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January
2020

Norfolk & Norwich Pulmonary Fibrosis Support Group

Winter Meeting – 11th January 2020

We started the new decade with an early January meeting and, as always, I was delighted to see so many of you at the Willow Centre. The group has been running for six years now and each meeting reaffirms my belief that it was necessary. As I hope you all know, you are very welcome to join the monthly Breathe Easy meetings at St Faiths, as well as our quarterly group, but the advice there is more general, as the majority attending will not have fibrosing lung disease. I felt a separate group with a focus on this specific group of lung conditions might be of benefit, to provide you with information and support, and six years on, with average attendance of around 60 each meeting, that still seems to be the case.

I hope you all feel that the meetings and the newsletter are useful, not just for information but, perhaps as importantly, for the opportunity to connect with others who really know what it is like living with lung fibrosis. Many of the tips we share with you come from the experiences of the patients and relatives shared over many years working in Respiratory Medicine.

I and my colleagues in the Norwich ILD service appreciate the feedback you give to us – the group gives us an opportunity, outside the clinic setting, to hear and try to understand what pulmonary fibrosis is like for you, so that we can improve the care we offer you. We really value your input into the medical student teaching sessions, which ensure that pulmonary fibrosis is on the radar of our future doctors, and the research studies which add to the national and international evidence base.

In the last decade, there has been considerable progress in the world of pulmonary fibrosis. We have seen the introduction of the first two effective anti-fibrotic treatments, the establishment of charities focused on the needs of those living with PF and increased attention from respiratory clinical teams and researchers. At the major respiratory conferences now, there are multiple educational sessions on interstitial lung disease and well-attended symposia in the main conference halls. There is a growing network of doctors, nurses and other health professionals interested in ILD.

We still have much to learn about how best to diagnose and treat ILD, what works and doesn't and what really matters to all of you. We want to provide you with the best care that we can and see you as partners in that process. Let us know what helps you, get involved where you can with the charities working on your behalf, think about telling your story to your MPs, perhaps, and consider participating in research.

I always think Spring is a hopeful time, with new shoots promising warmer weather on the way, and I think we have reason for some hope in the field of ILD too.

Best wishes, Sandra

Pulmonary Fibrosis: what does it mean?

Sandra Olive
Consultant Respiratory Nurse, NNUH

We thought it might be useful to revisit this topic. This January, our support group has been meeting for 6 years and we welcome new members each meeting, plus a number of people who join the mailing list for the newsletters but can't easily get to the meetings. Interstitial Lung Disease (ILD), which is the term we use to include a number of different types of pulmonary fibrosis, remains an area of medicine where we are continuing to learn things each year.

Medical terms are often a bit unintelligible and the specialism of ILD is no different! I am sure you have all seen a range of long medical diagnoses and abbreviations on your copy of the letters that go to your GP and this can seem quite confusing.

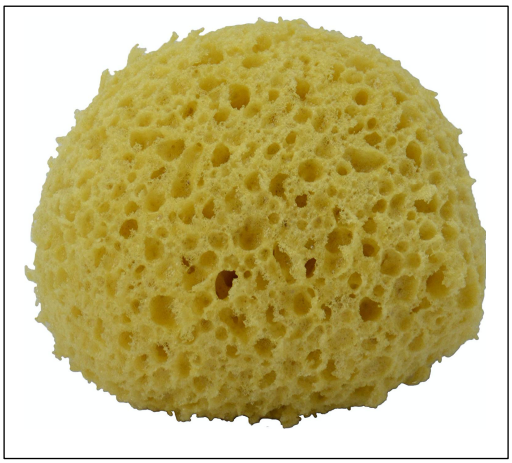
The commonest ILD is Idiopathic Pulmonary Fibrosis (IPF) and this is probably the condition that gets the most publicity. But there are many other conditions that fall under the ILD umbrella and the progression of symptoms varies enormously between individuals.

