

2014 ILFA Service of Prayer and Reflection

This year, the ILFA ecumenical prayer service will take place on Saturday 4th October at 3pm in the Mater Hospital Chapel, North Circular Road, Dublin. All are very welcome to attend.

We are delighted that the Reverend Vanessa Wyse Jackson will lead the service, during which, a candle lighting ceremony will take place to remember lung fibrosis patients who have passed away. If you would like your loved one remembered at the ceremony, please contact ILFA on 086 871 5264 or by email at info@ilfa.ie by no later than 12th September.



European IPF Patient Charter



EUROPEAN IPF
Patient Charter

The new European IPF Patient Charter is now available. ILFA has worked with other IPF patient advocacy groups to develop a European Charter to support more standardised care and equal access to diagnosis, treatment and after care options for those with IPF in Europe. The charter has been endorsed by medical experts who are actively involved in IPF management. The key elements of the Charter are:

1. Ensure early and accurate diagnosis
2. Irrespective of age, provide equal access to care, including medication and lung transplantation
3. Promote a holistic approach to standardise IPF management
4. Ensure that comprehensive and high quality information about the condition is available
5. Increase access to palliative care and end-of-life care, with support for both patients and families.

The European IPF Patient Charter is supported by IPF healthcare experts and patient organisations, along with industry, united in an unprecedented European-wide call to action to address this under-recognised fatal chronic disease and to ensure consistent and fair access to quality care across European countries.

The Charter needs your support. To sign the charter,



please go to www.ipfcharter.org, where you can read it in full, get more information and add your electronic signature supporting its adoption. You can also find out more information about the charter at our own website, www.ilfa.ie or on our Facebook page, www.facebook.com/ILFAIreland.

World IPF Week: World IPF Week will take place from 28th September to 5th October. As part of the international recognition of the week, the EU IPF Patient Charter will be launched in Brussels. Events will be taking place in Ireland to mark the week, too, so please keep an eye on our website and Facebook pages for more information.

Date for your Diary: Dublin Patient Information Day, October 18, 10am.
For full details, see page 6 of this newsletter.

Research update

There is no cure for Idiopathic Pulmonary Fibrosis (IPF) however research into new and existing drug treatments is continuing. In May 2014, the New England Journal of Medicine, a highly respected medical journal, published encouraging results from clinical trials on two drugs that slow down the rate of IPF progression; Pirfenidone and Nintedanib. The results of these studies offer significant hope for drug treatment options that may benefit patients with mild to moderate IPF.

The editorial of the New England Journal of Medicine described the results for the two clinical trials as “a major breakthrough for patients with IPF”. The clinical trials were carried out for a period of 1 year in patients with mild to moderate IPF who had to meet strict medical conditions in order to be included in the studies. The effects of the drugs are unknown for patients with more advanced or severe IPF or in those who develop acute exacerbations (a rapid worsening of fibrosis).

The results of the 2 studies are summarised below.

1) A Phase 3 Trial of Pirfenidone in Patients with Idiopathic Pulmonary Fibrosis. King TE et al. New England Journal of Medicine 2014 May 29;370(22):2083-2092.

This study was called the “Assessment of Pirfenidone to Confirm Efficacy and Safety in Idiopathic Pulmonary Fibrosis” or the “ASCEND” trial.

Pirfenidone, a drug with anti-fibrotic (anti-scarring) actions that slows down the progression of IPF, was shown to improve patient outcome in three previous international research studies involving patients from the USA, Europe, Australia and Japan. Pirfenidone is now a licensed drug in many countries, including Ireland, and can be prescribed by hospital consultants. However, the Food and Drug Administration (FDA) in the USA had reservations about the results of the initial

clinical trials and asked for more medical evidence before agreeing to give Pirfenidone a licence. The ASCEND trial was designed and carried out for this reason.

A total of 555 patients diagnosed with mild to moderate IPF took part in the clinical trial that lasted for 1 year; 278 patients were treated with Pirfenidone tablets three times a day in divided doses. A further 277 patients received placebo tablets (tablets made of sugar that would have no medical effect on IPF) three times a day. Neither the patients or doctors who took part in the study knew who received the Pirfenidone and who received the placebo.

