

# ILFA Newsletter



**Patrons:** Brian O'Driscoll, Charlie Bird, Senator Fergal Quinn, Andrea Corr.

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Irish Lung Fibrosis Association  
[www.ilfa.ie](http://www.ilfa.ie)

## ILFA Christmas cards

**Have you ordered your Christmas cards yet?**

If not, please consider buying some of ILFA's Christmas cards this year. Cards come in a pack of 8, with 4 different designs in each pack. One pack costs €6 or 2 packs cost €10. If you, your family or friends would like to buy some cards, please get in touch with Denise by emailing [info@ilfa.ie](mailto:info@ilfa.ie) or by phone, 086 871 5264.



## EU IPF Charter Launch

On 29 September, a diverse group of patient groups, medical experts, politicians and civil servants gathered in Brussels to launch the 1st EU Charter on IPF. This event followed a year of co-operation between European patient groups and medical experts working to identify common issues affecting people suffering from IPF and the people who care for or treat them.

This European effort culminated in the formulation of the 1st EU Charter with its call for consistent and equal access to care and treatment across the whole of the European Union.

Co-hosted by MEPs from Bulgaria, France and Italy, the delegates represented various EU countries. ILFA was represented by Liam Galvin who met with Irish MEPs, Mairead McGuinness and Sean Kelly, to highlight the IPF charter, as well as specific national issues such as access to medical cards. Many MEPs signed the charter at the launch.

ILFA contributed more than four thousand signatures from members of the public, all calling for the charter to be implemented. This sends a strong message from Ireland to the EU about how IPF affects patients, with more than 35,000 European citizens being diagnosed each year.

Following the event, an open letter from those MEPs supporting the charter was sent to a summit meeting of the EU Chief Medical Officers in Rome on the 6th of October for their consideration. This event was the first step in a focussed campaign by ILFA working with other patient groups, drug companies and politicians, to ensure that the EU, the European Centre for Rare Diseases and other organisations work towards all the recommendations of the charter. We look forward to these recommendations becoming best practice in all the Union nations.

## ILFA Service of Prayer and Reflection

The ILFA Service of Prayer and Reflection took place on 4th October 2014. Sincere thanks to Reverend Vanessa Wyse Jackson, Fr Vincent Xavier, Canon Charles Mullen and Sr. Margherita Rock who participated in the ecumenical service in the beautiful and peaceful Mater Misericordiae Hospital Chapel. Unfortunately Fr Brian McKay was unwell and unable to join us for the occasion but he sent everybody his best wishes.



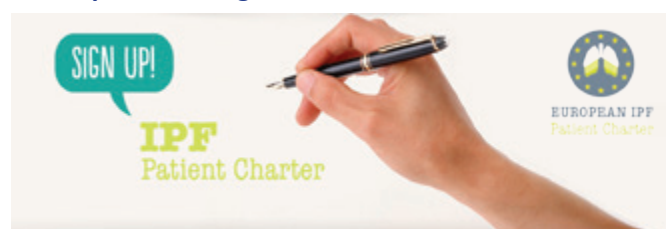
The service was very beautiful and significant for all those in attendance and indeed for those who could not join us but knew that we had gathered as a community to remember their loved ones and their own needs. Harry Carpendale treated everyone to beautiful harp music and Canon Charles Mullen played the organ to accompany the hymns. After the service, a lovely tea was served in the Pillar Room of the Mater Hospital and the get-together was a great chance for people to socialise and enjoy some tasty treats.

Reverend Vanessa Wyse Jackson, Rathgar Methodist Church, who led this year's service wrote "The third Service of Prayer and Reflection for those associated with ILFA took place this year on 4th October in the chapel of the Mater Misericordiae Hospital in Dublin. Truly ecumenical in nature, the service was conducted by clergy from the Catholic, Church of Ireland and Methodist traditions and a wide range of people attended, some of whom had travelled great distances to be with us. It was a most meaningful occasion as we remembered those who had passed on and also acknowledged the wonderful work of ILFA as it continues to meet the ongoing needs of all those who come seeking practical support, advice and encouragement."

## IPF World Week

**IPF World Week took place from 29th September to 5th October**

In Brussels, Liam Galvin of the ILFA Committee met with Irish MEPs, Mairead McGuinness and Sean Kelly, to discuss the EU IPF Patient Charter, the challenges of diagnosing IPF and the needs of IPF patients. The EU charter was signed by the MEPs following its official launch. If you have not yet had a chance to sign the charter, please do so at [www.ipfcharter.org](http://www.ipfcharter.org)



In Dublin, we had a photo shoot to mark the occasion with a number of our support group members in attendance. The sun shone for some beautiful photos and it was a lovely way to mark IPF World Week.