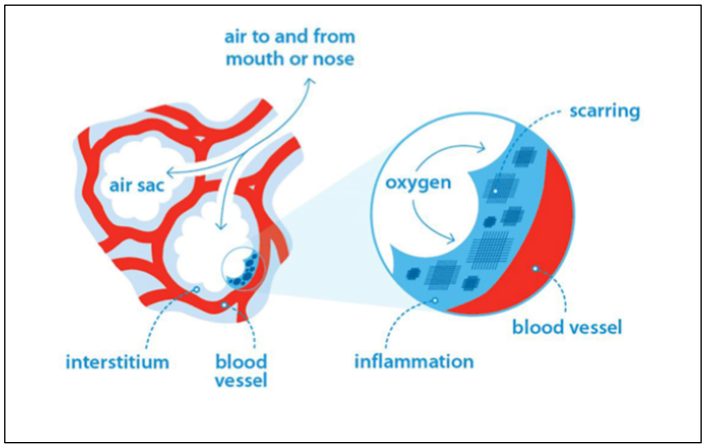
These conditions remain relatively uncommon; in an average GP practice of around 8,000 patients there are likely to be just a handful of people with diagnosed ILD, compared to around 250 people with a diagnosis of Chronic Obstructive Pulmonary Disease (COPD). Early symptoms are non-specific and common – typically cough and breathlessness on exertion, and many people accept these early symptoms initially as being a result of getting older and perhaps being a bit less fit – so early diagnosis can be difficult.

What do we mean by 'interstitial'?

Our lungs are made up of delicate 'spongy' tissue which, when healthy is very stretchy and able to expand and shrink as we breathe. Tubes lead from the nose and mouth down the main windpipe and then branch into smaller and smaller tubes (a bit like the roots of a plant get finer and finer), which transport the air we breathe in to tiny air sacs throughout the lungs.

The 'interstitium' is the supporting structure between the airways and air sacs. The lungs have a very good blood supply with a fine network of small blood vessels (capillaries) into which the oxygen from the air we breathe in is absorbed into the blood stream and carried off round the body to all our tissues and body organs.





In ILD it is the fine supporting tissue of the lungs that becomes affected by inflammation or scarring (fibrosis), reducing the stretchiness of the lungs and making it harder for oxygen to transfer across the interstitial barrier into the bloodstream.

In some conditions the damage can come on quite quickly (for example, in response to exposure to something breathed in) but very often, the damage happens slowly at a microscopic level and it is only as the damage affects more of the lung tissue, that symptoms begin to be noticed.

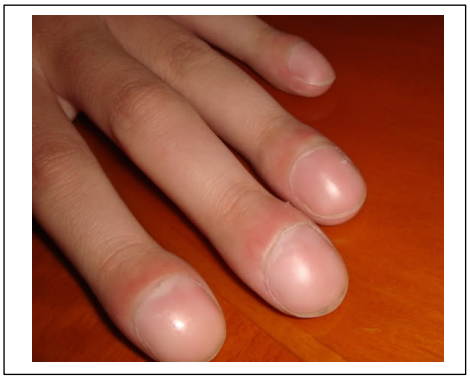
How do we make a diagnosis?

As many of you will know, it can take some time before we can make a definite diagnosis of what type of ILD we think you have.

I think of it as a bit like trying to do a jigsaw puzzle – we have to try to put all the pieces together so that we can see the picture. We don't have the helpful picture on the lid of the box to guide us and sometimes it is not possible to find every single piece, so that the picture is easy to see.

Most people first go to see their GP because they have noticed that they are getting more out of breath, often noticing things like the stairs being a bit harder work, or not being able to keep up with others where they used to be able to. Some people notice a cough, often a dry, irritating tickly cough, sometimes with some phlegm. Sometimes it is a cough that starts with a cold or a chest infection and then never quite goes away.