During the clinical trial, all the patients had regular physical examinations, lung function tests and 6-minute walk tests to monitor their progress. The results of all the tests were analysed using statistics to determine if there was a genuine difference between the patients who received the Pirfenidone and those who received the placebo treatment.

The results showed that Pirfenidone treatment for 1 year had a positive effect and slowed the rate of IPF development. Treatment also improved lung function tests and improved exercise capacity for some patients, and was associated with fewer deaths. The combined data from Pirfenidone studies showed most patients receiving Pirfenidone had a survival advantage.

The drug’s side effects were mostly minor and included nausea, diarrhoea, fatigue, cough, weight loss and minor changes in liver function tests. Some patients had to stop treatment with the drug because of the severity of the side effects. Overall, Pirfenidone was considered to be a safe and effective treatment for patients with mild to moderate IPF. The ASCEND results will be reviewed by the FDA.

2) Efficacy and Safety of Nintedanib in Idiopathic Pulmonary Fibrosis. Richeldi L et al. New England Journal of Medicine 2014 May 29;370(22):2071-2082.

This study consisted of two international clinical trials called “INPULSIS 1” and “INPULSIS 2” that were carried out to determine the usefulness and safety of Nintedanib in patients with mild to moderate IPF. Nintedanib, originally developed as an anti-cancer drug, can block the action of a number of growth factors (naturally occurring substances in the body that regulate cell growth) that are involved in the progression

for IPF

of IPF. A previous clinical trial (called the “TOMORROW” trial) suggested that Nintedanib could reduce the rate of lung function decline, result in less acute exacerbations of IPF and help maintain quality of life in IPF patients.

The INPULSIS studies took place over a 12 month period in 24 countries. Overall 1,066 patients diagnosed with mild to moderate IPF took part in the 2 studies which ran at the same time; 515 patients were included in the INPULSIS 1 study and 551 patients took part in the INPULSIS 2 study. In both studies, some patients were treated with Nintedanib tablets and some received placebo tablets (tablets made of sugar that would have no medical effect on IPF) twice a day. Neither the patients or doctors who took part in the study knew who received the Nintedanib and who received the placebo.

During the clinical trial, all the patients had regular physical examinations and lung function tests to monitor their progress. Patients also completed a survey about their quality of life at the start and at the end of the clinical trials. The results of all the tests were analysed using statistics to determine if there was a genuine difference between the patients who received the Nintedanib and those who received the placebo treatment.

The results for both studies showed that Nintedanib treatment for 1 year had a positive effect and reduced the decline in lung function which is consistent with a slowing of IPF progression. The results on the length of time to the development of an acute exacerbation and the impact on quality of life between the INPULSIS 1 and 2 studies differed and were inconclusive. There were some benefits for patients however the results were not statistically significant and more research is needed.

Side effects with Nintedanib were common and included diarrhoea, nausea, vomiting, cough, loss of appetite, and changes in liver function tests. Some patients had to stop treatment because of the severity of the drug’s side effects. Overall, Nintedanib was considered to be a safe and effective treatment for patients with mild to moderate IPF. Nintedanib has yet to be licensed by the European Medicines Agency and the FDA.

Patient Support Groups

CORK:

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

**Contact Dave at
087 286 5297**

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 information about setting up a Patient Support Group in your area, please contact ILFA on

086 871 5264

Fundraising News

ILFA's supporters have been busy this summer! We have been very lucky to have many fundraisers taking part in many different events to promote our work and raise the valuable funds on which ILFA relies.



Women's Mini Marathon

The Flora Women's Mini Marathon is always an important event for ILFA, and this year was no different. More than 40 ladies took up the challenge to run, jog and walk 10km around the streets of Dublin on the June Bank Holiday weekend. Participants included the fantastic "Troy's Trotters", Antoinette O'Dowd, Kathleen Troy, Amanda Dillon, Megan Troy, Roisin O'Dowd, Hannah Chapman, Mary Troy Ryan, Rebekah Troy, Claire Finlay, Jackie Fogarty, Nancy Ryan and Helen O'Leary; as well as Helen Durney, her daughter Helen, Pauline Mooney and Geraldine Browne who continued their support both with their mini marathon day out, and a coffee morning to raise funds and awareness for ILFA.