Special thanks to Eileen O'Sullivan, Joan Doyle and Matt Cullen for participating in the photos.

## Charles Lock

It is with great sadness that we remember Charles Lock, who passed away in October. Charles and his wife Claire Tunissen (who is an ILFA Committee member) were the Irish representatives to the group which developed the EU Patient Charter and worked so hard to ensure its endorsement by MEPs. Charles was a great friend to and supporter of ILFA and IPF patients. All of us who were lucky enough to know Charles will miss him greatly. Ar dheis Dé go raibh a anam.

## Cork Memorial Mass

The Cork memorial mass was held on the 25th September, in Saint Joseph's Church, Little Island, Cork. The celebrant was Fr. John Paul Hegarty (Parish priest of Glounthaune). Mass was offered for the deceased members of the Cork Support Group, and was attended by their families and friends.

After mass, families lit candles for their loved ones, then chatted for a while outside church yard, before heading off to the Elm Tree Restaurant, which is the meeting spot for the support group (details on page 3).





## Irish IPF Patient Charter

In 2015, ILFA with your help, will work to develop a National IPF Patient Charter for Ireland. We would like to bring together a selection of patients, carers, family members and healthcare professionals to share their experiences of IPF, with a view to identifying opportunities for improved patient care in the future. If you would like to be part of this panel please let us know by emailing Denise at [info@ilfa.ie](mailto:info@ilfa.ie) or phoning 086 871 5264. We will be in touch with interested parties early in 2015.

## Christmas Swim

If you are brave enough to take to the water to raise valuable funds for ILFA, we would be delighted to see you at Sandycove Pier, Co Dublin for a bracing dip on St Stephen's Day, 26th December at 12.30 pm. Afterwards, all will be welcome to join us at Fitzgerald's Pub for warming refreshments.



## PATIENT SUPPORT GROUPS

### Cork

The Cork Support Group meets at 11am on the last Thursday of every month at The Elm Tree, Glounthane. Contact Anne on 087 985 4587

### Kerry

The Kerry Support Group meets at 3pm on the last Saturday of every month in the Carlton Hotel, Tralee. Contact Adrian at 087 241 4004

### Midlands

The Midlands Support Group meets at 3pm on the first Wednesday of every month in the lobby of the Tullamore Court Hotel. Contact Tom at 086 150 0970

### Dublin

The Dublin Patient Support Group meets at 11am on the first Tuesday of the month in the Carmelite Community Centre, Whitefriar Street Church, Aungier Street, Dublin 2. Contact Paddy at 087 412 8612

**If you would like to talk to someone about setting up a Patient Support Group in your area, please contact ILFA at 086 871 5264**

## European Heart and Lung Transplant Championships



Huge congratulations to Paddy O'Mahony, an IPF lung transplant patient, who was one of nine athletes representing Ireland at the European Heart and Lung Transplant Championships in Vilnius, Lithuania, in July. The Irish athletes paid real tribute to their

donors, winning a total of 24 medals - six gold, ten silver and eight bronze. Paddy did us proud, bringing home a bronze medal for golf. He also participated in table tennis and cycling events. We look forward to more sporting success at the next games.

# IPF Research Update from the ERS Congress 2014

This year the European Respiratory Society (ERS) Congress took place in Munich, Germany, from 6th to 10th September. 22,000 doctors, nurses, scientists and healthcare workers from all over the world attended the congress, making it one of the largest medical conferences in the world. The ILFA delegates were Denise Dunne and Nicola Cassidy and they were joined by Irene Byrne, Senior Physiotherapist at the Mater Misericordiae University Hospital.

This year the congress programme contained many sessions dedicated to Idiopathic Pulmonary Fibrosis (IPF) including two evening symposia that were packed to capacity with over 300 people in the audience. When there were no seats left, people sat on the floor of the auditorium such was their interest in learning more about the latest research and developments in IPF. One doctor commented on greatness of having a large audience, contrasting it to the audience of 15 people who attended his lecture on IPF at the 1996 ERS Congress in Stockholm.

Another leading respiratory doctor proclaimed that "2014 was the year of IPF". This excitement reflected the upbeat mood of the audience following the publication of much anticipated results from three clinical trials that were announced in May 2014.

Two of these clinical trials (one with the drug Pirfenidone and one with Nintedanib) were successful and the medications under investigation slowed down the progression of IPF in patients with mild to moderate fibrosis over a 1-year period. A third clinical trial showed that the drug N-acetylcysteine (also called NAC) was ineffective for IPF when used alone as a treatment. During the congress, there were many talks about the finer details of the recent research results for Pirfenidone and Nintedanib, ongoing research studies and future plans.

