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INTERSTITIAL LUNG DISEASES: A RETROSPECTIVE CASE SERIES ANALYSIS IN A SPECIALIZED CENTER

Sutbayeva N.
2nd-year Internal Medicine Resident, Department of Internal Medicine, Research Institute of Cardiology and Internal Diseases, Almaty, Kazakhstan
Spabek A.
2nd-year Internal Medicine Resident, Department of Internal Medicine, Research Institute of Cardiology and Internal Diseases, Almaty, Kazakhstan
Kanatkyzy G.
2nd-year Internal Medicine Resident, Department of Internal Medicine, Research Institute of Cardiology and Internal Diseases, Almaty, Kazakhstan
Kartbayeva A.
2nd-year Internal Medicine Resident, Department of Internal Medicine, Research Institute of Cardiology and Internal Diseases ***Almaty, Kazakhstan***
Khabiyeva M.
2nd-year Resident, Department of Internal Medicine, Research Institute of Cardiology and Internal Diseases, ***Almaty, Kazakhstan***
Kuantaykyzy A.
2nd-year Resident, Department of Internal Medicine, Research Institute of Cardiology and Internal Diseases, Almaty, Kazakhstan
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Abstract

Background: Interstitial lung diseases (ILD) comprise a heterogeneous group of diffuse parenchymal lung lesions characterized by progressive fibrosis and respiratory failure. Their diagnosis and management in a multidisciplinary hospital are clinically challenging due to a broad etiological spectrum and frequent patient polymorbidity.

Objective: To conduct a retrospective analysis of a series of 10 clinical cases of ILD of various etiologies hospitalized in a specialized center, evaluating demographic, clinico-functional, and therapeutic characteristics.

Materials and Methods: A retrospective analysis of the medical records of 10 patients with verified ILD (ICD-10 codes: J84.8, J84.9, M34.8) was performed. Data analyzed included demographics, etiology, chest CT results, spirometry, oxygen saturation, and comorbidities. All data were fully de-identified.

Results: Of the 10 patients, 6 (60%) were men and 4 (40%) were women; the mean age was 54.6 ± 20.4 years (range 3–74 years). The identified etiologies were: ILD-COPD overlap (3 patients, 30%), RA-associated ILD (2 patients, 20%), idiopathic/unspecified ILD (2 patients, 20%), post-infectious ILD (2 patients, 20%), and SSc-associated ILD (1 patient, 10%). Among patients with available spirometry data ($n=5$), the mean FVC was $52.2 \pm 14.4\%$ and FEV_1 was $50.3 \pm 3.9\%$. Grade II–III respiratory failure was identified in 4 out of 10 patients (40%). Pulmonary hypertension was diagnosed in 3 patients (30%). The most common comorbid conditions were: AH (60%), HF (50%), COPD (30%), CAD (30%), and PH (30%).

Conclusion: The case series analysis demonstrates marked clinical heterogeneity of ILD, a high frequency of cardiovascular comorbidity and pulmonary hypertension, as well as the necessity for a multidisciplinary approach to managing this patient category. The findings align with current international ATS/ERS/JRS/ALAT recommendations.

Keywords: interstitial lung diseases, clinical case series, retrospective analysis, pulmonary fibrosis, respiratory failure, rheumatoid arthritis, systemic sclerosis, pulmonary hypertension, COPD, spirometry.

INTRODUCTION

Interstitial lung diseases (ILDs) constitute a broad and heterogeneous group of diffuse parenchymal lung diseases, comprising over 200 nosological forms with varying pathogenic mechanisms but shared clinical, radiological and functional manifestations [1, 2]. The common pathomorphological substrate is inflammation and/or fibrosis of the lung interstitium, which inevitably leads to progressive respiratory failure (RF) and a reduced quality of life for patients [3].

According to the 2013 ATS/ERS/JRS/ALAT international classification, ILDs are subdivided into idiopathic interstitial pneumonias (IIPs), ILDs associated with systemic connective tissue diseases (SCTDs), hypersensitivity pneumonitis, occupational ILDs, and other forms [4]. Among IIPs, idiopathic pulmonary fibrosis (IPF) occupies a special place, characterised by a UIP pattern on high-resolution CT and an extremely poor prognosis: the median survival is 2–5 years without specific antifibrotic therapy [5, 6].

