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INTERSTITIAL LUNG DISEASE

Interstitial Lung Disease ILDPAK
Right step to a better breath

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6 YEAR ILD PAK REGISTRY REPORT

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Interstitial Lung Disease

6 Year Registry Report

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MESSAGE

The Pakistan Chest Society has reasons to feel proud of two recent important contributions in the understanding of Interstitial lung diseases in Pakistan.

In April 2016, the PCS ILD Guideline Committee finalized a robust "Guideline Document on the Diagnosis and Management of ILD in Pakistan" which was circulated in the 2016 PCS Chest Con in Lahore. This is a comprehensive document which would help our fellows in learning to understand the HRCT and workup of ILDs especially IPF.

The second contribution is the PCS motivated ILD PAK Registry that has a great potential of integrating extensive information data on the subject. It makes use of more than a hundred variables available in the software of its electronic registry that is able to provide access to a large number of sites across the country.

The present six year registry report will give you a close and interesting insight of the status and profile of ILD from a few centers of one large city. The sites in KPK and lower Punjab would be coming up with their reports very soon. The next biennial registry report would hopefully present a larger geographic canvas and maybe more interesting results.

I am proud of the enduring spirit of the researchers involved in this project.

Prof. Arshad Javaid

President Pakistan Chest Society

Chairman PCS Guideline Committee

PREFACE

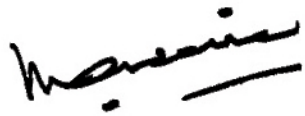
Epidemiological studies have always been vitally important in assessing the extent of a disease in a population group and identifying its various facets and proclivities so that better management strategies could be devised.

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 multicenter 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 this registry was presented earlier in scientific conferences (ERS 2012 Vienna, ACCP-SEPAR 2014 Madrid, CWC 2016 Shanghai) [10-12]. This is the first comprehensive 6 year 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.



Dr Mosavir Ansarie
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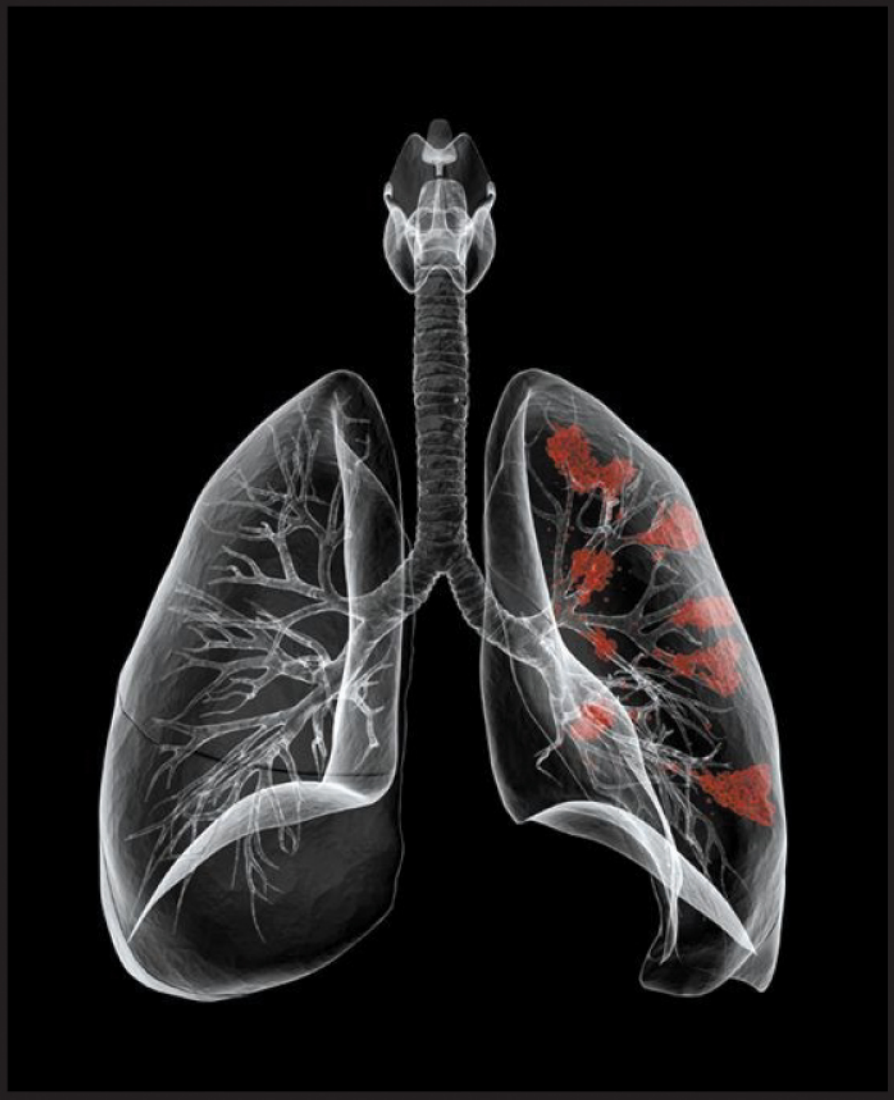
SECTION 1



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.

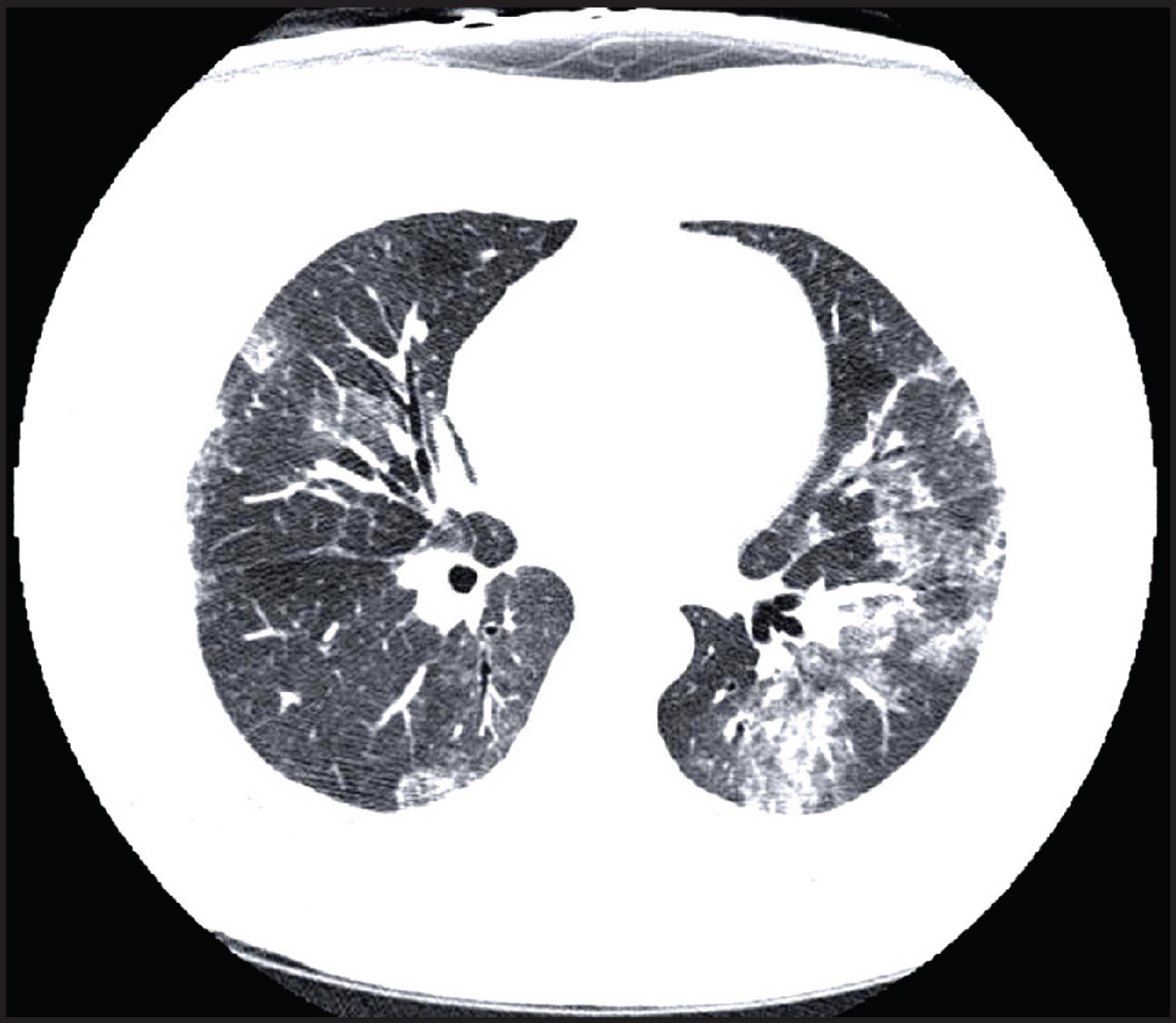
He can now depute a responsible person to fill in the online proforma according to the guidelines and the inclusion criteria. The responsible person will do this after obtaining written informed consent from the patient on the prescribed English or Urdu consent form. After uploading the data online, 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

SECTION 2



Results

ILD Epidemiology

(n=325)

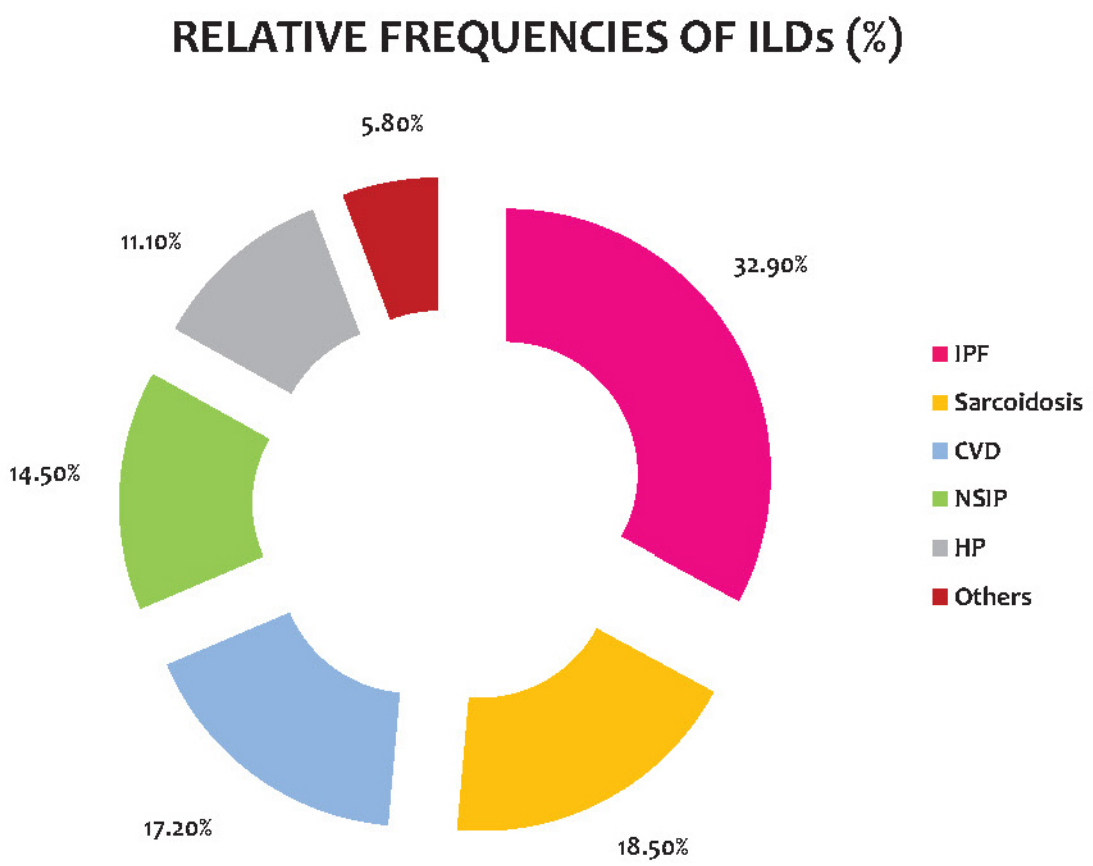


Figure 1: Percentage of different Interstitial Lung Diseases in the Pakistani Populace

Figure 1 shows that the heaviest burden of ILD was contributed by 32.90% of IPF, second was 18.50% of Sarcoidosis, third was 17.20% of CVD Associated, fourth was 14.50% of NSIP, fifth was 11.10% of HP and the rest was provided by 5.80% of other ILDs which are COP, AP, LAM, LIP, RB--ILD and drug induced.

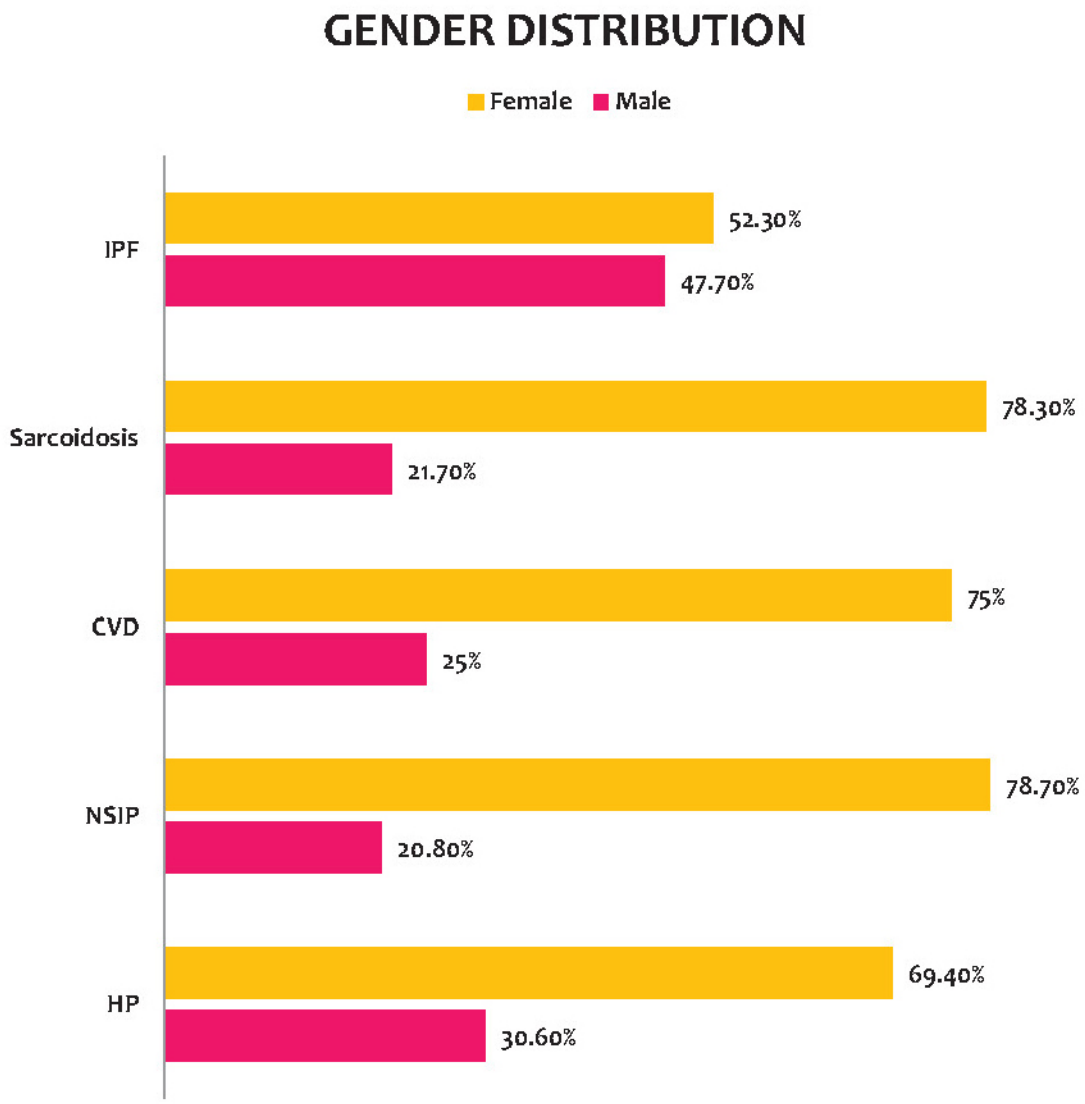


Figure 2: Gender wise distribution of ILD

Figure 2 shows that ILDs greatly burdened the female gender in relation to the males except in IPF where it was almost equal for the two genders. IPF burdened the females by 52.30% and males by 47.70%. Sarcoidosis prominently involved a big percentage of females 78.30% while males were 21.70%. CVD affected 75% females and 25% males while NSIP affected 78.70% women and 20.80% men.

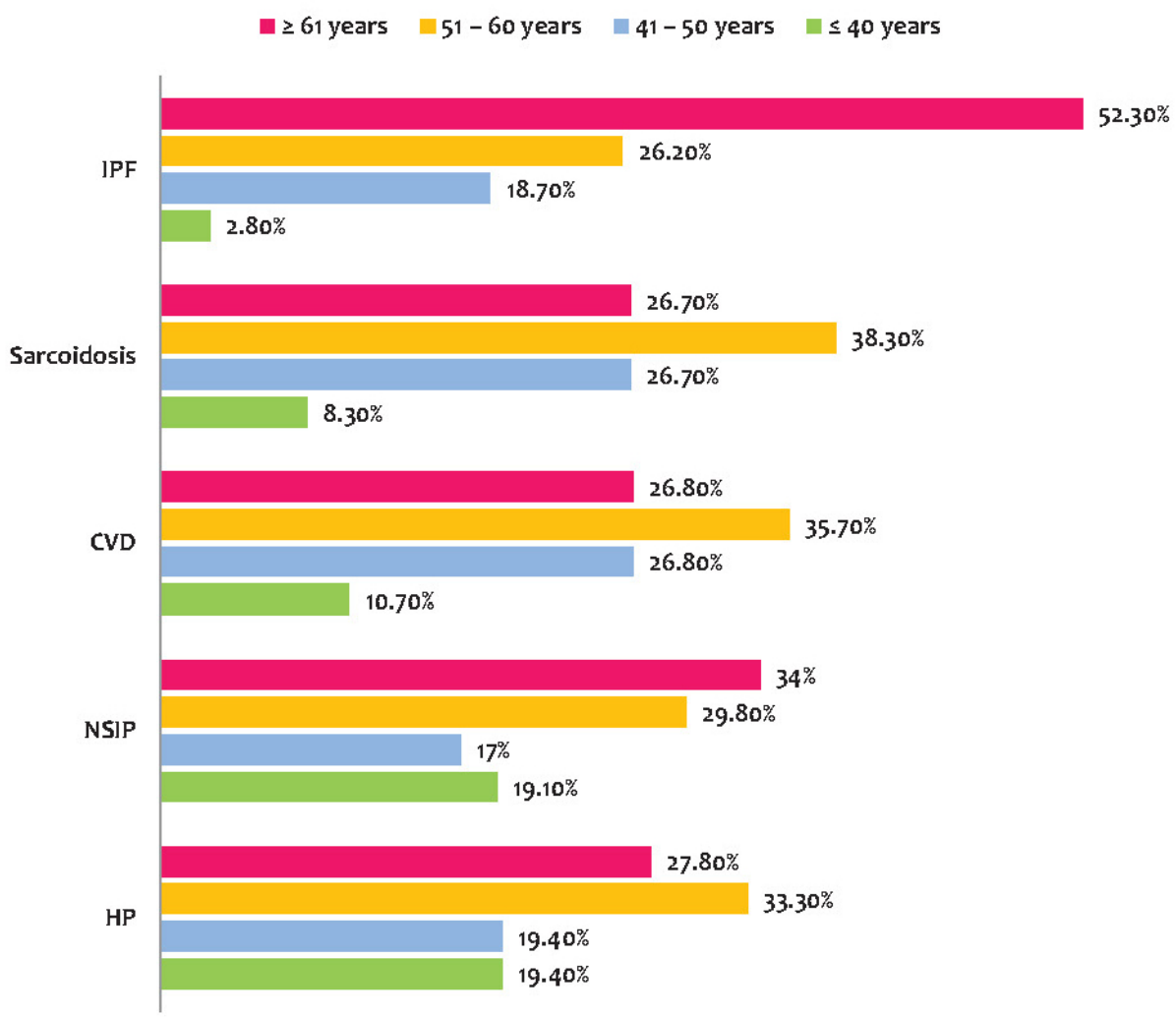


Figure 3: Distribution of ILD in relation with Different Age Groups

Figure 3 shows that IPF greatly affected the age group that was older than 60 years by 52.30%, then people of 51 – 60 years by 26.20%, 41 – 50 years by 18.70% and lastly people of and younger than 40 years, by 2.80%. Sarcoidosis substantially affected the age group of 51 – 60 years by 38.30%. The two groups older than 60 years & those between 41 – 50 years shared 26.70% each while younger than 40 years' group was 8.30%. CVD affected the age group of 51 – 60 years by 35.70% the two groups older than 60 years as well as of 41 – 50 years by 26.80% each while those younger than 40 years were 10.70%. NSIP posed the heaviest burden on the age group of and older than 61 years by 34%, then on people of 51 – 60 years by 29.80%, people of and younger than 40 years by 19.10% and 41 – 50 years by 17%. HP mainly affected the age group of 51 – 60 years by 33.30%, then people of and older than 61 years by 27.80% and both age groups 41 – 50 and younger than 40 years were 19.40% each.

FAMILIAL INCIDENCE

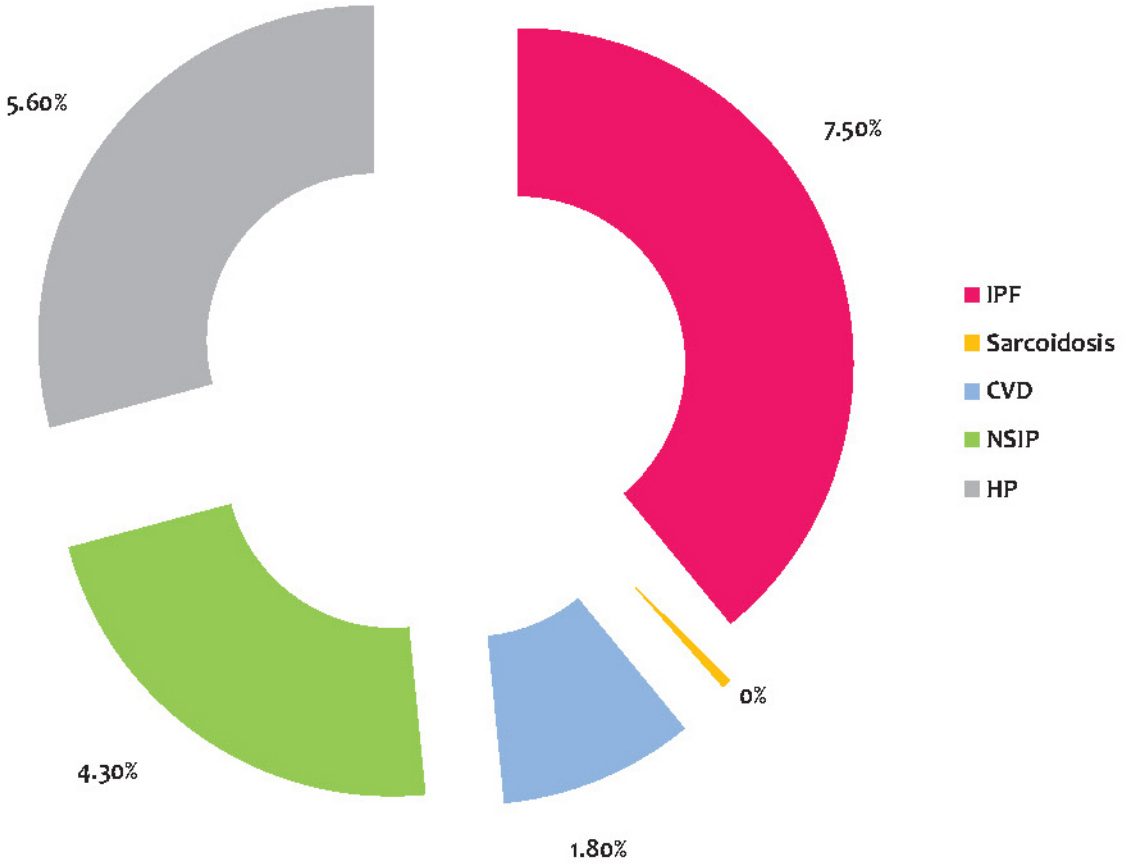


Figure 4: Interstitial Lung Diseases incidence in families

Figure 4 shows that a family history was positive in 7.50% of IPF, HP had 5.60%, NSIP 4.30% and CVD with 1.80%. No familial incidence was noted in Sarcoidosis.

SMOKING HISTORY

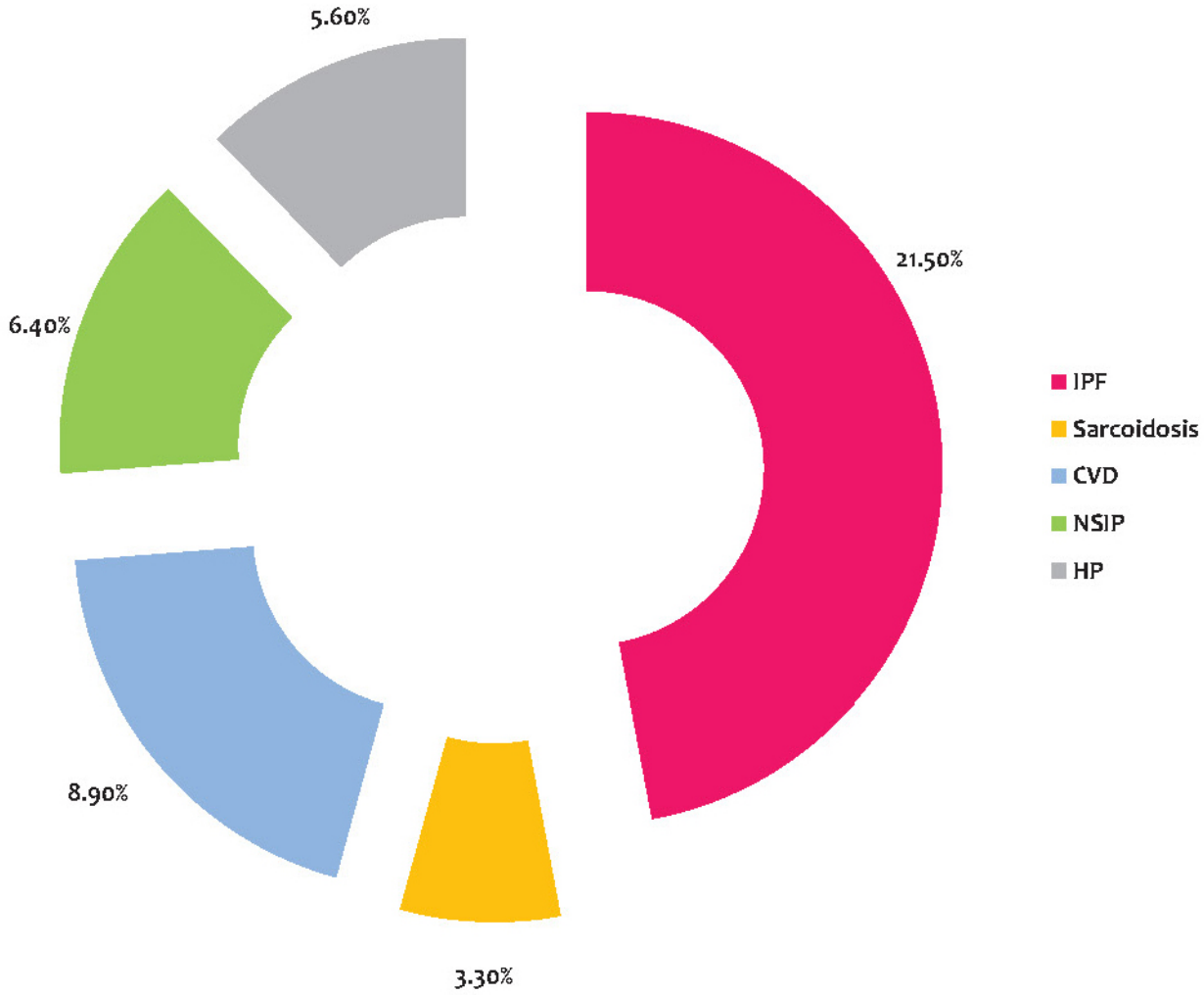


Figure 5: Distribution of Smokers in Interstitial Lung Diseases

Figure 5 shows that among the ILD affected populace, specifically involved in smoking the highest incidence was in IPF 21.50%. CVD 8.90%, NSIP 6.40%, and HP 5.60% while in Sarcoidosis 3.30% were smokers.

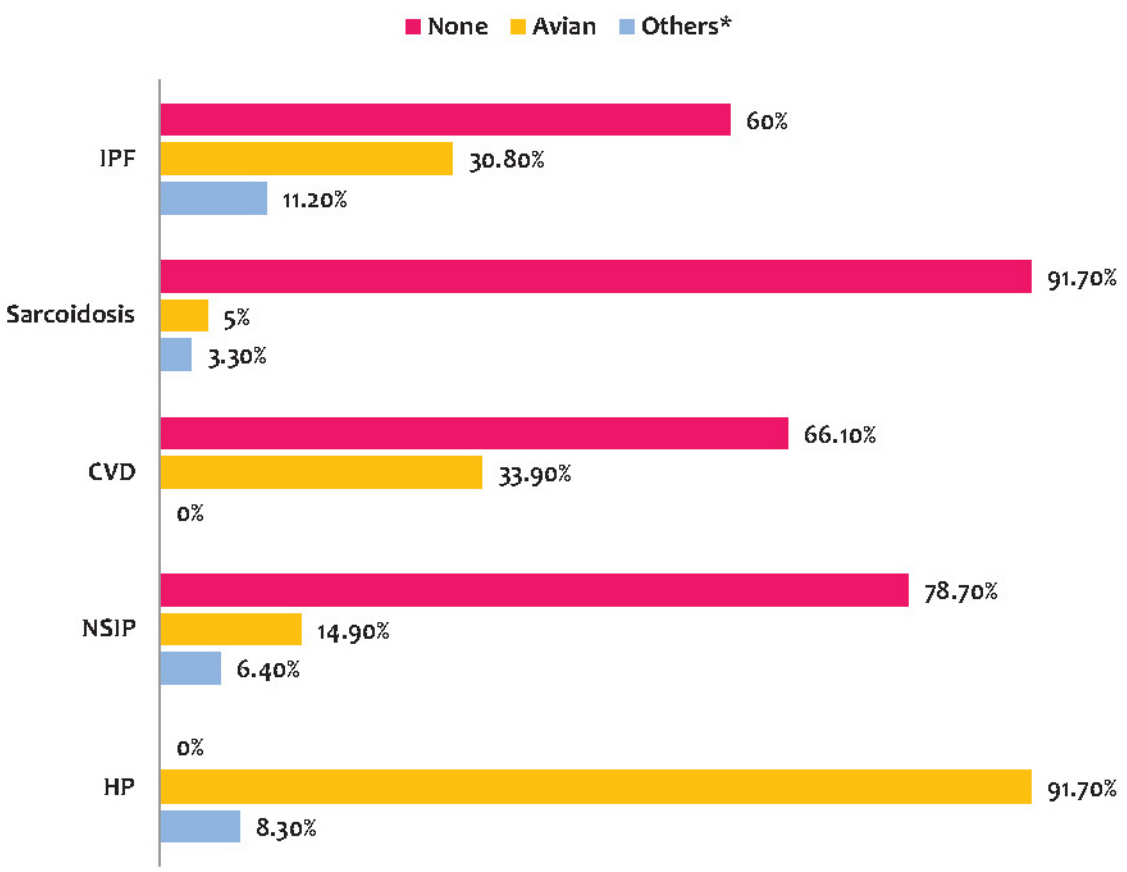


Figure 6: Exposures in Different Interstitial Lung Diseases

Figure 6 shows that 60% of the IPF cases had no exposure while 30.80% had avian exposure and 11.20% had others. 91.70% of the Sarcoidosis cases had no exposure, 5% had an avian exposure while 3.30% had others. 66.10% of the CVD had no exposure and 33.90% had avian exposure. 78.70% of the NSIP cases had no exposure, 14.90% had avian exposure whereas 6.40% had others. 91.70% of the HP cases had an avian exposure while 8.30% had others.

Others include agricultural, industrial, earthen and chemical exposures.

OCCUPATION

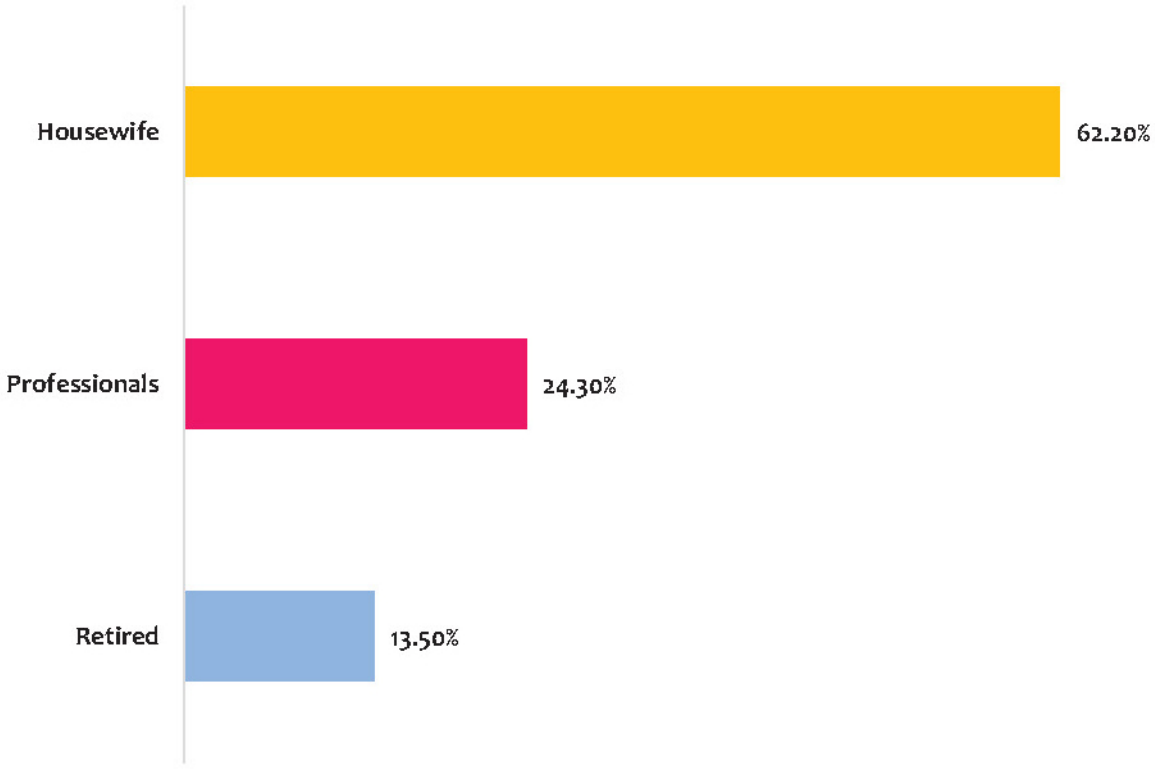


Figure 7: Occupational breakdown of ILD Cases

Figure 7 shows that 62.20% of ILD patients were housewives 24.30% were professionals while 13.50% were retired persons.

GEOGRAPHICAL LOCATION

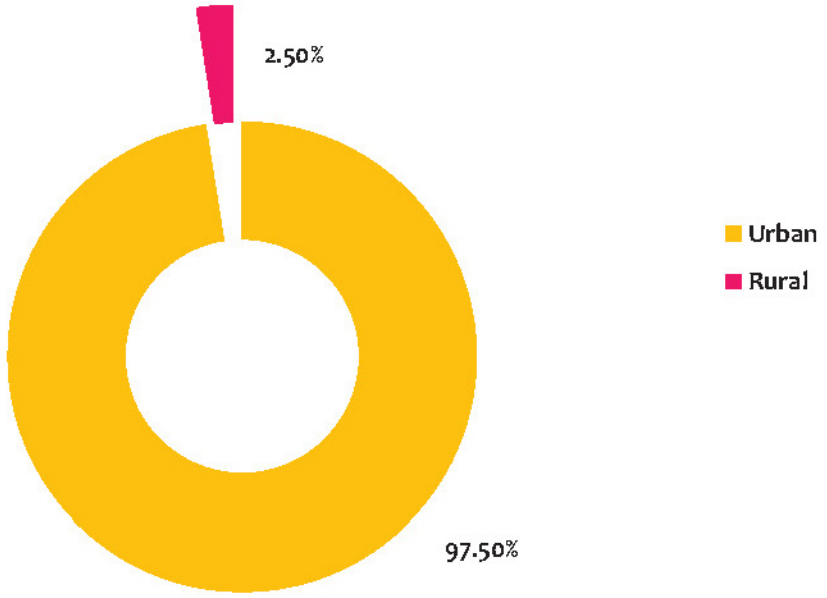


Figure 8: Disease Reporting in Geographical Locations

Figure 8 shows that the reporting ILD patients had an urban background in 97.50% while only 2.50% were from rural areas, with respect to data collected.

ETHNIC GROUPS IN ILDs

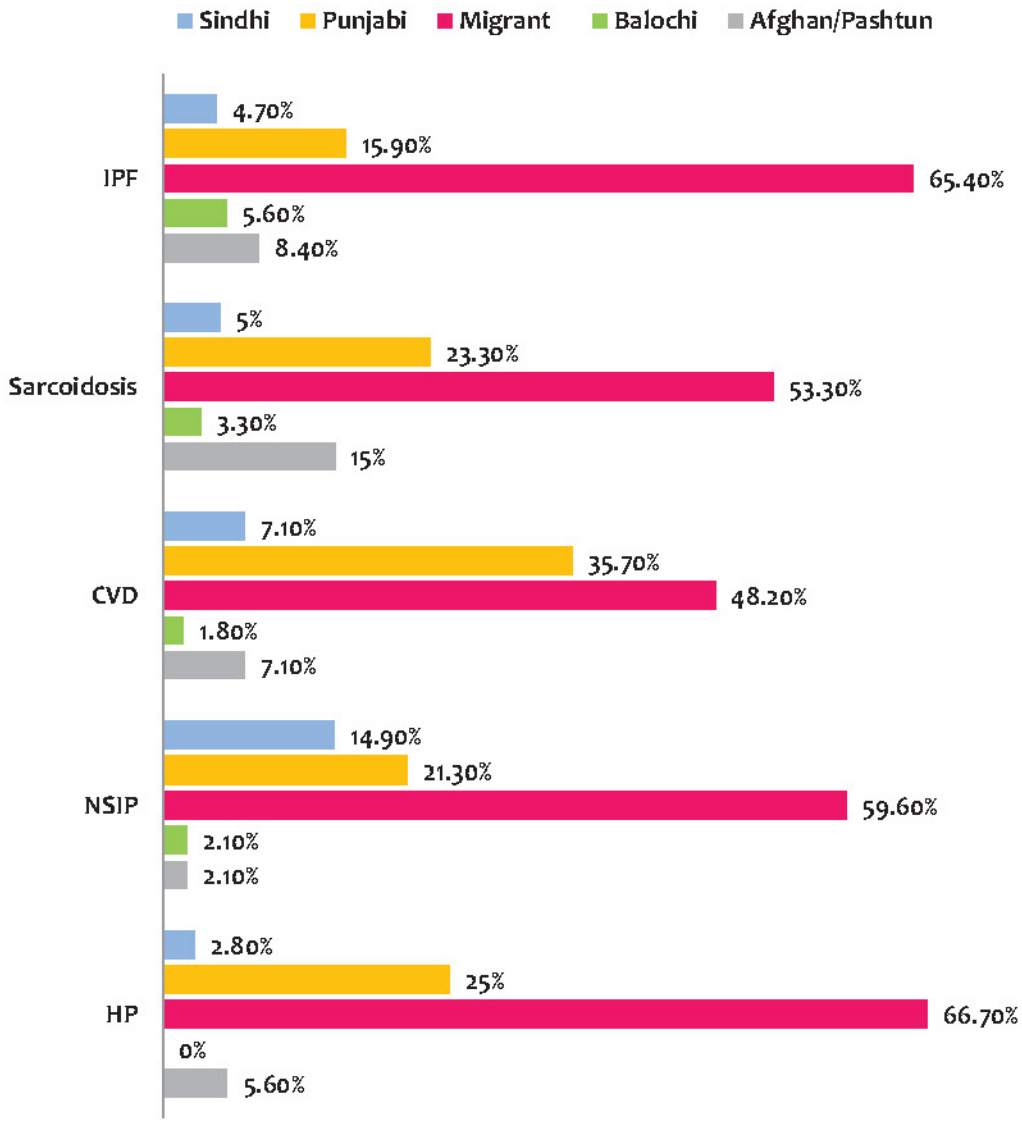


Figure 9: Frequency of Ethnic Groups in Different Interstitial Lung Diseases

Figure 9 shows that among reporting ethnic groups migrants pre dominated in all ILDs followed by Punjabis. Likewise, 65.40% of the IPF cases were of Migrants while 15.90% of Punjabis, 8.40% of Afghans and Pashtuns, 5.60% of Balochis and 4.70% of Sindhis. 53.30% of the cases of Sarcoidosis were reported by Migrants while 23.30% were by Punjabis, 15% by the Afghans and Pashtuns, 5% by the Sindhis and 3.30% by Balochis. 48.20% of CVD mainly affected Migrants while 35.70% of Punjabis, 7.10% of Sindhis as well as 7.10% of Afghans and Pashtuns and 1.80% of Balochis. Similarly, 59.60% people affected from NSIP were Migrants while 21.30% were Punjabis, 14.90% were Sindhis, and 2.10% were Balochis and 2.10% of Afghans and Pashtuns. 66.70% HP cases were of Migrants while 25% were Punjabis, 5.60% were Afghans and Pashtuns and 2.80% were Sindhis.

Migrant includes people originally from Central Sub-Continent, Southern Sub-Continent & Western Sub-Continent.

Results

ILD Clinical Features

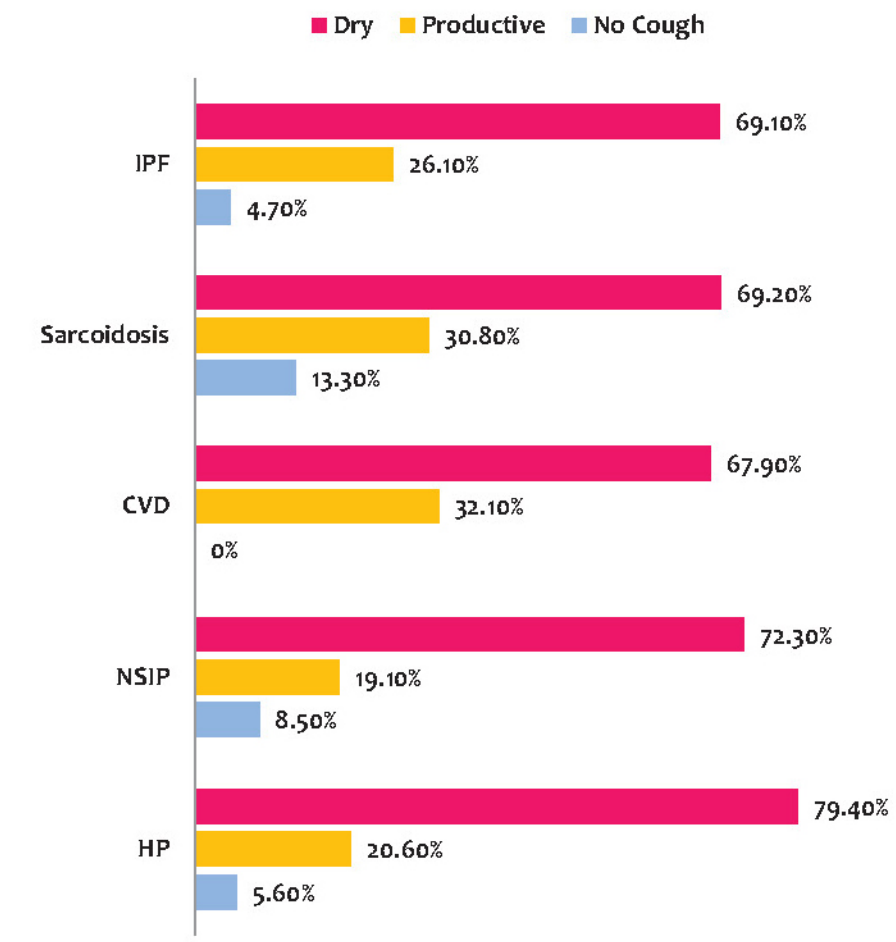


Figure 10: Features of Cough in Different Interstitial Lung Diseases

Figure 10 shows that the cough in Interstitial Lung Diseases was mainly characterized as a dry cough. Similarly, 69.10% of the IPF patients reported dry cough whereas 26.10% reported productive cough and 4.70% had none. 69.20% patients of Sarcoidosis had dry cough whereas 30.30% had productive cough and 13.30% had none. Similarly, 67.90% CVD patients reported dry cough whereas 32.10% reported productive cough. Likewise, 72.30% of the NSIP patients reported dry cough while 19.10% reported productive cough and 8.50% had none. 79.40% of the HP patients reported dry cough whereas 20.60% reported productive cough and 5.60% had none.