## Pirfenidone

Pirfenidone is a drug that slows down the rate of lung fibrosis and improves survival for IPF patients with mild to moderate disease. It is only recommended for IPF and cannot be used for treating other types of lung fibrosis. Pirfenidone was approved for use (licensed) by the European Medicines Agency in 2011 and can be prescribed by respiratory consultants for patients who meet strict criteria.

After a drug is licensed, further research studies are carried out to determine the long term safety profile of the medication and to detect any new side effects in patients receiving treatment. Ongoing studies have shown that most patients tolerate Pirfenidone quite well but side effects can occur, as with all medications. The side effects of Pirfenidone can include stomach upset,

rash following sunlight exposure and rarely changes to liver function tests. Side effects usually occur within 6 months of starting the medication but very few patients have had to stop taking Pirfenidone completely. Experience has shown that the best way for doctors to manage the side effects of Pirfenidone is to:

- educate patients on how to prevent these effects;
- to adjust the dose of medication when a side effect develops;
- to stop the medication for a short time if necessary;
- to re-start the drug when the side effect has resolved.

These steps help improve the tolerability of Pirfenidone and ensure that patients can continue on the medication and benefit from its actions. Ways to reduce the impact of side effects with Pirfenidone include advising patients to take the tablets with food or at intervals over the course of a meal, and wearing a hat and sun-block while outdoors.

Preliminary results from an ongoing study called the "RECAP" study have shown that long term treatment with Pirfenidone for up to 5 years is safe and tolerable and has a good survival benefit. An estimate of the survival rate was calculated as 69% for patients taking Pirfenidone for over 4 years.

It is important to remember that these results are based on the experiences of large numbers of patients around the world rather than looking at individual cases. Furthermore no patient has perfectly stable IPF, even when they are on medication.

## Nintedanib

Nintedanib has recently been shown to slow down the rate of IPF progression in IPF patients with mild to moderate disease. It works by modifying the activity of growth factors (naturally occurring substances in the body that are necessary for growth and normal function) in the lung. The clinical called INPULSIS 1 and INPULSIS 2 were carried out for 1-year and patients with mild to



moderate IPF. Patients on Nintedanib had a lower rate of decline in the lung function tests which is consistent with a slowing down of their lung fibrosis.

The combined results for the INPULIS 1 and 2 clinical trials also revealed a trend towards an improved quality of life and a reduced rate of acute exacerbations (rapid worsening of IPF) in patients on Nintedanib but the results were not statistically significant (a mathematical calculation used to show consistency) because of the low numbers involved.

To validate these results, combinations of results (including the impact of missing results for lung functions tests due to the death of a patient or withdrawal from the study due to illness) and variations for the rate of decline for lung function tests, were analysed using different mathematical calculations. When all of the information was scrutinised using strict research methods, the results were reliable and proved that Nintedanib was an effective treatment for IPF patients with mild to moderate disease.

Nintedanib is being assessed by the European Medicines Agency and may be licensed for treating patients with mild to moderate IPF in the coming months.

### **Biological and genetic research**

There were several talks about the biological and genetic aspects of IPF. IPF is a very complex disease with many cell reactions involved in the disease process. In order for IPF to develop, there must be a genetic susceptibility (weakness or vulnerability) and a trigger (or triggers) to start the process of fibrosis (scarring) in the lungs and cause it to carry on out of control.

There are a number of genes that are believed to be involved in IPF development. While some people with the gene abnormality may be more susceptible to developing IPF, this does not automatically mean they will develop the condition. The triggers thought to be implicated include viruses, bacteria, tobacco smoke or an inhaled chemical or particle but at the moment the exact cause of IPF is not known.

When lung tissue from a healthy adult is examined under the microscope, there are subtle cell changes present that are related to the natural aging process. In order to learn more about the development of IPF and the different pathways involved, experiments need to be adapted to look at "aged lungs" (older lungs) rather than using the "young lung" model that was used in the past.

Studies looking at the presence of different bacteria in the lungs are ongoing to see if there is an increased risk of disease progression associated with specific bacterial

strains. Results to date show that there is a wide variety of different bacteria and viruses found in the lungs of IPF patients but the type of bacteria detected varies depending on which part of the lung is looked at and how samples are obtained.

It is hoped that genetic and molecular research will help identify the early processes involved in IPF development and this in turn will help scientist to better understand and predict the response to medications and treatments.

### **ILFA 2000 Steps a Day Poster Discussion**

This year ILFA was selected to discuss our work on the 2000 Steps a Day Exercise Challenge for Lung Fibrosis Patients in an educational session called "Latest insights in physical activity, exercise testing and muscles".