IPF in rheumatoid arthritis (RA-IPF) is one of the most common extra-articular manifestations of RA, occurring in 10–30% of patients undergoing CT scans [7]. EI in systemic sclerosis (SS-EI) is characterised by a predominance of the NSIP pattern and occurs in 35–70% of patients with SS, being the leading cause of mortality in this condition [8, 9]. The ILD-COPD overlap syndrome presents particular diagnostic challenges,

as restrictive changes are masked by underlying obstruction, which significantly complicates the assessment of the severity of pulmonary impairment [10].

Pulmonary hypertension (PH), which develops as a complication of IPF, falls into WHO Group 3 and is associated with a markedly poor prognosis — the 5-year survival rate for IPF+PH does not exceed 25–30% [11]. A wide range of comorbid conditions — cardiovascular disease, diabetes mellitus, chronic kidney disease — significantly complicates the use of both antifibrotic drugs (nintedanib, pirfenidone) and immunosuppressive therapy [12, 13].

In Kazakhstan, as in other countries with economies in transition, epidemiological data on IBD are limited, and the organisation of specialised multidisciplinary care is still in its infancy [14]. Publications of case series are of significant value for the establishment of registries, the analysis of clinical practice, and the development of clinical guidelines at the national level [15].

The aim of this study is a retrospective analysis of a series of 10 clinical cases of IBD of various aetiologies, verified at a specialised centre (JSC 'NCKIV', Almaty), with an assessment of demographic characteristics, the aetiological spectrum, clinical and functional indicators, the frequency of comorbid conditions, and the therapeutic strategies employed.

MATERIALS AND METHODS

Study design. A retrospective descriptive case series study. Medical records were analysed for 10 patients hospitalised for IHD or with a confirmed diagnosis of IHD between 2024 and 2025 inclusive.

Inclusion criteria. The analysis included patients: (1) with a verified diagnosis of ILD according to ICD-10 codes (J84.8, J84.9, M34.8 with pulmonary involvement); (2) with available chest CT scan data or a clinically and radiologically confirmed diagnosis; (3) with available data on comorbidity, severity of chronic obstructive pulmonary disease (COPD) and treatment received.

Exclusion criteria. Patients with an unverified diagnosis, insufficient medical documentation, or suspected malignant lung tumours as the primary cause of changes on CT were excluded.

Data collection. The following data were extracted from medical records: gender, age, diagnosis (primary and comorbid), ICD-10 code, severity of respiratory failure (RF 0–III according to clinical and functional criteria), spirometry results (FEV_1, Tiffeneau index), peripheral blood oxygen saturation (SpO_2), radiographic/CT characteristics, immunological test results (ANA, SCL-70, RF, CRP), as well as treatment regimens used. All patients' personal data have been fully de-identified in accordance with bioethical requirements: patients have been assigned serial numbers (Patient No. 1 – No. 10).

Aetiological classification. All cases of ILD were classified according to aetiological principles: (1) ILD associated with CSD (RA, SSc) ; (2) COPD-overlap; (3) post-infectious ILD (following COVID-19, tuberculosis, viral-bacterial pneumonia); (4) idiopathic/unspecified ILD; (5) ILD with possible occupational aetiology.

Functional assessment. The severity of hypoxemia was assessed based on clinical criteria and SpO_2: Hypoxemia 0 — SpO_2 ≥95%, Hypoxemia I — SpO_2 90–94%, Hypoxemia II — SpO_2 75–89%, Hypoxemia III — SpO_2 <75% or requirement for mechanical ventilation. Spirometry data were interpreted according to the 2022 ATS/ERS criteria. A restrictive pattern was defined as FEV_1/FVC ≥0.70 and a FVC <80% of predicted.

Pulmonary hypertension. PH was diagnosed when the pulmonary artery systolic pressure (PASP) was >35 mmHg as determined by echocardiography, in accordance with the 2022 ESC/ERS criteria (type 3 — due to lung pathology).

Statistical analysis. Due to the small sample size (n=10), the statistical analysis is descriptive in nature. Quantitative variables are presented as the mean ± standard deviation (M ± SD) or the median with interquartile range (Med [Q1; Q3]) in cases of non-normal distribution. Qualitative variables are presented as absolute values and percentages. The normality of the distribution was tested using the Shapiro-Wilk test.

