Interstitial Lung Diseases in the Idiopathic Inflammatory Myopathies

Intira Masayavanich M.D.
Fellow-in-Training
Division of Respiratory Disease and Tuberculosis, Department of Medicine
Faculty of Medicine Siriraj Hospital, Mahidol University

Introduction
Idiopathic inflammatory myopathies or myositis (IIM) are heterogenous disorders characterized by varying degrees of muscle weakness and inflammation. Lungs are the most common extramuscular involvement in IIM including respiratory muscle weakness, pulmonary hypertension, interstitial lung disease, and pleural effusion. Interstitial lung disease (ILD) is the hallmark of pulmonary involvement that causes significant morbidity and mortality. Classification of Idiopathic inflammatory myopathies

The major subgroups in adult IIM are dermatomyositis (DM), polymyositis (PM), and inclusion body myositis (IBM). Several diagnostic criteria for IIM have been proposed as shown in Table 1. The Bohan and Peter’s criteria is the most widely used, but have several limitations including no external validation with the estimation of sensitivity or specificity and lack of specific criteria for exclusion of other forms of myopathies. Recently, The European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) published the classification criteria using multidisciplinary consensus that showed higher sensitivity and specificity for diagnosis of major subgroups of IIM.

Types of pulmonary involvement
Pulmonary involvement in IIM has various symptoms and signs that can be classified as direct pulmonary involvement (e.g. ILD and pulmonary hypertension) and indirect pulmonary complications (e.g. infection and respiratory muscle weakness) that are listed in Table 2. Pulmonary involvement is common in PM and DM, but IBM has no direct pulmonary involvement. ILD is the most common pulmonary involvement that can cause significant morbidity and mortality. This article will focus on ILD-associated with PM and DM.
### Table 1  Diagnostic criteria for idiopathic inflammatory myopathies

<table>
<thead>
<tr>
<th>Features</th>
<th>Features (except inclusion body myositis)</th>
<th>Dalakas and Hohlfeld⁷</th>
</tr>
</thead>
<tbody>
<tr>
<td>2. Elevated serum muscle enzymes</td>
<td>2. Elevated serum creatinine kinase level</td>
<td>2. Elevated serum muscle enzymes</td>
</tr>
<tr>
<td>3. EMG consistent with myopathy</td>
<td>3. Other laboratory criteria (EMG, MRI, or MSAs detected in serum)</td>
<td>3. Muscle biopsy</td>
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<tr>
<td>5. Typical rash</td>
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</table>

#### Diagnosis

<table>
<thead>
<tr>
<th>Definite PM: 1-4 criteria present</th>
<th>Definite PM: 1 without rash + 2 + 4</th>
<th>Definite PM: criteria 1, 2 with muscle biopsy showing inflammation with CD8/MHC-I complex and no vacuoles</th>
</tr>
</thead>
<tbody>
<tr>
<td>Probable PM: Any 3 of 1-4 present</td>
<td>Probable PM: 1 without rash + 2 + 3 (1 of 3) + 4</td>
<td>Probable PM: criteria 1 and 2 with muscle biopsy showing MHC-I expression without T cells or vacuoles</td>
</tr>
<tr>
<td>Possible PM: Any 2 of 1-4 present</td>
<td>Probable DM: 1 + 4 or 3 (1 of 3)</td>
<td>Probable DM: no rash + typical biopsy</td>
</tr>
<tr>
<td>Definite DM: Rash + any 3 of 1-4</td>
<td>Amyopathic dermatomyositis, possible dermatomyositis sine dermatitis, non-specific myositis, immune-mediated necrotizing myopathy diagnostic criteria: not shown</td>
<td>Definite DM: rash + muscle biopsy</td>
</tr>
</tbody>
</table>

ENMC, The European Neuromuscular Center; PM, polymyositis; DM, dermatomyositis; EMG, electromyography; MRI, magnetic resonance imaging; MSA, myositis-specific autoantibodies; ADM, amyopathic dermatomyositis

### Table 2  Pulmonary involvement in idiopathic inflammatory myopathies

<table>
<thead>
<tr>
<th>Pulmonary involvement</th>
<th>Diseases or conditions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Direct pulmonary involvement</td>
<td>• Interstitial lung disease</td>
</tr>
<tr>
<td></td>
<td>• Pulmonary arterial hypertension</td>
</tr>
<tr>
<td></td>
<td>• Pulmonary hypertension due to chronic hypoxemia</td>
</tr>
<tr>
<td>Indirect pulmonary complication</td>
<td>• Pulmonary infection</td>
</tr>
<tr>
<td></td>
<td>• Respiratory muscle weakness</td>
</tr>
<tr>
<td></td>
<td>• Drug-induced lung disease</td>
</tr>
<tr>
<td></td>
<td>• Lung cancer</td>
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</table>

