…

- The consensus statement of ICS & NCCP on management of interstitial lung diseases outline the following management protocols in HP:
  - **Oral corticosteroids**
  - **Azathioprine**
  - **Mycophenolate (MMF).**

MANAGEMENT OF HYPERSENSITIVITY PNEUMONITIS (HP) contd…

- **Oral corticosteroids for 4–12 weeks are an appropriate treatment option for patients with acute/subacute HP** with monitoring of lung function parameters and side effects.
- Prolonged use of oral steroids, azathioprine, and MMF should be based on clinical response and tolerance.
- **In patients with acute HP, there is improvement in symptoms and lung functions with oral corticosteroid. However, this benefit is not sustained over long term.**

MANAGEMENT OF HYPERSENSITIVITY PNEUMONITIS (HP) contd…

- **There is a lack of evidence pertaining to duration and dose of corticosteroids for long-term therapy in HP.**
- Prolonged use of corticosteroids and other immunosuppressants should be prescribed after weighing benefit of individual response and side effects associated with the drugs.

MANAGEMENT OF HYPERSENSITIVITY PNEUMONITIS (HP) contd…

- **Inability to identify the inciting antigen is associated with worse survival.**
- Thereby, every effort should be made to identify the inciting antigen.

MANAGEMENT OF CONNECTIVE TISSUE DISEASES ASSOCIATED ILD (CTD-Associated ILD)

- CTD may be associated with pleura-parenchymal disease or sometimes interstitial tissue involvement of the lung.
- The ILD in CTD may either be an initial manifestation or may manifest after a long-standing autoimmune illness.

MANAGEMENT OF CONNECTIVE TISSUE DISEASES ASSOCIATED ILD (CTD-Associated ILD) contd…

- **The prevalence of CTD-ILD is highest in systemic sclerosis (SSc) and the least in systemic lupus erythematosus (SLE).**
- **An acute presentation of ILD is more common in SLE and polymyositis/dermatomyositis (PM/DM).**
- The management protocols of individual connective tissue lung diseases associated with ILD as per the consensus statement of ICS and NCCP are as follows

MANAGEMENT OF CONNECTIVE TISSUE DISEASES ASSOCIATED ILD (CTD-Associated ILD) contd…

(I)RHEUMATOID ARTHRITIS ASSOCIATED INTERSTITIAL LUNG DISEASE (RA-ILD): contd…

- **Corticosteroids may be used in the treatment of RA-ILD.** They are anti-inflammatory drugs which help suppress disease activity leading to improvement in symptoms and lung functions.
- **Cyclophosphamide, mycophenolate mofetil, and rituximab may be used in the treatment of RA-ILD in case of no response to corticosteroids.**

MANAGEMENT OF CONNECTIVE TISSUE DISEASES ASSOCIATED ILD (CTD-Associated ILD) contd…

(I)RHEUMATOID ARTHRITIS ASSOCIATED INTERSTITIAL LUNG DISEASE (RA-ILD): contd…

- Role of other drugs in RA patients who develop ILD:
  - **Methotrexate should be discontinued in patients of RA diagnosed with ILD.**
  - **Leflunomide can be continued in patients diagnosed with RA-ILD.**
  - **Other antitumor necrosis factor (TNF) agents may be used cautiously.**

MANAGEMENT OF CONNECTIVE TISSUE DISEASES ASSOCIATED ILD (CTD-Associated ILD) contd…

(II)SCLERODERMA ASSOCIATED INTERSTITIAL LUNG DISEASE (SSC-ILD):

- **Low-dose steroids may be continued in the treatment of SSC-ILD.**
- **High-dose steroids should be avoided in scleroderma as it is associated with risk of renal crisis.**
- **Immunosuppression and secondary infection are a dreaded complication of these drugs.**
- Effect lasts till the drugs are taken and there is no long lasting benefits.

MANAGEMENT OF CONNECTIVE TISSUE DISEASES ASSOCIATED ILD (CTD-Associated ILD) contd…

(II)SCLERODERMA ASSOCIATED INTERSTITIAL LUNG DISEASE (SSC-ILD) contd… :

- **Treatment in SSC-ILD may be initiated in cases with progressive disease with either cyclophosphamide or mycophenolate mofetil.**
- Mycophenolate mofetil has better tolerability and lesser side effects, though more expensive.
- **Azathioprine is an alternate drug for maintenance therapy in SSC-ILD.**

MANAGEMENT OF CONNECTIVE TISSUE DISEASES ASSOCIATED ILD (CTD-Associated ILD) contd…

(II)SCLERODERMA ASSOCIATED INTERSTITIAL LUNG DISEASE (SSC-ILD) contd… :

- **Rituximab could be considered in patients with refractory scleroderma.** It should be administered at tertiary care level after evaluating for the pros and cons of treatment.
- **Nintedanib, an anti-fibrotic drug has been shown to reduce the annual decline in lung functions associated with SSC-ILD.** However, there was no advantage on the other manifestations of the SSC.

