ILD-PAK-PCS Registry Report 2016-18

INTERSTITIAL LUNG DISEASE

Interstitial Lung Disease ILDPAK
Right step to a better breath

VOLUME II
DOI XXXXXXXXXX
ISSN 2518-2250

PAKISTAN CHEST SOCIETY
STRIVING FOR PULMONARY CARE

ILD PAK PCS Report 2016-18

Interstitial Lung Disease

Registry Report

2016 - 2018

www.ildpak.com

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ISSN 2518-2250

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Published by

Edited by

Sponsored by

December 2018

Printed in Pakistan

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Acknowledgment

ILD PAK PCS Registry acknowledges time, effort and expertise provided by the following specialists.

Resource Group

Radiology
Rasheed Ahmed (ARC)
Waseem Memon (AKU)
Shafiq Ahmed (SZ)
Afshan Qureshi (KEMU)

Rheumatology
Mehfooz Alam (LNH)
Saliha Ishaq (AKU)
M Saeed (SCH)

Surgery
Saulat Fatemi (AKU)
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Documentation
Anum Fatima

CONTENTS

MESSAGE FROM PRESIDENT

Pakistan Chest Society

Research has an important role in the development and promotion of any society. PCS had initiated the project of ILD registry involving all big cities of Pakistan. Today as President of Pakistan Chest Society, I am proud to announce that we have successfully published the second report of Registry on Interstitial Lung Disease. This was a difficult task but the tireless efforts especially by Dr. Mosavir Ansarie and Prof. Ali Bin Sarwar Zubairi led to the accomplishment of this project. The data of this registry will help in understanding the pattern of ILD in Pakistan and in turn help in streamlining the management of ILD.

Regards,

Professor Nisar Rao
President
Pakistan Chest Society

MESSAGE FROM CHAIRMAN STEERING COMMITTEE

ILD PAK PCS Registry

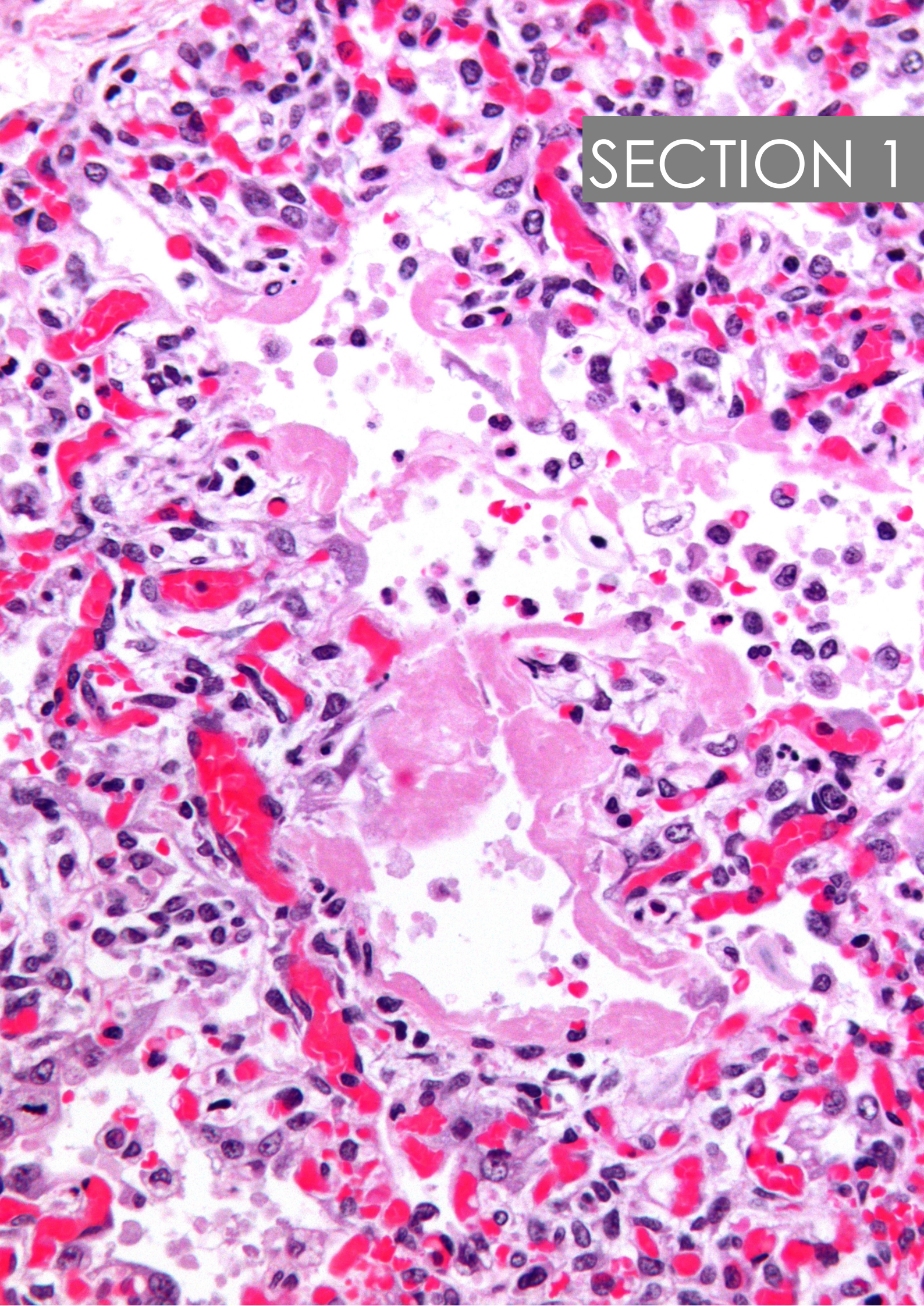
I am very excited on the publication of multi-centre prospective registry of interstitial lung disease from Pakistan. The previous report published in June 2016 provided insight from 3 centres of the largest city of Pakistan. This second report from the ILD-PAK PCS Registry provides the latest information from the rapidly expanding data on interstitial lung disease (ILD) from Pakistan. The registry has been steadily gaining momentum with 10 hospitals entering data and contains over 700 patients in last 2 years. As information on ILD continues to build, the picture of the prevalence, the natural history and outcomes will become clearer. The registry allows centres to monitor and improve the data as recommended by the Steering committee.

The Pakistan Chest Society (PCS) is committed to ILD-PAK registry and we hope to expand the ILD registry program to other centres of the country. I would like to encourage my colleagues and friends in centres who see patients with ILD to consider joining the registry.

I am extremely thankful to the entire team who helped in preparation of this report, to all those who contributed in data collection, to the steering committee members and in particular to Professor Arshad Javaid, Dr. Mosavir Ansarie and Mr. Jaffer Bin Baqar who continued to supervise this important venture throughout the journey.

Regards,

Professor Ali Bin Sarwar Zubairi
Chairman Steering Committee
ILD PAK PCS Registry


SECTION 1

OVERVIEW

Epidemiological studies have always been vitally important in assessing the extent of a disease in a population group and identifying its various facets and proclivities so that better management strategies could be devised.

Historically, a population-based registry differentiating between various types of ILDs established in New Mexico in 1994 was a landmark study in this respect[1]. Later in 2001, a comparison of registries established in three European countries- Belgium, Germany and Italy highlighted the similarities and dissimilarities between them suggesting that a global registry was perhaps necessary[2-4]. Considering that this data emanated from European countries alone with possible geographical differences a local registry was set up in 2008 in Karachi, Pakistan by the name of ILD PAK Registry.

Since 2011, however, after the consensus document from ERS/ATS/JRS/ALAT, a greater recognition of the need of registries has led to the establishment of new registries at a national level in various countries like Spain, Greece, Germany, India and Australia, in that order[5-9]. At this point of time ILD PAK was converted into a web based electronic national registry providing access for data entry to various pulmonology centres across the country. Generally, there remains a great paucity of literature reporting ILD in Asia with no other multicentre registry in Pakistan.

The differences between this and other studies may reflect differences among country variations in the multidisciplinary methods, applied diagnostic measures and data reporting. It however attempts to consolidate valuable demographic and clinical information on ILDs into a national database to enable better understanding of these chronic conditions. It also attempts to enhance the quality of collected data across the country, and by this process develop networking among those of our clinical scientists who have interest in the field of ILD.

Some data from volume 1 of this registry was presented earlier in scientific conferences (ERS 2012 Vienna, ACCP-SEPAR 2014 Madrid, CWC 2016 Shanghai)[10-12]. This comprehensive report in line with the overall objectives of the ILD PAK registry and the PCS National Guideline which reiterates the importance of multi city data entry into the national ILD registry[13]. The Registry will hopefully serve as an important resource for future clinical and applied research leading to improvement in standards of patient care.

WHY REGISTRIES?

A registry is a comprehensive document that records data and keeps track of a specific sub population of patients with a specific condition within stipulated time boundaries.

A registry must have specific objectives and focus on these while following a methodology devised to ensure quality data entry and updating.

A registry facilitates in a variety of ways. It can collect epidemiologic and clinical information regarding the number of individuals with diseases, treatments outcomes. Alternatively, registries invite people to register and be contacted regarding prospective participation that can be helpful in further clinical analysis and trials.

OBJECTIVES OF ILD PAK REGISTRY

1. To record the epidemiology of ILD in Pakistan.
2. To determine the relative frequencies of ILD with Demographic distribution in Pakistan.
3. To record the clinical features, associated risk factors and co-morbidities in ILD population in Pakistan. To record exposure histories and relate identifiable causes of ILDs in local environment.
4. To record the survival and mortality rate of ILD patients in Pakistan.

UTILITY OF THIS REGISTRY DATA

- Provision of essential information on baseline demographic features such as age, gender, race, location, occupation, exposure and smoking status as well as details of significant associations and co morbidities.
- Provision of information on the incidence and prevalence of different ILDs in various geographical locations of Pakistan.
- Determining the status of disease recognition from the appearance of first symptoms to the time of diagnosis and the delay in specialist referral and diagnostic measures in different areas of the country.
- Determining the status of knowledge about the disease in general practices and their preference of treatment given at the time of patient referral and their inclusion in the registry.
- Provision of a data bank of clinical profiles of Pakistani ILD patients who may be available for research and clinical trials in future.
- Provision of access to comprehensive information of patients available to respective investigators for publication individually and jointly with other investigators. This data will enable them to determine differences in disease behaviour in different regions of the country and harmonize the management on a national scale enabling a periodic review of the national guidelines.

AN INTRODUCTION TO ILD

The Interstitial lung diseases are a heterogeneous group of pulmonary disorders, classified together because of similar clinical, radiologic, physiologic or pathologic features[14,15]. Pathophysiologically, these can be categorized on the basis of diffuse involvement of pulmonary parenchyma that can be acute or chronic with variable degree of lung fibrosis[16,17]. These are often described as diffuse parenchymal lung disease (DPLD) because the disease process is not limited to interstitium of the lungs and may involve alveolar spaces, acini and the bronchioles[18]. The clinical classification of ILD includes four clinically distinct groups; (i) ILD of known association such as Hypersensitivity Pneumonitis (HP) or Collagen Vascular Associated Disease (CVD), (ii) Granulomatous ILD like Sarcoidosis, (iii) Rare ILDs such as Pulmonary Langerhans Cell Histiocytosis and (iv) Idiopathic diseases where etiology in a majority of cases remains unidentified and such ILD types include most importantly idiopathic pulmonary fibrosis, non-specific interstitial pneumonias, sarcoidosis and so on[19-21].

