

Pulmonary Interstitial Functional Unit

The Functional Lung Interstitial Unit (UFIP) has a multidisciplinary committee of clinical experts, radiologists, pathologists, thoracic surgeons, rheumatologists and respiratory pathophysiologists who study diffuse interstitial lung disease (MPID), and is coordinated by Dr. María Molina-Molina from the Pulmonology Service (Dr. Salud Santos Pérez), and attends to more than 1000 patients with these diseases annually.

The activities of the UFIP focus on the diagnosis and therapeutic treatment of excellence of the different interstitial lung entities. The most frequently evaluated types of MPID are: idiopathic pulmonary fibrosis (IPF), pulmonary fibrosis associated with systemic autoimmune diseases, induced FP, sarcoidosis, and hypersensitivity pneumonitis. It also includes many other less prevalent pathologies, such as different idiopathic interstitial pneumonias (NINE, NID, BR / EPID, NIA, NIL, pulmonary fibroelastosis), eosinophilic (NEA, NEC), cystic (lymphangiomyomatosis-LAM and cell histiocytosis). Langerhans cells), pulmonary microlithiasis or pulmonary proteinosis. UFIP participates in clinical trials that provide the option of combined antifibrotic treatment and coordinates as a promoter of clinical trials for lymphangiomyomatosis-LAM (LORALAM) and post-COVID Pulmonary Fibrosis (FIBROCOVID).

UFIP is a supporting Partner of the ERN-Lung ILD group (pending CSUR accreditation when the call opens and actively participates in national and international academic training activities.

Committee of Experts of the Multidisciplinary Functional Unit of Pulmonary Intersection

Pneumology Service

Dra. María Molina-Molina
Dra. Vanesa Vicens
Dra. Lurdes Planas
Dra. Guadalupe Bermudo
Dr. Guillermo Suarez
Dr. Jaume Bordas
Dra. Rosa López
Josep Palma (Enfermeria DUE)
Cristina Subirana (Enfermeria DUE)
Eva Rubio (Enfermeria DUE)
Dra. Ana Montes (Bióloga)
Dr. Carlos Machahua (Biólogo)
Leonor García (Gestor Casos, Administrativa)

Rheumatology Service

Dr. Javier Narvaez

Radiodiagnosis Service

Dr. Patricio Luburich
Dra. Belen del Rio
Dr. Santiago Bolívar
Dr. Héctor Ignacio Jofre

Thoracic Surgery Service

Dr. Ignacio Escobar
Dr. Francisco Rivas
Dr. Ivan Macià

Anatomical Pathology Service

Dr. Roger Llatjós
Dr. Eduard Dorca
Dra. Rosa María Penin Dr. Matias Guiu

Rehabilitation Service

Dra. Rosa Planas

Psychological Support

Dr. Francisco Gil (Psico-Oncología)

Clinical Laboratory

Nuria Lletja

Clinical Genetics

Dra. Lurdes Planas

Transversal healthcare activity

- Monographic consultation on interstitial pathology (5 blocks per week, including asbestos and familial pulmonary fibrosis and EPID associated with rheumatoid arthritis (RA)).
- Pneumology Day Hospital (health education, emergency care, urgent evaluation, oxygen titration by exertion, initiation and control of antifibrotic therapy or clinical trials).
- Complementary examinations (fibrobronchoscopy, cryobiopsy-RBA, PFRs, WT6m, chest CT scan, PET-CT, EBUS).
- Hospitalization.
- Day Hospital (Floor 1, Module A, Box A17).
- Multidisciplinary Lung Interstitial Committee (first and third Tuesday of the month).
- Laboratories; genetic study, cell response evaluation.
- Clinical trials of new drugs.
- Research: clinical and basic (IDIBELL).
- Psychological Support: Dr. Francisco Gil (ICO).

Online attention: ufip@bellvitgehospital.cat

Program of activities

- Informative talks aimed at patients, relatives, professionals and interested people (Community Health Education program) that take place in the auditorium of the Bellvitge University Hospital.
- **#Twitter: UFIP – Patients.**
- Nordic Walking.