Your GP will have asked about your symptoms – when they started, what brings them on, what helps or makes it worse. They are also likely to ask some general questions about your medical history and things like whether you have smoked. Although ILD can occur in non-smokers, it is more common in people who have smoked and in that first brief assessment it can be difficult to work out whether the symptoms are the result of a smoking related condition like COPD/emphysema or another disease. Your GP is likely to have asked for some routine blood tests to rule out other reasons for breathlessness, like anaemia.



Examination initially may not find anything very distinctive. Sometimes there is clubbing of the fingers – the picture on the right is a more extreme example – but it is not always present, may be much less obvious and can be present in non-lung conditions too.

Carefully listening to the chest through a stethoscope may pick up some fine 'crackles' at the end of the breath in but these can be very subtle and it can be difficult to distinguish the typical crackles of ILD from other causes such as infection or heart failure.

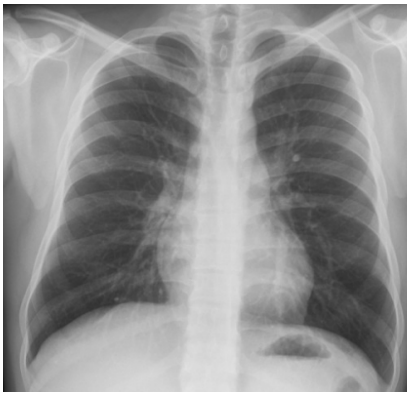
Most GP surgeries have access to basic breathing tests, which can tell if there is narrowing of the tubes of the lungs (which obstructs the air flowing out of the lungs on the out breath) or if the lungs appear smaller than expected (restriction of the volume of air breathed in and out). The reduced stretchiness of the lungs in ILD causes the lung volumes to be restricted but this can be difficult to spot in early disease, particularly in an ex-smoker when there may be some airway narrowing too.

The GP may have tried some inhalers to see whether they helped the symptoms – they might help to open up narrowed airways but they don't make any difference to inflammation or scarring in the lung tissue itself. It is often when these inhalers haven't helped that people get referred to a hospital respiratory team for more detailed assessment.

At the first respiratory outpatient appointment you will be asked a lot more questions – some of it you may well feel you have been asked about before but it is important for us to try to gather as many clues as we can from your family history, early life, working life, pets and hobbies, earlier illnesses. We all breathe many things in over the years and for most of us they don't cause illness but we know there are some exposures or occupations that carry a higher risk for lung disease; coal dust, asbestos, or exposure to silica dust in stone are all known to increase the risk of lung scarring (pneumoconiosis, asbestosis, silicosis) but also working in environments with long term exposure to metal working fluids, birds, moulds and shellfish processing can cause problems. There are probably other less common exposures where there is not enough evidence to say with certainty that there is a link and you may have seen some of the press around vaping – e-cigarettes are likely to cause less damage than continuing to smoke tobacco but we don't know yet what the long term effects on the lungs will be. Some ILDs are known to be associated with auto-immune conditions – where the body's immune system works against itself – such as Rheumatoid Arthritis and scleroderma or can be a side effect of some medications used to treat other medical conditions.

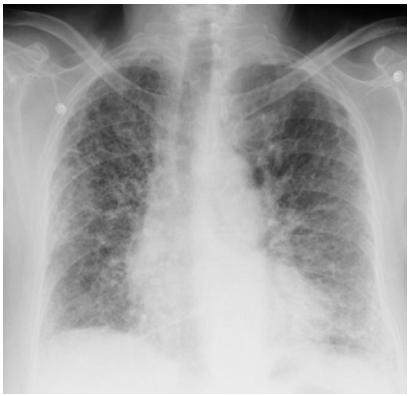
Tests and more tests!

If you hadn't already had a chest x-ray, you will certainly have had one when you come to the respiratory department. In early ILD, there may not be much to see – the image produced is a 2-dimensional picture and may not look abnormal. Even when it does, we can't tell from the x-ray what is causing the abnormality.



