Boehringer Ingelheim

Pulmonary Fibrosis

ACADEMY FOR NURSES

4 and 10 October 2023

On 4 and 10 October 2023, Boehringer Ingelheim hosted the third virtual Pulmonary Fibrosis Academy for Nurses. The event was truly global, with over 400 attendees from 31 countries.

The meeting was co-chaired by specialist interstitial lung disease (ILD) nurses and pulmonologists. The meeting featured presentations and discussions on pulmonary fibrosis by specialist ILD nurses and pulmonologists, as well as experts from the fields of rheumatology, radiology, nutrition, and psychology.

The meeting revisted and expanded on key learnings from the 2021 and 2022 Academies for Nurses, featuring a varied programme of plenary presentations, Q&A sessions, panel discussions, and audience polls.

Objectives of the meeting

- Review the diagnosis and monitoring of idiopathic pulmonary fibrosis (IPF) and other fibrosing ILDs such as **connective tissue disease-associated ILDs (CTD-ILDs), including the role of nurses in guiding patients through diagnosis**
- Gain insights into the comprehensive management of patients with pulmonary fibrosis
- Explore effective communication during difficult conversations with patients and holistic approaches to palliative care

This newsletter presents highlights from each day of the Pulmonary Fibrosis Academy for Nurses including, key learnings from each plenary presentation and insights from Q&A sessions and panel discussions.

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Claudia Valenzuela

Pulmonologist, Hospital Universitario de la Princesa, Spain

An update on IPF and other progressive fibrosing ILDs

Claudia opened the meeting with a presentation on the diagnosis, clinical course, and complications of IPF and progressive pulmonary fibrosis (PPF). Claudia discussed the shared pathogenesis of ILDs and the fact that diagnosis is challenging, requiring laboratory tests, physical examination, and imaging.[1] **Almost a third of patients with fibrotic ILDs will develop PPF,** which has a similar clinical course to IPF.[2] We learned how common symptoms such as dyspnea, cough, and anxiety impact the quality of life of patients. A comprehensive approach to the treatment and care of patients with IPF and PPF is essential, and **nurses are vital in implementing this care with other members of the multidisciplinary team (MDT)**.

Interstitial lung disease:
Diseases that affect the tissue around the alveoli, known as the interstitium; in some cases, ILDs also affect the inside of the alveoli

Pulmonary fibrosis:
Refers to the scarring or thickening of the lungs' interstitium, a key characteristic of fibrosing ILDs

Progressive fibrosing ILDs:
ILDs in which pulmonary fibrosis becomes progressive, with worsening respiratory symptoms, a decline in pulmonary function, or increased extent of fibrosis on high-resolution computed tomography (HRCT)

Progressive pulmonary fibrosis:
A term adopted by the ATS/ERS/JRS/ALAT 2022 clinical guidelines; it is defined by similar parameters to those of progressive fibrosing ILD (a combination of worsening respiratory symptoms, decline in lung function, and radiological progression) but is used exclusively for patients with progressive fibrosing ILDs other than IPF[3]

Anna-Maria Hoffmann-Vold

Rheumatologist, Oslo University Hospital, Norway

An introduction to CTD-ILDs

Anna followed with an overview of CTD-ILDs. The prevalence of CTDs and CTD-ILDs varies widely and informs screening recommendations. Owing to the high prevalence of ILD in patients with systemic sclerosis (SSc), it is recommended that all patients with SSc are screened. Identifying patients with rheumatoid arthritis (RA) who should be screened for ILD is more challenging because of the high prevalence of RA and lower prevalence of ILD, but algorithm- and risk-factor-based screening methods are emerging.[4-6] Anna highlighted that patients with CTD-ILDs can experience disease progression and extra-pulmonary manifestations, and **nurses are integral to identifying disease progression in their patients and supporting them in all aspects of their disease**.[7,8]

Pulmonary function tests (PFTs) are not required as frequently when IPF is stabilized with treatment.

Smoking is a problem for patients with ILD worldwide, and preventative measures need to be taken. Before the pandemic, PFTs were performed every 3 months, but now, with home monitoring, we are able to patients more frequently

Delayed diagnosis has a major impact on patients with CTD-ILDs as the presence of ILD is associated with increased mortality and symptoms that inhibit patients' lives

> ***Can we refer 10,000 patients with RA a year for ILD screening? No, we cannot. It is clear that we need better tools than just referring everyone***
>
> Anna-Maria Hoffmann-Vold