ADM: Rash without muscle weakness
Epidemiology

The prevalence of ILD-associated with PM/DM is ranging from 17 to 36% \cite{11-14}. This variation is due to by a lack of standardized criteria and screening for ILD and the limitation of retrospective studies \cite{10}. The most common type of ILD in IIM patients is non-specific interstitial pneumonia (NSIP) follow by organizing pneumonia (OP) and usual interstitial pneumonia (UIP) as shown in Table 3.

Table 3  Interstitial lung disease in idiopathic inflammatory myopathies

<table>
<thead>
<tr>
<th>Type</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nonspecific interstitial pneumonia (NSIP)</td>
<td>+++</td>
</tr>
<tr>
<td>Organizing pneumonia (OP)</td>
<td>++</td>
</tr>
<tr>
<td>Usual interstitial pneumonia (UIP)</td>
<td>++</td>
</tr>
<tr>
<td>Acute interstitial pneumonia (diffuse alveolar damage)</td>
<td>+</td>
</tr>
<tr>
<td>Lymphocytic interstitial pneumonia (LIP)</td>
<td>+/-</td>
</tr>
</tbody>
</table>

+++ common; ++, fairly frequent; +, occasional; +/-, rare

Pathogenesis

The pathogenesis of ILD-associated with PM/DM remains unknown. It is generally accepted that genetically susceptible individuals and the exposure to some environmental factors like viral infection, smoking, inhalation of organic and inorganic dust may play a role. There were associated with human leukocyte antigen (HLA) class II, HLA-DRB*03, HLA- DQA1*05, and HLA-DQB1*02 irrespective of myositis subtype or anti-Jo-1 autoantibodies \cite{10}.

Immunohistochemistry on muscle biopsy provides some data about immune-mediated mechanisms \cite{1}. In polymyositis, CD8+ T-cells are usually found with diffuse cytotoxic effect leading to muscle cell necrosis predominantly within the endomysium of healthy-appearing, non-necrotic muscle fibers expressing major histocompatibility complex (MHC) class I antigen. In DM, B-cells and CD4+ T-cells were found in perivascular areas and complement on the endothelial cell wall of endomysial vessels which may be responsible for complement-mediated microangiopathy in patients with

Clinical presentation

Clinical presentation is variable including asymptomatic (25%), subacute or chronic ILD (58%), and rapidly progressive ILD (17%) \cite{2,13}. Progressive dyspnea, non-productive cough and decreased exercise tolerance are the most common symptoms that can precede (19%), occur concomitantly with (42%), or present after (39%) the diagnosis of PM/DM \cite{2,10,13}. However, these symptoms are non-specific to ILD because they can either present in patients with pulmonary involvement other than ILD, e.g. respiratory muscle weakness and pulmonary infection \cite{16}. Physical examination usually reveals velcro crackles with or without a sign of respiratory distress or central cyanosis depending on the severity of disease. Older age (≥45 years old), joint symptoms, and positive anti-Jo-1 at onset predict the presence of ILD in IIM patients \cite{17}.

Asymptomatic or occult ILD

Patients do not have respiratory symptoms. ILD is identified on radiological imaging, including plain radiographs or high-resolution computed tomography of the chest (HRCT) or abnormal pulmonary function tests \cite{16}. 

DM \cite{15}.
Subacute or chronic ILD

Chronic form of ILD was defined as a slowly progressive presentation with gradual deterioration over more than 3 months\(^\text{18}\). Patients with slow progressive pattern had a better survival rate than patients presented with rapidly progressive ILD regardless of the underlying IIM\(^\text{18}\).

Rapidly progressive ILD

Rapidly progressive ILD is an acute interstitial pneumonia (AIP) that develops in several weeks or a few months\(^\text{10}\). The presentation includes fever, dyspnea and rapid progression to acute respiratory failure with abnormal chest X-ray. The acute and aggressive presentation is more frequently seen in dermatomyositis, clinically amyopathic dermatomyositis (CADM), and hypomyopathic dermatomyositis\(^\text{2,12, 20-21}\). This form of aggressive ILD is more resistant to treatment with intensive immunosuppressive therapy and has a very poor prognosis with high mortality\(^\text{2,10,13}\). A meta-analysis demonstrated that the presence of anti-MDA5 autoantibodies in DM patients increased the risk of developing rapidly progressive ILD with a sensitivity of 77% and a specificity of 86%\(^\text{16, 22}\). The detection of anti-aminoacyl-tRNA-synthetase (anti-ARS) autoantibodies seems to be a protective factor against this presentation\(^\text{23}\).