The physiological, clinical, histopathological and radiological findings help in the diagnosis of ILD among varied groups. There are multiple factors among which age has been identified as a significant risk for the development of certain types like IPF[22]. A gender predilection towards males is also noted in the case of IPF[23]. There are also other associated risks of development of ILD such as history of smoking in IPF and exposure to metal, wood dust, avian antigens and certain drugs as in HP. Factors like Gastroesophageal Reflux Disease (GERD) and genetic background have also been linked to the risk of development of IPF[24-27].

Usual presenting symptoms include difficulty in breathing, especially on exertion and persistent dry cough along with specific auscultatory findings[28], but these may vary. In addition, systemic and extra thoracic clinical features help to guide towards the diagnosis of ILD. Diagnostic investigations include imaging studies such as chest x-ray and High Resolution Computed Tomography (HRCT) of the chest[29]. Each type of ILD manifests a different pattern on the HRCT[30,31]. Pulmonary Function Tests (PFTs) typically show a restrictive pattern of ventilatory defect[32]. However, in case where diagnosis is ambiguous, a biopsy of the lung tissue is warranted[33,34]. Ideally, the diagnosis of ILD should be based on a multidisciplinary approach involving a pulmonologist, radiologist and histopathologist[21,23,35].

Current therapy includes new anti-fibrotic agents aimed at slowing the progression of the disease[36,37]. Many clinical trials are underway, raising hopes of having better treatment options in the future for patients with fibrosing ILD.

ILD PAK REGISTRY METHODOLOGY

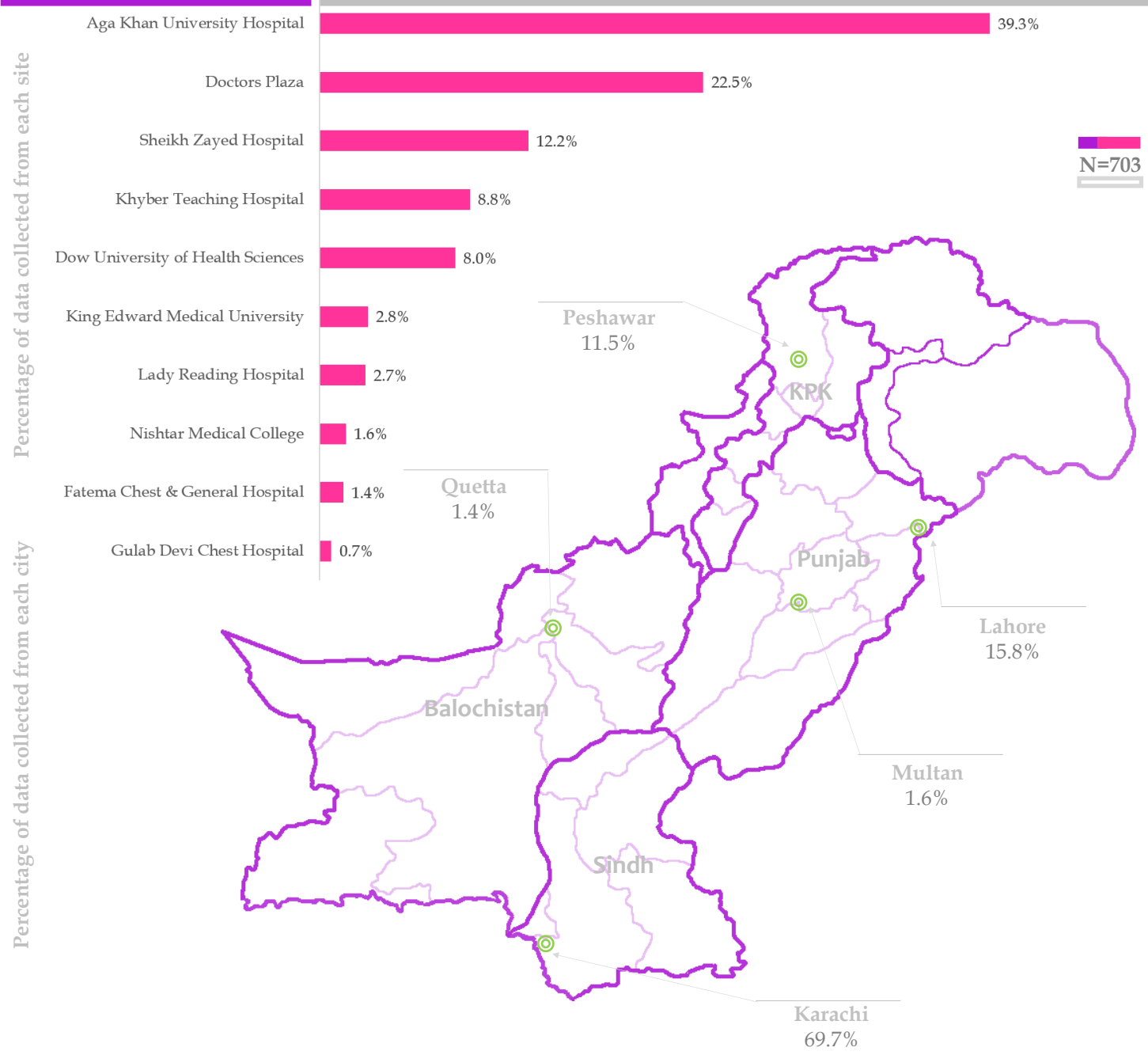
The ILD PAK Registry is an electronic database of clinical details and diagnostic investigations of patients suffering from Interstitial Lung Disease. A clinician who participates in the registry is requested to obtain approval from his respective institutional Ethics Review Board. The coordinating consultant pulmonologist is then designated as one of the Principal Investigators.

He can now depute a responsible person to fill in the online proforma according to the guidelines and the inclusion criteria. The responsible person will do this after obtaining written informed consent from the patient on the prescribed English or Urdu consent form. After uploading the data online, the supporting documents (such as PFT reports, HRCT scans, ECHO reports and so on) should be emailed to registryinfo@ildpak.com. The coordinating pulmonologist maintains a hard copy of the proforma after filling in the relevant details.

He will require a written HRCT and histopathology opinion (where applicable) from a radiologist and/or histopathologist, trained in reading HRCT and histopathology slides, relevant to ILD. It is understandably preferable to designate one or two radiologists and histopathologists to maintain uniformity in diagnostic criteria. The opinions of the additional radiologists and histopathologists can be obtained in cases of doubt, and the HRCT and biopsy specimens may be reviewed by senior radiologists/ histopathologists available in the resource pool. In such a case, all original documents will be returned after review.

A follow up form is also present in the registry to record follow up visits with investigations and medications. In case of any unforeseen event such as the death of the patient, or loss to follow up for more than six months, the participating centres will be responsible to update the registry.

SITES PARTICIPATING IN ILD PAK REGISTRY



The Aga Khan University Hospital

DOCTORS PLAZA

نشتر میڈیکل یونیورسٹی

SUMMARY

The registry data contributes significant information regarding the differential frequencies of various types of ILD. IPF/UIP was recorded as the most common type (34.3%), followed by iNSIP and HP (17.6% each). This is closely followed by CTD-ILD (15.8%) and Sarcoidosis (9.4%). With respect to gender distribution of ILDs, data showed that it mostly affected the female gender except in IPF/UIP.

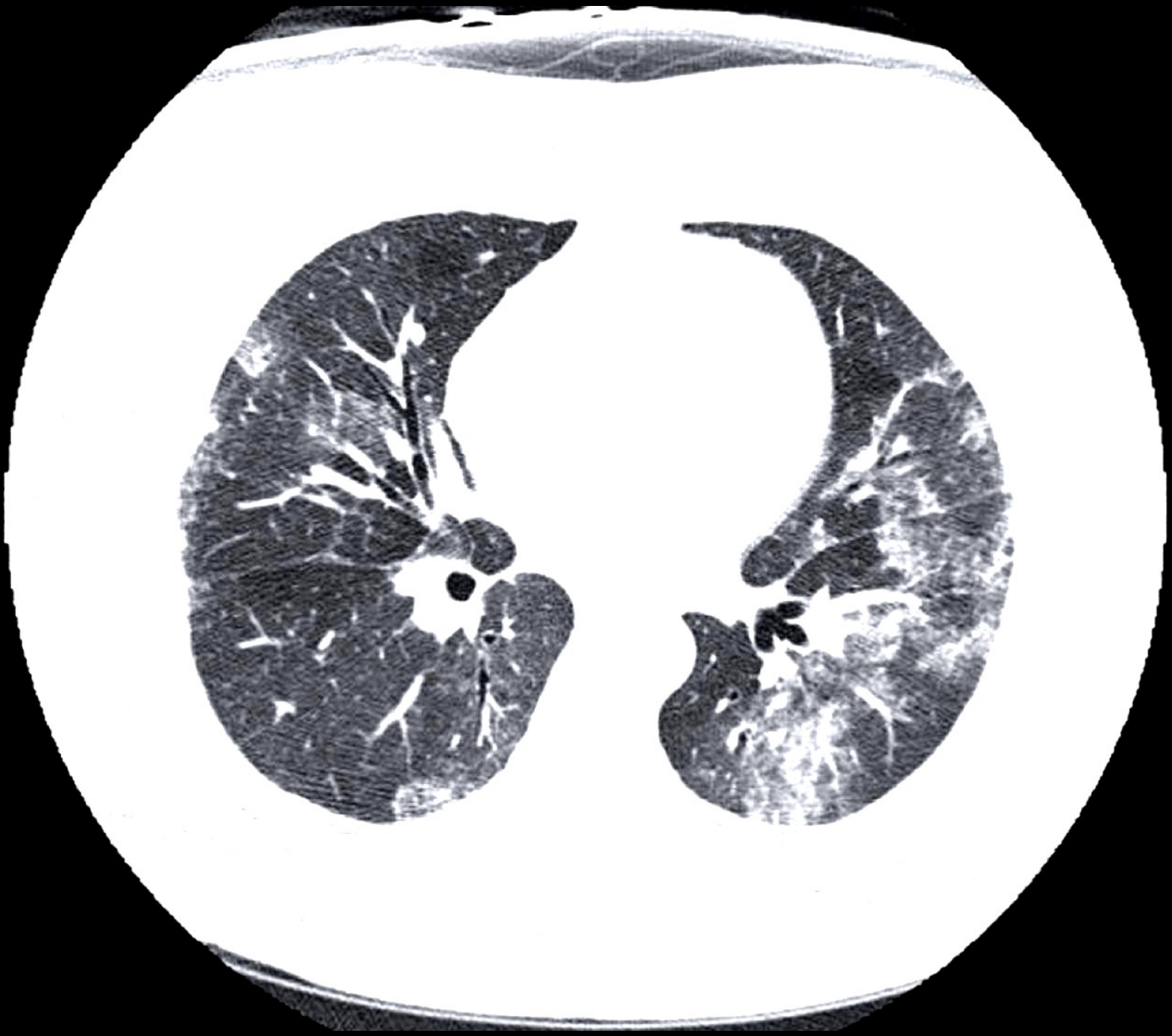
Data demonstrates that majority of cases of IPF were seen among those older than 60 years followed by 51 – 60 years. iNSIP was also noted mostly in the age group between 51 – 60 years whereas Sarcoidosis, HP and CTD-ILD were reported mostly in those who were younger than 50 years. Familial incidence was found to be positive mostly in cases of IPF/UIP and iNSIP.

Our results show that smoking history was reported more with the IPF/UIP than the rest of the ILD types. Avian was the overwhelming exposure in HP (79%) while it was also seen in other types (less than 18%). Agricultural, industrial, earthen and chemical exposures had less than 5% contribution in all ILDs. Majority of patients in our registry had an urban background (88.9%).

Cough was the most common symptom of ILD with 92.9% occurring in IPF/UIP patients followed by other types. Severe dyspnea (at rest) was observed more in fibrotic ILDs like IPF/UIP, chronic HP and iNSIP.

