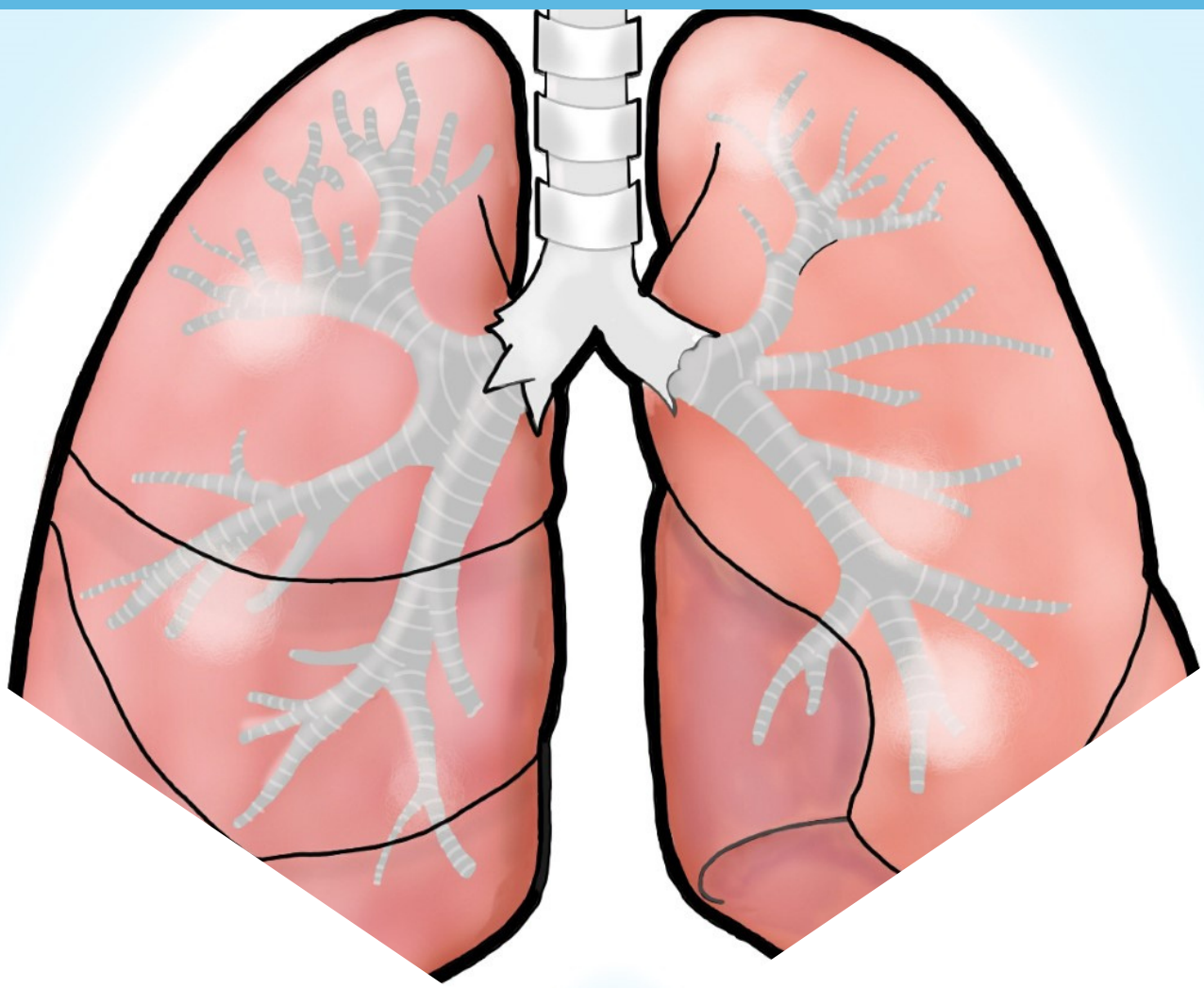


A PATIENT'S GUIDE TO

PULMONARY FIBROSIS

SECOND EDITION

Great Staff - Great Care - Great Future

UNIVERSITY OF HULL

Pulmonary Fibrosis Trust

NHS
Hull University Teaching Hospitals
NHS Trust

CONTENTS

WHAT IS PULMONARY FIBROSIS (PF)?

Pulmonary - LUNG **F**ibrosis - SCARRING

HOW DOES PF AFFECT THE LUNGS?

- Thickening of the lower parts of the lung (alveoli)
- The lungs stiffen as a result of tissue scarring, making deep breathing more difficult
- Scar tissue reduces the amount of oxygen that leaves the lungs and enters the bloodstream
- Lung size and capacity is reduced

Detailed information about this is in figure 1 (see page 4).