A poster describing the pilot study carried out to assess the usefulness of the ILFA exercise challenge was prepared by Nicola Cassidy from ILFA and Irene Byrne, Senior Physiotherapist from the Mater Misericordiae University Hospital, who jointly developed the novel exercise challenge. The poster described the development of the exercise programme and how it was successfully trialled and assessed by a group of 15 lung fibrosis volunteers. The feedback from the IPF patients was hugely positive and led to the production of the ILFA 2000 Steps a Day Challenge literature and resources.

Irene Byrne also gave a 5 minute presentation to the audience and answered questions about the ILFA exercise programme. Professor Sally Singh (Head of Pulmonary and Cardiac Rehabilitation and Consultant Clinical Scientist at University Hospital Leicester), who chaired the session, commented that the ILFA 2000 Steps a Day Exercise Challenge was "terrific" and "a really nice programme" and gave us plenty of encouragement.

There was great interest from doctors, nurses, physiotherapists, and researchers about the 2000 Steps exercise programme. ILFA received requests from healthcare professionals from Portugal, Germany, Switzerland and the Czech Republic to learn more with a view to developing a similar programme for lung fibrosis patients in their countries.

### **And finally ...**

The 2014 ERS Congress was a great success and highlighted many advances in IPF research and developments. The coming years promise exciting and dynamic developments to help our understanding of this complex disease.

ILFA's attendance at the 2014 ERS Congress was kindly supported by an educational grant from InterMune.



# Fundraising Update

**As always, we are very grateful to our committed fundraisers who give their time and energy to raise vital funds and awareness for ILFA. If you have an event or fundraising idea, please do get in touch with us, we are always happy to help fundraisers in whatever way we can.**



## Half and Mini Marathons

Our runners and racers were out supporting us in the last few months. Noreen Ring and Aileen Swords, a mother and daughter team, took part in the Cork City Mini Marathon, as did friends and relatives of the Byrne family – Angela, Ellen and Breda Byrne, Catherine and Jacinta O'Sullivan, Julie, Tracy, Christine and Dympna Sheahan and Eileen Kearney. Fiona Hedderman and her sister in law Caroline Hedderman took part in the Limerick Women's mini-marathon.



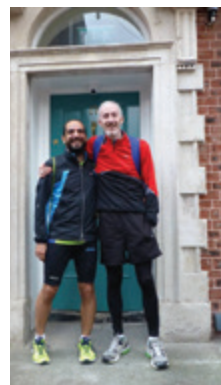
Tom Erangey undertook a half marathon in Charleville, Co Cork, his second running challenge for ILFA this

year. Eamon McGrath completed the Dublin half marathon in a very creditable time. Eamon told us, "what made it so special for me though was, firstly the amount of sponsorship that I raised and which is still coming in, and the amount of support and well wishes that I got from colleagues, friends and family in the run up to and on the day of the run itself."



## Dublin City Marathon

Alex Sayve travelled from Switzerland to take part in this year's Dublin City Marathon, in memory of his father Pierre. This was Alex's second time running the Dublin City Marathon, and this year he also represented his good friend Andrew Grehan, who unfortunately could not take part in what would have been his 7th marathon for ILFA, in memory of his aunt, Denise. Helen Matthews also undertook the marathon for ILFA as did Hugh McFadden. Congratulations to you all for such a phenomenal achievement. The ongoing dedication of our fundraisers is immensely appreciated by all at ILFA.



## Great North Run

Conor Jennings flew the ILFA flag across the Irish Sea, taking part in the Great North Run in Newcastle. "The run went fantastic!! It was a surreal occasion; there were over 56,000 people there which was simply incredible. Plus, it was actually a really sunny day in Newcastle - I think I was more shocked by that! The last few miles were tough, but I kept remembering the support that everyone at ILFA and those who donated gave me going into the race, and that really helped drive me on over the finish line."





# Pat Casey Memorial Cycle

On the 13th of September, 26 cyclists set out from the Cork City Ford Centre on Forge Hill on a 150Km cycle to Limerick and back in aid of ILFA and the Cork University Hospital.

The cyclists and helpers who graciously donated their time and effort started arriving from 9:30am. The car park in front of the dealership was awash with colour as men and women performed the safety checks on their bikes for the long haul. By 10am all the cyclists had arrived and the support teams had all provisions and equipment ready. The event was officially started by Canon Murphy of Ballyphehane church, Daniel, Pat's son and members of the Casey Family. They were led to the road by members of the Cork City Fire Brigade and Togher Pipe Band and two members of An Garda Siochana Motorcycle Traffic Division who accompanied the cyclists for a while. Unfortunately their names were not supplied, so we cannot thank them specifically but a general thanks to the assistance of an Garda Siochana is deserved. We would like to give a special thanks to the members of the Pipe Band, Caroline Dineen, Bertie McCurtain, Martin Walsh and Noel McCarthy.