RESULTS

The study included 10 patients. Of these, 6 (60%) were men and 4 (40%) were women. The mean age of the patients was 54.6 ± 20.4 years (range 3–74 years). Excluding the paediatric patient (Patient No. 1), the mean age of the adults was 58.9 ± 12.9 years, which corresponds to data from international IHD registries [16]. The majority of patients (70%) were in the 35–74 age range.

By aetiological classification, patients were distributed as follows: ILD-COPD overlap (Patients No. 5, 7, 8) — 3 cases (30%); IPF associated with RA (Patients 2, 3) — 2 cases (20%); idiopathic/unspecified IPF (Patients 4, 6) — 2 cases (20%); post-infectious ILD (Patients No. 1, 10, with signs of a UIP pattern in the latter) — 2 cases (20%); ILD in SLE (Patient No. 9) — 1 case (10%). In Patient No. 10, immunological screening (SCL-70, ANA, ANCA, dsDNA) was negative, which, in the presence of a UIP pattern on CT, meets the criteria for idiopathic pulmonary fibrosis (IPF) according to the 2022 ATS/ERS/JRS/ALAT guidelines [5].

Stage 0 pulmonary fibrosis was recorded in 2 patients (20%); Stage I pulmonary fibrosis — in 3 patients (30%); Stage I–II pulmonary fibrosis — in 1 patient (10%); Stage II pulmonary fibrosis — in 3 patients (30%); Grade III DN was recorded in 1 patient (10%). Thus, severe DN (Grades II–III) was identified in 4 out of 10 patients (40%), indicating late diagnosis and/or high severity of the pathological process in this sample.

Spirometry results are available for 5 patients (50%). Among them: the mean FEV_1 was 52.2 ± 14.4% (range 38–74%), mean FEV_1 was 50.3 ± 3.9% (range 46–56%), and the mean Tiffeneau index (FEV_1/FEV_1) was 97.0 ± 27.8% (range 72–135%). A restrictive pattern (FEV_1/ FEV_1/FVC ≥0.70 with reduced FVC) was identified in 3 out of 5 patients. An obstructive component (FEV_1/FVC <0.70) was recorded in 2 out of 5 patients (Patients No. 2 and No. 8), which corresponds to a mixed pattern of pulmonary functional impairment. Severe restriction (FEV_1 38%) in Patient No. 10 is an unfavourable prognostic marker in ILF [6].

PH (WHO class 3) was diagnosed in 3 patients (30%): chronic cor pulmonale with class III PH in Patient No. 6; class III PH in Patient No. 10 (PA pressure 40 mmHg on echocardiography); PH as a complication of ILD in Patient No. 9. The presence of PH is associated with a significant deterioration in functional status and prognosis [11, 17].

Among the verified CT patterns: UIP pattern ('honeycomb lung', traction bronchiectasis) — 1 case (Patient No. 10); bilateral interstitial changes with fibrosis — 3 cases (Patients No. 6, 7, 9); polysegmental interstitial pneumonia — 2 cases (Patients 1, 4); linear pneumofibrosis — 1 case (Patient 2); bronchiectasis with fibrosis — 1 case (Patient 8). CT data were unclear or unavailable for 2 patients.

All 10 patients had ≥1 significant comorbidity. The mean number of comorbid conditions per patient was 3.8 ± 1.7. The most common were: hypertension — 6 (60%), heart failure — 5 (50%), COPD — 3 (30%), CHD — 3 (30%), pulmonary hypertension — 3 (30%), autoimmune thyroiditis/hypothyroidism — 2 (20%), RA — 2 (20%). A detailed breakdown of comorbidities is presented in Table 3.

Various therapeutic strategies were employed depending on the aetiology of IBD. Immunosuppressive therapy (methotrexate, leflunomide, mycophenolate mofetil, glucocorticoids) was used in 5 patients with SIBD or a post-infectious component. Antifibrotic therapy (pirfenidone, nintedanib) was considered in 2 cases (Patients No. 7, 10), but was limited by severe comorbidities. Bronchodilators (DHA, DBA, combined IGC/DBA) were used in 3 patients with IZL-COPD overlap. Long-term oxygen therapy was prescribed for Patient No. 10 (DN II, reduced SpO_2). Patient No. 1 (paediatric case) received immunotherapy and systemic GCS.

Table 1.