Mean forced vital capacity (FVC% predicted) was least compromised in Sarcoidosis (66.4%) and most compromised in chronic HP (50.4%) amongst all ILDs. Pulmonary Hypertension (PH) and Gastroesophageal Reflux Disease (GERD) had significant association with ILDs. Hypertension, Diabetes and COPD were the common comorbidities. Combined pulmonary fibrosis with emphysema (CPFE) was recognized as a distinct entity in 4.6%.

SECTION 2



Results
Epidemiology

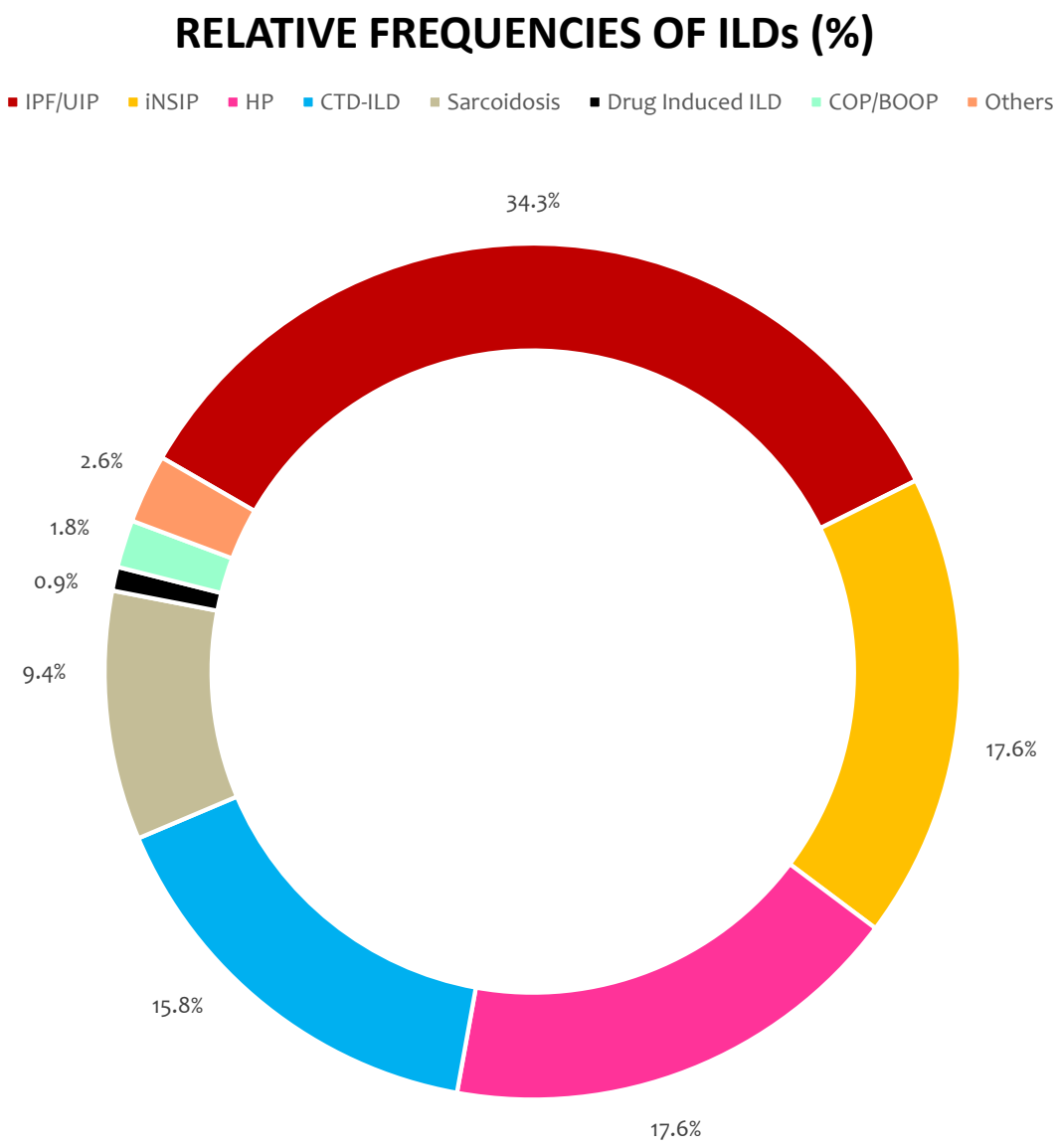


Figure 1: Percentage of different Interstitial Lung Diseases in the Pakistani Populace

Figure 1 shows that the highest burden of ILD was contributed by 34.3% of IPF/UIP, followed by 17.6% of iNSIP and HP each. This is closely followed by the CTD-ILD 15.8% and Sarcoidosis 9.4%.

GENDER DISTRIBUTION

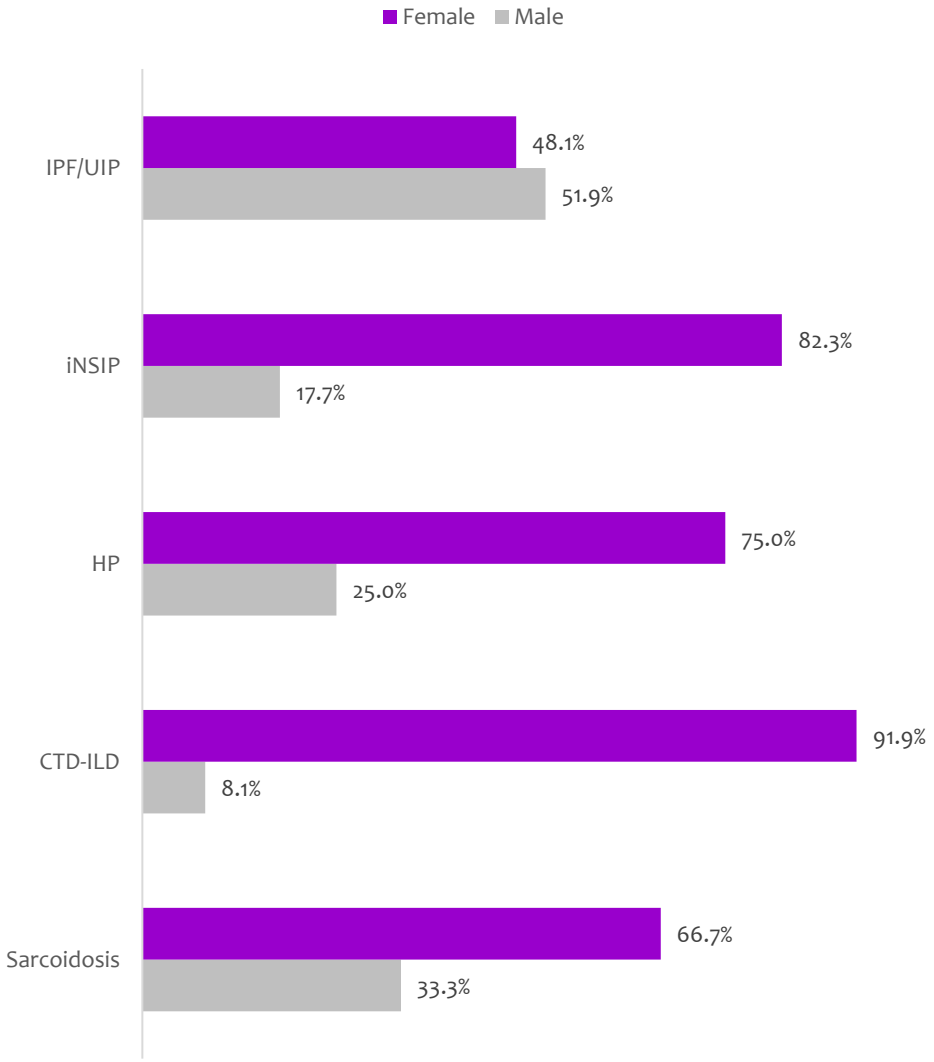


Figure 2: Gender distribution of ILD

Figure 2 shows that ILDs mostly affected the female gender in comparison with the males except in IPF/UIP.

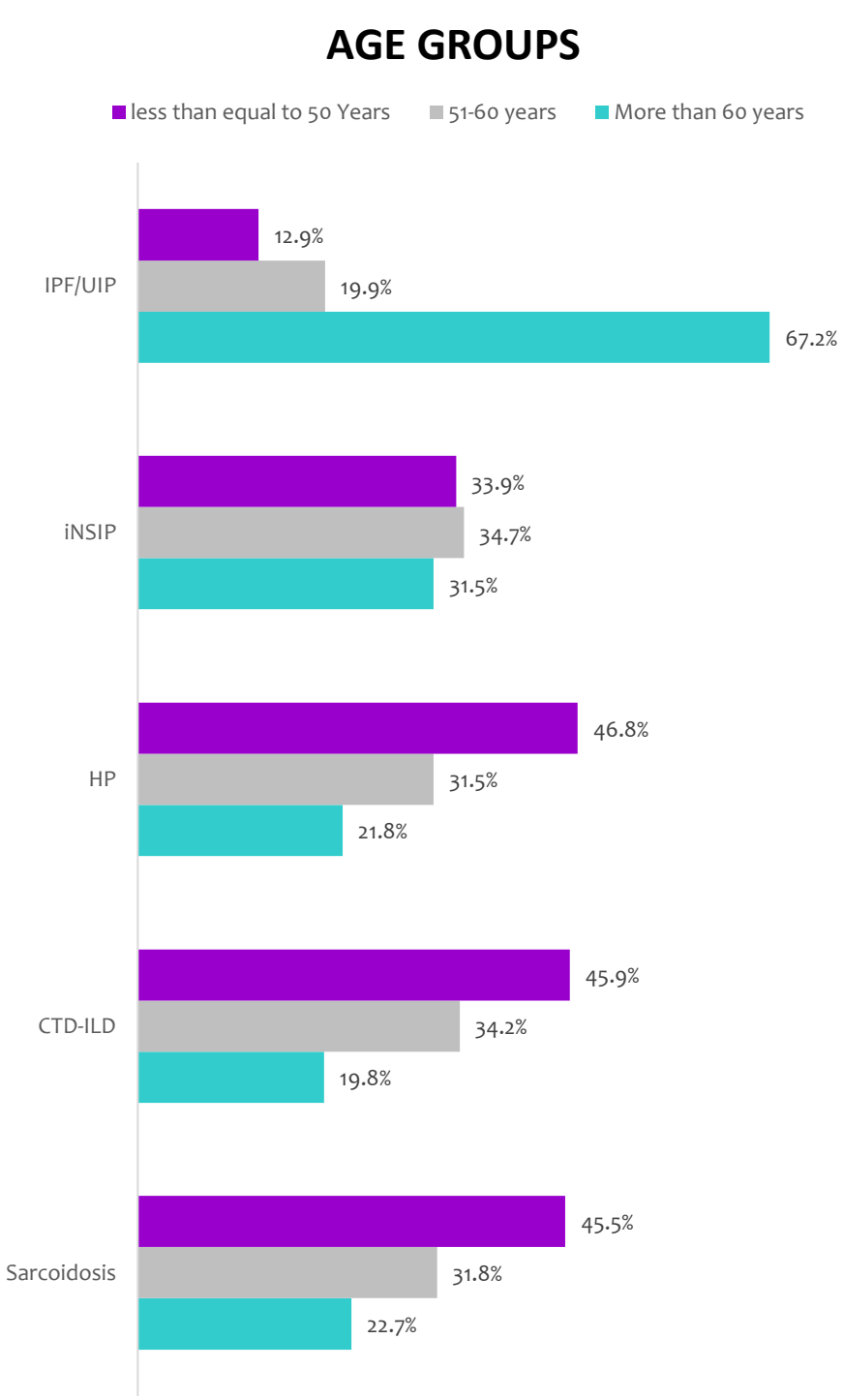


Figure 3: Distribution of ILD in relation with Different Age Groups

Figure 3 shows that majority of cases of IPF seen among those older than 60 years followed by those between 51 – 60 years. iNSIP was also noted mostly in the age group between 51 – 60 years. Sarcoidosis, HP and CTD-ILD were reported mostly among those who were younger than 50 years.