The weather was with the cyclists the whole way up, extra water stops had to be provided and the event went off without a hitch. The participants stayed overnight in the South Court Hotel in Limerick and were looked after fantastically. They returned the following day on a 100km homeward trip and all 26 arrived home safe again to a reception in the Viaduct

Bar and Restaurant who once again looked after us spectacularly.

The event organisers would like to thank all participants.

**Cyclists:** Lorcan Aherne, Tim O'Sullivan, Brian O'Sullivan, Graham Desmond, Tim Finn, James Wilson, Maria Tracey, Johathon Wilson, Larry O'Donoghue, Paul O' Keefe, Damien O'Keefe, Martin Davis, Keith Doyle, Alan Jones, Paul Tobin, Fergal Quinlan, Ray Leahy, Daniel Leahy, Jamie O'Connell, Karol Horgan, Billy Cronin, Simon Carruthers, Ashling Murphy, Catriona O'Connor and Brian Aherne.

**Drivers and Safety Crew:** Pat Kelly, Mark O'Shaughnessy, Martin Casey, Alan O'Riordan, Anthony Forde, Christy Hollybrook, Jim Wilson and Aileen O'Sullivan. Billy O'Connell (Paramedic)

**Catering:** Mary Dilworth, Anne Casey, Daniel Casey, Brian Mahar. A special thanks to Brian's children, Andrew and Christine Mahar who were a great help on the day, and Noreen Aherne. Thanks to Anne and Daniel Casey for reporting on the event.



# ILFA Patient Information Day, Dublin, October 2014

The ILFA Patient Information Day took place on Saturday 18th October in Dublin.

Terence Moran, ILFA Chairman welcomed everybody to the event.

Our first speaker, **Professor Jim Egan** from the Mater Hospital, was known to many of the audience. Prof Egan spoke about what IPF is and how it is currently treated. There are a number of ways in which to manage the condition, specifically, exercise and oxygen, lung transplant and medications. The issues around exercise, oxygen and transplant were dealt with in more detail by other presenters later in the meeting. Prof Egan talked about the importance of lung transplants for people with IPF and acknowledged the huge gift that is organ donation and how we owe so much to donors and their families.

Prof Egan described recent innovations in drug development, specifically the potential availability of two drugs which help slow the progress of IPF for some patients. One of these medications, Pirfenidone, has been licenced in Ireland and the EU and is being prescribed to some patients. A second medication, called Nintedanib, is currently being assessed by the European Medicines Agency. Last week Nintedanib was approved for use in the United States. Clinical studies of the two drugs showed they both slow the progress of IPF in patients with mild to moderate disease, although the exact mechanisms are not known.

Professor Egan also launched the new ILFA patient information leaflet called "The treatment of Idiopathic Pulmonary Fibrosis".

**Irene Byrne**, Senior Physiotherapist at the Heart and Lung Transplant Unit in the Mater Hospital, Dublin, gave a presentation entitled "Turning over a New Leaf". She spoke about the importance of recognising and acknowledging breathlessness and how this can help limit the the fear experienced when breathlessness strikes. She also discussed the cycle of breathlessness and how being short of breath causes us to reduce our activity, which in turn causes us to become unfit and more afraid of breathlessness with further activity. Irene discussed the importance of patients pushing themselves during their activity to a level of moderate breathlessness. When breathlessness does strike, there are tactics to cope with it, and Irene outlined these. The STALL technique is a

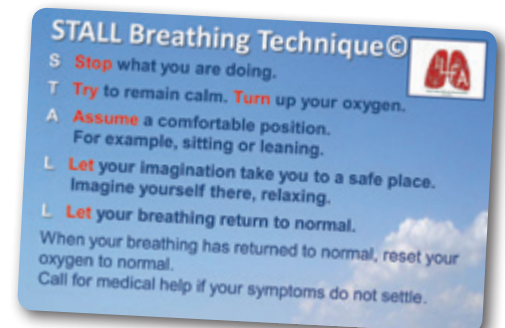
simple method of reducing the stress of being short of breath and helps patients to cope and regain control of their breathing.

- The first step is to **Stop** what you are doing.
- **Try** to remain calm and **Turn** up your oxygen to help ease your breathing.
- **Assume** a comfortable position. This may be sitting, leaning against a wall or table, or supporting yourself in a more comfortable position which allows you to breathe more easily.
- **Let** your imagination take you to a safe place where you can relax.
- **Let** your breathing come back to its natural state, but get medical help if your breathing does not improve. Irene showed everyone the STALL card which ILFA can provide, and suggested that patients and their family members keep these useful cards close to hand. Contact ILFA to get a STALL card for your wallet.