Demographic and clinical characteristics of patients

No.	Sex	Age (years)	ILD type / etiology	Main comorbidities	RF	SpO_2 (%)	FVC (%)	Main therapy
1	F	3	Bilateral interstitial (viral-bacterial) polysegmental pneumonia; MIS-C	Iron deficiency anemia, DIC, CMV+EBV infection, GERD stage III, secondary immunodeficiency, CIDP	III	88	n/a	Dexamethasone, immunotherapy (Bioven), antibiotics
2	F	61	RA-associated ILD (J84.8)	RA (since 2023), COPD (since 2013), 40-year smoking history	0	n/a	51	Methotrexate 12.5 mg SC once weekly
3	F	59	RA-associated ILD (J84.8)	RA (since 2015–2016), hypertension (up to 180 mmHg), AIT, COVID-19 (2021, CT-3, 80% involvement)	0	97	Restrictive type	Leflunomide 20 mg/day, Methylprednisolone 4 mg/day
4	F	47	Unspecified ILD (J84.9); bilateral polysegmental interstitial pneumonia	Occupational chlorine exposure (36 years), pulmonary emphysema	I	n/a	48	Symbicort 160/4.5 μg, Berodual as needed
5	M	62	ILD (J84.8) + COPD group E, exacerbation	CAD, stable angina class II, post-stenting (RCA), HFpEF 58%, hypertension stage 2, chronic hepatitis C, asthma, type 1 diabetes, CKD stage 3b	I	n/a	n/a	Comprehensive therapy of comorbidities
6	M	65	ILD (J84.8); chronic cor pulmonale; pulmonary hypertension group III	CAD, post-infarction cardiosclerosis (2021), PCI (RCA), CABG (LIMA-LAD, SVG-RCA), atrial fibrillation (tachysystolic), HFmrEF 50%, hypertension stage 2	II	n/a	n/a	Anticoagulants, cardiotropic therapy; evaluation of antifibrotic therapy

7	M	63	Fibrotic ILD (J84.8) + COPD group E; onset after ARVI (Feb 2020)	Hypertension stage 1 (risk 3), HFpEF 51%, weight loss 20 kg in 6 months	II	n/a	n/a	LAMA/LABA; systemic glucocorticoids; evaluation of nintedanib
8	M	35	Post-inflammatory residual changes; bilateral bronchiectasis (J84.8)	Pulmonary tuberculosis (2008–2011), chronic bronchitis since childhood; FVC 74%, FEV_1 46%	I–II	n/a	74	Antibiotics, bronchodilators, postural drainage
9	M	37	Progressive fibrosing ILD in systemic sclerosis (M34.8); ANA(+), SCL-70 (246)	Systemic sclerosis with skin, vascular (Raynaud's), GI (esophagopathy) involvement; RF 79, CRP 27	I	n/a	n/a	Mycophenolate mofetil 500 mg BID, Methylprednisolone 8 mg/day, Pentoxifylline, Nifedipine
10	M	74	Fibrotic ILD (J84.8); UIP pattern; honeycombing; traction bronchiectasis; PH class III	Hypertension stage 3, critical aortic stenosis (AVA 0.85 cm^2), MR grade 2, HFpEF 55% → 46%, CT-confirmed fibrosis	II	n/a	38	Oxygen therapy; pirfenidone under consideration; TAVI planned

Note: RF – respiratory failure; n/a – not available; AH – arterial hypertension; CHD – coronary heart disease; HF – heart failure; IDA – iron deficiency anaemia; DIC — disseminated intravascular coagulation; CIDP — chronic inflammatory demyelinating polyneuropathy; GERD — gastro-oesophageal reflux disease; COPD — chronic obstructive pulmonary disease; BA — bronchial asthma; AF — atrial fibrillation; RA — rheumatoid arthritis; AIT — autoimmune thyroiditis; SSc — systemic sclerosis; MPM — mycophenolate mofetil; GCS — glucocorticosteroids; ICS — inhaled GCS; LTAA/LTAB — long-acting anticholinergics/β_2-agonists.

Table 2.