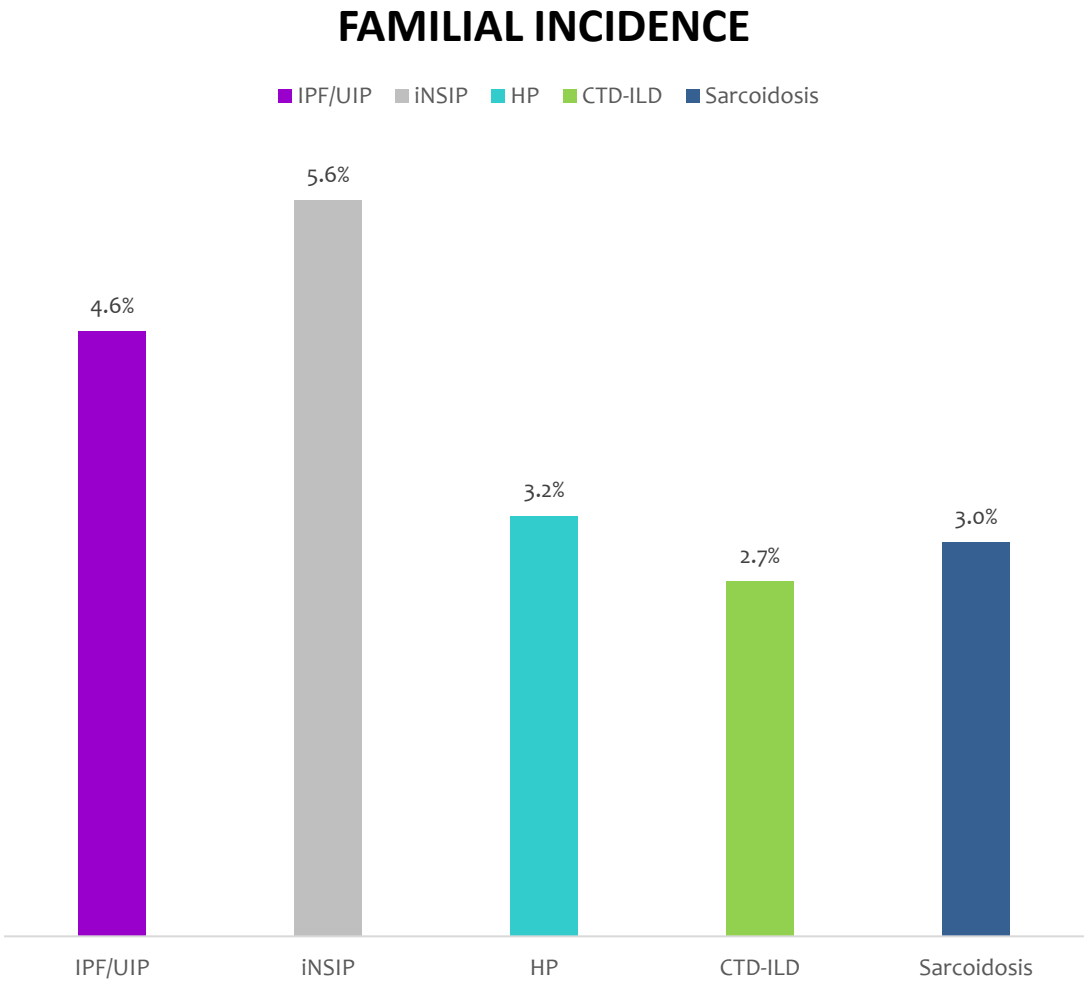


Figure 4: Interstitial Lung Diseases incidence in families

Figure 4 shows that a family incidence rate was positive in 5.6% of iNSIP, 4.6% in case of IPF/UIP, 3.2% in HP, 3.0% in Sarcoidosis and 2.7% in CTD-ILD.

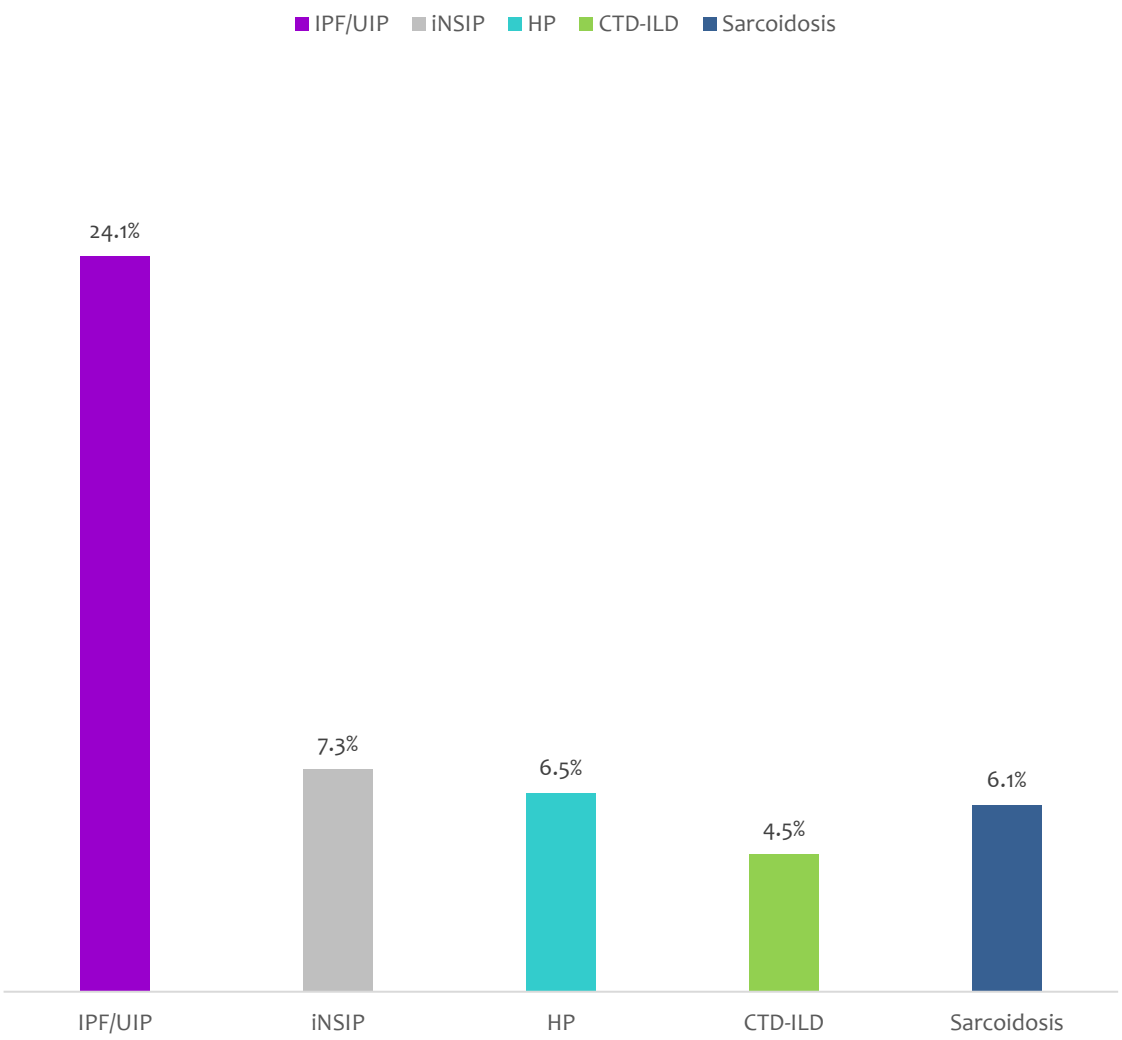


Figure 5: Distribution of Smokers in Interstitial Lung Diseases

Figure 5 shows that among the ILD affected populace those having smoking history were at highest incidence of IPF/UIP with 24.1% followed by iNSIP with 7.3%, HP at 6.5%, Sarcoidosis at 6.1% and CTD-ILD at 4.5%.

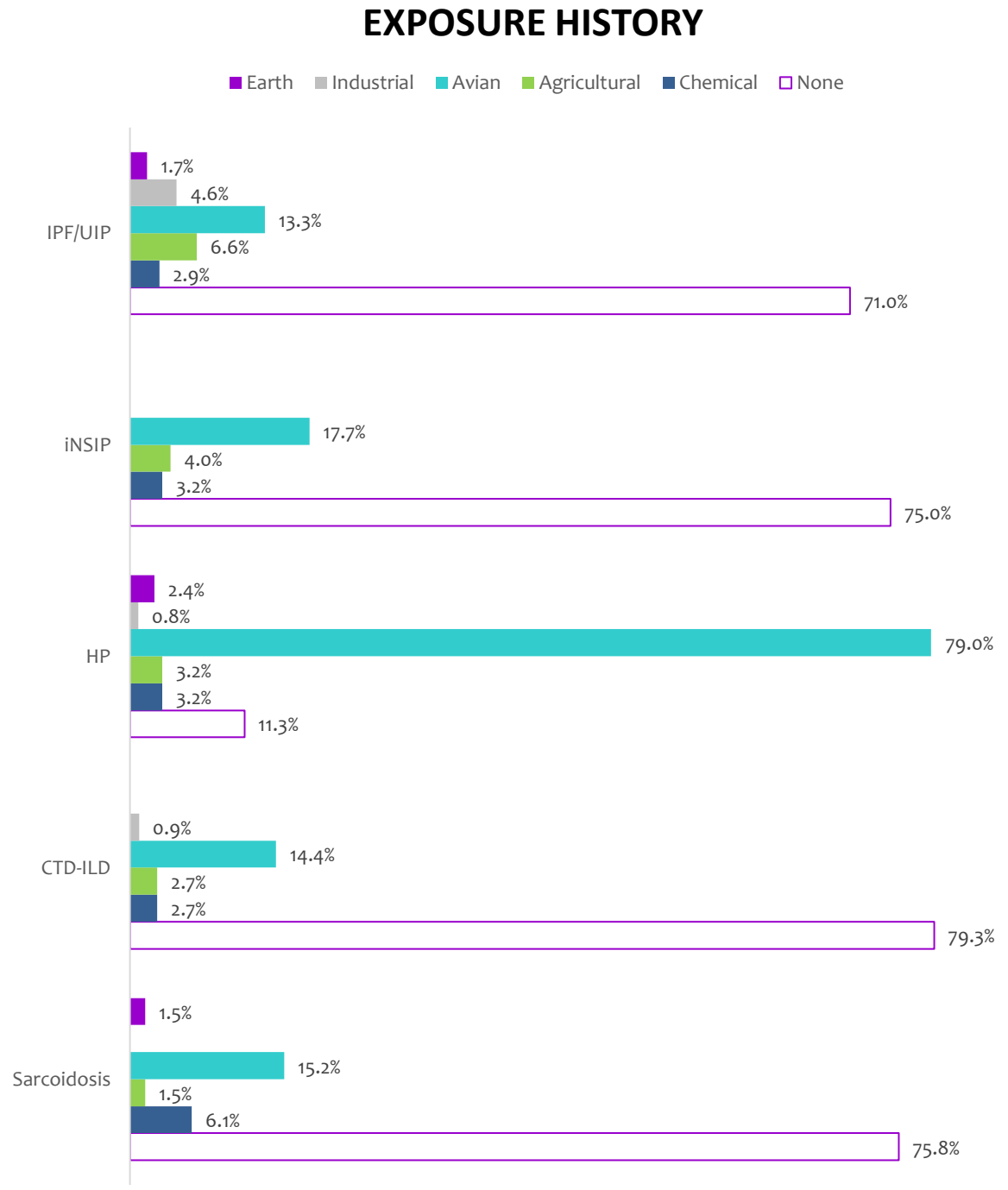


Figure 6: Exposures in Different Interstitial Lung Diseases

Figure 6 shows that no exposures was noted in majority of IPF/UIP with 71.0%, iNSIP with 75.0%, CTD-ILD at 79.3%, Sarcoidosis at 75.8%. However, in HP significant number of cases are due to avian exposure with 79.0%.