Irene also talked about oxygen, its use and oxygen prescriptions. She emphasised that oxygen is a medication

that can only be prescribed by your doctor. It is important that patients use oxygen when they need it as it will help support breathing as well as the vital organs that keep the whole body healthy. If you find that you need more oxygen cylinders (than the usual prescription for 6 cylinders per month) to allow you to leave the house and carry on comfortably with your normal activities, ask your doctor to prescribe more oxygen. The oxygen companies can only deliver what is prescribed for you by your doctor. Speak with your oxygen supplier to find out more about the range of oxygen equipment that would be best suited to your needs.

The final part of Irene's talk was about exercise and its importance in helping to improve breathlessness and overall health. Doing exercise in small





amounts throughout the day is as useful as doing it all at once. Some of Irene's recommendations are to walk up and down the hallway at home, step on the spot while the kettle boils or go to a shopping centre to walk when the weather is bad. The ILFA 2000 Steps a Day Challenge pack is especially designed for lung fibrosis patients. It is available from ILFA and can help you to gradually increase your exercise activity and tolerance. Irene stressed the importance of using mobility and other aids to help patients live life to the full, even while using oxygen. She highlighted ways of breaking up tasks or pacing activities through the day to make sure they don't drain our energy too much.

Irene's final messages were:

- Exercise should be a basic part of your life and is as important as your medications or oxygen.
- We can't "store" the benefits of exercise so we need to keep it up every day and not waste energy on things that aren't important to us.
- We must challenge and reward ourselves for our efforts, and remember that some days will be less good than others.
- Finally, it is important to talk to your doctor, nurse or physiotherapist before you start an exercise programme or if you have any concerns about what you are doing.

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**Pam Martin**, an IPF patient, spoke about her experience of lung fibrosis and how exercise helps her to keep well. Pam was diagnosed in late 2009 with IPF. At that time she was a regular swimmer and led a reasonably healthy lifestyle. She never smoked, she ran the ladies mini marathon for years and played sports regularly. While on holidays in 2009, Pam began to feel unwell and following unsuccessful treatments with a number of antibiotics, further investigations led to her diagnosis of IPF. Pam was given instructions in exercise by Irene, who was her physiotherapist in the Mater Hospital. Pam was quite unwell for the first year and a half after her diagnosis, but in spite of this, started a pulmonary rehabilitation class. She found it tough at first, but she persevered and pushed herself. Soon, she was increasing her exercise to more difficult levels and using different machines, under supervision. In the four years since starting her pulmonary rehab, Pam feels better than ever. She is no longer using constant oxygen and she has not had to stay in hospital in more than 17 months. She puts it down to three things – staying positive, working hard at keeping fit and the correct dosage of her medication, Pirfenidone. Last month Pam was on holiday in Spain, swimming in the sea and only needed

her oxygen on the airplane. Pam also gets great support at the Dublin IPF support group and she encouraged others to attend a local support group if there is one available.

Pam also highlighted the difficulties she had with her insurance company when she put in a claim for critical illness cover. The insurance company refused to recognise IPF as a critical illness but Pam persevered and eventually won her case after taking it to the Health Ombudsman, Pam recommended that all patients check their insurance cover and fight for their rights if necessary.

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The last speaker was **Zita Lawlor**, a transplant co-ordinator in the Mater Hospital, Dublin. Zita spoke about transplants in the Mater Hospital and how transplant co-ordinators support patients and their families. Zita talked about the process of referral to the unit, how patients are assessed to go on the transplant waiting list, how the list is managed, and what happens at the time of a potential lung transplant.

Patients are referred to the transplant team at the national lung transplant centre by their respiratory consultant. Once referred, they are given an appointment at the Mater Hospital for a review and assessment by the transplant team. The transplant team is a group of healthcare professionals in a wide range of fields including surgeons, nurses, transplant co-ordinators, physiotherapists, social workers and many others. At the assessment, a wide range of tests are done including blood tests, x-rays, scans and information gathering by talking to the team members. There are four main questions that the transplant team considers when assessing each patient for the list. They are:

- 1) Are this person's lungs sick enough to need a transplant?
- 2) Is the rest of the body "well enough" to undergo the lung transplant?
- 3) Is there adequate family support for the patient before and after the transplant?
- 4) Is there informed consent for the operation to go ahead?