The x-ray on the left is a normal x-ray with the white bones visible and the large white area of the heart in the centre but the lungs look pretty dark.

On the right, you can see white 'fluffy' areas in both lungs, which is what we see when there are changes in the interstitium (typically scarring or fibrosis).

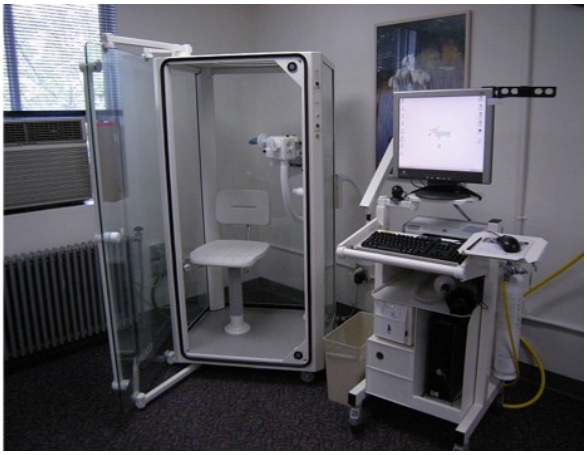


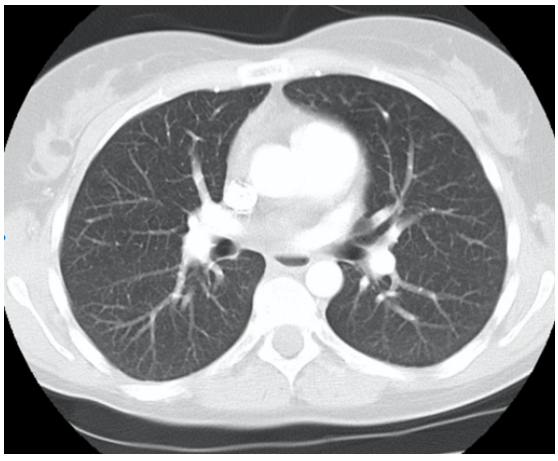
You are also likely to have had more detailed measurements of your lung function – these tell us something about how well the lungs are working and in what way they are working less well than they should be. As you know, they are hard work! It is important that the lung physiologists and nurses doing the tests get the best results they can, otherwise the information may be inaccurate and could result in extra unnecessary tests or just not help with completing that jigsaw puzzle of clues.

Some of the tests are done in the 'box' – which many people find a bit difficult. This gives us information about the volume of air that can be moved in and out of the lungs and of how well the lungs can transfer oxygen, which is really useful, particularly when we are trying to work out the correct diagnosis when we first see you but also for monitoring changes that can't always be picked up on an x-ray.

You will have had more blood tests too – these look for some of the less common causes of ILD, like those related to auto-immune conditions.

Sometimes GPs arrange for a high-resolution computerised tomography (HRCT) scan; if not, this will be arranged as it gives us much more information than the plain x-ray. The scanner is like a large 'Polo mint' shaped machine, which takes multiple pictures in 'slices' across the body provide a more 3D image of the organs in the chest. Pictures are taken with a full breath in and a full breath out, which makes it possible to see if air gets 'trapped' in the lungs.





On a normal CT scan the lungs look quite evenly grey in colour, with the airways, major blood vessels and heart showing up as lighter or white structures. In this image on the left, you are looking from the feet, with the backbone at the bottom of the picture and the breastbone at the top, above the heart.

Where there are interstitial changes in the lungs, the appearances show abnormal white areas and the radiologists look for specific patterns that indicate scarring, or fibrosis, like 'honeycomb' areas or airways pulled open by the scarring, called 'traction bronchiectasis'.