Antisynthetase syndrome

The antisynthetase syndrome is characterized by the presence of one of antisynthetase antibodies in combination with fever (43%), polyarthritis (62%), myositis (57%), ILD (70%), mechanic’s hands (28%), and Raynaud’s phenomenon (47%) as shown in Table 4\(^\text{1}\). This syndrome occurs in up to one-third of patients with PM and DM. ILD is found in up to 70% of patients with antisynthetase syndrome and may precede the other symptoms\(^\text{10}\). Joint manifestations range from polyarthralgia to destructive polyarthritis of the hands, wrists, elbows, and knees. Erosive joint disease is uncommon\(^1\). 5-8% of antisynthetase syndrome manifests as overlap diseases with other connective tissue diseases, e.g. rheumatoid arthritis, systemic lupus erythematosus, scleroderma, and Sjögren’s syndrome\(^\text{24}\). ILD dominates the prognosis of antisynthetase syndrome which is associated with more than 40% mortality. Anti-Jo-1 and anti-PL are autoantibodies to histidyl-tRNA-synthetase which is the most common autoantibodies found in 60-80% of the antisynthetase syndrome. The other autoantibodies are found in 20-40%: anti-PL7 (10-15%), anti-PL12 (5-10%), anti-EJ, anti-OJ, and anti-KS (5-15%)\(^\text{10}\). Anti-PL7 and anti-PL12 seem to be associated with isolated pulmonary fibrosis or arthritis\(^\text{25}\).

Table 4 Proposed criteria for myositis associated anti-tRNA synthetase antibody\(^\text{1}\)

<table>
<thead>
<tr>
<th>Features</th>
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<tbody>
<tr>
<td><strong>Positive serological tests for anti-tRNA synthetase antibody plus one major involvement:</strong></td>
</tr>
<tr>
<td>• Evidence of overt or hypomyopathic myositis by Bohan and Peter criteria*</td>
</tr>
<tr>
<td>• Evidence of interstitial lung disease according to ATS criteria</td>
</tr>
<tr>
<td>• Evidence of articular involvement**</td>
</tr>
<tr>
<td><strong>Or two minor involvement:</strong></td>
</tr>
<tr>
<td>• Unexplained persistent fever</td>
</tr>
<tr>
<td>• Raynaud’s phenomenon</td>
</tr>
<tr>
<td>• Mechanic’s hands</td>
</tr>
</tbody>
</table>

* Elevated creatine phosphokinase (CPK) levels, myalgia, proximal muscle weakness, positive muscular biopsy, electromyographic triad of myositis or MRI muscular edema

** Symmetrical inflammatory arthralgia or overt arthritis
Anti-melanoma differentiation-associated gene 5 (MDA5)-related ILD

In 2005, anti-MDA5 autoantibody, originally called anti-CADM-140, was firstly described in 8 patients with CADM of whom 50% had developed rapidly progressive ILD\(^{25-27}\). The anti-MDA5 autoantibodies are specific to DM which may be found in 7-13% of DM patients. Significant myositis could be found in only 20% of patients. Skin manifestations are digital ulcers which usually located over the Gottron’s papules, digital pulps, periungual area, or other sites of dermatomyositis rash. Other manifestations include panniculitis, arthritis (80%), Raynaud’s phenomenon (45%), and mechanic’s hands (80%). The screening for ANA may be negative. Other laboratory investigations are a high level of serum ferritin, alpha-glutamyl transpeptidase, and lower CK values at presentation\(^{27}\).

Several studies have reported that anti-MDA5 had a strong association with rapidly progressive ILD and resulted in poor survival\(^ {25,28}\). The survival rate of patients as low as 54% at 6 months. An autoantibodies titer level >500 U/mL is associated with treatment resistance is a higher risk of short-term death from acute respiratory failure. High serum ferritin (> 1500 ng/ml) has also been associated with poor survival\(^ {27}\).