It has been estimated that **70,000** people in the UK are affected by PF [1]

FIGURE 1 TERMINOLOGY

Bronchioalveolar stem cell (BASC)	**Ciliated cells**	**Club cells**	**Alveolar type I (ATI) cells**
Possible role in lung regeneration	Remove dust, bacteria and mucus up to the back fo the throat	Protect lung tissue by secreting proteins	Exchange oxygen and carbon dioxide between alveoli and blood
ATII cells	**Macrophage**	**Neutrophil**	**Endothelial cells**
Involved in immune response	A cell which removes unwanted microscopic particles	A type of immune cell which helps to fight infection	Line the interior surface of vessels
Extracellular matrix	**Fibroblasts**	**Interstitium**	**Bronchial smooth muscle**
Network of molecules which provide support to surrounding cells	Maintain the structural framework of organs	Tissue between lung alveoli and the bloodstream	Induces airway narrowing and contributes to inflammation

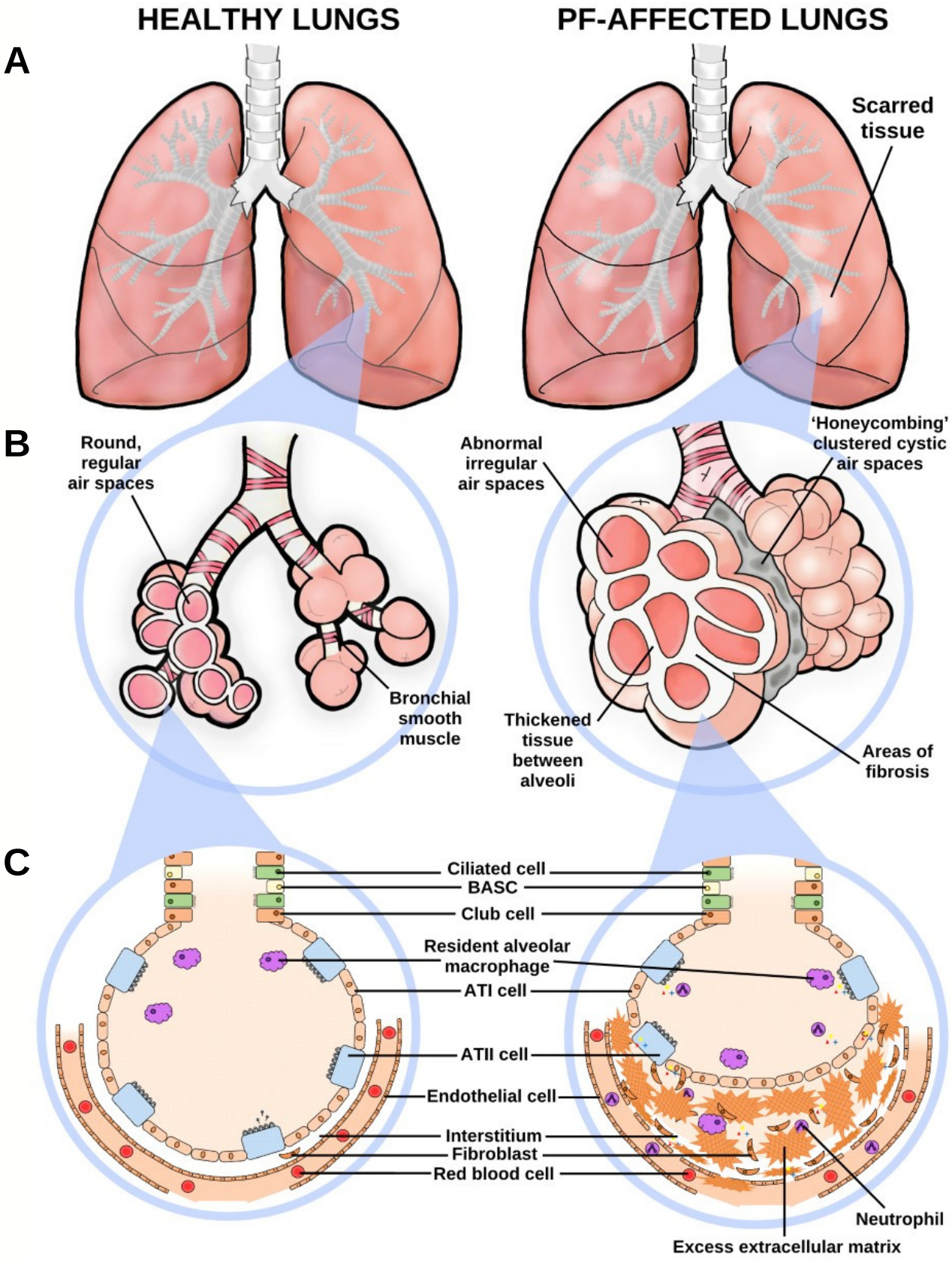


Figure 1. Structural and cellular changes seen in fibrotic lungs damaged by pulmonary fibrosis (PF). (A) Healthy lungs on the left and PF affected lungs showing areas of fibrosis alternate with areas of normal lung on the right. (B) PF causes the tissue between the alveoli to thicken and the airspaces become irregular with a 'honeycomb' appearance, right. Compared to healthy 'normal' alveoli, left. (C) In the PF affected lungs, the lining cells are damaged with the build up of extracellular matrix and fibroblasts, as well as, more macrophages and neutrophils.