OCCUPATION

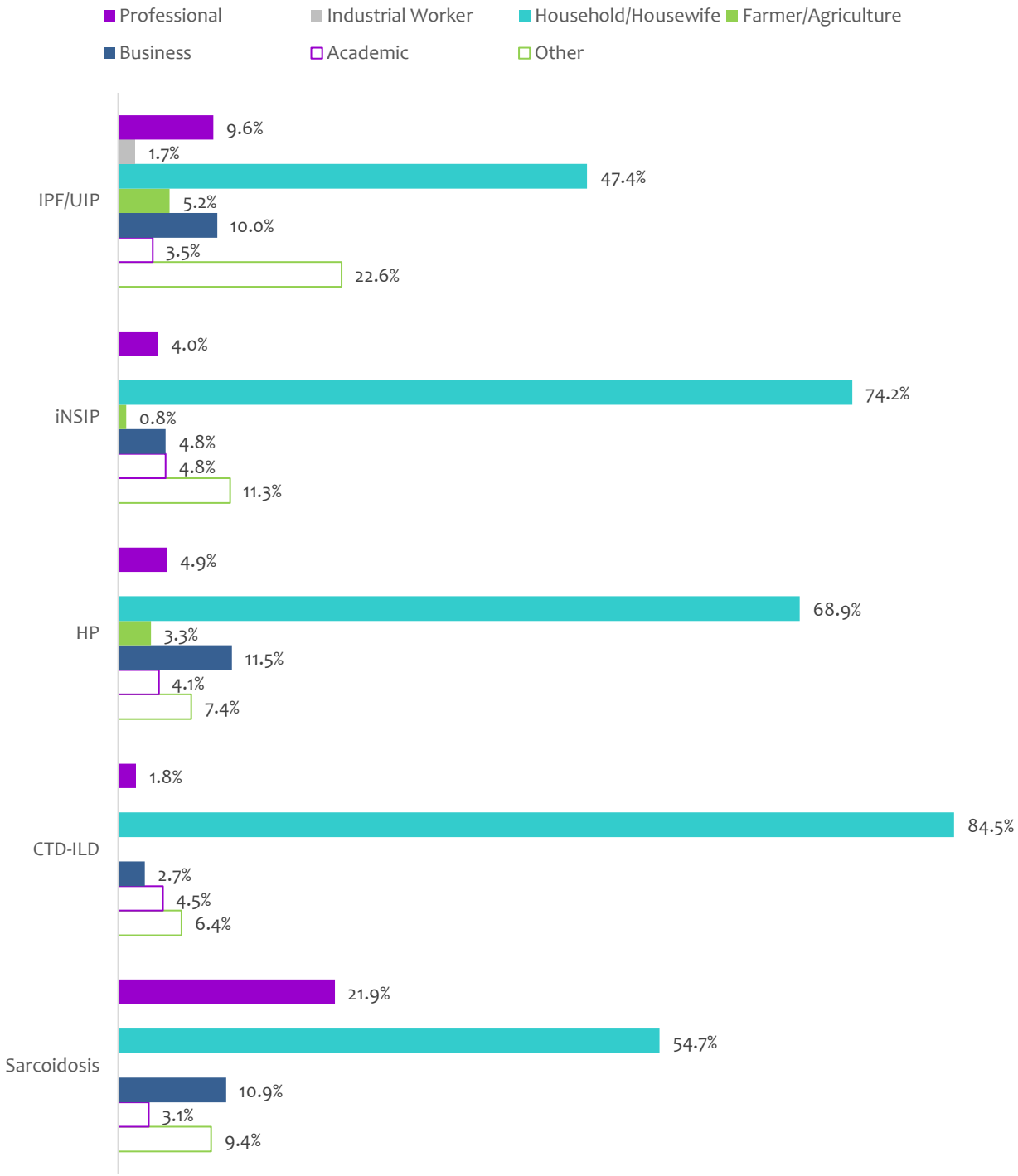


Figure 7: Occupational breakdown of ILD Cases

Figure 7 shows a predominance of affected housewives in comparison with the professionals, academician, agriculturalist and industrial workers.

GEOGRAPHICAL LOCATIONS

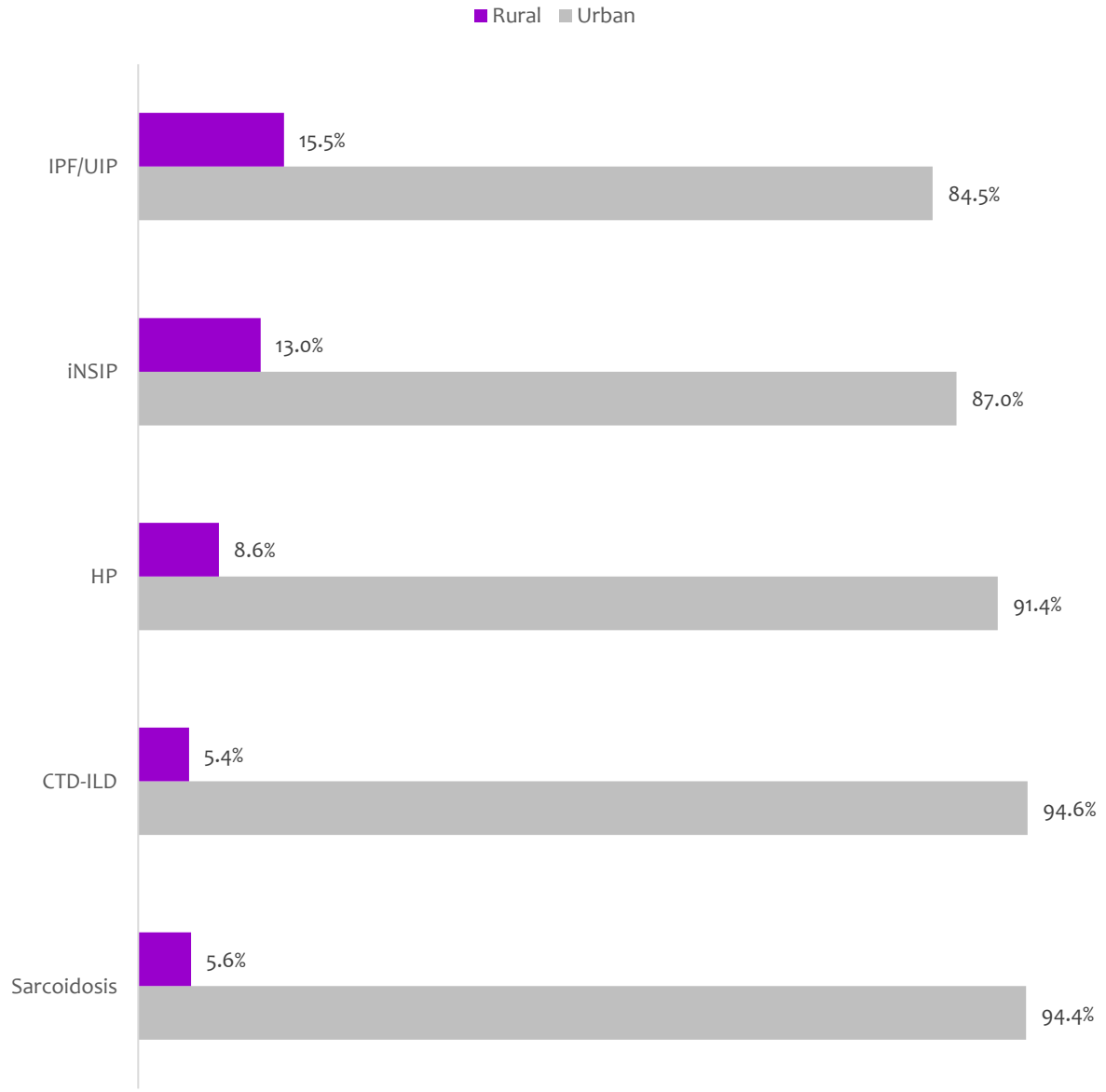


Figure 8: Disease Reporting in Geographical Locations

Figure 8 shows predominant affectees were from the urban areas in comparison with the rural areas.

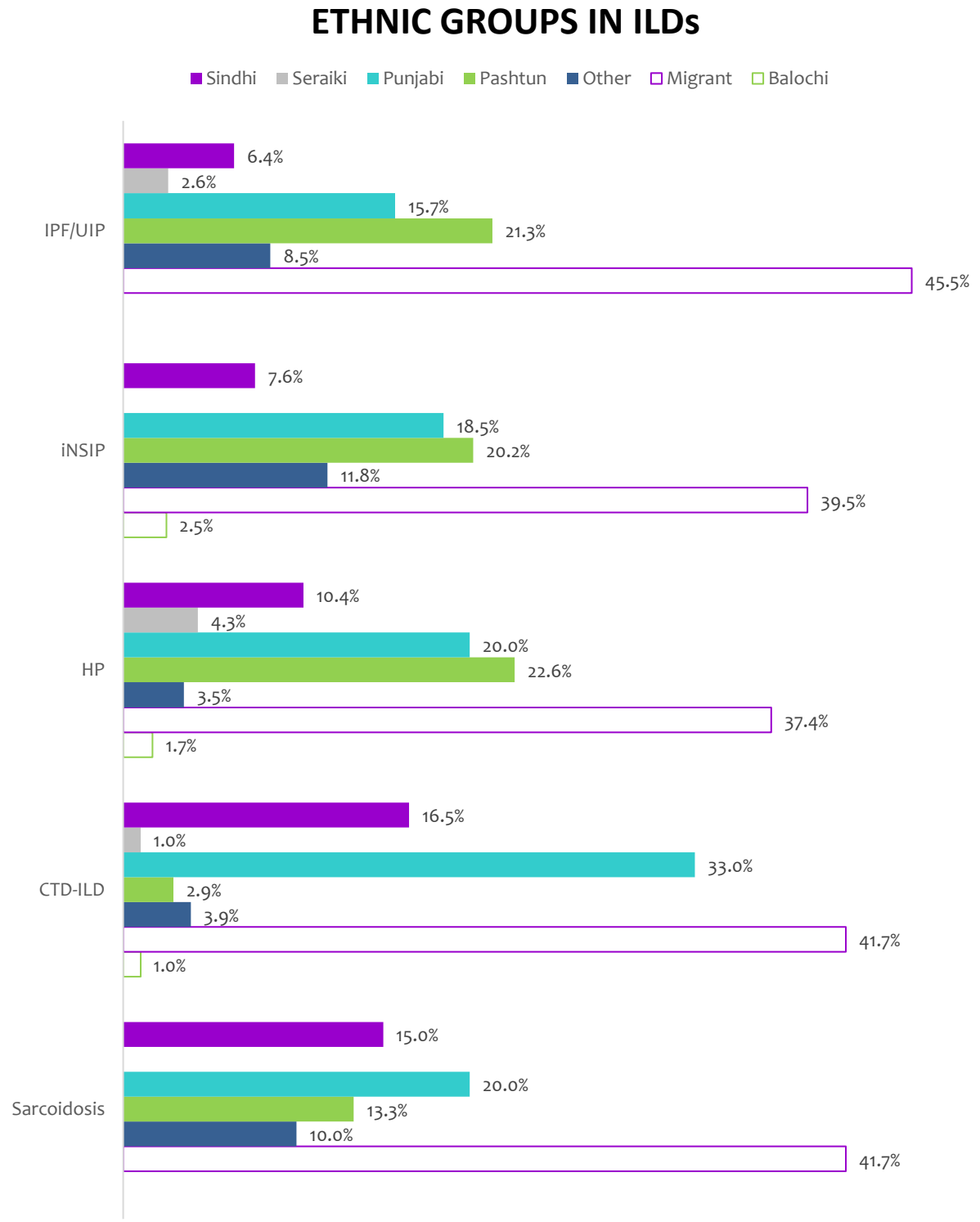


Figure 9: Frequency of Ethnic Groups in Different Interstitial Lung Diseases

Figure 9 shows that among reporting ethnic groups migrants pre-dominated in all ILDs followed by Pashtoons and Punjabis. Likewise, 45.5% of the IPF cases were seen in Migrants while 15.7% of Punjabis, 21.3% of Pashtuns and 6.4% of Sindhis. Migrants include people originally from Central, Southern and Western region of Sub-Continent.