**Investigation**

**Chest imaging**

Chest X-ray is helpful but has limited sensitivity and specificity in the diagnosis of ILD. HRCT is more sensitive in the detection and characterization of ILD. The most common HRCT findings include consolidation, reticulation, ground-glass opacities, and peribronchovascular thickening as shown in Figure 1B and Table 5.\(^ {1}\) Honeycombing, traction bronchiectasis, and bronchiolectasis are less frequently found (Figure 1A)\(^ {1,11}\). A combination of reticulation and area of consolidation is usually found in acute to subacute onset of ILD.

HRCT patterns at diagnosis can predict the prognosis of patients. Dominant consolidation is generally responded well to corticosteroids and immunosuppressive agents. Dominant ground-glass opacities and/or reticulation without honeycombing, subpleural bands and traction bronchiectasis are often associated with severe ILD and a possibility of deadly outcome\(^ {1}\).

**Figure1** High-resolution computed tomography. A. Patient with usual interstitial pneumonia (UIP) associated with polymyositis, predominant traction bronchiectasis and honeycombing with reticulation; B. Patient with nonspecific interstitial pneumonia (NSIP) associated with scleroderma-polymyositis overlap syndrome, predominant ground-glass opacities with subpleural sparing, reticulation, traction bronchiectasis, and esophageal dilatation.

**Pulmonary function tests (PFTs)**

PFTs are a sensitive but non-specific test in the diagnosis of ILD but they can be used for evaluation of disease severity, course of disease and treatment response. Patients typically demonstrated a restrictive ventilatory defect and decreased diffusion capacity for carbon monoxide (DLCO). PFTs should be carefully interpreted due to a potential coexistence of respiratory muscle weakness and ILD which PFTs also show.
Table 5  Common HRCT and histopathologic patterns

<table>
<thead>
<tr>
<th>Pattern</th>
<th>HRCT findings</th>
<th>Histopathology</th>
</tr>
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<tbody>
<tr>
<td>Organizing pneumonia (OP)</td>
<td>Consolidation (unilateral or bilateral), usually peripheral, subpleural, or peribronchovascular</td>
<td>Foci of granulation tissue in alveoli and their ducts</td>
</tr>
<tr>
<td>Non-specific interstitial pneumonia (NSIP)</td>
<td>Subpleural and bibasilar ground-glass opacities with some traction bronchiectasis and reticulation</td>
<td>Temporally and geographically homogenous infiltration of lung interstitium by inflammatory cells with moderate collagen deposition, preserved lung architecture, and scarcity of fibroblastic foci and honeycombing</td>
</tr>
<tr>
<td>Usual interstitial pneumonia (UIP)</td>
<td>Subpleural and bibasilar honeycombing with reticulation and traction bronchiectasis</td>
<td>Spatially and temporally heterogenous collagen deposition within the lung, architectural destruction, fibroblastic foci, honeycombing, and moderate inflammatory cell infiltration</td>
</tr>
<tr>
<td>Acute interstitial pneumonia</td>
<td>Diffuse ground-glass opacities with consolidation with or without reticulation and traction bronchiectasis on background</td>
<td>Diffuse interstitial inflammation with edema and hyaline membrane production</td>
</tr>
</tbody>
</table>

HRCT, high-resolution computed tomography.

restrictive ventilatory defect from chest wall restriction and diffusion defect from basal lungs atelectasis\(^{11}\). FVC <60% predicted at the time of diagnosis is associated with poor survival.

**Bronchoscopy**

Bronchoscopy with bronchoalveolar lavage (BAL) is useful for the exclusion of occult infection and other diseases resembling ILD. Transbronchial lung biopsy (TBLB) may be helpful in the diagnosis of organizing pneumonia and ruling out other causes e.g. infection and malignancy but it has a limited role in the diagnosis of NSIP or UIP due to inadequate lung tissue.

**Surgical lung biopsy**

Surgical lung biopsy (SLB) in the diagnosis of ILD remained controversial.

**Histopathology**

Obtaining lung tissue for the diagnosis of ILD is challenging. According to the limitation of TBLB, transbronchial cryobiopsy (TBCB) and surgical lung biopsy (SLB) may play an important role in obtaining larger tissue for histopathological diagnosis. However, TBLB and SLB are more invasive procedures with reported major complications, e.g. pneumothorax, bleeding, acute exacerbation, and death. Currently, we recommend that TBLB and SLB should be performed only in patients with clinical and HRCT uncertainty.
The most common histology is NSIP and OP. Other histologic subtypes include usual interstitial pneumonia, diffuse alveolar damage, and lymphocytic interstitial pneumonia\textsuperscript{2,11,18}. Several histopathologic patterns correlate with HRCT findings, therefore, a tissue biopsy may not be necessary if there are typical findings on HRCT as shown in Table 5\textsuperscript{2}.

