Patient Journey
Idiopathic Pulmonary Fibrosis (IPF) - Interstitial Lung Disease (ILD)

1. First symptoms

IPF symptoms develop over time and can vary. The most common one is shortness of breath (dyspnoea) along with:

- dry chronic cough
- gradual, unexplained weight loss
- presence of typical "Velcro Sounds"
- loss of appetite
- fatigue, tiredness and generally feeling unwell
- aching joints and muscles

Need: Symptoms look mostly like a common disease. This often leads to late and/or wrong diagnosis, delaying access to specialised care.

Ideally: Refer patient to specialist after primary care physician considers signs and symptoms. Further tests may include: lung auscultation, chest x-ray or HRCT scan and patient history/risk factors like smoking, pollution, environment, chronic viral infection, gastro-oesophageal reflux, genetic predisposition...).

2. Diagnosis

Tests performed at specialist clinic:

- Lung auscultation
- Pulmonary function test (PFT)
- Six-minute walk test
- Chest X-ray
- Blood tests
- High-resolution computed tomography (HRCT)
- Bronchoscopy for trans-bronchial biopsy
- Broncho alveolar lavage (BAL)
- Genetic counselling
- Lung biopsy

Need: Accurate and timely diagnosis preferably starting with instant referral upon presentation at the primary care practitioner, access to high quality diagnostic testing preferably at a recognized centre of expertise with a full multi-disciplinary team. Carefully exclude other interstitial lung disease (ILD).

Ideally: Diagnosis most accurate with multi-disciplinary ILD team of experts. Offer patients printed clear and concise information and refer to a local support group.

3. Treatment

There are two available drugs. It is recommended to combine them with other treatments, regular check-ups and medical examinations. Psychological support is recommended in case of progressive chronic disease.

Two views on when to start treatment:

- Immediately upon diagnosis
- Delayed until worsening of the condition to avoid possible side effects. Research is still being published.

Need: The following treatments should be available to every patient: Drug and non-drug treatments, pulmonary rehabilitation, oxygen therapy, psychological support, palliative and end-of-life care, symptom management, managing cough.

Ideally: Drug treatments available and reimbursed upon diagnosis. Patients & carers involved with the multi-disciplinary team deciding whether to start or delay treatment.

4. Surgery

Lung transplantation for IPF is available for patients who pass a rigorous assessment process. Early conversation on it is often seen as a priority due to the chronic and progressive nature of the disease.

Lung transplantation is a complex and high-risk procedure.

Only 5% of all IPF patients qualify for a lung transplant.

Need: Transplantation is the only surgical intervention which can reverse the progression of IPF, improve quality of life and life expectancy. 30% of lung transplants worldwide are performed on IPF patients. Criteria to quality for lung transplantation are strict.

Ideally: Patient qualifies for lung transplantation and by this can improve their quality of life as well as their life expectancy.

5. Follow Up Care

Use to detect disease progression

Need: Patient to be informed of upcoming clinical trials. Check for ongoing trials on IPF that the patient may qualify for. More information at ClinicalTrials.gov (worldwide) or the EU Clinical Trials Register.

Ideally: Physician or patient support groups keeping patient well informed. Some hospitals have dedicated specialist nurses who provide regular clinics and or helplines so that patients can discuss problems and receive support.

European Reference Network | Respiratory Diseases (ERN-LUNG)

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