Sven Gläser

Pulmonologist, Vivantes Kliniken Berlin, Germany

Understanding spirometry results

Sven provided a detailed exploration of spirometry, explaining why and how we measure patients' lung function and how the results are translated into patient care. He showed us how to interpret a spirometric curve and put the current definition of PPF in the perspective of PFT readings, eg a 5% decrease in forced vital capacity (%) predicted over 1 year may only translate into a few milliliters of volume loss. Sven emphasized that assessments should be extremely stringent – ≥3 consecutive readings with <5% variability – and correlating readings with patient symptoms can help to identify disease progression. **Educating patients on interpreting spirometric curves can motivate them to perform regular PFTs, including by themselves with home spirometry.**

Helmut Prosch

Radiologist, Medical University of Vienna, Austria

The role of HRCT in pulmonary fibrosis

Helmut provided insights into the use of imaging in the diagnosis and monitoring of patients with fibrosing ILDs. HRCT is the gold standard for diagnosing and monitoring ILD; lung biopsy is not always performed because of associated risks, and chest X-ray is insufficiently sensitive to accurately identify the clinical features of pulmonary fibrosis. Helmut explained the technical details of HRCT, such as the optimal thickness of scan slices, and showed how radiologists identify key features of lung fibrosis, such as honeycombing. **As patients can be worried about radiation exposure with HRCT, Helmut emphasized that the radiation dose from modern HRCT can be very low**.

Diffusing capacity of the lungs for carbon monoxide (DL_{co}) is a very sensitive measure, but it is also very vulnerable to external influence and may require repeated testing. For example, if the patient passes a construction site and inhales dust, or smokes before their test, their DL_{co} levels may worsen

If a patient with pulmonary fibrosis is deteriorating, X-ray can be useful for identifying other causes, such as pneumonia

Nelleke Tak

ILD Specialist Nurse, Erasmus University Medical Center, Netherlands

The nurse's role in guiding patients through diagnosis

Nelleke explained that although the pulmonologist talks through diagnostic test results to patients, the nurse may discuss additional tests, outcomes, or treatments with them, and we gained practical advice on how these can be communicated. Nelleke emphasized that **nurses support the patient throughout their diagnostic journey, providing accessible information from the first diagnostic test to treatment follow-up**. Delivery of patient care may be optimized through a personal care plan. Finally, we heard insights from Erasmus Medical Center about follow-up care, such as how nurses call patients 2 weeks after a medication is started and provide advice on other non-pharmacologic therapies.

Two-thirds

of meeting attendees first see their patients after they have visited the pulmonologist, while a third see them at treatment initiation

Time

In a word cloud poll of attendees, 'Time' was the most common answer to the question, 'What are the challenges in communicating with patients with ILDs?'

Q A **Nurses have a very close professional relationship with pulmonologists; nurses often see patients together with the pulmonologist, but also separately as needed. If a patient has questions about other departments or disciplines, the nurse will contact them on behalf of the patient and help with referrals. The extent of nurses' collaboration with other professionals will depend on what the patient needs**

Roundtable discussion on the MDT: Optimizing communication between nurses and other healthcare professionals (HCPs) in diagnosis and care

The last session of Day 1 was a multidisciplinary roundtable discussion. Claudia began the session by presenting an overview of the structure and roles of the MDT before asking the panel to share their personal experiences and thoughts on how and when the nurse is involved in the MDT.

Day 1 panelists discussing optimal integration and communication in the MDT

Panel, clockwise from top right: Yasmin Gür (ILD specialist nurse), Claudia Valenzuela (pulmonologist), Geraldine Burge (ILD specialist nurse), Nelleke Tak (ILD specialist nurse), Sven Gläser (pulmonologist), Sarah Agnew (ILD specialist nurse), Helmut Prosch (radiologist)

Key points discussed by the panel:

- The MDT is different in every country and at every center. Some countries have ILD referral units, others do not
- The nurse is often closer to the patient than the pulmonologist, but while joining an MDT may be beneficial for nurses, not every center can offer this
- In some centers, ILD specialist nurses have the option to join multidisciplinary discussions when there is time. While they do not have an active role in the discussion about diagnosis, they can advocate for the patient and provide information, such as issues with treatment or deterioration, for appropriate management decisions

> ***At the diagnostic MDT, we have a very important role, and I learned everything from there. It's the most wonderful educational opportunity; I have seen more scans and pathology than many junior doctors***
>
> Geraldine Burge

Holistic management of patients with pulmonary fibrosis: From initial interventions to end-of-life care

DAY 2

Marlies Wijsenbeek

Pulmonologist, Erasmus University Medical Center, Netherlands

Supporting patients who have been prescribed pharmacologic treatment for pulmonary fibrosis