DYSPNEA

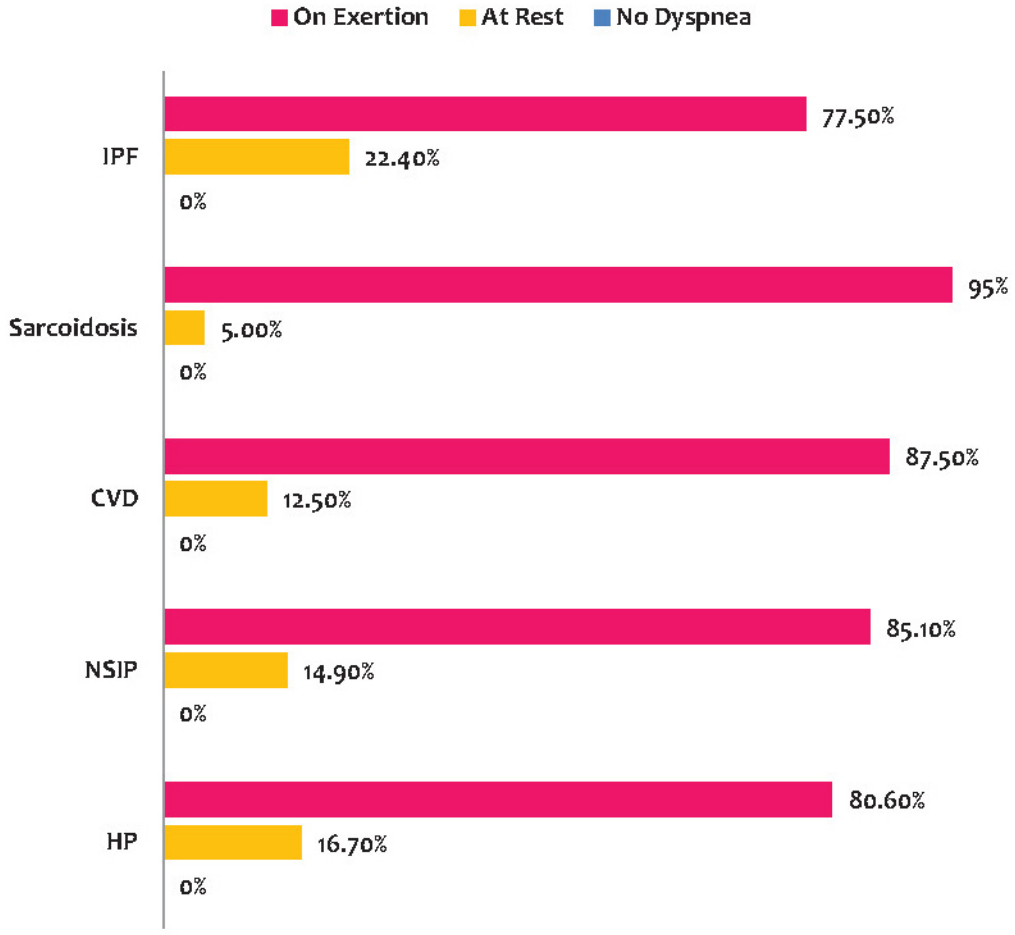


Figure 11: Dyspnea in Different Interstitial Lung Diseases

Figure 11 shows that characteristically dyspnea triggered on exertion in ILD patients. IPF had 77.50% cases with dyspnea on exertion and the largest group 22.40% who were dyspneic even at rest. Sarcoidosis had dyspnea on exertion in 95% cases and even at rest in 5%. CVD had dyspnea on exertion in 87.50% of the cases and even at rest in 12.50%. NSIP had dyspnea on exertion in 85.10% and even at rest in 14.90% cases. HP had dyspnea with exertion in 80.60% cases and even at rest in 16.70% cases.

CREPITATIONS

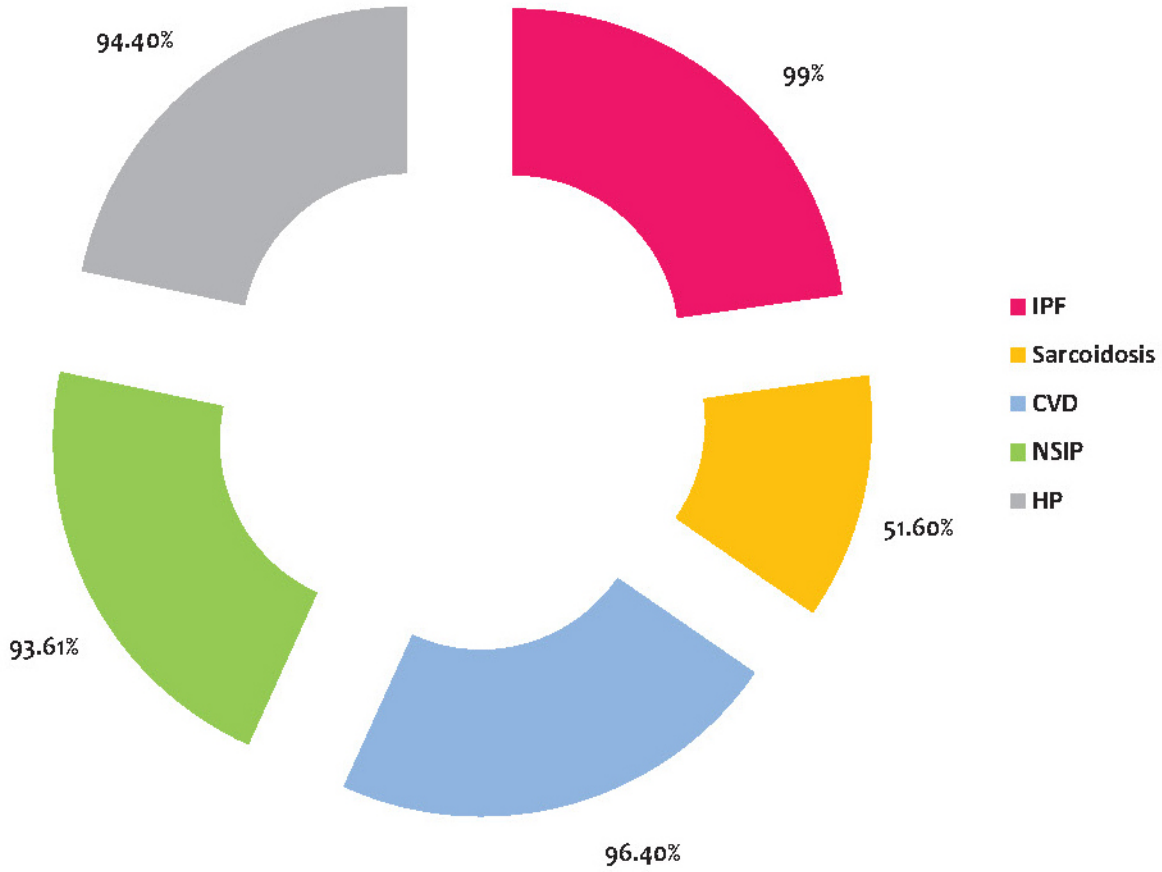


Figure 12: Crepitation's in Patients with Interstitial Lung Disease

Figure 12 shows that crepitation's were observed in 99% cases of IPF, 96.40% of the CVD, 94.40% of HP, 93.61% of NSIP and much less 51.60% of Sarcoidosis.

CLUBBING

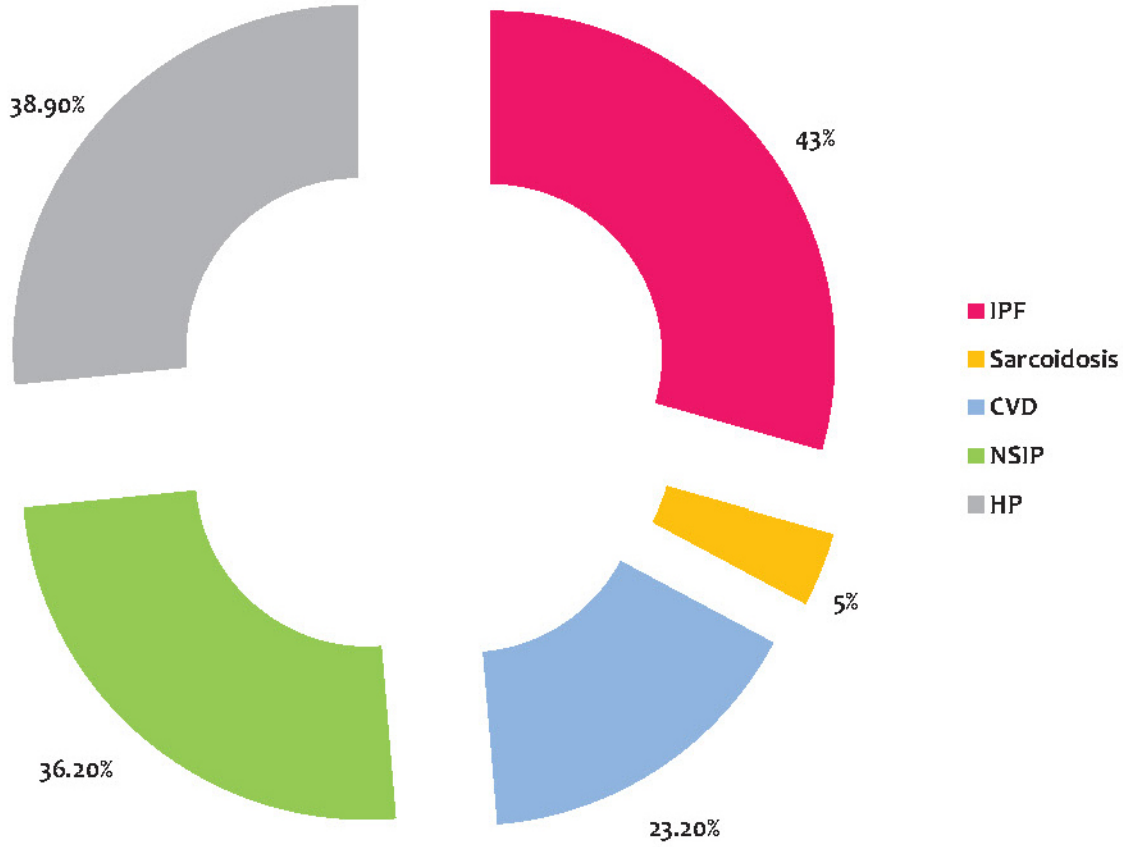


Figure 13: Clubbing in Patients with Interstitial Lung Disease

Figure 13 shows that clubbing was observed in 43% cases of IPF, 38.90% of HP, 36.20% of NSIP, 23.20% of the CVD and the least in 5% of Sarcoidosis.