**Autoantibodies**

The screening for ANA may be negative. Further testing for myositis-specific autoantibodies (MSA) and myositis-associated autoantibodies (MAA) is necessary. ILD is commonly found in patients with antisynthetase syndrome. DM patients with positive antisynthetase antibodies are more likely to have ILD than antibody-negative DM (94\% and 23\%, respectively) and more likely to require higher and prolonged doses of immunosuppressive agents. The most common antisynthetase antibody is anti-Jo-1 is the most common antisynthetase antibody which strongly associated with the presence of ILD (89\%)\textsuperscript{11}.

**Serum biomarkers**

Several serum biomarkers have been studied, e.g. glycoprotein Krebs von den Lundgen-6 (KL-6), cytokeratin 19 fragment, and surfactant protein A and D, which showed variable results. Currently, there is limited data about the role of serum biomarkers in the diagnosis of ILD associated IIM and disease progression\textsuperscript{2,29-30}.

**Treatment**

1. **Pharmacologic treatment**

Corticosteroids is the mainstay treatment of IIM. However, there are no established treatment regimens for ILD associated with IIM\textsuperscript{10-11}. Immunosuppressive agents should be considered when ILD is severe or rapidly progressive in order to reduce the side effects of corticosteroids and improve the response to treatment\textsuperscript{18}.

**Corticosteroids**

Corticosteroids are the first-line treatment of IIM with a starting dose of 0.75 to 1 mg/kg/d equivalent to prednisolone with a slowly tapering based on clinical response. Approximately half of patients have a good clinical response to initial corticosteroid therapy. Corticosteroids monotherapy achieved favorable responses in 37.5\% of PM-ILD but only 8.3\% in ILD with DM patients\textsuperscript{26}. The overall 2.5-year survival rate of DM-ILD was 58\% and the 5-year survival of PM-ILD patients was 81\%. A combination with immunosuppressive drugs is recommended for steroid sparing or increased efficacy, particularly in rapidly progressive ILD\textsuperscript{10}.

**Azathioprine**

Azathioprine is commonly used in ILD associated with IIM as a corticosteroid-sparing agent or maintenance therapy, solely or following induction with cyclophosphamide. Dosage ranges from 2-3 mg/kg/day\textsuperscript{2,11}.

**Cyclophosphamide**

Cyclophosphamide can be administered orally or monthly intravenous pulse in combination with corticosteroids. According to side effect profiles, cyclophosphamide is commonly used in rapidly progressive ILD and refractory ILD. Several case studies have demonstrated its potential efficacy in treating ILD associated with IIM. Dosage ranges from 1-2 mg/kg/day per oral or monthly intravenous pulse 300-800 mg/m\textsuperscript{2} at least 6 times in combination with prednisolone 0.5-1 mg/kg/day\textsuperscript{2,18}. 

**Methotrexate**

Methotrexate is used as an adjunctive treatment in arthritis and myositis in PM/DM. However, there is controversy about the efficacy of methotrexate in the treatment of ILD associated IIM\(^\text{10}\). Methotrexate should be used with caution due to a known association with idiosyncratic drug-related hypersensitivity pneumonitis\(^\text{11}\). It is difficult to distinguish worsening symptoms after drug initiation between the manifestation of ILD and drug-induced lung disease\(^\text{11}\). It is usually administered orally at a dose of 0.2-0.3 mg/kg weekly.

**Calcineurin inhibitors**

Cyclosporin A and tacrolimus are the T-cell- and IL-2 production inhibitor\(^\text{29}\). Several case series have demonstrated that maintenance with cyclosporin A and tacrolimus may be an appropriate choice for early, slowly progressive, and non-diffuse ILD because of their safety profile and efficacy on stabilized lung functions\(^\text{2,29}\). Cyclosporin A dosage is 2-5 mg/kg/day adjusted for trough level of 100-200 ng/ml. Tacrolimus dosage depends on the trough level (5-20 ng/mL)\(^\text{2}\).