Results
Clinical features on presentation

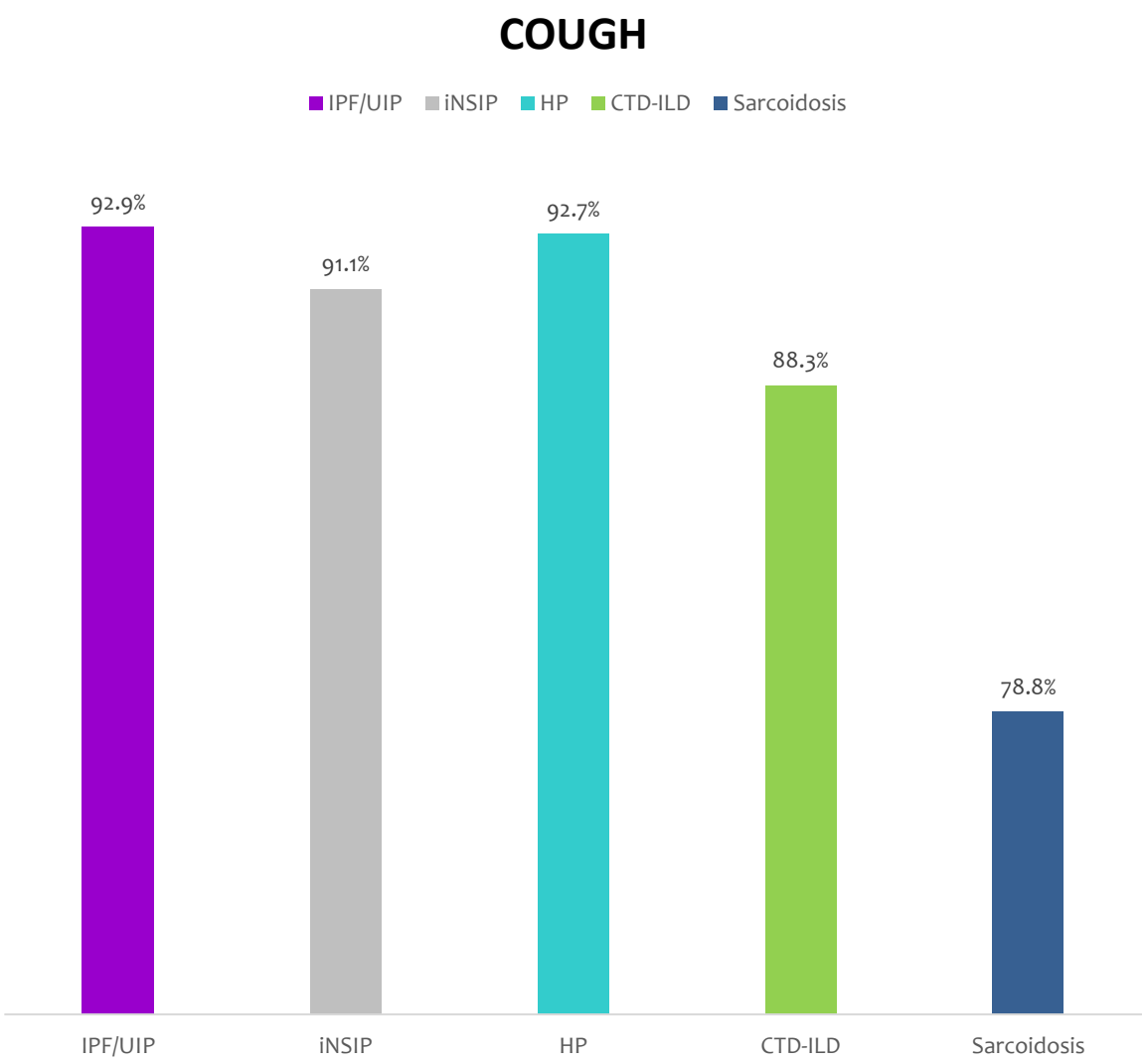


Figure 10: Cough in Different Interstitial Lung Diseases

Figure 10 shows that interstitial Lung Diseases was mainly characterized by cough with 92.9% occurrence among IPF/UIP patients, 92.7% in HP patients & 91.1% among the iNSIP group. While 88.3% and 78.8% patients belonging to CTD-ILD and Sarcoidosis groups respectively.

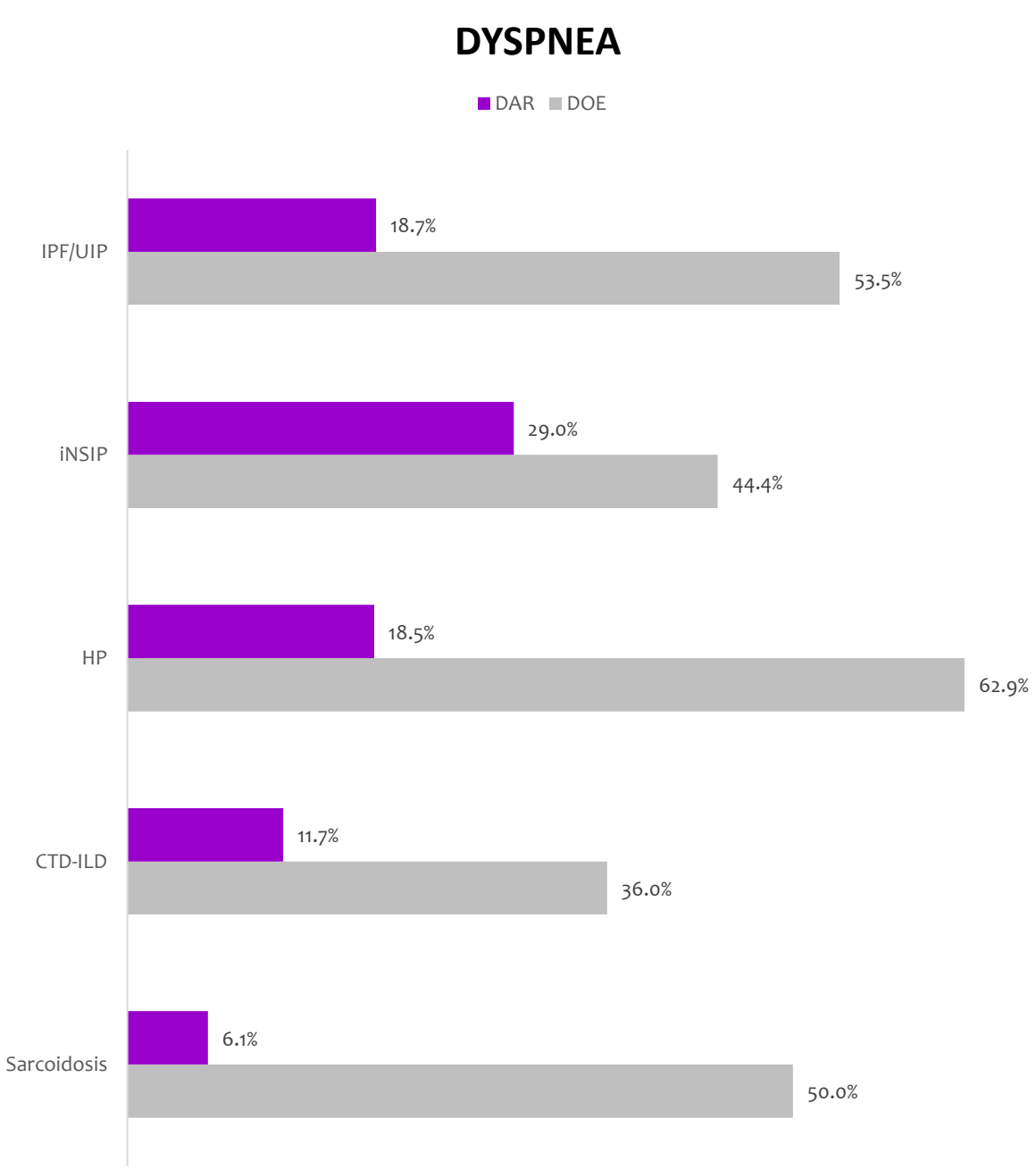


Figure 11: Dyspnea in Different Interstitial Lung Diseases

Figure 11 shows that characteristically dyspnea triggered on exertion in ILD patients. IPF/UIP had 53.5% cases with dyspnea on exertion (DOE) and 18.7% who were dyspneic even at rest (DAR). iNSIP had DOE in 44.4% and DAR in 29.0% cases. HP had DOE in 62.9% cases and DAR in only 18.5% cases. CTD-ILD had DOE in 36.0% of the cases and DAR in 11.7% and Sarcoidosis had DOE in 50.0% cases.