Spirometric and functional parameters

Patient No.	**Sex**	**Age (years)**	**FVC (%)**	**FEV_1 (%)**	**FEV_1/FVC (%)**	**Ventilatory pattern**	**Respiratory failure grade**
1	F	3	n/a	n/a	n/a	No data available (age-related limitation)	RF III
2	F	61	51	56	80	Restrictive (+ obstructive component)	RF 0
3	F	59	n/a	n/a	n/a	Restrictive pattern	RF 0
4	F	47	48	49	121	Restrictive	RF I
5	M	62	n/a	n/a	n/a	COPD + restriction (obstructive)	RF I
6	M	65	n/a	n/a	n/a	Restrictive + pulmonary hypertension	RF II
7	M	63	n/a	n/a	n/a	Restrictive + fibrosis	RF II
8	M	35	74	46	72	Obstructive (moderate)	RF I–II
9	M	37	n/a	n/a	n/a	Restrictive (SSc-ILD)	RF I
10	M	74	38	50	135	Severe restrictive (UIP pattern)	RF II
Total (n=5*)	**—**	**Mean age 52,3**	**52,2 ± 14,4**	**50,3 ± 3,9**	**97,0 ± 27,8**	***Patients with available spirometry data**	**—**

*Note: FVC — forced vital capacity; FEV_1 — forced expiratory volume in 1 second; Tiffeneau index; n/a — not available; respiratory failure; SSc — systemic sclerosis. *The mean value was calculated only for patients with available numerical spirometry data (n=5).*

Table 3.

Prevalence of comorbid conditions in the study group (n=10)

Comorbid condition / disease	Number of patients (n=10)	Frequency (%)
Arterial hypertension	6	60
Heart failure (any class / EF)	5	50
Rheumatoid arthritis (RA)	2	20
COPD (diagnosed)	3	30
Bronchial asthma	1	10
Coronary artery disease (any form)	3	30
Systemic sclerosis (SSc)	1	10
Diabetes mellitus	1	10
Atrial fibrillation	1	10
Chronic kidney disease	1	10
Chronic viral hepatitis C	1	10
Autoimmune thyroiditis / hypothyroidism	2	20
Pulmonary hypertension (confirmed)	3	30
History of pulmonary tuberculosis	1	10
Severe COVID-19 with lung involvement	1	10
Occupational exposure (chlorine)	1	10
Iron-deficiency anemia	1	10
Cardiac valve disease (aortic stenosis / mitral insufficiency)	1	10

Note: AH – arterial hypertension; PH – pulmonary hypertension; MI – mitral insufficiency; AS – aortic stenosis.

DISCUSSION

This retrospective analysis of a series of 10 clinical cases of IHD enables us to characterise the actual clinical management of this patient group within a specialised multidisciplinary centre. The data obtained demonstrate marked heterogeneity in the nosological spectrum, severe comorbidity and a significant proportion of patients with moderate to severe renal impairment.

The demographic profile of the sample (mean age 54.6 years, 60% male) generally corresponds to the profile of hospitalised patients with acute myocardial infarction in specialised centres. According to data from international registries (EUROLD, INPULSIS), the mean age of patients with ILF is 65–70 years, whereas for ILD within the context of SCD it is 45–60 years [16, 18]. The presence of a paediatric patient (aged 3 years) with ILD in the context of severe viral-bacterial pneumonia and MIS-D highlights the uniqueness of the clinical setting. MIS-C (multisystem inflammatory syndrome in children) and associated interstitial lung changes represent a pressing nosological issue in the post-pandemic period, as highlighted in paediatric guidelines [19].

The most common aetiological group in this series was IBD-COPD overlap (30%), which corresponds to data from real-world clinical practice, where the combination of destructive and fibrotic changes is a common and prognostically unfavourable phenomenon [10, 20]. In patients with this pattern, FEV1 values may be pseudonormal due to the mutual offsetting of obstructive and restrictive components, which creates diagnostic difficulties and necessitates mandatory high-resolution CT to verify interstitial changes.

IPF in RA was recorded in 2 patients (20%). Both cases are characterised by a long history of RA and the use of basic anti-inflammatory therapy (methotrexate, leflunomide). It should be noted that methotrexate itself can cause drug-induced pneumonitis, which requires careful differential diagnosis [7]. The presence in Patient No. 2 of both RA and COPD against a background of 40 years' smoking history clearly demonstrates the difficulties in attributing pulmonary involvement.

The sole case of SSc-ILD (Patient No. 9) presents a typical clinical picture: a young man with a long history of Raynaud's syndrome, positive ANA and high titres of SCL-70 anti-topoisomerase antibodies (246), progressive pulmonary fibrosis and a need for immunosuppression (mycophenolate mofetil, corticosteroids). SCL-70 positivity is the most significant predictor of ILD in SSc [8, 9]. The use of mycophenolate mofetil is consistent with the results of the SLS-I and SLS-II studies, which demonstrated its efficacy comparable to that of cyclophosphamide in SSc-ILD, with a better safety profile [21].