Putting it all together…

Once all the tests have been done, your case is discussed at our weekly Multidisciplinary team (MDT) meeting. An expert panel of consultant respiratory doctors, consultant radiologists, consultant thoracic surgeons and an ILD specialist nurse meet and the clinician you saw in the clinic (usually the respiratory consultant but not always) presents the 'story' of your symptoms and background, whilst your x-rays and scans are reviewed. There is good evidence that putting more heads together like this helps to get a more accurate diagnosis and to ensure every angle has been considered. The meeting agrees a diagnosis if possible and recommend a management plan. Sometimes it is not possible to make a final diagnosis and the meeting may suggest more tests, such as a biopsy if possible, or a period of monitoring. Sometimes, what happens over a few months tells us more about the most likely diagnosis and may be the most appropriate course of action.

What does this mean for you?

It is really important that we work out as well as we can what condition you have, as the treatment and future management is not the same for all ILDs. We do realise that the waiting for tests, results and follow up appointments is difficult and that the uncertainty presents lots of challenges for you and your loved ones. We would like to be able to support you better with this and our aim is for you to have contact with an ILD nurse specialist early on in your contact with the NNUH ILD service. At the moment, we know we don't manage this for all of you but I hope you feel you can get in touch if you don't understand the information in the letters to your doctor or are not clear about what is happening.

What about treatment?

For some people, regular monitoring is the most appropriate plan and you will usually have regular follow up appointments in the respiratory clinics.

If you are diagnosed with a condition where most of the changes are thought to be the result of inflammation, such as hypersensitivity pneumonitis (e.g. from exposure to birds, moulds or some other environmental dust or an unknown cause) or an auto-immune related condition, treatment aims to 'dampen down' the immune system and reduce inflammation. This is usually with several weeks of oral steroid tablets (Prednisolone) in the first place and perhaps the addition of another immunosuppressant medication like Methotrexate, Azathioprine or Mycophenolate. These are given to keep on top of the lung condition with the least side effects from steroids, which are a risk if you are on long term Prednisolone at higher doses. Steroids can cause indigestion, make you hungry and at risk of weight gain, disturb sleep, cause mood changes, increase blood pressure and upset blood sugar control. In the longer term, they can also cause bone-thinning, muscle wasting, eye problems, bruising and skin thinning and can affect the body's essential natural steroid production. Quite a long list of possible problems! But they do remain a useful and effective drug, we just have to use them as sparingly as possible and give some additional medication to reduce the risks.

The commonest fibrotic lung condition, idiopathic pulmonary fibrosis (IPF), is the only condition at the moment for which two anti-fibrotic medications are licensed for in the UK. These are Pirfenidone, which has been available since 2013 and Nintedanib, since 2016. These medications cannot cure the fibrosis but have been shown to slow the rate the fibrosis progresses, on average halving the rate of decline in one of the lung function test measurements (called the FVC). Unfortunately, they are unlikely to make you feel better, rather we hope they slow the getting worse. We are only able to prescribe them in the UK when FVC is less than 80% of what is predicted for you. Many people are quite stable for some time, with little change in their symptoms or breathing tests, but it is important that we follow you up regularly so that we pick up promptly when things start to change and you might be eligible for treatment. Both drugs can cause sickness, diarrhoea and some other less common side effects and both need close monitoring with regular blood tests.

Alongside possible medication, it is just as important that we consider non-medication treatments – keeping active is vital and many people benefit from attending a Pulmonary Rehabilitation programme, which is a structured exercise and education course over several weeks for people with breathlessness. Symptom control clinics, run in this region by the specialist palliative care team, offer expert advice on managing difficult to cope with symptoms like cough and breathlessness.

A handheld fan can be very helpful to relieve episodes of breathlessness and we have some available in the clinic, provided by the support group. If you want to know more about these other treatments and services that we can refer you to, do ask when you come to clinic.

If your oxygen levels are low, oxygen therapy may be helpful for symptom relief and to help you to keep as active as you can, though it doesn't help when oxygen levels are normal.

Support!