While the sun didn't split the stones, the day was warm and dry, and a good day for fast times and even some personal bests in the ILFA contingent!

Sincere thanks to everyone who took part in the 2014 Flora Women's Mini-Marathon. We are extremely grateful to you for your ongoing support and for raising almost €12,000 for ILFA. It was great to see so many ILFA t-shirts among 40,000 participants representing so many charities. Well done to everyone – we hope you are very proud of your achievements and we really hope to see you again next year!

Organ Donation and IPF Awareness Cycle

Paddy O'Mahony, lung fibrosis patient and transplant recipient, organised a two-route cycle challenge in Dublin on 25th May, one 40km route and the other 100km. The cycle was dedicated to the memory of Elio Lodola, IPF patient and father of Brendan and Mario who worked with Paddy in putting the event together.

ILFA Patrons Senator Feargal Quinn and GAA Star Michael Darragh Macauley were on hand to sound the starting bell and see the cyclists off from the Dublin Fire Brigade's O'Brien Institute in Marino after some rousing speeches. The Dublin Fire Brigade and particularly Pat Fleming, Chief Fire Officer, very kindly supported the event, offering their technical expertise and unfailing support in marshalling, catering, entertainment and facilities. The O'Mahony family members who weren't cycling were busy organising the registration desks and ensuring the cyclists were well fed on their return.

One of the key objectives for the cycle was to increase awareness of IPF and organ donation, which was achieved through significant media coverage on tv, in print and on radio. **Paddy O'Mahony** appeared on the couch at Ireland AM along-side **Terence Moran (Chairman of ILFA)** to talk about his experience of lung



transplant and the difference it has made to his life.

The cycle was also a very important fundraiser for ILFA. We are hugely grateful to everyone who gave so much time, effort and expertise to support a really great day.

Hell and Back

Denise Dunne, our Development and Management Officer, undertook the gruelling Hell and Back course in Kilruddery House, Wicklow, on 14th June. The 12km course is billed as one of the toughest physical challenges on the calendar, and it lives up to its name, with participants having to climb a 6 foot wall just to get to the start line! From electric shocks to a chest-deep river crossing, cargo nets and a hip deep swamp, getting to the end of the course was the only objective, and avoiding a mud bath was impossible.

The camaraderie was a highlight of the day, with participants helping to pull and push other competitors over, under and through obstacles, and impromptu teams were formed and disbanded over and again. Luckily the sun shone so the cold water didn't bother people too much. With participants of all ages, shapes and sizes, it's definitely a day to remember!

Table Quiz

Huge thanks to **Lorraine Ryan** and her dedicated team of amazing volunteers who not only undertook the Great Limerick Run in May, but also held a Table Quiz in The Spotted Dog pub in Limerick in July. This was the third year for the hugely popular Table Quiz and Lorraine and her friends deserve huge credit for their great commitment and generosity. A great night was had by all with music by DJ Will, a raffle with great prizes and tasty food. Sincere thanks to all the local businesses and community who supported Lorraine in her fundraising activities.

Other activities

We had a number of runners and walkers in the Cork half and full marathons and Great Limerick Run this year, including **Tom Erangey, David Levy, Claire Corry and Siobhan Ryan**. We are delighted our sporting stars are covering the country!

Nicola Cassidy, ILFA committee member undertook a trio of challenges this year, climbing Croagh Patrick, completing the 40km cycle in Dublin and the Women's Mini-Marathon over the course of 3 weeks. One of our youngest fundraisers, **Dylan O'Brien aged 8**, took part in the Great Ireland Run on 6th April. He ran his personal best on the day, taking a full 5 minutes off his time from when he started training, which is something any of our runners would aim for!

Patient Information Days

ILFA hosts two patient information days each year. This year, the first meeting was held in Belfast on Saturday 5th April. We had a wide and very interesting range of speakers who offered unique and thought provoking insights. After being welcomed by our Chairman, Terence Moran, attendees were addressed by Joe Brolly, the GAA All-Star and pundit and organ donor champion. Joe spoke about his own experience of being a living donor and the importance of access to treatments of all kinds. He highlighted how so few patients in the North (approximately 15 out of 150) have been prescribed Pirfenidone, the only drug available which has been shown to slow the progress of lung fibrosis. He also talked about the importance of telling loved ones our preferences about organ donation, to ensure those who do want to offer the gift of life have their wishes understood and adhered to.

The second speaker of the day was Dr Joe Kidney, a Respiratory Physician at the Mater Hospital, Belfast. Dr Kidney described various symptoms and how diagnosis can be made by the treating medical team. He also spoke about the very



Announcing the next Patient Information Day

The next ILFA Patient Information Day will be held on Saturday 18th October in the Crowne Plaza Hotel, Santry, Dublin. The information day will begin at 10am and conclude at approximately 1pm with light refreshments.

Patients, carers and family members are all very welcome to attend as these sessions are always a useful opportunity to meet the experts as well as others going through similar experiences. Speakers on the day will include a physiotherapist; a medical social worker; a transplant co-ordinator; a respiratory physician; and patient who has experience of a pulmonary rehabilitation programme.

ILFA's Patient Information Days are free of charge. For further information, please contact Denise at info@ilfa.ie or by phone on 086 871 5264.



Pictured launching the new Patient and Carer Information leaflets were: (back l-r) Terence Moran, Dr Joe Kidney, Prof Jim Egan, (front l-r) Matt Cullen, Joe Brolly and Teresa Cullen

high prevalence of IPF in Northern Ireland, particularly in the Belfast area. This may be accounted for by traditional heavy industries in the area, linen mills, ship building, and power-stations. This highlights the importance of doctors taking a full employment history of patients when diagnosing IPF to identify any prolonged periods of exposure to industrial dusts or chemicals which may have triggered the condition.

The use of oxygen and the importance of exercise were discussed by Respiratory Nurse Specialists, David Moore and Noreen Baxter. David talked about pulmonary rehabilitation (a structured exercise programme designed for patients with lung conditions) to support patients and the emotional impacts that can be triggered by breathlessness and the exertion of daily life. Noreen addressed the practicalities of life with oxygen and how to make best use of the options available to patients.

Robert Early spoke to the meeting about his family's experience of organ donation, when 20 years ago Robert's father died suddenly. He was a fit and healthy man, and in spite of the tragedy of losing him at a young age, the Early family took solace from the knowledge that his organs were transplanted into 5 recipients. The touching story of the Early family's generosity moved everyone attending, particularly given Robert's obvious pride in his father's legacy.

The final speaker of the session was Prof Jim Egan who addressed three areas: organ donation, transplantation and access to IPF medication. Prof Egan urged those attending to write to their public representatives to campaign for better access to Pirfenidone as well as to challenge the perceptions around age limits for organ transplantation.

The meeting ended with a Question and Answers sessions where various topics which had been touched on in the presentations were addressed in more detail.

For more information on the Belfast Patient Information Day and more detailed reports on each of the presentations, please see www.ilfa.ie.

Annual report from the National Organ Donation and Transplantation Office

The first annual report from the National Organ Donation and Transplantation Office (NODTO) was launched in May. In 2013, there were 86 organ donors in Ireland and 294 organ transplants were performed at the three transplantation centres: the National Heart and Lung Transplant Programme at the Mater Misericordiae Hospital, the National Renal and Pancreatic Transplant Programme at Beaumont Hospital and the National Liver Transplant Programme at St Vincent's University Hospital.

In Europe, the leading countries for organ transplantation were Croatia, Spain, Belgium, Malta and Estonia. Ireland was in 16th place out of 31 countries. In order to improve Ireland's performance and increase the rates of organ donation and transplantation, the report sets out a number of targets for 2014 transplant activity, including a public awareness campaign.

In the last 5 years (2009 to 2013), 63 lung transplants were carried out at the Mater University Hospital with an average of 12 lung transplants per

year. 2013 was the most successful year for lung transplants since the beginning of the National Lung Transplant Programme in 2006 and last year 32 patients received a lung transplant. The target for 2014 is to achieve 30 lung transplants.

The average 5-year survival rate for a patient following a lung transplant was 81% compared to 51% in the United Kingdom and 55% internationally.