Patient No. 10 represents the most clinically severe case in the series: ILF with a UIP pattern on CT (honeycomb lung, traction bronchiectasis), severe restriction (FEV1 38%), pulmonary hypertension (PA pressure 40 mmHg) and critical aortic stenosis (AVA 0.85 cm^2), making the use of antifibrotic therapy and surgical intervention associated with high risk. The coexistence of ILF and significant heart disease requires a multidisciplinary discussion involving a pulmonologist, cardiologist and cardiac surgeon [5, 22]. Discussion of the possibility of TAVI (transcatheter aortic valve implantation) in this patient is warranted in light of the high surgical risk associated with open surgery.

The high prevalence of cardiovascular comorbidities (hypertension 60%, heart failure 50%, coronary artery disease 30%, PH 30%) in the present series corresponds to data from large IHD registries and is explained by common pathogenetic mechanisms: systemic inflammation, oxidative stress and hypoxaemia [12, 17]. Progressive hypoxaemia in severe IHD is the leading mechanism in the development of stage II–III PH and right ventricular failure. The prevalence of PH identified in our series (30%) slightly exceeds the

data from epidemiological studies (15–25% in mild IHD) [11], which may reflect the selective nature of the sample from a specialist centre.

Functional parameters in the group of patients who underwent spirometry indicate a significant reduction in FEV_1 (mean 52.2%), which, in combination with CT lung function data, corresponds to moderate and severe restriction. It should be emphasised that a normal or only slightly reduced FEV_1 value does not rule out severe IPF with concomitant emphysema (IPF-COPD overlap) [10]. The sole obstructive pattern with minimally altered FEV_1/FEV_1 ratio and normal FEV_1 (Patient No. 2) indirectly confirms this phenomenon.

The therapeutic approaches in the study group reflect the current consensus on the management of IBD. Immunosuppressive therapy (methotrexate, leflunomide, MFM, GCS) is indicated for SIBD and a number of forms of post-infectious IBD. Antifibrotic agents (nintedanib, pirfenidone) are first-line therapy for ILD and progressive fibrotic ILD [5, 23]. Nintedanib is also approved for SSc-ILD (SENSCIS study) and for ILD-COPD with a progressive phenotype [24]. The use of these drugs in clinical practice is limited by their high cost, severe comorbidity and the need for liver function monitoring.

Study limitations. The main limitations of this analysis are: (1) a small sample size (n=10), which does not allow for statistically significant comparisons; (2) a retrospective design with incomplete data (no spirometry in 50% of patients); (3) the absence of data on histological verification of the diagnosis for the majority of patients; (4) heterogeneity of observations in terms of diagnosis, age and severity. Nevertheless, a series of clinical cases is a valuable tool for the initial characterisation of the patient population and for generating hypotheses for subsequent prospective studies [15].

CONCLUSION

A retrospective analysis of a series of 10 clinical cases of ILD at a specialist multidisciplinary centre allows the following conclusions to be drawn.

1. The aetiological spectrum of ILD in the study group is characterised by marked heterogeneity: the most common subgroups were ILD-COPD overlap (30%), SSc-associated ILD (RA + SSc, 30%) and idiopathic/unspecified ILD (20%).

2. Severe respiratory failure (grade II–III) was identified in 40% of patients, indicating late presentation and/or high disease severity in this sample.

3. Functional parameters (FVC 38–74%, FEV_1 46–56% in those examined) indicate moderate to severe impairment of lung function, consistent with the clinical picture.

4. The high prevalence of cardiovascular comorbidities (hypertension — 60%, heart failure — 50%, coronary artery disease — 30%) and pulmonary hypertension (30%) complicates therapeutic decisions and requires a multidisciplinary approach.

5. Modern therapeutic strategies (anti-fibrotic therapy for ILD, immunosuppression for SLE-associated ILD) were applied in accordance with international guidelines, but were limited by significant comorbidity in a number of cases.

The data obtained justify the need to establish a regional IHL registry in Kazakhstan, to standardise the diagnostic algorithm with mandatory CT-lung and multidisciplinary discussion, and to increase the availability of antifibrotic drugs for patients with progressive fibrotic IHL.

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