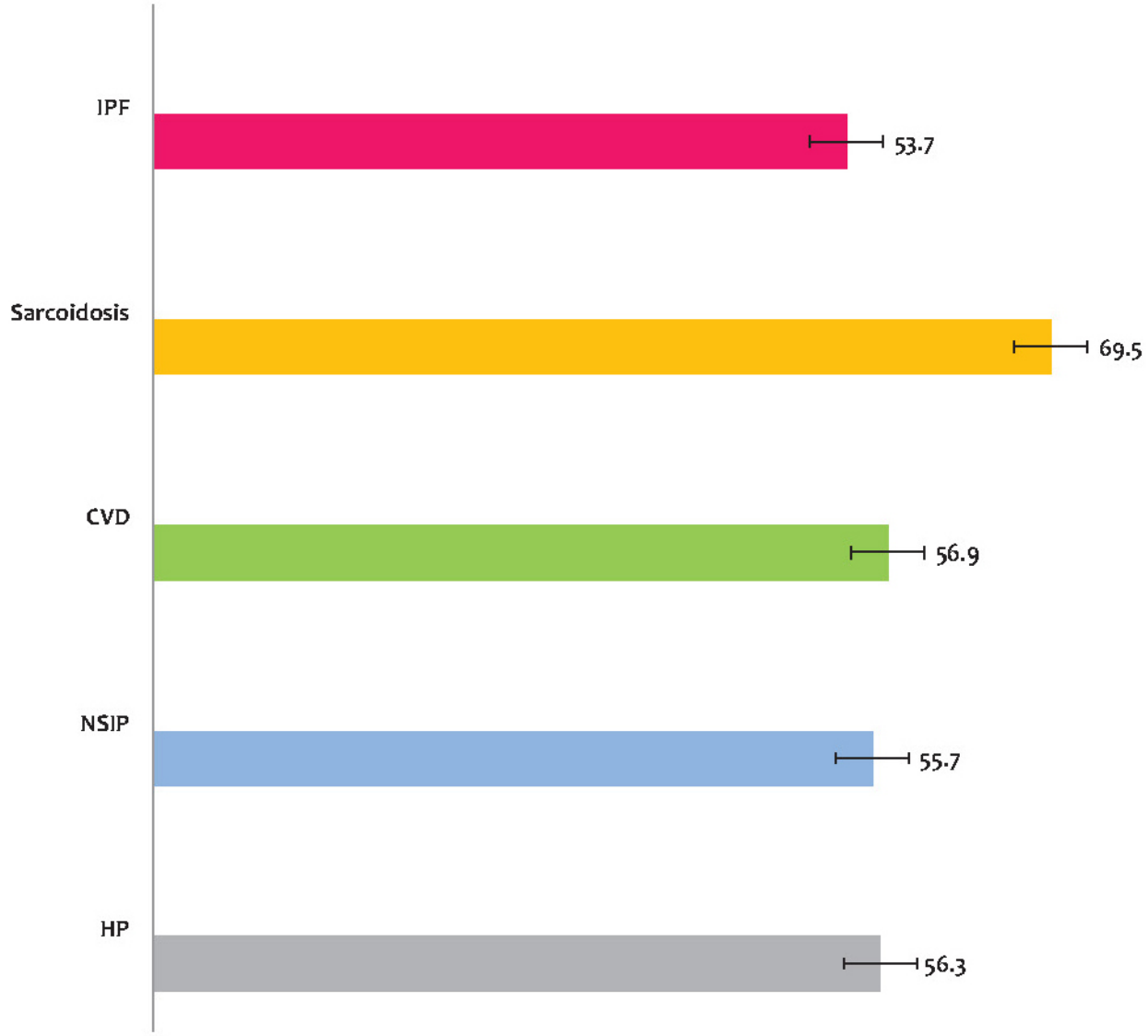


Figure 14: Mean Forced vital capacity *(% of predicted)* in the Patients of Interstitial Lung Diseases

Figure 14 shows that FVC (as % of predicted) was 53.7± 19.1 (Mean ± SD) in the case of IPF while it was 69.5±22.3 (Mean + SD) in the case of Sarcoidosis. It was 56.9±21 (Mean + SD) in the case of CVD and similarly It was 55.7±22.2 (Mean + SD) in NSIP and 56.3±17.7 (Mean + SD) in the case of HP.

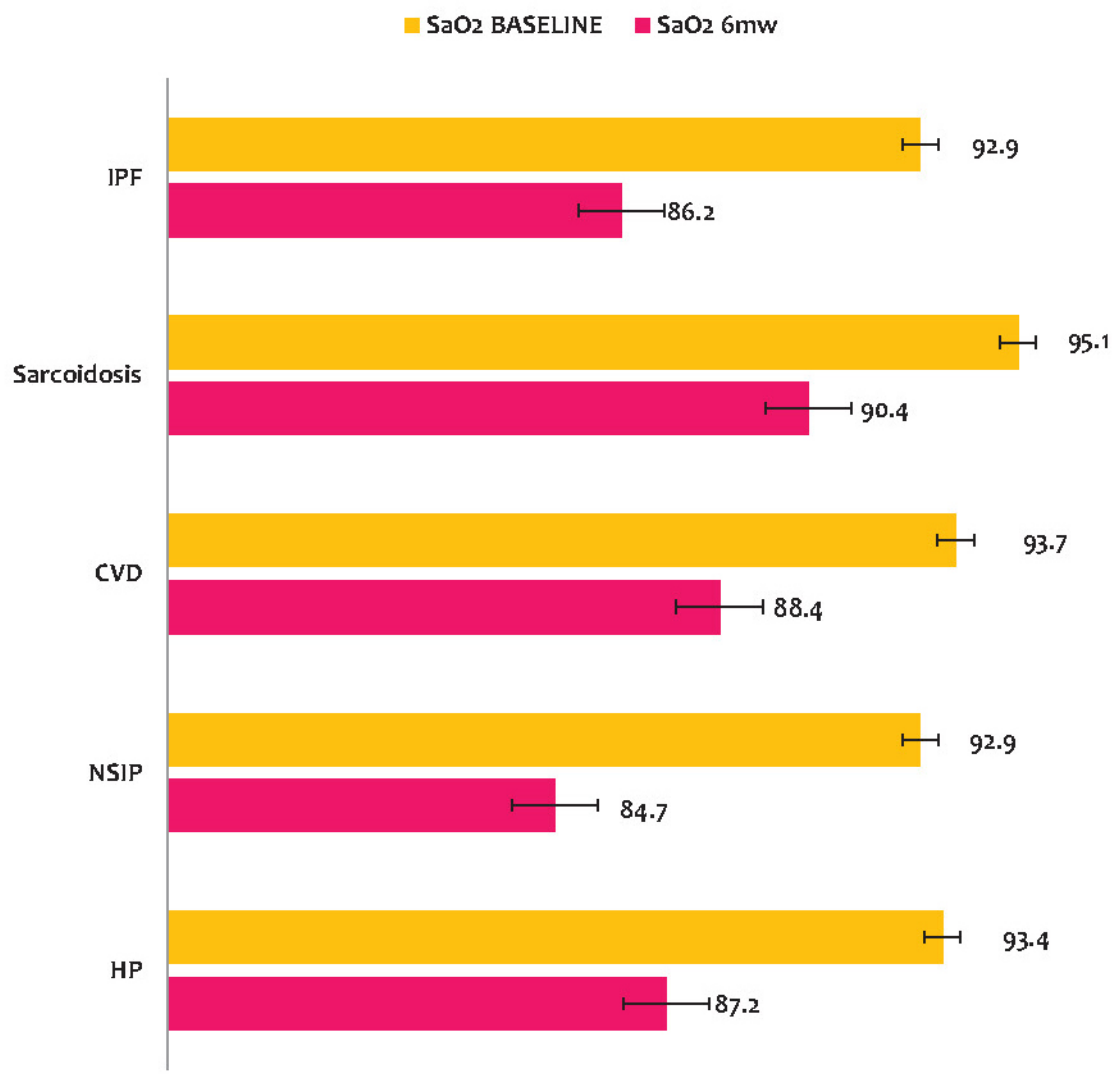


Figure 15: *Mean oxygen saturation at rest & exercise with standard deviation*

Figure 15 shows that the oxygen saturation at 6MW in IPF was 86.2±8.1 (Mean ± SD) while at baseline it was 92.9±5.1 (Mean ± SD). O2 saturation at 6MW in Sarcoidosis was 90.4±5.4 (Mean ± SD) while at baseline it was 95.1±3.1 (Mean ± SD). O2 saturation at 6MW in CVD was 88.4±6.5 (Mean ± SD) while at baseline it was 93.7±4.2 (Mean ± SD). Oxygen saturation at 6MW in NSIP was 84.7±9.2 (Mean ± SD) while at baseline it was 92.9±6.4 (Mean ± SD). Oxygen saturation at 6MW in HP was found to be 87.2±7.6 (Mean ± SD) while at baseline it was 93.4±8.7 (Mean ± SD).

ASSOCIATIONS

Figure 16: Association of ILD with Pulmonary Hypertension (PH) and Gastroesophageal Reflux Disease (GERD)

Figure 16 shows that 41.10% of the IPF cases were associated with PH and 32.70% with GERD. 15% of the Sarcoidosis cases were associated with PH and 23.30% with GERD. 17.40% of the CVD cases with PH and 23.30% with GERD. 27.70% of the NSIP cases were associated with PH and 29.80% with GERD. 22.20% of the HP cases with PH and 22.20% with GERD.

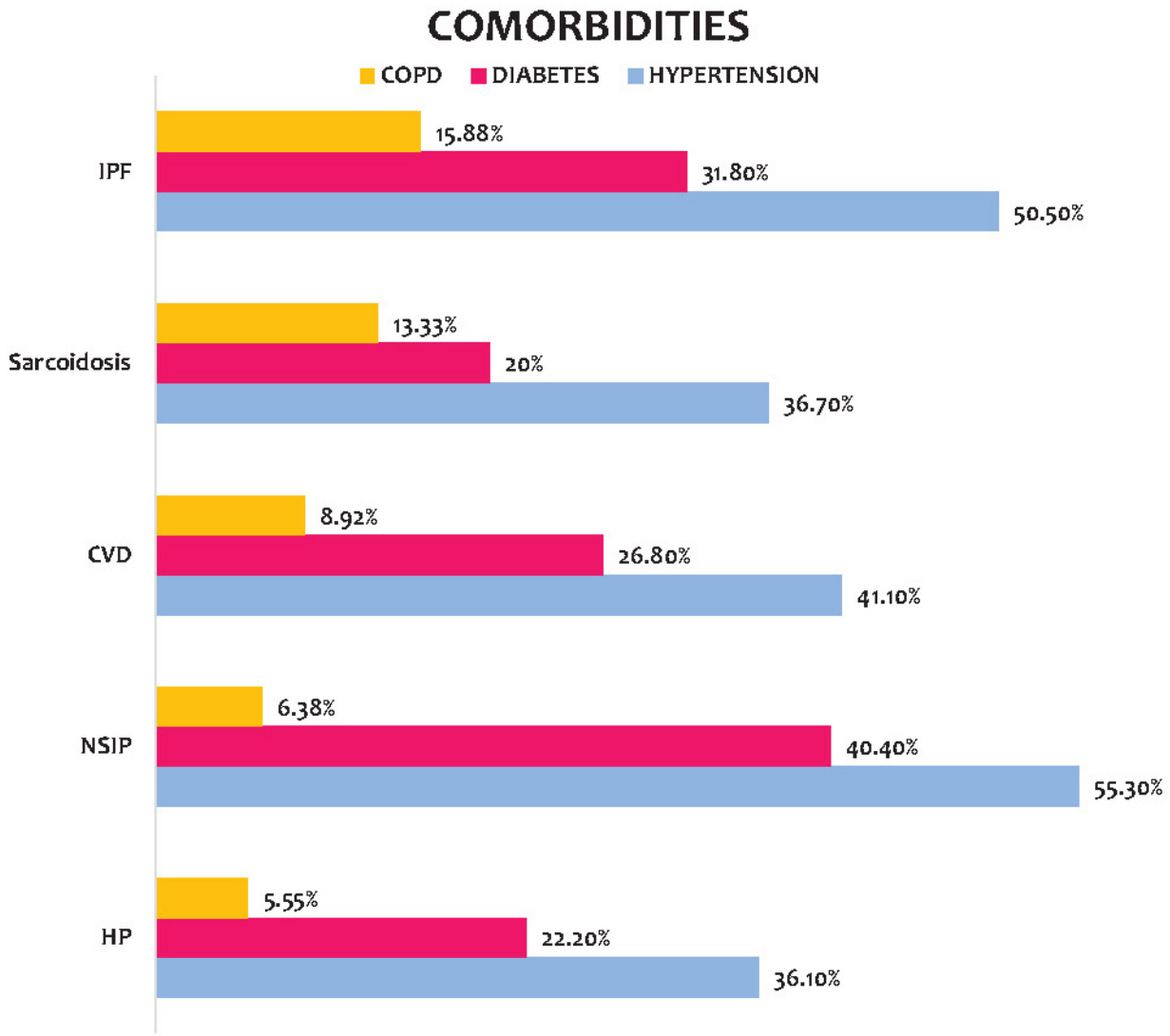


Figure 17: Comorbidities with Interstitial Lung Disease

Figure 17 shows different comorbid conditions that had been observed in the patients of ILD where Hypertension tended to be the most common. IPF also had hypertension in 50.50% of the cases along with Diabetes in 31.80% and COPD in 15.88%. Likewise, Sarcoidosis had hypertension in 36.70% of the cases along with Diabetes in 20% and COPD in 13.33%. Similarly, CVD had hypertension in 41.10% of the cases along with 26.80% of diabetes and 8.92% COPD. Again, hypertension was observed in 55.30% along with 40.40% of diabetes and 6.38% of COPD in the NSIP. Hypertension had been observed in 36.10% along with 22.20% of diabetes and 5.55% of COPD in HP.