CREPITATIONS

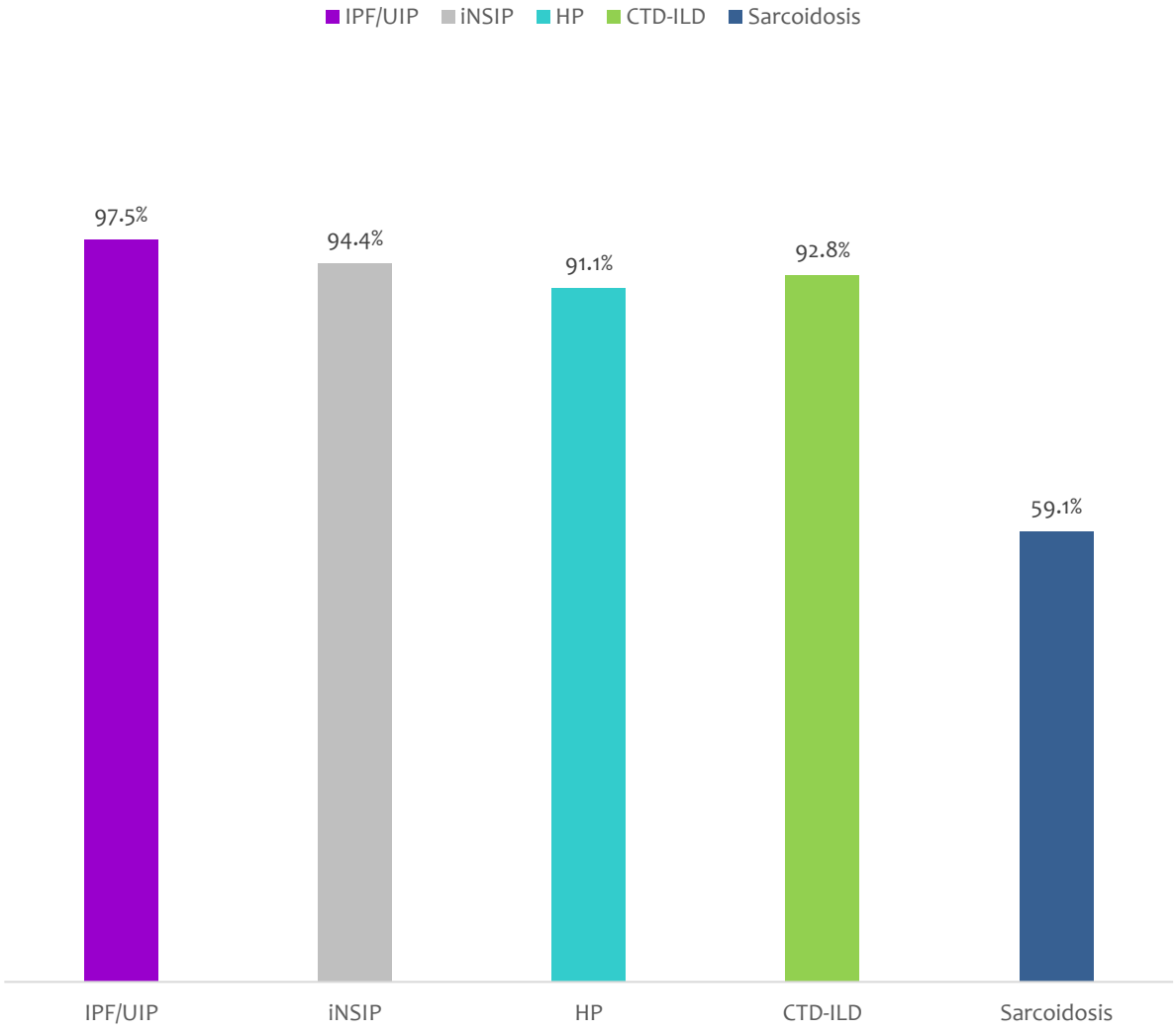


Figure 12: Crepitation's in Patients with Interstitial Lung Disease

Figure 12 shows that crepitations were observed in 97.5% cases of IPF/UIP, iNSIP 94.4%, CTD-ILD 92.8%, HP 91.1% and 59.1% among Sarcoidosis patients.

CLUBBING

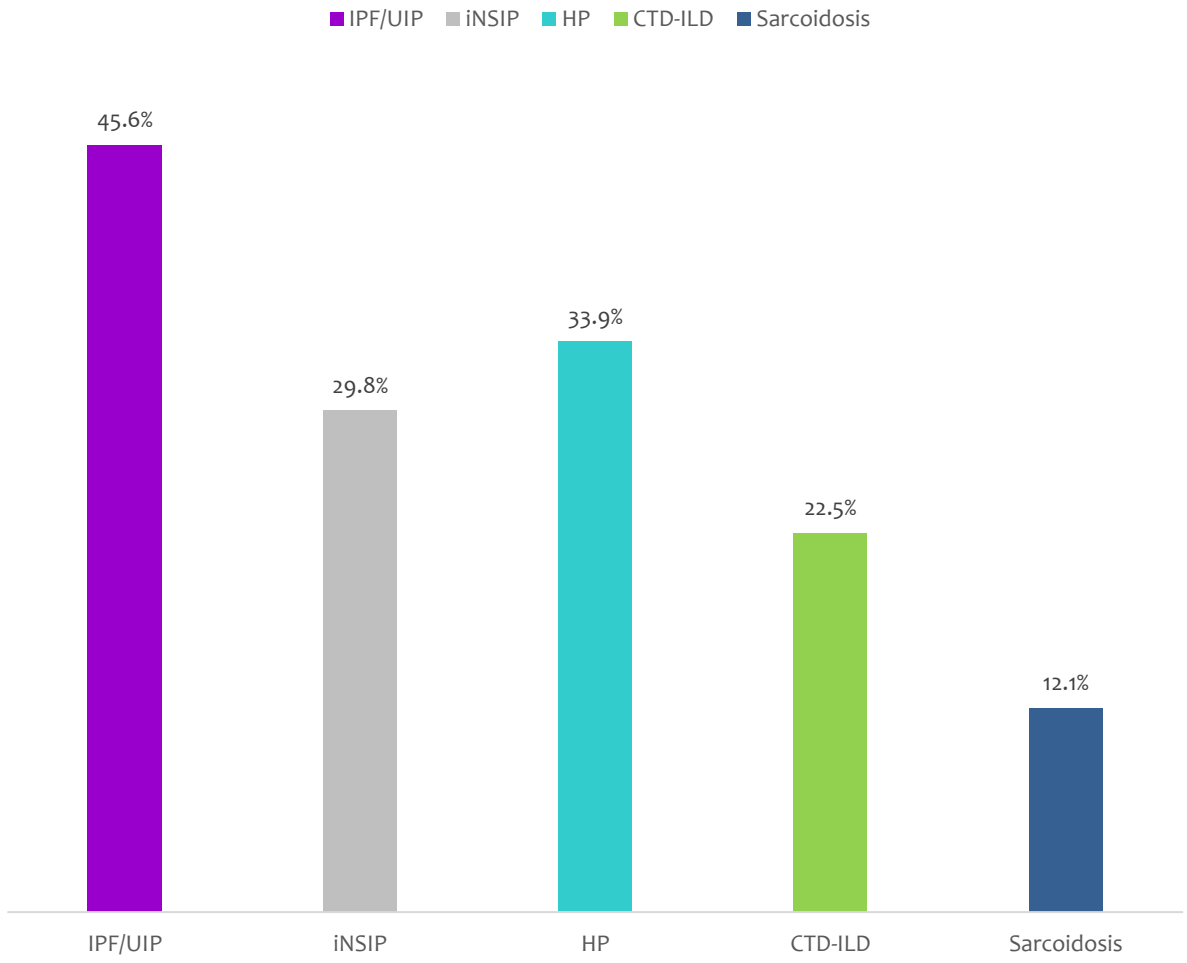


Figure 13: Clubbing in Patients with Interstitial Lung Disease

Figure 13 shows that clubbing was observed in 45.6% cases of IPF/UIP, 33.9% of HP, 29.8% of iNSIP patients, 22.5% of the CTD-ILD and the least in Sarcoidosis i.e. 12.1%.

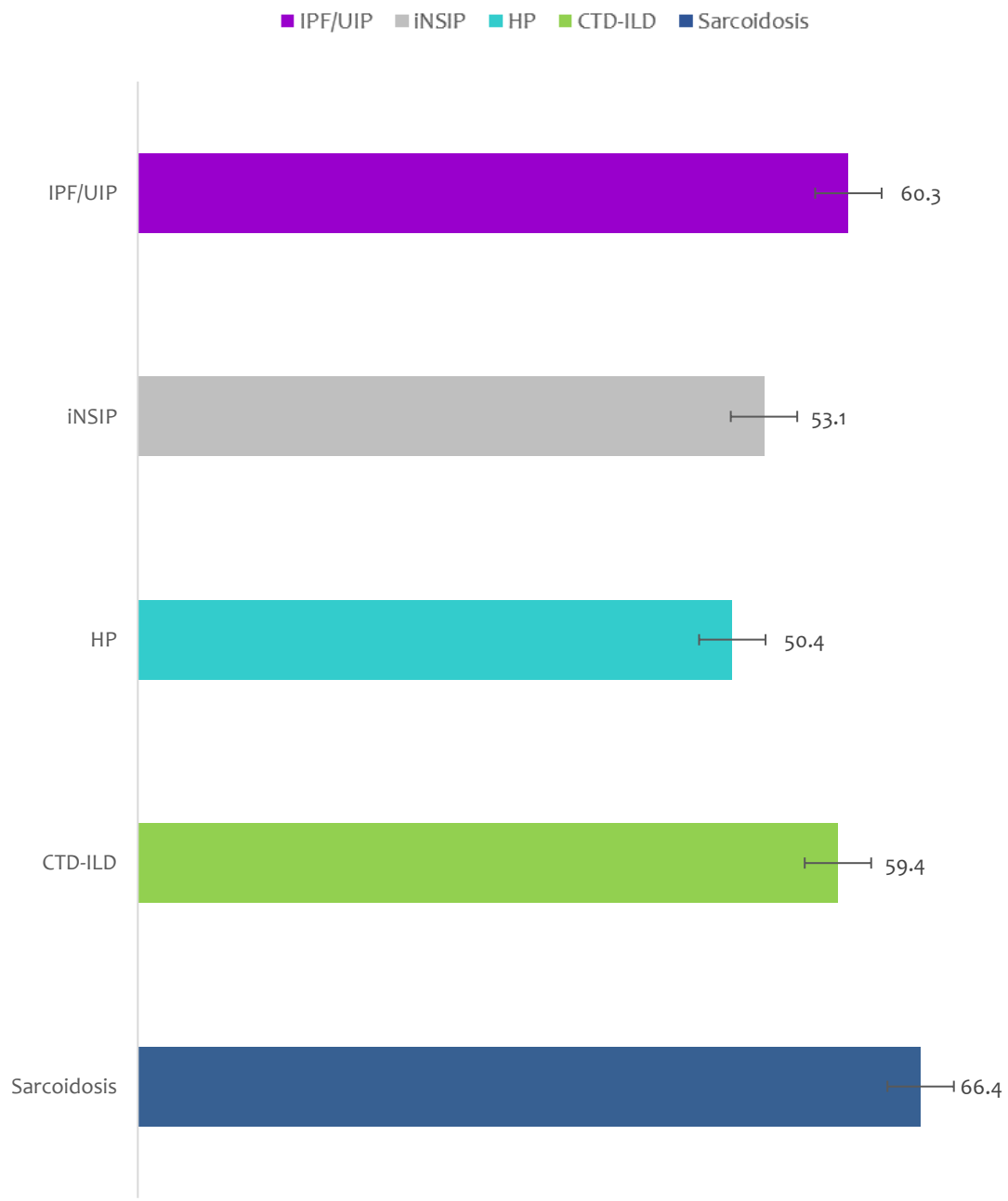


Figure 14: Mean Forced vital capacity (% of predicted) in the Patients of Interstitial Lung Diseases
Figure 14 shows that FVC (as % of predicted) was 66.4±22.3 (Mean ± SD) in the case of Sarcoidosis while it was 60.3± 19.1 (Mean ± SD) in the case of IPF/UIP. It was 59.4±21 (Mean ± SD) in the case of CTD-ILD and similarly it was 53.1±22.2 (Mean ± SD) in iNSIP and 50.42±17.7 (Mean ± SD) in the case of HP.

