British Thoracic Society

The British Thoracic Society Interstitial Lung Disease Registry Programme

Annual Report 2013/14

The first report from the BTS Idiopathic Pulmonary Fibrosis (IPF) and Sarcoidosis Registries

"BTS is leading the way in harmonising the capture of valuable demographic and clinical information on IPF and sarcoidosis patients onto a national database to enable better understanding of these chronic conditions. The data will establish indicators of best practice and help us to enhance the quality of care for patients with IPF and sarcoidosis across the country.'

Professor Monica Spiteri, chair of the BTS Lung Disease Registry Steering Committee

The BTS Lung Disease Registry for Idiopathic Pulmonary Fibrosis and for Sarcoidosis launched in February 2013. More than 25 NHS hospitals are now actively participating in the BTS Registry. This is the first of a series of annual reports from the British Thoracic Society on the IPF and Sarcoidosis Registries.

In line with the overall objectives of the British Thoracic Society, the Lung Disease Registry has been designed to provide a means of national longitudinal data collection on patients with IPF and Sarcoidosis. The aim is to provide an easily accessible system for prospective datasets in a large number of patients so that the public health and epidemiological status of these conditions in the UK can be established. The Registry will also serve as an important resource for future clinical and applied research.

The incidence of IPF is increasing in the UK, and sarcoidosis, the commonest interstitial lung disease, accounts for around one third of the interstitial lung diseases seen in specialist respiratory clinics.

However, little is known about the cause of these conditions. It is intended that the BTS Registry will provide vital information on the natural history, mode of referral, management and clinical outcomes of these diseases, and in due course, lead to improvement in standards of care for patients.

The BTS Idiopathic Pulmonary Fibrosis (IPF) Registry

Idiopathic pulmonary fibrosis (IPF) is a progressive lung disease of unknown cause characterised by variable degrees of inflammation and scarring. The aetiology of IPF is not known, although possible risk factors may include infectious agents, gastro-eosophageal reflux and genetic factors. The true incidence of IPF is unknown, but it is estimated that annually there are 5000 new cases of IPF or an incidence rate of 7-9 per 100,000. Prevalence rate is estimated to be 15-25 per 100,000 but increases with age giving a current UK prevalence of approximately 15,000. Prevalence remains relatively low because unfortunately about 5000 patients with IPF per annum die of their disease.

The reported median survival in IPF is 3 years from diagnosis; it has a poorer prognosis than cancer of the colon, breast or ovary. Only 20% of patients survive to 5 years post-diagnosis.

Importantly it is now clear that the incidence of IPF is rapidly climbing with a 35% increase in diagnosed cases between 2000 and 2008. The average age at diagnosis is 70 years, but the rising incidence has been shown not to be a consequence of an ageing population.

The disease poses significant challenges for clinicians: diagnosis of IPF requires expert integration of clinical, radiological and, when available, pathological data. Diagnostic precision is critical to distinguish IPF from other interstitial lung diseases that may respond for example to corticosteroid therapy. IPF does not respond to steroids.

Until recently best supportive care was the only realistic treatment option open to many IPF patients. This includes tailored oxygen therapy (short-burst, long-term domiciliary and ambulatory), pulmonary rehabilitation, and expert palliative care input. Lung transplantation has also been available. This was the only proven treatment to improve survival in IPF patients, but it is often only feasible for a minority of patients and many that are listed do not make it to transplant. IPF patients have the highest mortality of any group waiting for lung transplants.

In July 2013 the first NICE-approved novel drug to treat IPF in the UK became available - called Pirfenidone. Evidence suggests this drug slows disease progression and has an impact on survival. Use of Pirfenidone is currently limited to mild/moderate disease only, and if patients progress significantly whilst on drug it has to be stopped as defined by NICE criteria. It does not appear to help all patients - some progress despite the drug but it is a good start to IPF treatment and very welcome. Other promising drugs are also now in the pipeline so more treatment options may be available in the near future.

Many questions remain unanswered about this devastating disease. There is still much to learn about IPF and how its care is delivered across the NHS. Furthermore as novel therapeutic agents become available for IPF, the precise patient groups that would benefit from these drugs will need to be defined. Data collected through the Registry will start to address these questions.

The BTS Sarcoidosis Registry

Sarcoidosis is a multisystem disease which may affect many organs at any one time, with lung involvement in around 90% of patients. Sarcoidosis is the commonest interstitial lung disease (ILD), typically accounting for around one third of the ILD seen in a specialist respiratory clinic. The cause remains unknown; both genetic and environmental factors have been implicated.

Prevalence ranges from 3 (in white populations) to 47 (in African Americans) per 100,000 population in North America, and 64 per 100,000 population in Scandinavia. Average incidence from radiographic population screening programmes in continental Europe is 10 per 100,000 population. UK general practice data suggest an incidence of around 3 per 100,000 person-years, similar to figures derived from New Mexico and Japan but lower than North American or European estimates. The disease is not only more prevalent in ethnic minorities such as Black and Afro-Caribbean, but these groups also suffer more severe disease and a higher mortality.

Sarcoidosis is more common in females; incidence peaks between the ages of 20 and 50 years with a smaller peak after the age of 60. There are no exact European data on working days lost due to sarcoidosis,

but many patients with active disease are unable to work because of exertional dyspnoea or other symptoms such as fatigue and joint pains. The course of sarcoidosis varies greatly; there is a high rate of spontaneous remission but chronic disease may occur in up to 30% of patients. To date it remains difficult to predict which patients will develop chronic, progressive severe disease and how best to manage these patients.

Complications associated with sarcoidosis and increased mortality include pulmonary hypertension; fungal infections and aspergillomas, opportunistic infections resulting from immunosuppressive therapy; chronic fatigue when present may be persistent and incapacitating. Overall mortality in large case series is around 5%, most commonly due to severe parenchymal disease causing pulmonary fibrosis, as well as cardiac and neurological involvement. When required, treatment usually includes systemic corticosteroids with or without other immunosuppressive agents. Long term oxygen may be required; lung transplantation is reserved for those who have failed to respond to maximal therapy and is limited by organ availability.

What will Registry data be used for?

The BTS Registry datasets for IPF and Sarcoidosis record information on the clinical features of the condition in patients presenting to respiratory clinics and allows follow-up data to be added at regular intervals.

The data collected comprise basic demographics including age, gender, occupation, residence and smoking status, as well as details of significant co-morbidities, the delay between onset of symptoms and specialist referral, lung function and disease-specific diagnostic metrics and treatments.

Information collected through the Registry will enable us to characterise the largest number of patients with IPF and with sarcoidosis ever profiled in this country. In addition to providing essential information on baseline demographics, we anticipate that the data will facilitate sharing of best practice, benchmarking and clinical audit. In due course, this information will also provide an invaluable resource for NHS healthcare commissioners and providers alike as they seek to obtain and offer the best services for these challenging diseases and to improve services as well as patient experience.

Data about each hospital's patients are immediately available to the participating clinicians in those centres for use in audit and service development - data can be exported for further analysis and a range of graphical reports are also available to download in real time.

Patients who consent to participation in the Registry are also asked whether they are happy to be contacted about future research studies and their consent is recorded. The Registry will thus provide a hitherto unparalleled cohort of well-characterised patients with IPF and sarcoidosis who are eligible for inclusion in research investigations, including clinical trials of new treatments.

BTS aims to publish information about the entire cohort on an annual basis in future; these data will allow us to determine regional variations in disease behaviour and care delivery, and importantly to work with all key stakeholders to improve and harmonise the management of IPF and sarcoidosis patients across the NHS.

We encourage all colleagues from across the UK to participate so we can ultimately improve the care and outcome in IPF and sarcoidosis.

Acknowledgements:

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The BTS Interstitial Lung Disease Registry Programme has HRA ethical approval - REC reference: 12/EE/0381

The following organisations are currently participating in the BTS Lung Disease Registry – our thanks to all involved:

Aintree University Hospital NHS Foundation Trust

Croydon Health Services NHS Trust

Gloucestershire Hospitals NHS Foundation Trust

Harrogate and District NHS Foundation Trust

Heart of England NHS Foundation Trust

Hull and East Yorkshire Hospitals NHS Trust

Imperial College Healthcare NHS Trust

King's Health Partners (Kings, Guys & St Thomas ILD service)

Newcastle upon Tyne Hospitals NHS Foundation Trust

Norfolk and Norwich University Hospital NHS Foundation Trust

North Bristol NHS Trust

Northern Devon Healthcare NHS Trust

Nottingham University Hospitals NHS Trust

Oxford University Hospitals NHS Trust

Papworth Hospital NHS Foundation Trust

Peterborough & Stamford Hospitals NHS Foundation Trust

Royal Devon & Exeter Foundation NHS Trust

Royal Wolverhampton NHS Trust

Sheffield Teaching Hospitals NHS Foundation Trust

Taunton & Somerset Foundation NHS Trust

University Hospital of North Staffordshire NHS Trust

University Hospital of South Manchester NHS Foundation Trust

University Hospital Southampton NHS Foundation Trust

Aberdeen Royal Infirmary, NHS Grampian

Betsi Cadwaladr University Health Board

If you would like to know more about the BTS Lung Disease Registry – see the details on our website at:
http://www.brit-thoracic.org.uk/audit-and-quality-improvement/bts-lung-disease-registry-programme/

And if you want to get started: contact BTS on 020 7831 8778 or by email to registry@brit-thoracic.org.uk

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