WHAT ARE THE RISK FACTORS FOR DEVELOPING PF?

Whilst some people are genetically at an increased risk of developing PF, there are also environmental risk factors that can impact the development of PF such as breathing in smoke or dust.

BIOLOGICAL RISK FACTORS

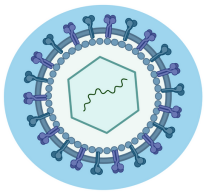
Genetics

- Some genes have been identified to increase the risk of developing PF
- There is currently no testing available to determine who may be susceptible to developing PF

Age

- The average age of diagnosis is 65 years
- PF is rare in people under the age of 40

Viral Infections

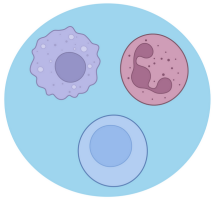


- Exacerbations of PF can be triggered by viruses, other infections or trauma/surgery

Gastroesophageal Reflux

- The spillover of stomach chemicals into the lungs may lead to scarring
- Elevating the head of your bed may reduce reflux

Autoimmune Disease



- The immune system attacks it's own cells in autoimmune disease
- Autoimmune diseases such as rheumatoid arthritis (RA) and scleroderma are commonly associated with PF
- RA causes inflammation of the joints

Sex

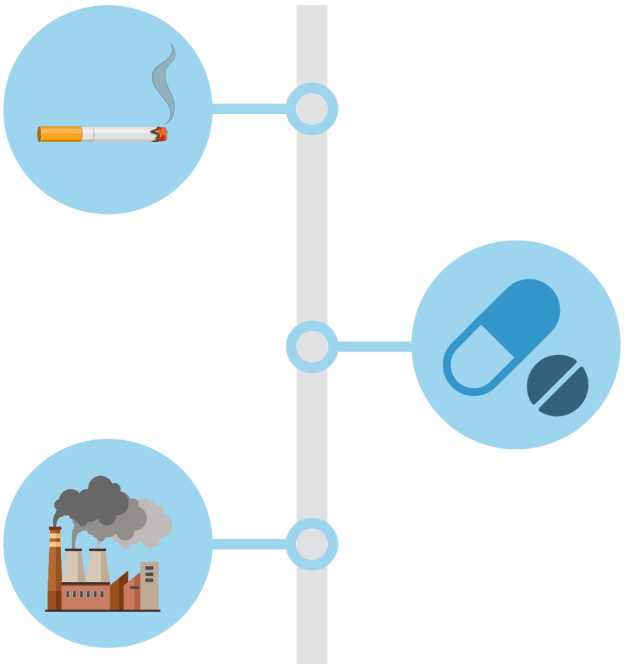
- Men are more commonly affected by PF than women

ENVIRONMENTAL RISK FACTORS

Cigarette smoking:

- Brings in inflammatory cells to the lungs from the blood
- Leads to inflammation and damages lung tissue

Industrial exposure:
Irritation of the lungs and increased tissue scarring can be caused by asbestos, organic dust from agriculture, metal and wood



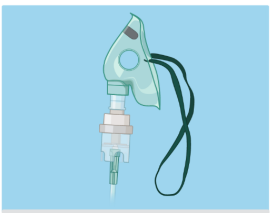
Medication/drugs:

- Cancer treatment such as radiotherapy and chemotherapy can cause damage to the lung tissue
- Some medication and drugs have been associated with PF disease

What if the cause of PF is unknown?

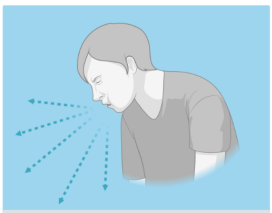
Idiopathic PF (IPF) refers to PF that develops because of an unknown cause

WHAT ARE THE SYMPTOMS OF PF?

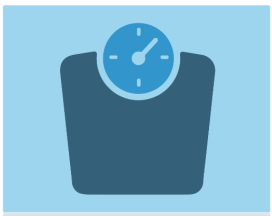


Progressive difficulty in breathing

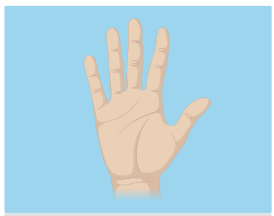
Tiredness



Persistently dry or mildly mucus productive cough



Loss of appetite and weight



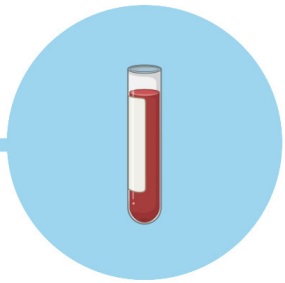
Rounded and swollen fingertips (clubbing)

HOW IS PF DIAGNOSED?