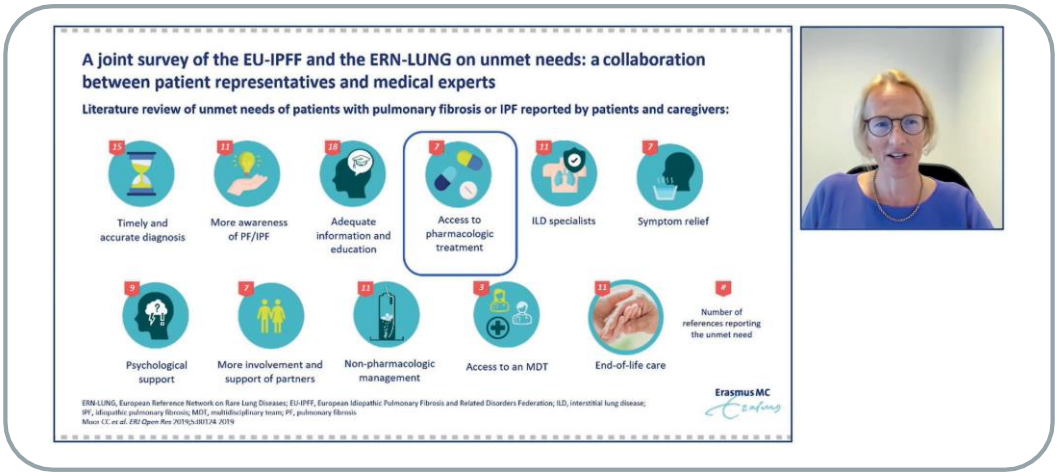
Marlies started Day 2 by revisiting the studies that led to the approval of nintedanib for IPF, SSc-ILD, and chronic fibrosing ILDs with a progressive phenotype, as well as pirfenidone for IPF.[9–12]
We also heard that while immunosuppressants are sometimes used to treat fibrosing ILDs other than IPF, there is little evidence to support their off-label use. Marlies **shared her insights into how pulmonologists may use published treatment guidelines to guide their treatment decisions**.[13] She emphasized the importance of monitoring for infections in patients receiving immunosuppressants and gastrointestinal side effects in patients taking antifibrotics, and that adaptations should be made so that the patient can tolerate and stay on the treatment.

Q A

It is important that we continue to have trials and collaborate with patients in order to advance the field and find treatments that not only slow disease progression but stop and reverse it

Recurrent, short breaks in antifibrotics, eg 1-2 weeks, can help with management of side effects such as diarrhea. The break could show whether a symptom is a side effect or part of the disease. The side effects of nintedanib seem to be similar for patients with IPF and other ILDs

Marlies presenting her insights into treatment guidelines for fibrosing ILDs



Sarah Agnew

ILD Specialist Nurse, Aintree University Hospital Trust, UK

Non-pharmacologic treatments for pulmonary fibrosis

Sarah emphasized that a holistic approach addressing all symptoms through both pharmacologic and non-pharmacologic therapies is vital for patient care. She highlighted that **cough is frequent and usually severe** in patients with IPF or PPF, interacts with other comorbidities, and **impacts patients' lives, eg causing social isolation because of the stigma of coughing following the COVID-19 pandemic**. Sarah described coping strategies for breathlessness and cough and highlighted that new approaches, including potential pharmacologic therapies, are being investigated. She stressed the importance of tailoring ambulatory oxygen therapy to each patient and understanding how this can impact a patient's lifestyle.

> ***If we don't focus on getting patients' breathing, diet, nutrition, and sleep right, how can we expect them to take on complex drugs? It's like trying to put a roof on a house when you haven't built the walls***

Sarah Agnew

Lorenzo Bergami

Strategic Nutrition Center, Italy

Managing nutrition in patients with ILDs

Lorenzo explained nutritional management and how the nutritional status of patients is assessed. Malnutrition and obesity are common in patients with IPF and PPF and associated with decline in muscle function, which can negatively affect patients' clinical outcomes. **Lorenzo outlined dietary interventions that can improve nutritional status and manage symptoms such as breathlessness, gastroesophageal reflux disease, and loss of appetite**, eg avoiding foods that cause gas and bloating. He also suggested foods and drinks to consume or avoid to manage treatment side effects such as diarrhea. Lorenzo explained that we should consider more than just body mass index when assessing nutritional health.

Around

70 %

of attendees think online home monitoring of symptom scores could be useful in daily practice

Gizal Nakshbandi

MD, PhD student, Erasmus University Medical Center, Netherlands

Digital tools for patients with ILDs

Gizal presented insights into how digital tools are reshaping the management of ILDs. She described how **online home monitoring can facilitate early diagnosis, help with symptom and side-effect monitoring, evaluate responses to therapy, and improve patient engagement and quality of care**. An online home-monitoring programme developed by Erasmus Medical Center provides patients with real-time access to home spirometry results, video consultations, an information library, and a platform for reporting patient outcomes. Wearable technology and social media can be helpful but involve challenges such as data loss and a lack of technological savviness in patients. Barriers to implementing and upscaling home monitoring in regular care include structural reimbursement, technological infrastructure, data protection, and ethical implications.