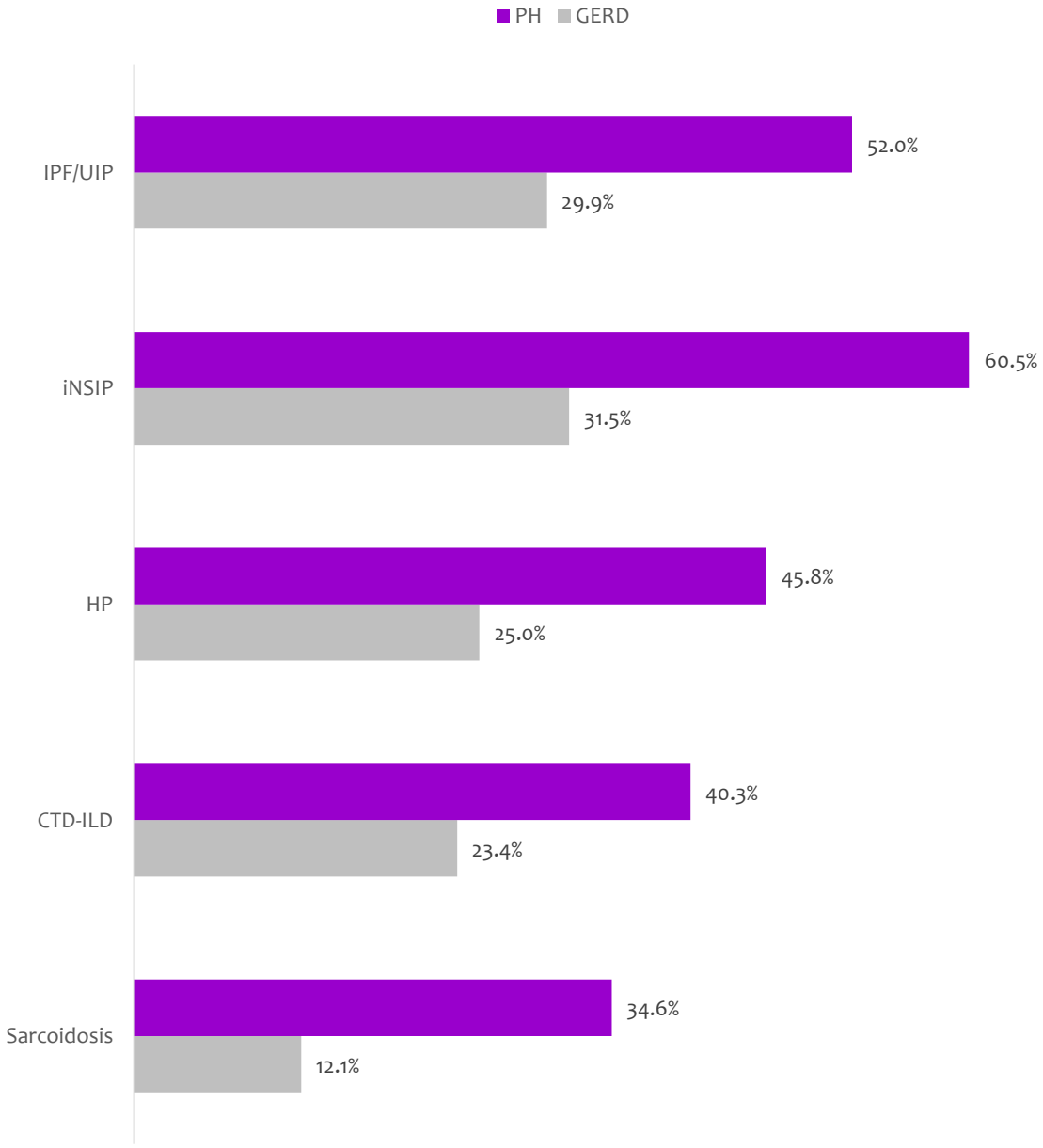


Figure 15: Association of ILD with Pulmonary Hypertension (PH) and Gastroesophageal Reflux Disease (GERD)

Figure 15 shows that 52% of the IPF/UIP cases were associated with PH and 29.9% with GERD. 60.5% of the iNSIP cases were associated with PH and 31.5% with GERD. 34.6% of the Sarcoidosis cases were associated with PH and 12.1% with GERD. 45.8% of the HP cases with PH and 25% with GERD. 40.3% of the CTD-ILD cases with PH and 23.4% with GERD.

COMORBIDITIES

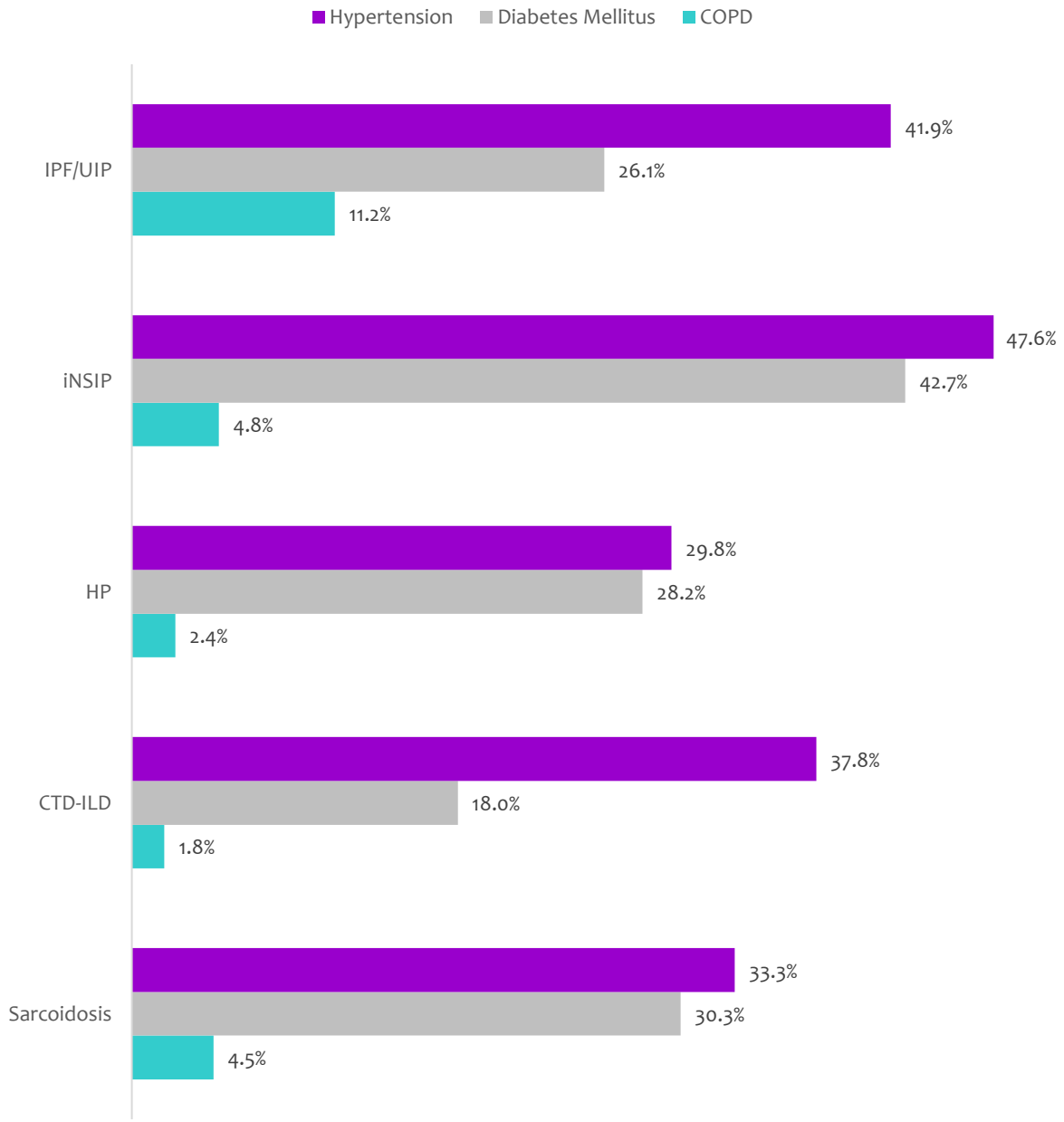
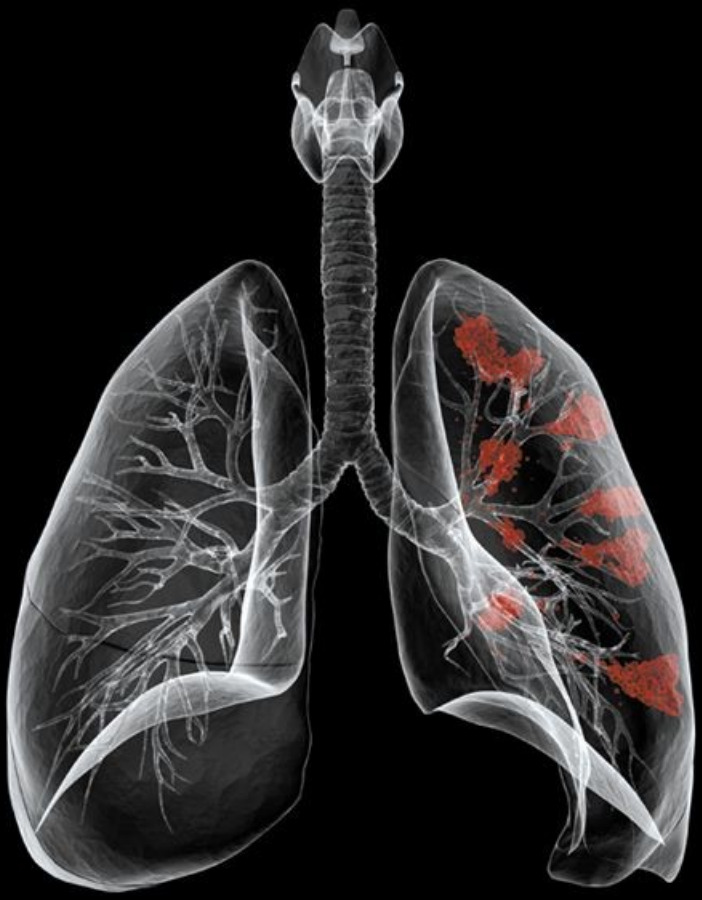


Figure 16: Comorbidities with Interstitial Lung Disease

Figure 16 shows different comorbid conditions that had been observed in the patients of ILD where hypertension was the most common. IPF/UIP had hypertension in 41.9% of the cases while Diabetes and COPD in 26.1% and 11.2% respectively. Likewise, Sarcoidosis had hypertension in 33.3% of the cases, Diabetes in 30.3% and COPD in 4.5%. Similarly, CTD-ILD patients had hypertension in 37.8% of the cases along with 18% of diabetes and 1.8% COPD. Again, hypertension was observed in 47.6% along with 42.7% of diabetes and 4.8% of COPD in the iNSIP patients. Hypertension had been observed in 29.8% along with 28.2% of diabetes and 2.4% of COPD in HP.

SECTION 3



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ABBREVIATIONS

AP	Alveolar Protienosis
COP	Cryptogenic Organizing Pneumonia
COPD	Chronic Obstructive Pulmonary Disease
CVD	Collagen Vascular Diseases
DMARDs	Disease Modifying Anti Rheumatic Drugs
DPLD	Diffuse Parenchymal Lung Disease
FVC	Forced Viral Capacity
GERD	Gastroesophageal Reflux Disease
GPA	Granulomatosis with Polyangiitis
HP	Hypersensitivity Pneumonitis
HRCT	High Resolution Computed Tomography
ILD	Interstitial Lung Disease
IPF	Idiopathic Pulmonary Fibrosis
LAM	Lymphangioleiomyomatosis
LIP	Lymphocytic Interstitial Pneumonia
MCTD	Mixed Connective Tissue Disorder
iNSIP	Idiopathic Non-Specific Interstitial Pneumonia
PFT	Pulmonary Function Tests
PH	Pulmonary Hypertension
RA	Rheumatoid Arthritis
RB-ILD	Respiratory Bronchiolitis-ILD
SLE	Systemic Lupus Erythomatosis
SS/SCL	Systemic Sclerosis/Scleroderma

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