SYMPTOMS

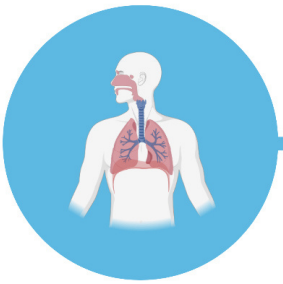
Stethoscope: 'Velcro' like crackles are heard when listening to the lungs with a stethoscope

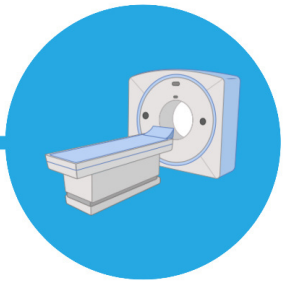
COMMON



Blood test: Blood from the vein is drawn to identify antibodies

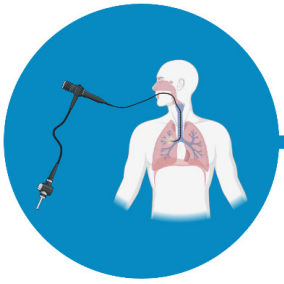
Lung function tests: Determine how well lungs function by measuring lung size and airflow in and out of the lungs



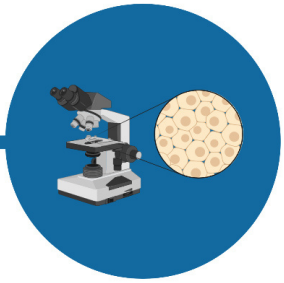


High resolution computed tomography (HRCT): Creates detailed images of the inside of the chest and the characteristic PF pattern is shown as honeycombing

Bronchovascular lavage: A small flexible tube (bronchoscope) is used to sample cells from the lower part of the lungs (alveoli) for examination by a pathologist



RARE



Thoracoscopic lung biopsy: A thin tube is placed to extract small samples of the lung from multiple lobes for testing in a laboratory

MULTIDISCIPLINARY TEAM MEETING

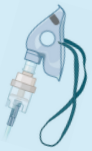
HOW IS PF MANAGED AND MONITORED?

Pulmonary rehabilitation

- An exercise programme designed to reduce breathlessness by teaching breathing techniques and improve general fitness level

Medication

- There are two drugs which slow down disease progression by reducing lung scarring: nintedanib and pirfenidone
- This medication is only currently available to patients diagnosed with IPF



Oxygen therapy

- Supplemental oxygen is used to treat low oxygen level (hypoxaemia) in the blood caused by scarring via a nasal cannula or a face mask
- Ambulatory oxygen therapy is given/taken additionally during exercise and everyday living activities
- As the disease progresses (severe hypoxaemia) you may require constant oxygen supplementation

Stomach acid reduction

- Omeprazole and lansoprazole are medications which reduce the production of stomach acid and antacids neutralise stomach acid

Cognitive behavioural therapy/anti-depressants

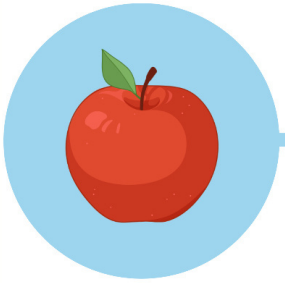
- To manage possible anxiety/depression caused by the PF diagnosis

Lung transplantation

- Transplant of one or both lungs from a donor
- Only 5% of PF patients qualify for the procedure for a number of reasons including their age and other health problems

WHAT CAN I DO?

Maintain a healthy diet and weight:
By eating balanced meals which include fruit, vegetables and fibre and reducing saturated fats, salt and sugar



Stay relaxed:

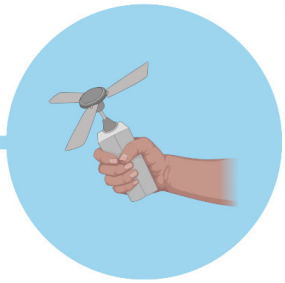
- Avoid stressful situations which can cause excessive oxygen consumption
- When you feel anxious or stressed perform yoga exercises to aid your breathing

Aid and adapt your home:

- Long-handled dustpan
- Bath grab bars
- Shower/bath seat
- Raised toilet seat
- Stair lift
- Portable oxygen
- Zimmer frame

Use a handheld fan:

- To help recover from episodes of breathlessness
- