Written evidence submitted by Action for Pulmonary Fibrosis (DEL0263)

Action for Pulmonary Fibrosis (APF)

1. APF is the main UK charity supporting pulmonary fibrosis patients and their families and funding research to finding a cure for this devastating disease.

2. We are a patient-led organization. Our Board of Trustees comprises patients, family members and leading doctors and we facilitate a network of 80 patient support groups across the UK.

3. We work with a growing community of patients, families, researchers and health care professionals. We provide personalized support to patients and their families and raise awareness of pulmonary fibrosis through campaigning, fundraising and education. We are committed to funding research to improve quality of life for people living with pulmonary fibrosis today and tomorrow.

About Pulmonary Fibrosis

4. Pulmonary Fibrosis is an incurable lung disease in which scars are formed in the lung tissues. It is a devastating and debilitating condition characterized by increasing breathlessness, disability and death. It affects 70,000 people in the UK.

5. The most common form of the disease is idiopathic pulmonary fibrosis (IPF), which has a life expectancy after diagnosis of 3-4 years. This is worse than most major cancers. Over 30,000 people in the UK live with IPF and over 5,000 people annually die from the disease. It generally occurs in people over 50 years of age and affects men more than women. Unlike other respiratory disease, it is spread evenly through all sections of society.

6. We do not know the precise cause of IPF, but scientists believe it is triggered, in people with a genetic precondition, by exposure to dust, pollution and cigarette smoke. The incidence of IPF is increasing rapidly at 2-3% annually. The reason for this is not clear.

How NHS care is organised

7. Pulmonary fibrosis patients are diagnosed and treated at district general hospitals and tertiary-level specialist ILD centres. In England, there are 23 specialist centres, each staffed by ILD consultant physicians and one or more specialist ILD nurses. Scotland, Wales and Northern Ireland do not designate specific hospitals as specialist centres but there are leading hospitals in each nation, which are regarded as ILD centres of excellence.

8. Most pulmonary fibrosis patients are seen at out-patient's clinics every 3 or 6 months and are cared for, in between, at GP practices. Pulmonary fibrosis patients sometimes suffer with severe lung infections or other conditions, including pulmonary embolism, requiring admission to hospital. Such exacerbations often result in an irreversible loss of lung function.

Issues in NHS care

9. The main issues facing people living with pulmonary fibrosis are:

Late diagnosis and misdiagnosis by GPs. APF conducted a survey of about 800 IPF patients in 2018. It found that 35% of patients are misdiagnosed by their GP (for example with heart failure or asthma), which delays referral to hospital respiratory specialists. Some GPs seem unaware of the signs and symptoms of pulmonary fibrosis.

Long waiting times for hospital appointments. Long waiting times for hospital appointments also delay diagnosis and treatment. Fewer than 50% of IPF patients get the diagnosis within 6 months of their first visit to their GP and for 20% of patients it takes over 2 years.

No clear pathway to diagnosis and treatment. Although IPF and other forms of pulmonary fibrosis have a worse prognosis than most major cancers[1], patients do not receive the same level of care as cancer patients. Cancer patients have a clear pathway designed to ensure timely and accurate diagnosis and treatment. They must start treatment within 62 days of a GP's referral and are provided access to a specialist nurse. The timeline for pulmonary fibrosis is less strict – patients only have to be seen within 18 weeks (126 days). IPF patients who attend one of the 23 specialist centres in England generally have access to a specialist nurse, but this is often not the case at district general hospitals.

Access to anti-fibrotic medicines. NICE has approved two anti-fibrotic drugs for use in IPF, which slow the progression of the disease and extend life. Unfortunately, doctors can only prescribe these medicines to patients whose lung function has declined to 80% of expected value[2]. This means that many thousands of early stage patients with a higher than 80% lung function cannot be given the drugs, even though they would work for them. The UK is the only country in Europe, where this is the case. In other countries, patients may be prescribed these drugs immediately after diagnosis.

Feeling ignored by the NHS. Although patients and their families are generally happy with the care provided at specialist centres, they are concerned that NHS often ignores them. Recent examples include:

- The NHS Long-Term Plan. Despite the fact that ILD was the 4th biggest respiratory killer, when it was published, neither ILD or pulmonary fibrosis is mentioned in the plan. As a result, ILD patients were often not included in follow-on activities.
- Government shielding guidelines. ILD was not included in the list of diseases which make patients extremely vulnerable to Covid-19 and hence ILD patients did not immediately receive shielding letters or get access to food deliveries. The advice was eventually changed, as a result of efforts of the British Thoracic Society, senior doctors and APF. Most ILD patients received their letters in mid-to-late April – a month or more after the government guidance was first published.
- Access to pulmonary rehabilitation (PR). In some parts of the UK, ILD patients are refused access to PR (exercise and education) classes because they do not have a diagnosis of COPD. When they do get accepted onto courses, they find the education component is focused on the needs of COPD patients and is of limited use to them. Despite the call in NICE Quality Standard 79 for disease specific PR courses, very few are offered.

10. Pulmonary fibrosis patients frequently ask: **why does NHS so often ignore a group of 70,000 seriously ill patients?**

Impact of Covid-19 on pulmonary fibrosis patients

[1] Only lung cancer and pancreatic cancer will kill you quicker than IPF.
[2] Given the persons, age, gender and height.

Anxiety due to late screening letters

11. Pulmonary fibrosis patients knew the government had issued shielding guidelines on 21st March and would be sending letters to patients. When the letters did not arrive, patients and their families were confused and anxious. Hungry for more information, they flooded our support line, GP surgeries, specialist centres and general hospitals looking for information. For some patients it took two months to receive their shielding letter, which provides a lifeline to food and medicine.

Out-patients appointments.

12. Pulmonary fibrosis is a terminal lung disease. Regular assessments and testing (for example, blood tests and lung function tests) are a critical part of a person's care. They allow symptoms to be managed and help people live well with the time they have left.

13. When the NHS stopped all non-essential clinics and tests, people living with pulmonary fibrosis were offered phone or video calls to their ILD teams. Although better than nothing, this led to increased anxiety, especially for people living with moderate or advanced symptoms. They worry that, without hospital tests, they cannot assess, for example, how their disease is progressing; whether their medications were having adverse impacts on their kidneys and other organs; or whether they needed to start on ambulatory oxygen or increase their oxygen prescription to protect their heart and other organs. Many pulmonary fibrosis patients know they may have less than two years to live. For them, not knowing how their disease is progressing is very worrying.

Shielding.

14. People living with pulmonary fibrosis are becoming increasingly anxious because they understand that people who are shielding may have to continue to do so for a year or more, until a vaccine is discovered. When your life expectancy is under two years, having to isolate from friends and family, especially grandchildren, if you have them, is very hard.

15. Many people with pulmonary fibrosis are over 70 and are isolated because they live on their own, do not have access to the internet, with no close friends or family. Their lifeline of a monthly support group meeting, hospital visit or seeing a neighbour is now out of reach. Their general feeling of isolation is made worse because few people in the general public know about pulmonary fibrosis, what it means, or the seriousness of the disease.

16. People living with pulmonary fibrosis understand they will be among the last people able to return to normal life. Sadly, some people will not live long enough to make that return.

Transplant patients.

17. The 60-70 pulmonary fibrosis patients on lung transplant waiting lists are very worried about the reduction in the number of transplant operations being undertaken and the implications for them. The fall is due to fewer organs being available, a shortage of ICU beds and some transplant professionals being assigned to Covid-19 wards. Most of the five centres have discussed the problem with patients and explained that those who were stable and not in urgent need would be temporarily suspended from the active waiting list. APF understands that the five centres are now gearing up for a return to normal.

Recommendations

18. Being initially left of the extremely vulnerable list has left many people with pulmonary fibrosis fearful and angry about the future. They want assurance that they will be listened to and their needs will be addressed.

19. In view of this, APF recommends that the NHS should:

- **Give a high priority to restarting ILD out-patient's clinics** so that people living with pulmonary fibrosis can be given their regular tests and have face-to-face consultations with a consultant or other member of their ILD team.
- **Ensure ILD services are adequately resourced**, especially if greater demands are placed on clinics by post-Covid patients.
- **Guarantee priority access to vaccines** for those ILD and other shielding patients with a limited life expectancy so that they can enjoy some normal life with their families.
- **Build back better and introduce a care pathway** for pulmonary fibrosis patients similar to the cancer care pathway with the same timelines.

20, We would also like to request that **NICE urgently reviews its restriction on prescribing of anti-fibrotic medicines**, in the light of emerging evidence that people on these drugs live longer, and urgently makes these drugs available to all people living with IPF.

May 2020