MANAGEMENT OF CONNECTIVE TISSUE DISEASES ASSOCIATED ILD (CTD-Associated ILD) contd…

(III) SJOGREN'S SYNDROME:

- The treatment of Sjogren's syndrome and ILD remains empiric, since no controlled studies have been performed.
- **In symptomatic patients with Sjogren's syndrome and ILD and functional deterioration, the standard treatment is oral prednisone in the dosage of 1 mg/kg/day with subsequent tapering, continued for at least 6 months.**
- The patients who do not improve with systemic glucocorticoids should received azathioprine, cyclophosphamide and cyclosporine.

MANAGEMENT OF CONNECTIVE TISSUE DISEASES ASSOCIATED ILD (CTD-Associated ILD) contd…

(IV) DERMATOMYOSITIS AND POLYMYOSITIS (DM & PM):

- **Treatment of ILD in patients with myositis almost always requires glucocorticoids, regardless of whether pulmonary or muscle disease is the predominant feature of the patient's illness.**
- No controlled trials of glucocorticoids have been undertaken; about 50% myositis patients with ILD respond to corticosteroids.

MANAGEMENT OF CONNECTIVE TISSUE DISEASES ASSOCIATED ILD (CTD-Associated ILD) contd…

(IV)DERMATOMYOSITIS AND POLYMYOSITIS (DM & PM) contd…:

- **Since ILD is a fatal disease, a second agent is added from the beginning of therapy in patients with DM or PM complicated by ILD**.
- **Options for this second agent in patients with mild ILD include azathioprine and methotrexate; azathioprine is preferred because of it's favourable side effect profile.**
- In patients with moderate to severe ILD cyclophosphamide is given.

MANAGEMENT OF CONNECTIVE TISSUE DISEASES ASSOCIATED ILD (CTD-Associated ILD) contd…

(IV)DERMATOMYOSITIS AND POLYMYOSITIS (DM & PM) contd…:

- **In patients with inexorable disease rituximab and intravenous immunoglobulin (IVIG) are the most commonly used treatment options.**
- The commonly employed drugs for CTD related pulmonary diseases are listed in Table-3.

TABLE-3
COMMON DRUGS EMPLOYED FOR TREATMENT OF CONNECTIVE TISSUE DISEASE-RELATED LUNG DISEASES

- Corticosteroids, cyclophosphamide, methotrexate and azathioprine, and biological agents are the main drugs used for CTD – related lung diseases
- Rheumatoid arthritis-related interstitial lung disease responds to corticosteroids and methotrexate
- Corticosteroids are not useful in scleroderma-related lung disease; cyclophosphamide is the drug of choice
- Acute lupus pneumonitis responds to corticosteroids
- Collagen vascular disease-related OP/CP has good prognosis and responds to corticosteroids
- Polymyositis-dermatomyositis-related lung disease is fatal; corticosteroids and cyclophosphamide should be used

MANAGEMENT OF SARCOIDOSIS

- The consensus statement of ICS & NCCP on management of interstitial lung diseases outline the following drugs for management in sarcoidosis:
- **Corticosteroids**:
  - **Corticosteroids improve dyspnea score, lung functions and radiology in sarcoidosis.**
  - **Corticosteroids have no significant benefits for asymptomatic stage 0/1 sarcoidosis.**

MANAGEMENT OF SARCOIDOSIS contd…

Corticosteroids contd…

- Moreover, the effects last as long as they are used.
- There is no long-term benefits in term of lung functions.
- Immunosuppression is another concerning feature not only for corticosteroids but for other immunosuppressants as well.

MANAGEMENT OF SARCOIDOSIS contd…

- **Methotrexate:**
- **Azathioprine:**
- **Leflunomide:**
- **Hydroxychloroquine:**

may be tried in patients not responding to oral steroids or with associated steroid toxicity

- **Infliximab:** may be tried after carefully weighing the risk benefit ratio in patients with refractory pulmonary sarcoidosis

MANAGEMENT OF SARCOIDOSIS contd…

- ***Management Protocols in Sarcoidosis:-***
  - **Observe patients without pharmacological interventions in patients who are asymptomatic stage 0/1 pulmonary sarcoidosis.**
  - **Treat patients with symptomatic stage 1 and all stage 2,3 and 4 pulmonary sarcoidosis with oral corticosteroids.**


Thank
you