Results for the 10-year survival rates are not yet available as the lung transplant programme only started in 2006. The remarkable 5-year survival

outcome is a real measure of the success of the Irish lung transplant programme and is a credit to the skills of the surgeons, doctors, nurses and people caring for transplant recipients.

The NODTO Annual Report was launched by the Minister for Health, Dr James Reilly and he paid tribute to the families of the organ donors for their enormous generosity.

Have the conversation about organ donation with your family today.

ILFA submission on Medical Card Eligibility

In June, the Department of Health announced a review of the medical card eligibility process. An expert panel was convened and submissions were sought from individuals and groups with an interest in the review. ILFA asked our website, Facebook and Twitter followers to let us know your views, and with these in mind, we made a submission to the expert panel. The submission can be seen on our website. We will continue to work to have the views of lung fibrosis patients heard on the very important issues around access to medical cards. We would be delighted to hear from you with any views you have.

Please contact Denise by email on info@ilfa.ie, or post at PO Box 10456, Blackrock, Co. Dublin.

Fergus Goodbody Memorial Lecture

The Irish Lung Fibrosis Association (ILFA) was founded in 2002 in the memory of Fergus Goodbody. As part of our work to increase knowledge and awareness of IPF among the medical community, ILFA hosts the Fergus Goodbody Memorial Lecture. In previous years, this keynote educational address was delivered by Professor Ron du Bois (2012) and Professor Hal Collard (2010), both international experts in IPF.

This year, we are delighted that the lecture, "State of the Art Treatments for IPF", will be given by Professor Luca Richeldi, from the University of Southampton, on the evening of 9th October, in the Royal College of Physicians, Dublin. The event is aimed at medical professionals working in respiratory medicine and we look forward to it being a great success.

ILFA Patient and Caregiver surveys

Thank you to everyone who completed our patient and caregiver surveys. We have had a good response, both online and by post. We appreciate the effort that has gone into responding and we thank you most sincerely for sharing your experiences.

We are currently analysing the information from the surveys and we will be developing a report which will be circulated and available on our website in due course.

Patient Information Leaflets



QUIZ TIME

Questions:

1. Which metal is heavier, silver or gold?
2. The All Ireland Senior Football Championship is held for a trophy named after whom?
3. What is the largest mammal in the world?
4. In 1951 which film was shot on location in Cong, County Mayo and directed by John Ford?
5. Which is the sport where you could be out "leg before wicket" or "hit a six"?
6. When is Bloom's Day celebrated?
7. What nickname is given to Kilkenny sporting teams?
8. What record did Irish bobsledder Aoife Hoey set at the 2010 Olympics?
9. Armagh 'The Orchard County', is known for which fruit?
10. What did Lir turn his wife Aoife into after he discovered what she had done to his children?

- Answers:**
1. Gold
 2. Sam Maguire
 3. Blue whale
 4. The Quiet Man
 5. Cricket
 6. June 16th
 7. The Cats
 8. She was the tallest woman competitor (6'4").
 9. The apple
 10. A vulture



ILFA has a number of useful leaflets which together make up our Patient Information Pack. They are a helpful resource for both patients and carers of people with lung fibrosis. If you would like any of these leaflets, or a complete Patient Information Pack, please do get in touch by phone at 086 871 5264 or by email at info@ilfa.ie and we will be happy to send them to you, free of charge.

- "Getting the most out of your hospital visit"
- "Tips for Caregivers"
- "What is Pulmonary Fibrosis?"
- "Weight Management and Nutrition for Pulmonary Fibrosis"
- "Get Moving with ILFA! ILFA 2000 Steps A Day Challenge"
- "ILFA Join Us! Get Involved!"
- "STALL" Card and "Medical Alert" Card

ILFA Merchandise

Are you having a fundraising event for ILFA? We have a range of merchandise from balloons and pens to key rings that we can provide to support your fundraising efforts. And for anyone getting married who would like something different for a wedding favour, please do get in touch as we have a selection of items which are proving popular!



Find us on Twitter and Facebook – ILFA Ireland

Tel: 086 871 5264 (general enquiries)

Tel: 086 057 0310 (fundraising enquiries)

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