AFEFPI. Association of Relatives and Patients of Idiopathic Pulmonary Fibrosis

- Association of Relatives and Patients of Idiopathic Pulmonary Fibrosis
- Interview with Dr. Molina and Alicia Boquete



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What is Idiopathic Pulmonary Fibrosis (IPF)?

It is a specific chronic form of progressive fibrosing interstitial pneumonia, of unknown cause, that occurs primarily in adults, limited to the lungs.

It accounts for 50-60% of idiopathic interstitial pneumonias, and the estimated prevalence is 13-20 patients / 100,000 inhabitants. Between 5-20% of cases have familial aggregation (familial pulmonary fibrosis). The most common symptoms are progressive suffocation and dry cough. Semiologically, the existence of dry bivalent crackles in auscultation stands out.

Diagnosis requires having ruled out all causes of pulmonary fibrosis as a first requirement. Currently, the identification of a NIU pattern in the chest CT scan, in the absence of other causes, allows diagnosis without the need for surgical lung biopsy.

FPI in the COVID 19 era:

- Use of masks, hand washing, social distance.
- COVID19 vaccination (Pfizer or Modern). Vaccination is recommended because COVID19 in IPF increases mortality and is considered high risk.

Activities performed

1. Sharpening in REIT. What we do? That means? How do we treat?

Date: March 21, 2013, at 3:30 pm Speaker: Dra. Vanesa Vicens

Targets:

- Educate the patient to identify when he may have an exacerbation and what to do
- Determine a series of diagnostic and therapeutic parameters to be performed in a standardized way for this type of pathology
- Prevent: Annual influenza vaccination and gastroesophageal reflux measures

2. Maintain physical capacity and exercise. Nordic Walking Program



3. Antifibrotic treatment

IPF has for many years been not only a minority pathology but also an orphan of treatments focused specifically on inhibiting disease progression. Advances in translational research that have led to a better understanding of the pathogenesis of IPF have led to the opening of a new era focused on inhibiting disease progression; "The anti-fibrotic era." In this sense, although at the moment there is no therapeutic strategy to improve global survival, in 2011 the first drug to inhibit the progression of the disease was approved in Europe; the pirfenidone. In addition, there are multiple antifibrotic clinical trials currently underway, and it is possible that in the next decade we may aspire to truly deliver better survival with effective antifibrotic therapeutic combinations. First of all, the introduction of pirfenidone forces us to approach the therapeutic approach more specifically. Many patients will have to live with this drug and surely with others who may come, with the benefits they bring and the possible side effects.

- [Living with Nintedanib and Perfenidone](#)

Comprehensive treatment

Comprehensive Patient Support Program (PSPi). The validation and evaluation of the effect of a home support program that includes accompanying the patient, physical activity with a specialized physiotherapist, oxygen control, and potential doubts or side effects of antifibrotic treatment have been led. The home team is part of Esteve Teijin, coordinated with the UFIP, and has been shown to improve the quality of life (ERS 2020, ERS 2021, article pending publication).

Ambulatory Oxygen Therapy in Pulmonary Fibrosis

One of the most difficult areas to treat in IPF is when ambulatory oxygen therapy is needed. On the one hand, desaturation in the REIT effort is often not associated with a feeling of suffocation in the patient. In addition this fact can appear while the patient is active even at work. This situation makes it difficult for the patient to take oxygen therapy correctly at the indicated times, and even more so when there is no scientific evidence of the benefit in these cases.

On the other hand, the oxygen therapy guideline and guidelines were determined at the time for mostly obstructive respiratory patients (mainly COPD), for whom the a priori requirements do not adapt to the specific conditions of IPF (flow). high, no autoflux,...). Lightweight portable oxygen devices do not allow autonomy for more than 2 hours at high flows, and those that allow 3-4 hours of autonomy are often heavy and difficult for these patients to carry. These and other associated issues are intended to be addressed in this blog.