# Chronic Interstitial Lung Disease and Sarcoidosis

08

## Key Points

- Sarcoidosis and idiopathic pulmonary fibrosis are amongst the most important of the 300-plus interstitial lung diseases.
- Approximately one in three cases of interstitial lung disease has a known cause
- The management of interstitial lung disease of known aetiology is, in the first place, prevention and cessation of the cause

## Background

Interstitial lung diseases (ILDs) result from damage to the alveoli (air sacs) leading to inflammation and fibrosis of the interstitium (tissue wall between the alveoli). There are more than 300 different diseases included in this category. As most ILDs are rare, accurate data on prevalence and mortality are scarce. The main symptom of ILD is progressive shortness of breath initially on exertion, and then at rest when the disease is more advanced[1].

A practical classification distinguishes ILDs of known cause from those of unknown cause. Of the major ILDs, approximately 35% have a known aetiology. These include external factors such as organic or inorganic material inhalation, drug reactions and infections. Included amongst those of unknown aetiology are sarcoidosis (ICD 10: D86) and idiopathic pulmonary fibrosis (ICD 10: J84.1)[1].

Among the 35% of major ILDs of known aetiology are pneumoconioses (see chapter 11), extrinsic allergic alveolitis (hypersensitivity pneumonitis) (see chapter 11), iatrogenic ILD caused by drugs and/or radiation and post-infectious ILD[1]. Included among the 65% of major ILDs of unknown aetiology are: sarcoidosis, idiopathic interstitial pneumonias (IIPs) ILD in connective tissue diseases (CTD) and in collagen-vascular disease[1].

Of the IIPs, the most common are: IPF with a histo-pathological pattern of usual interstitial pneumonia (~55% of IIPs), nonspecific interstitial pneumonia (~25% of IIPs), respiratory bronchiolitis ILD, occurring in smokers (~10% of IIPs), desquamative interstitial pneumonia (~5% of IIPs), cryptogenic organising pneumonia (~3% of IIPs), lymphoid interstitial pneumonia (~1% of IIPs) and acute interstitial pneumonia (~1% of IIPs)[1].

Some of these diseases, such as sarcoidosis and ILDs associated with connective tissue disease (CTD), also affect other organs and this may determine the prognosis to a greater extent than the lung dysfunction. Sarcoidosis often improves spontaneously without treatment[2]. To date, there are no curative treatments available for most ILDs. Lung transplantation offers hope for selected patients.

In this chapter, we focus on sarcoidiosis (ICD 10: D86) and "Other interstitial pulmonary diseases" (ICD 10: J84). This latter in the remainder of this chapter is called chronic interstitial lung disease. As the most common of these is idiopathic pulmonary fibrosis (ICD 10: J84.1), the latter term will be used with respect to hospitalisations.

## Incidence and Prevalence

The incidence and prevalence of chronic interstitial lung disease and idiopathic pulmonary fibrosis (IPF) in Ireland are not known. A national hospital based registry of idiopathic pulmonary fibrosis commenced in 2016. Based on international experience Ireland would expect an incidence of 400 new cases per year with a prevalence of 1,000 cases.

Ireland has the second highest prevalence of sarcoidosis in the world[4].

## Mortality

In 2015, there were 341 deaths due to chronic interstitial lung disease compared with 199 in 2007. For deaths from sarcoidosis the numbers in 2015 were 12 compared with 17 in 2007[3].

## Impact on health services

Data is not available at national level in Ireland on ILDs, nor is data available for those for whom respiratory aids and appliances, such as nebulisers and oxygen, are prescribed, either for the population as a whole or just those with general medical eligibility. Also lacking at national level is data on the impact of these diseases on GP services, Emergency Department services, and out of hours services. In terms of impact on hospital services, these conditions impact not just on inpatient hospitalisations but also on pulmonary rehabilitation services, Outpatient Departments and respiratory laboratories. National data is not available for these latter services by disease group. The impact on health services in terms of hospitalisations and bed usage is available from HIPE reporting acute publicly funded hospitals only.

In 2016, there were 324 day cases and 657 inpatient hospitalisations for idiopathic pulmonary fibrosis (0.7% of respiratory inpatient hospitalisations, 0.1% of all inpatient hospitalisations) with a usage of 6,452 inpatient hospital bed days (1.1% of respiratory inpatient bed days, 0.2 % of all inpatient bed days). Also in 2016, there were 524 day cases and 234 inpatient hospitalisations for sarcoidosis using 1,364 inpatient bed days.