Zita spoke about the importance of patients keeping themselves as healthy and active as possible in advance of a transplant, maintaining a healthy weight and being otherwise healthy. She also highlighted that patients must show that they take medications correctly, have good support from family and friends, and are emotionally prepared for the impact of such major surgery. When each patient is reviewed, the multi-disciplinary team meets to discuss their suitability for transplant.

This meeting has five possible outcomes for the patient – 1) listed for transplant; 2) further tests are required; 3) postpone listing because it is too early (in the course of their disease) or there is a question about some tests; 4) recommend another treatment or procedure before transplant listing; or 5) the patient is not suitable for transplant. Sometimes a person needs to be removed from the transplant list because they are no longer suitable, for example, due to worsening health or bad test results. This decision is made by the transplant team. Once a patient is on the active transplant list, they receive a weekly phone call from the transplant co-ordinator for a chat. The patient is asked to update the coordinator about any changes in their contact details, any health problems or questions, and to confirm transport details should a call for transplant come through. Patients' family members are not permitted to drive the patient to the hospital when called for a transplant, as it is believed to be too stressful. Instead, transport by ambulance is arranged.

Zita talked about what happens if a patient on the list becomes sick. Their first port of call should always be their GP or local A&E. It is important to be seen quickly and any health concerns or complications dealt with to ensure the patient is back to their best health as soon as possible. Zita recommended being treated close to home if at all possible, rather than coming all the way to the Mater Hospital. The medical team at the Mater will share any necessary information with the team at your local hospital to ensure you are well cared for.

Next Zita discussed what happens once a person on the waiting list gets a call. When lungs are offered for donation, the co-ordinators will ring a number of people on the transplant list and they will all be transported to the Mater Hospital by ambulance. Once there, blood tests are done to ensure the patient's blood is compatible with the donor's blood. This test is crucial as it gives a good indication of whether the lung or lungs could be rejected by the recipient's body. Unfortunately, not everyone who is called to the Mater Hospital will receive a lung transplant, and some people will be sent home if the operation does not go ahead for them. This may be for a number of reasons including: the donated lungs may not be suitable for transplantation; the blood tests for the patient and donor do not match; or the potential recipient is too unwell to undergo the surgery. New technologies are becoming available to help preserve donor lungs. This will help to increase the numbers of potential lungs that can

be transplanted. Zita stressed that donors and recipients are only matched by organ suitability, not by the length of time people are on the transplant waiting list.

Following a transplant it is possible for the recipient to write to the donor's family in the future to thank them. This is done anonymously. In order to protect the anonymity of both organ donors and recipients, the transplant team stresses that it is important not to post messages on social media about being called for a possible transplant.

The meeting ended with an interactive Questions and Answers session. Topics discussed included transplant procedures, living donors, and oxygen use.

Terence Moran, ILFA Chairman, concluded by thanking the audience for attending, Baywater for hosting an information stand, Boehringer Ingelheim for supporting the day and the speakers for their excellent presentations. Terence announced that the ILFA Christmas cards are now available to buy and suggested people may wish to take some of the ILFA information leaflets including the new leaflet, "The treatment of Idiopathic Pulmonary Fibrosis".

The ILFA Patient Information Day was kindly supported by Boehringer Ingelheim.

#### **PATIENT INFORMATION LEAFLET**

At the Patient Information Day, Prof Jim Egan launched the newest ILFA patient information leaflet, "The Treatment of Idiopathic Pulmonary Fibrosis". This leaflet provides an overview of the different options your medical team may suggest to help maintain your health and manage your condition. If you would like a copy of the leaflet, please do get in touch with us by phone or email.



Pictured are Caroline Daly, Pamela Martin and Professor Jim Egan



# Fergus Goodbody Memorial Lecture

ILFA was set up in 2002 in memory of Fergus Goodbody by his family, friends and Professor Jim Egan to facilitate research, education and support for people affected by Idiopathic Pulmonary Fibrosis. Every 2 years ILFA invites a leading expert in IPF to deliver the Fergus Goodbody Memorial Lecture to respiratory doctors and healthcare professionals to further their knowledge of IPF.



Speakers at the lecture were: Dr Mike Henry, Prof Jim Egan, Prof Luca Richeldi, Ms Donna Eaton and Dr Eoin Murtagh.

The 2014 Fergus Goodbody Memorial Lecture took place on October 9th in the Royal College of Physicians in Ireland, Kildare Street and was delivered by **Professor Luca Richeldi**, Professor of Respiratory Medicine and Chair of Interstitial Lung Disease at the University of Southampton.