Perhaps most importantly, getting a diagnosis of an ILD can be difficult and can feel life changing but, for most people, changes to what they can manage are fairly gradual and it should be possible to continue making the most of life, perhaps with some adaptations or by taking a bit more time. Ask questions, find out what you can do to help yourself and talk to those around you, so that they know what you can and can't manage. Recognise that you may have good and bad days and be kind to yourself if it is a bad day – you may just need to rest a bit and save some activities for a day when you have more energy. Talking to others who are living with similar symptoms can be helpful both for the person living with the condition and their family members – some people find online forums helpful and you are always welcome at any of the local support groups.

Sharon Moon, Fundraising Support Coordinator Action for Pulmonary Fibrosis

"My role within APF is to support fundraisers. We are so lucky as a charity to be approached by so many lovely people wanting to do something to either raise awareness, funds or both. In order to continue to raise awareness, campaign for better care, provide support to families affected and fund research into effective treatments we need to keep inspiring people to get involved."

APF are encouraging at least one person from each of the 75 PF support groups around the country to sign up to a fundraising event in 2020. It could be anything – from the more extreme, like sky-diving, biking or hiking, to baking a cake, holding a coffee morning or anything and everything in between.

Sharon can help with fundraising tips, merchandise, planning and ideas if you would like to take on a challenge or host an event. As a group, we plan to hold a fundraising event for our September meeting to mark Global PF awareness month – as yet to be decided! Let me know if you have any ideas and perhaps we can ask for Sharon's help! If you, or one of your family or friends, would like to raise funds for APF, you can contact Sharon direct via email at fundraising@actionpulmonaryfibrosis.org or via the APF office on 01733 475642.

Would you like to help train future doctors?

Norwich Medical School welcomes patient volunteers to help UEA medical students learn how to undertake consultations and physical examination. There are 3 respiratory sessions a year, plus sessions for the students' exams, led by NNUH respiratory consultants, so the UEA try to keep a pool of people they can call on. The students learn important skills, including listening to chest sounds, detecting subtle signs from clinical examination and taking a good medical history.

The sessions take place at the Bob Champion Research & Education Building and last up to 3 hours. Volunteers are offered reserved parking close to the building. You get reimbursed for your travel costs and are offered tea/coffee and biscuits. to add to their pool of people they can call. Those of you who have helped in the past always tell me it is an enjoyable experience and it is enormously valuable for our 'doctors of the future'.

If you are interested, do chat to your consultant or give Sandra a call, or you can contact Caroline Coombs, Medical School Administrator, direct on 01603 286618 or via email caroline.coombs@nnuh.nhs.uk

Group News

The Norfolk & Norwich Pulmonary Fibrosis Support Group has a membership of over 150 now. If you have picked up the newsletter in the respiratory department waiting room and would like to be added to the mailing list, please do get in touch on 01603 289654 and we will send you a membership form. You are very welcome to attend the meetings, with a family member or friend if you wish, but if you are not able to get to the group, I hope the newsletter is informative and helps you to feel less alone with this condition. If you are coming to the meeting on your own, please don't worry – a good few people do come alone; we are a friendly group and there are always people to chat to.

If you have an email address, we will send the quarterly newsletter and other group mailings by email, which saves some postage. If you would prefer a paper copy, please do let me know and I will make sure one is posted to you. Your name will remain on our mailing list unless you let me know that you wish to be taken off it.

Group Funds

As of 31st December 2019, the support group fund balance is £5016.84. We have £39.74 in petty cash at present. The raffle and donations from the January meeting came to £93.59.

We have provided pictures for the new quiet room in the Respiratory Outpatients Department and will make a large donation towards the furniture in the room (once the bill comes in!), a big part of which was from the generous donations of the family and friends of Mr Russell Warne, pictured below at the room opening. The room is completed and in regular use already and is making a real difference to the quality of experience we can offer – some of you may remember that we often had nowhere in the department to take some time to talk things through after a clinic appointment and this is a much nicer space to be in.