TREATMENT HISTORY AT TIME OF DIAGNOSIS

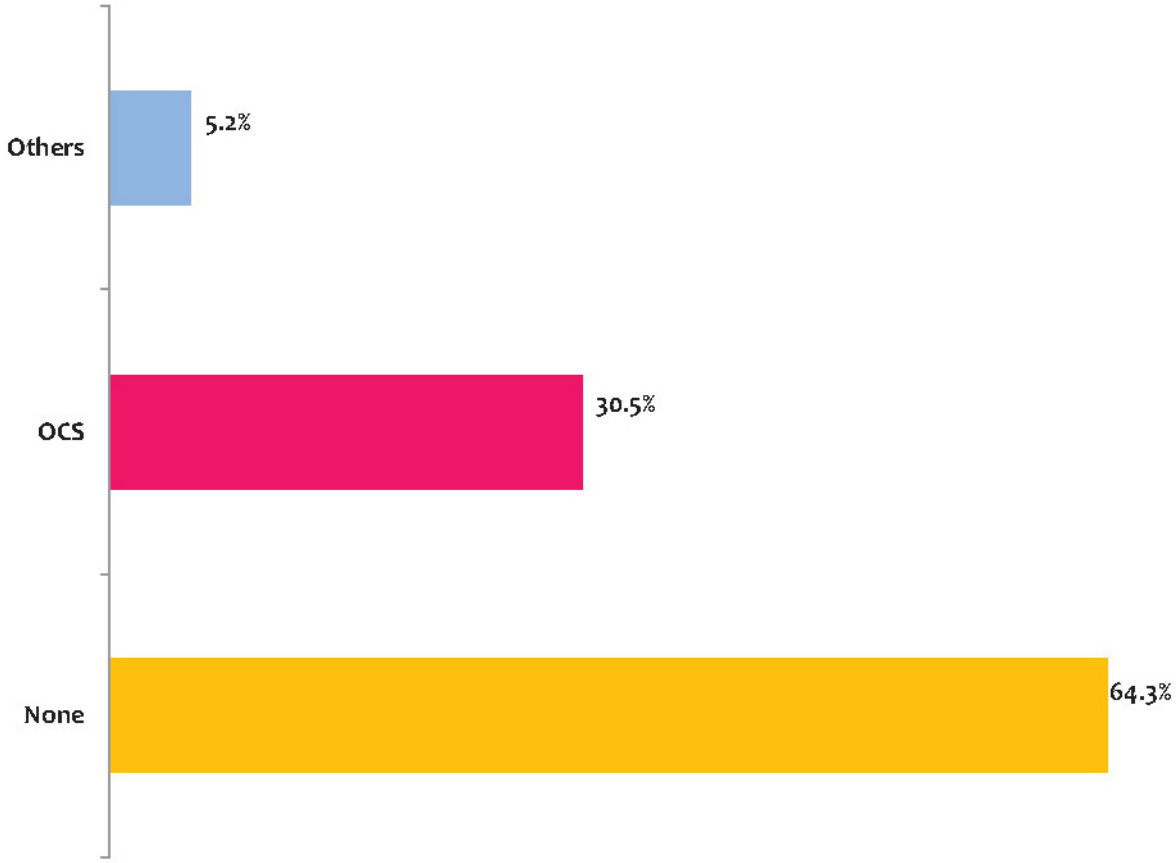


Figure 18: Treatment History of ILD Patients

Figure 18 Most of the patients, i.e. 64.30%, took no specific treatment in the past. 30.50% of the patients had been treated with random OCS and 5.20% with other immune suppressants and disease modifying anti rheumatic drugs(DMARDs).

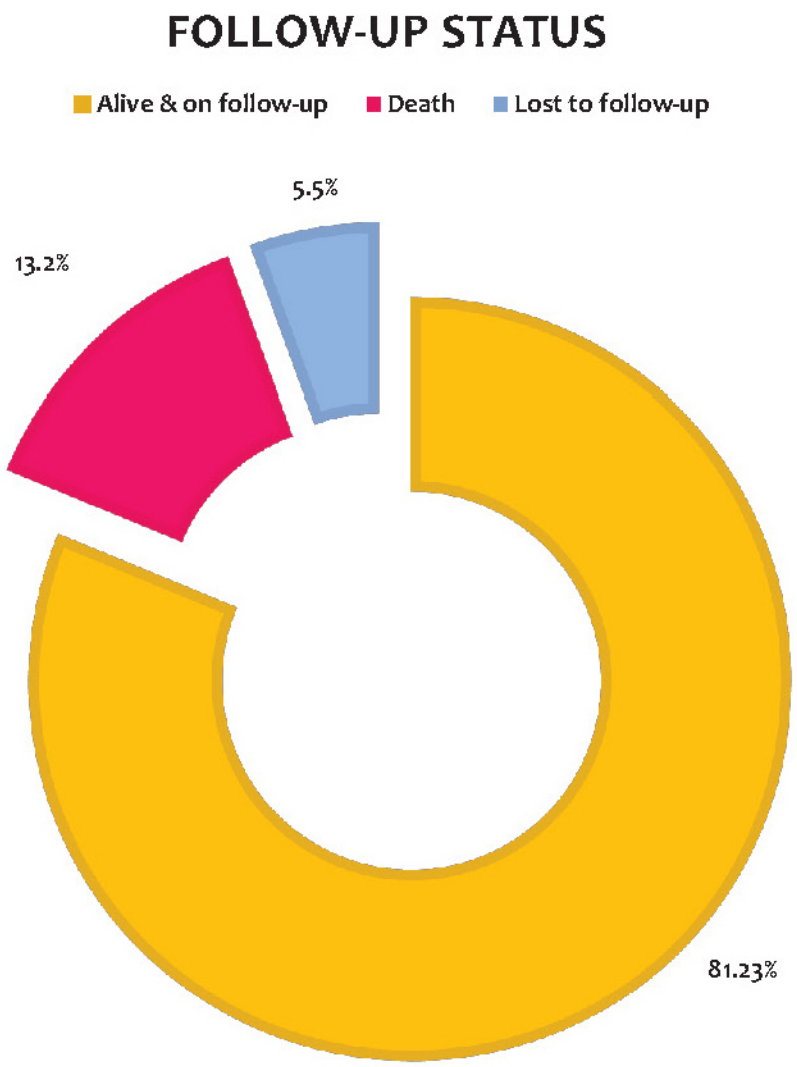


Figure 19: Follow-up status & reported Deaths in ILD registry

Figure 19 shows that majority 81.23% of ILD patients are alive and continued with follow-up, whereas 13.2% were dead 5.5% and identified as lost to follow-up.

MORTALITY

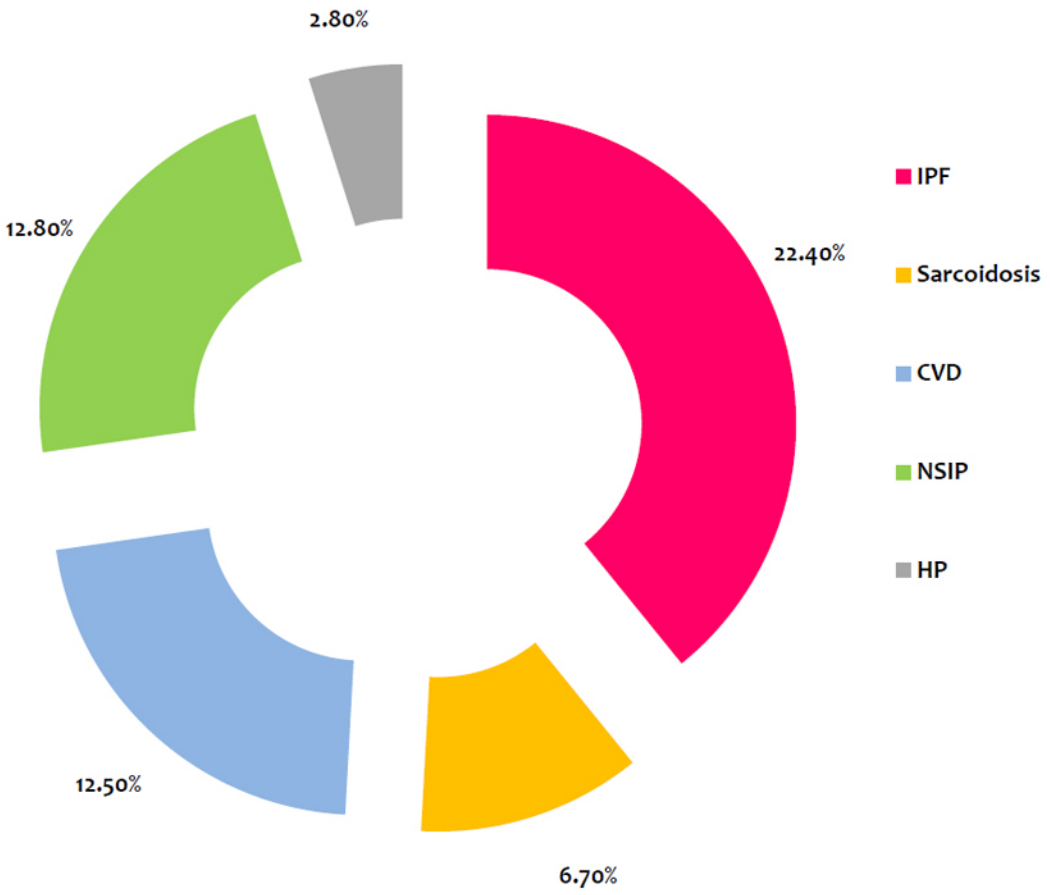


Figure 20: Mortality in Interstitial Lung Disease

Figure 20shows that highest incidence of mortality was observed in IPF with 22.40%, then in NSIP with 12.80%, CVD with 12.50%, Sarcoidosis with 6.70% and 2.80% in HP.