Walter Baile

University of Texas MD Anderson Cancer Center, USA

Navigating difficult conversations with patients

Walter, who developed the SPIKES model of delivering bad news to patients, shared his insights into how nurses can best support the patient and family when discussing difficult topics from diagnosis and prognosis to end-of-life care.[14] He described **key strategies to address strong emotions or difficult questions, such as giving the patient and family time to express how they feel and empathizing without providing false reassurance or attempting to 'make it better'**. He also explained how to use 'tell me more' phrases to clarify patients' feelings and needs. Nurses play a central role in helping the patient after they receive difficult news, and answering challenging questions is important in providing support.

Geraldine Burge

ILD Specialist Nurse, University of Birmingham NHS Trust, UK

Discussing palliative care with patients: The nurse's perspective

Geraldine set the scene for the Day 2 roundtable discussion on palliative care by explaining that palliative care should be an integral part of patient care in IPF and PPF to improve quality of life throughout the disease course, not just at the end of life. It can address symptoms and help patients to gain or maintain a sense of control, dignity, and self-worth. **Geraldine emphasized that cultural competence is very important in palliative care** as cultural background, faith, and healthcare beliefs influence how patients perceive palliative care. By taking the time to listen to and understand the beliefs of patients and families, nurses can help patients to discuss and accept palliative care.

Roundtable discussion on holistic approaches to palliative care

On Day 2, the roundtable discussion explored the panel's views and personal experiences of palliative care with their patients, with the panel sharing advice on how best to open up the conversation and implement palliative care.

Day 2 panelists discussing holistic approaches to palliative care

Panel, clockwise from top right: Walter Baile (psychologist), Geraldine Burge (ILD specialist nurse), Sarah Agnew (ILD specialist nurse), Nelleke Tak (ILD specialist nurse), Marlies Wijsenbeek (pulmonologist), Yasmin Gür (ILD specialist nurse), Lorenzo Bergami (nutritionist)

Key points discussed by the panel:

- To avoid misunderstanding around palliative care, it should be introduced early when patients are feeling well, eg at diagnosis, and patients should be asked what their understanding of the term is. Some patients will never be comfortable with the term 'palliative care' and may prefer 'supportive care' or 'symptom management'
- To decide when to discuss end-of-life care with patients, the 'surprise question', ie 'Would you be surprised if your patient dies in the next year? can be a helpful tool
- It can be very difficult for nurses and HCPs to decide when treatment is prolonging life or prolonging the dying process. Palliative care consultants can join the MDT to help determine the best decision for the patient
- When patients have lost their appetite, nurses should encourage, but never force, patients to eat. It is important to counsel the family not to pressure the patient to eat while being sensitive to the role of food in the family's culture

Open Q&A

The final session of the Academy was an open Q&A, addressed to all speakers of Day 2

Digital tools have become widely expected by patients and families. A conversation over the Internet can feel as genuine as one in person and is more convenient. Psychologically, patients feel empowered and uplifted by home monitoring as they can participate in their own care

If a patient with pulmonary fibrosis was to make only one change to their diet, they should reduce the amount of processed food they consume. This could be a simple 'game changer' as unprocessed foods are more nutrient dense and rich in minerals and vitamins

Patients who do not want to involve their loved ones/caregivers should be approached with sensitivity and care. Allow them to go at their own pace and enquire what they are concerned about and what the benefits are of not sharing with their loved ones

Feedback from attendees

> **Excellent programme. I wish we could do it in person. Great team, thank you!**

> **It was a very good setting for me, confirmation of my work and also new approaches**

> **Has been just very, very good!!!! Thank you very much. I appreciate very much to have been invited!**

The Pulmonary Fibrosis Academy in numbers

>400
attendees

31
participating countries

12
speakers and chairs

Key messages from the meeting

- **Diagnosis, treatment, and care throughout the disease course are enhanced when a multidisciplinary team is involved, including pulmonologists, rheumatologists, radiologists, nutritionists, and nurses, where possible**
- **A holistic, patient-centered approach to supportive care is recommended**
- **Digital tools are becoming more widely used and can help patients feel empowered in their own healthcare**
- Nurses are the main point of contact for patients with pulmonary fibrosis and play a key role in providing **information and supporting patients throughout the disease course, including end-of-life care**

We thank all panel members and attendees for their participation in the Pulmonary Fibrosis Academy for Nurses in 2023. We hope you enjoyed the meeting and will join us for another event in the future – Dr Josep Fortea Busquets, Global Medical Advisor, Boehringer Ingelheim International GmbH

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