**Mycophenolate mofetil (MMF)**

MMF has a potential efficacy in reversing progression or stabilization of disease activity in various connective tissue diseases associated with ILD including IIM. Several studies have demonstrated remission or stabilization of ILD in 80% of patients with chronic ILD. MMF in combination with corticosteroids is increasingly used due to its potential efficacy and safety profile. Recommended dose is 2,000-3,000 mg/day\(^\text{2}\).

**Plasmapheresis**

Plasma exchange is used to remove circulating autoantibodies, cytokines, and immune complexes. Case reports of efficacy of plasmapheresis in patients with antisynthetase syndrome who were refractory to corticosteroids and other immunosuppressive therapies\(^\text{18}\).

**Intravenous immunoglobulin (IVIG)**

IVIG is widely used for the treatment of numerous autoimmune diseases. The efficacy of IVIG in ILD associated with IIM is uncertain. There were some case series demonstrated an improvement of PFT and CT imaging following the treatment of IVIG in patients with refractory myositis\(^\text{2,18}\).

**Rituximab**

Rituximab is a monoclonal antibody targeting the CD20 protein. The effectiveness on stabilizing and/or improving the pulmonary function tests, including FVC, DLCO, and TLC, is 72%. The most benefit on pulmonary function was observed in patients with disease duration <1 year and acute onset of ILD. Patients with antisynthetase syndrome, mainly positive anti-Jo-1 and anti-Mi-2, were more likely to respond to rituximab therapy\(^\text{31}\). There is an upcoming trial which is the first randomized control trial to study the efficacy of rituximab versus cyclophosphamide for treatment of connective tissue disease-associated interstitial lung disease as first-line treatment in CTD-associated ILD\(^\text{32}\). The recommended dose is 1,000 mg intravenously on day 0 and day 14.

2. **Non-pharmacologic treatment and vaccination**

**Pulmonary rehabilitation**

Currently, there is no recommendation about the criteria for pulmonary rehabilitation in ILD patients. Recent data demonstrated that exercise can improve muscle strength and reduce impairment in patients...
with IIM\textsuperscript{33}. A prospective cohort study, which included patients with different types of ILD, described improvement in quality of life and 6-minute walk distance following pulmonary rehabilitation\textsuperscript{34}. Although no study has evaluated the efficacy of pulmonary rehabilitation specifically in patients with myositis related ILD, it is likely that they would benefit from therapy\textsuperscript{35}.

**Long-term oxygen therapy**

No clinical trial for the use of long-term oxygen therapy in patients with ILD associated with IIM specifically. However, several guidelines recommend the use of oxygen in ILD patients who have significant hypoxemia, defined by oxygen saturation of <88% or partial pressure of O\textsubscript{2} (PaO\textsubscript{2}) of <55 mmHg. Patients should be reassessed regularly and oxygen prescription should be adjusted as oxygen demand change.

**Vaccination**

Pulmonary infection can contribute to a worsening of symptoms and cause significant morbidity and mortality. There is no study evaluating the impact of vaccination on ILD patients. However, several guidelines recommend that ILD patients should receive influenza and pneumococcal vaccination.

**Lung transplantation**

There are very few published case reports or series of successful lung transplantation in patients with ILD related IIM\textsuperscript{36-38}. There is a slightly lower 1-year and 2-year survival rates in patients with ILD related IIM (67.5% and 56.3%, respectively) compared to those of IPF (72.7% and 66.3%, respectively) and those of CTD-associated ILD (77.7% and 68.2%, respectively)\textsuperscript{38}.

**Palliative and end-of-life care**

The objective of palliative care is to provide comfort to patients and caregivers and to reduce the burden of symptoms. Discussion about diagnosis and prognosis of disease between physician, patient, and caregivers should be undertaken once the diagnosis is made.

**Prognosis**

The predictors of poor outcome include acute presentation, neutrophilic alveolitis, initial DLCO <45%, FVC ≤60%, DM, microangiopathy, digital infarcts in DM/ADM, and histopathologic diagnosis of UIP\textsuperscript{2,19}. The survival of ILD associated with IIM was 94%, 90%, and 87% at 1, 3, and 5 years, respectively\textsuperscript{2}.

**Conclusion**

ILD is the most common pulmonary manifestation of IIM which causes significant morbidity and mortality. ILD associated IIM has various manifestations and clinical courses. Careful evaluation and investigation is necessary for diagnosis and treatment decision.

**References**

11. Connors GR, Christopher-Stine L, Oddis CV, Danoff SK. Interstitial lung disease associated with the idiopathic inflammatory myopathies: what progress has been made in the past 35 years? Chest 2010; 138:1464-74.