YEARS OF SURVIVAL

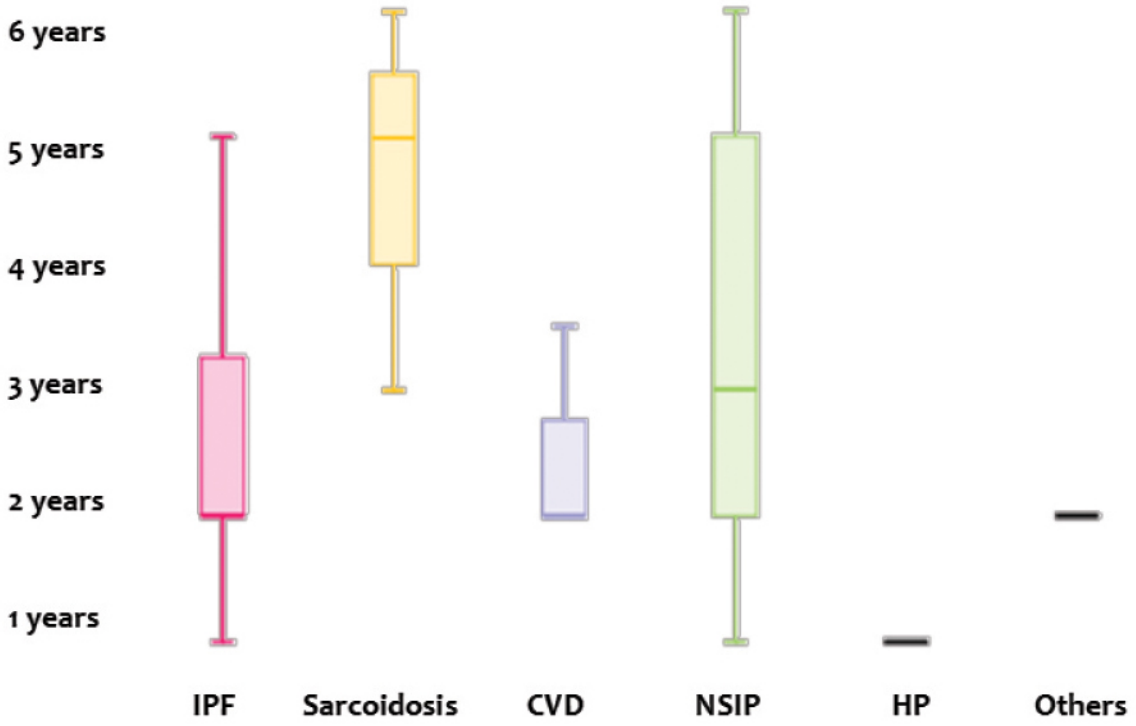


Figure 21: Years of Survival in the mortality cases of Interstitial Lung Diseases

Figure 21 shows the years of survival box-and-whiskers plot (showing the median, quartiles, minimum and maximum values) for ILD patients who died during the registry. *The median survival years for IPF was 2 (range: 1-5), for Sarcoidosis it was 5 (range:3-6), for CVD was 2 (range:2-3.5), for NSIP was 3 (range:1-6). There was only one reported death in HP which occurred within one year of diagnosis and one from the other groups.*

TIMELINE

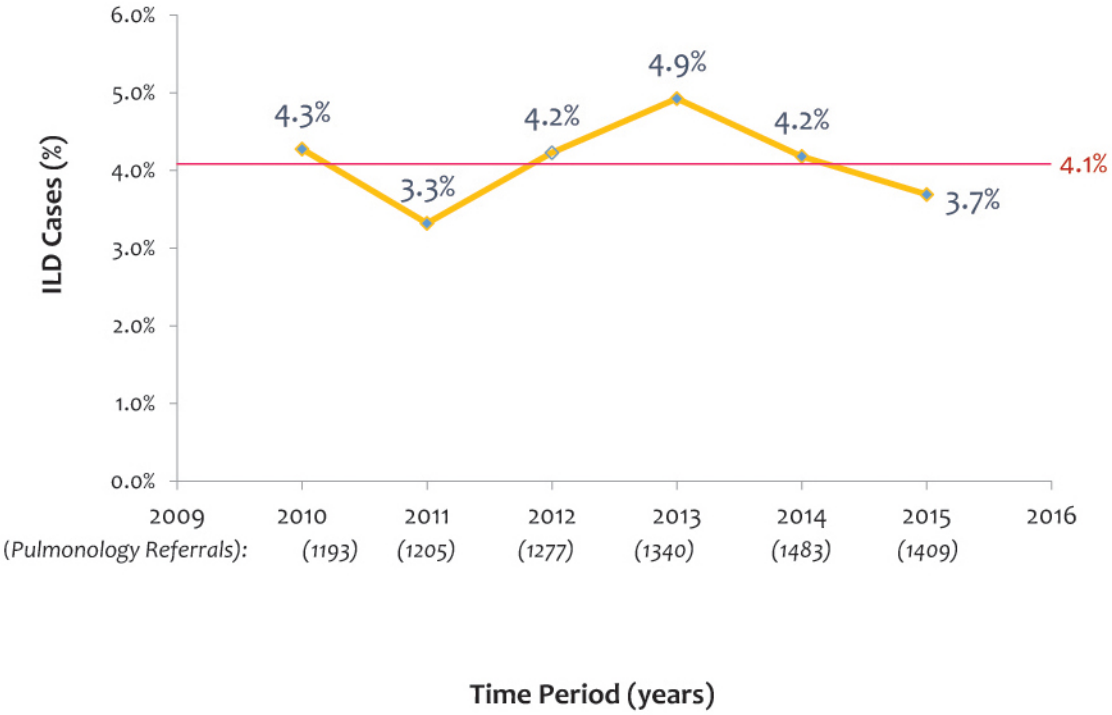


Figure 22: Annual Trend of ILD Prevalence amongst Pulmonary Referrals

Figure 22 shows the annual trend of ILD prevalence from the year 2010 to 2015 which on an average stood at 4.1%

INTERPRETATION AND DISCUSSION

The presented data provides an opportunity for an evidence based understanding of the local profile of interstitial lung disease and its risks in Pakistan. On the basis of Registry reporting, ILD-PAK shows that the most common cause of ILD is Idiopathic Pulmonary Fibrosis (32.90%), followed by Sarcoidosis (18.50%), Collagen Vascular Associated Lung Disease (17.20%), Non Specific Interstitial Pneumonia (14.50%), Hypersensitivity Pneumonitis (11.10%). The least common causes identified include miscellaneous class of ILD-Other ILDs (5.80%) comprising of rarer entities.

Our data highlights the significance of age in characterization of IPF and Sarcoidosis. In Sarcoidosis the majority of patients were under 60 years of age. Contrarily in IPF the majority of patients were above the age of 61 years. There was also a sizeable number of patients between ages 41 and 60 years suggesting that IPF in our population may also be considered in the age group less than 60 years. The overall age distribution in ILD differentials suggests that these are less common in the younger age group population and their prevalence increases with age.

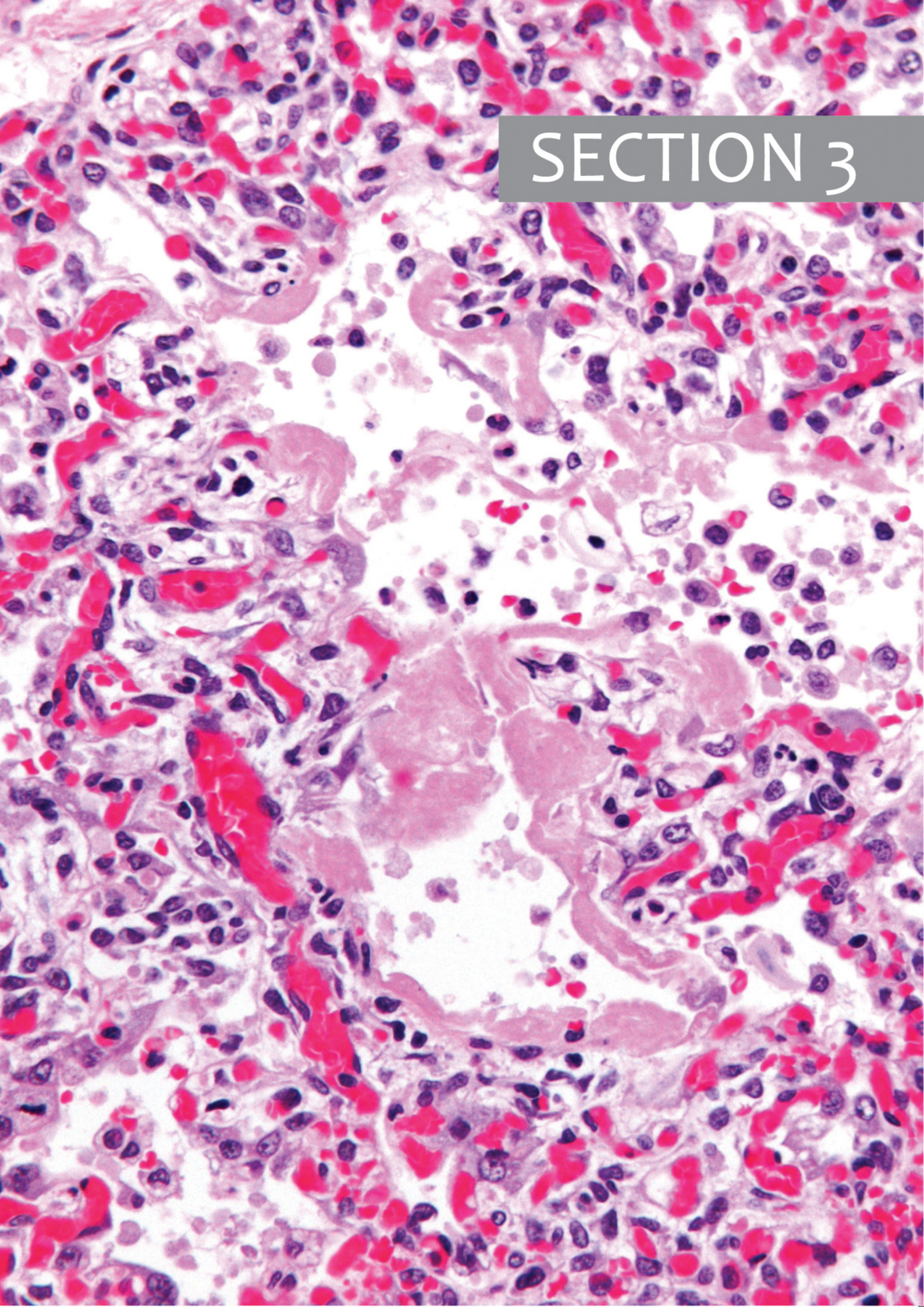
All the differentials of ILD greatly burden the female gender more than the males except the Idiopathic Pulmonary Fibrosis, which was found almost equal among both genders. A family history of affected siblings was also seen in Idiopathic Pulmonary Fibrosis more than in any other type. The results describe associated risk factors that may predict the clinical outcome and importance of management and prevention strategies. It is notable in this context that there is a significant association of PH and GERD with IPF as per our results. The data shows that almost a quarter of IPF patients presented with smoking history of greater than 25 pack years. All the differentials of ILD were found highest among Migrants, followed by Punjabis. However, it may not truly indicate that the disease is more common in migrants but because in our study these respondents were overwhelmingly more than any other ethnic group.

Among the different types of cough, dry cough was found most commonly among all the ILDs. Although exertional dyspnea was found to be very common among all patients, there was a substantial number who had dyspnea even at rest and who desaturated significantly on or less than six minutes' walk. The highest number of such patients had IPF. Clubbing was found most commonly among IPF followed by HP.

Crepitations were heard in almost all the ILDs although Sarcoidosis was one type with relatively less than others and IPF invariably had the Velcro type. The FVC was moderately reduced in all the differentials but relatively less in Sarcoidosis. The most frequently occurring co-morbid condition along with ILD was Hypertension followed by Diabetes, though these numbers may merely be a reflection of their prevalence in the general population. Among all the causes of ILD, highest mortality was found among the IPF cases and least among Sarcoidosis. The medium survival in mortality cases was of 2 years (Range 1 - 5 years) and in Sarcoidosis was 5 Years (Range 3 - 6 years).

CONCLUSION

To our knowledge, this is the first population based registry that attempts to provide an epidemiologic update of ILD trends in the recent years in a population segment of patients in Pakistan. The forte of this registry is its mandatory recording of HRCT and PFT investigations and the recording of exposure history and clinical manifestations without which no case was included. The electronic database support and direct data entry access to principal investigators across the country, along with the recently published PCS ILD National Guideline and this report will gather impetus towards the production and publication of further regional reports periodically. The PCS ILD PAK Registry nexus can thus function on a national scale as a standardized recording and tracking mechanism in trends of presentation, incidence, prevalence, morbidity, mortality and treatment evaluation in future.


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
NSIP	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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