Of the inpatient hospitalisations, 78.8% with idiopathic pulmonary fibrosis were admitted as an emergency. For those with sarcoidosis, the figure was 81.2%.

## Gender

In 2015, of the 341 deaths due to chronic interstitial lung disease (ICD 10: J84), 58.7% (200) were in males. In 2007, of the 199 deaths, 61.3% were in males. Of the deaths from sarcoidosis, in 2015, 58.3% (7) of the deaths from sarcoidosis were in males while in 2007 the figure was 53% (9) males[3].

Of those hospitalised in 2016 with sarcoidosis, 56% were males. Of those hospitalised with idiopathic pulmonary fibrosis in 2016, 63.5% were males.

## Age

In 2015, of the 341 deaths due to chronic Interstitial lung disease, 7.9 % (27) were aged 30-64 years, 22.6% (77) were aged 65-74 years, 45.7% (156) 75-84 years and 23.8% (81) 85 years or over[3]. Of the deaths from sarcoidosis in 2015, 7 (58.3%) were aged under 65 years of age. Of those admitted as inpatients to hospital in 2016, with idiopathic pulmonary fibrosis, 71% (465) were aged 65 years and over. Of the inpatient hospitalisations with sarcoidosis, 87% were aged 16-64 years.

## Regional variation

Robust data is not available at national level.

## Socio-economic analysis

Robust data is not available at national level.

## International Comparisons

In Europe registries of the epidemiology of different ILDs have been compiled in several countries. As many of these registries are of data reported by respiratory physicians, they have the disadvantage of not being population based. However, they do allow comparison of the relative frequencies of the different ILDs[1]. Sarcoidosis and IPF account for up to 50% of ILDs. In the UK, the annual incidence of sarcoidosis is 7 cases per 100,000 population which is consistent with the range of 5-40 per 100,000 reported by other northern European countries[2].

The UK prevalence of idiopathic pulmonary fibrosis is 50/100,000, as quoted in the UK document "Battle for Breath" which is more than double that previously reported by NICE of 15-20/100,000[2,5].

ILD, especially IPF, occurs in older subjects, while sarcoidosis occurs in young adults of both sexes and in women over 50 years of age[1]. In terms of hospitalisations, Irish figures are in line with this age trend. Internationally, IPF is more frequent in males and sarcoidosis is more frequent in women[1].

Mortality data for the majority of European countries shows differences between countries, which are partly real and partly may be due to differences in diagnostic and therapeutic strategies. The highest mortality rates in Europe (WHO) due to ILD, of 6.68 per 100,000 people, was reported in Ireland by WHO Europe in 2011, compared with the figure for the EU 28 of 2.2[1]. Among the most important ILDs, chronic ILD (ICD 10: J84) had the highest age standardised mortality rate, followed by ILD associated with connective tissue disease. The mortality rate for chronic ILD in most countries was less than 2 per 100,000, but in Ireland it was 5.49, in Malta 5.92 and the UK 4.02 compared with the average for EU 28 of 1.7. The age-standardised mortality rate for sarcoidosis as reported by WHO Europe in 2011, was 0.37 in Ireland and 0.31 in Denmark compared with EU 28 average of 0.1[1].

The WHO mortality data should be interpreted cautiously for reasons outlined above and also because for systemic diseases, such as sarcoidosis and connective tissue diseases (CTD), the WHO mortality data do not distinguish whether patients had related ILD or, if present, whether the ILD contributed to death; thus the mortality rates only partly reflect deaths due to ILD.

The 2015 report on the Global Burden of Disease reported that for ILD and pulmonary sarcoidosis, the number of total deaths between 2005 and 2015 increased globally by 51.8% to 121,800 (CI: 94,100-135,200) and the age standardised rate rose by 14.1% (CI: 4.1-20.9)in the same period[6].

The hospitalisations rate for ILDs across Europe (WHO) was highest in Austria, Denmark, Norway, Finland, Poland and Slovakia based on the WHO Hospital Morbidity Database (2011). However, no data was recorded for Ireland[1].

## References


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