Thanks to group member Karen Simmons who decided to ask Sainsburys and Tesco for a donation and managed to secure a £10 voucher from each – well done Karen! The October newsletter cost £140.00 to produced and we have spent less than £20 on cups and refreshments for the last couple of meetings.

Any donations can be made payable to the Pulmonary Fibrosis Support Group Fund 052, c/o Sandra Olive, Consultant Nurse, Dept of Respiratory Medicine, NNUH, NR4 7UY.

Your committee

Professor Andrew Wilson, Honorary President
Sandra Olive, Chair/Editor
John Stroud, Treasurer

Next Meeting – Saturday 28th March 2020

The NNUH Pulmonary Rehabilitation team will be joining us to talk about the importance of maintaining activity levels. We know this can be difficult with a lung condition but PR is specifically designed for people who are breathless and has been shown to improve activity levels and ability to cope with symptoms.

Keeping Active – Pulmonary Rehabilitation and more!

Keeping active is important for all of us, including when you are living with a long-term lung condition. Find out why muscle fitness matters, how PR helps and how to get on a PR programme.

All meetings at the Willow Centre, Cringleford, from 2-4pm.

Useful Information Sources

Websites:
There is a lot of information on the internet – some reliable and helpful, some less so, and it can be difficult to know what you should believe. The following are reliable sources and have lots of useful information and resources. Do ask if you read anything that needs clarifying.

www.actionforpulmonaryfibrosis.org
www.pulmonaryfibrosistrust.org
www.blf.org.uk (British Lung Foundation)
www.europeanlung.org/en (European Lung Foundation)
www.sarcoidosisuk.org (UK Sarcoidosis Charity)
www.angliaasbestos.co.uk (support groups for those living with asbestos related disease, run by solicitors specialising in asbestos claims)
www.breathingmatters.co.uk
www.pulmonaryfibrosis.org (US Pulmonary Fibrosis Foundation)

Action for Pulmonary Fibrosis: Advice Line 01223 785725
Aims to give a voice, at national level, to those affected by pulmonary fibrosis to influence policy, research and education. Raising awareness and supporting individuals through PF specific support groups, information and advice.

Pulmonary Fibrosis Trust: Advice Line 01543 442191
Aims to provide personal practical, emotional and financial support to those affected by pulmonary fibrosis. Can help with stairlifts, mobility scooters, oxygen away from home and more.

British Lung Foundation: Helpline 03000 030 555
Information and support for people living with or affected by any lung disease. Oversee a network of UK Breathe Easy Support Groups. Provide wide range of information on website and in leaflet/booklet form.

Breathe Easy Norwich *(contact Paula Browne, Respiratory Nurse Specialist – 01603 289779)*
Monthly meeting: First Tuesday of each month, from 2-4pm, function room at St Faiths Community Centre, Horsham St Faiths, NR10 3LF. Free to attend, no need to book, all very welcome. Bi-monthly newsletter.

Singing for Breathing
Informal singing group with Breathe Easy Norwich. Second and last Friday of each month at the Willow Centre, Willowcroft Way, Cringleford, NR4 7JJ, 1.00pm for 1.30pm start.

Respiratory Nursing Team, NNUH

Sandra Olive, Consultant Nurse, Respiratory Medicine – 01603 289654
Oxygen/Pulmonary Rehabilitation Specialist Nurses – 01603 289779

We have a team of specialist respiratory nurses who you may meet at Pulmonary Rehabilitation, in the hospital wards or outpatient clinics, or in the home oxygen service. We are based in the Department of Respiratory Medicine, Level 3 East, NNUH, NR4 7UY. You can contact us for non-urgent advice and any of the team will do their best to find the answers to your queries. Please bear in mind that the telephone lines usually go to an answerphone or secretary so please leave your name, some general information about your query and a contact number and we will get back to you, usually within 48 hours.