**Professor Jim Egan** from the Mater Misericordiae University Hospital chaired the meeting and welcomed the audience of doctors, surgeons, nurses and healthcare professionals to the event. Professor Richeldi, a key global leader in IPF, delivered the keynote lecture entitled “State of the art treatment for IPF”. Professor Richeldi discussed the historical aspects of treatment for IPF, the huge growth in the number of clinical trials for IPF taking place over the last two decades and the improved quality in research design with increased numbers of patients around the world taking part in clinical studies. The results from some clinical trials have proved negative and as a result of the lack of good evidence to support these treatments, they are no longer used. Next Professor Richeldi described the most recent advances in clinical research with medications that can slow down the progression of IPF; Pirfenidone and Nintedanib. He described the research studies for these drugs in detail including how patients were recruited to the clinical trials, the different methods

and definitions used to monitor disease progression and the challenges of analysing the results of the studies due to the complexity of IPF and the difficulties of looking at populations of patients with varied stages of IPF. Both Pirfenidone and Nintedanib slowed down the progression of IPF and this positive effect was noticed within 3 months of starting the medication. He also highlighted the importance of managing patient’s expectations as both medicines will slow down IPF but will not cure the disease, and patients’ lung function will continue to decrease but at a slower rate. He also discussed ongoing and future studies that may help with the monitoring, treatment and control of IPF. Professor Richeldi ended with a positive note, encouraging the audience to be proud of the considerable progress made in IPF in recent times, especially with the huge advances in treatment options for patients.

The other invited speakers at the educational symposium were;

- **Dr Mike Henry** (Cork University Hospital) who gave a lecture entitled “Treatment of pulmonary connective tissue disorders” in which he described the incidence and treatment of sub-groups of lung fibrosis patients who also have a connective tissue disease, mostly rheumatoid arthritis, scleroderma or dermatomyositis.

- **Ms. Donna Eaton** (Mater Misericordiae University Hospital) spoke about “Lung Transplantation for IPF patients”. The success of the national lung transplant programme in the Mater Hospital was described. Selected IPF patients have improved survival with a lung transplant and early referral to the transplant unit for assessment is vital. The benefits and reasons for deciding on a single lung or double lung transplant for IPF patients were discussed in detail. Ms. Eaton praised the multidisciplinary team in the Mater Hospital for their great achievements with lung transplantation and acknowledged the positive attitude and motivation of patients on the lung transplant waiting list.

- **Dr Eoin Murtagh** (Antrim Area Hospital) discussed “Establishing multidisciplinary teams for IPF in district general hospitals”. He described the challenges and opportunities to set up an IPF clinic in Northern Ireland and the important role that nurse specialists have supporting IPF patients.

There were lively interactive discussions following each of the presentations. Sincere thanks to Professor Richeldi and all the speakers for their time and valuable insights on IPF.

## Ways to donate to ILFA

Please consider supporting ILFA's work either by making a once-off donation, setting up a regular standing order with your bank or taking part in a fundraising event. Secure on-line donations can be made via ILFA's MyCharity page or we can provide you with a Donation Card.

For more information, please call 086 871 5264 or email [info@ilfa.ie](mailto:info@ilfa.ie)

## Keep in Touch

You can keep in touch with ILFA

- by phone on 086 871 6254 (general enquiries) or 086 057 0310 (fundraising enquiries)
- by email [info@ilfa.ie](mailto:info@ilfa.ie)
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## QUIZ TIME

1. How many pairs of ribs are normally in the human body?
2. In Charles Dickens' novel "A Christmas Carol", who was Scrooge's dead business partner?
3. Who wrote the classic novel "Les Miserables"?
4. Name the original eight reindeer from the "Twas the night Before Christmas" poem.
5. What was the name of Sherlock Holmes' housekeeper?
6. The character Jack Skellington appears in which 1993 Tim Burton film?
7. Which Irishman won the Tour de France in 1987?
8. What is New Year's Eve called in Scotland?
9. Which film was the first to feature a laser gun?
10. Who wrote "How the Grinch Stole Christmas"?

## Happy Christmas

On behalf of all on the ILFA committee, I would like to wish all of our supporters a very Happy Christmas. It has been a good year for IPF in Ireland, with the prospect of more treatment options on the horizon and the National Lung Transplant Unit reaching its 100th lung transplant recently. We are hugely grateful for the on-going fundraising and awareness raising work of our supporters. We will continue to work on your behalf in 2015 to improve access to treatments and services so desperately needed. Beannachtaí na Nollag.

**Terence Moran, Chairman**



Quiz answers: 1. 12; 2. Jacob Marley; 3. Victor Hugo; 4. Comet; 5. Mrs Hudson; 6. The Nightmare before Christmas; 7. Stephen Roche; 8. Hogmanay; 9. Goldfinger; 10. Dr Seuss

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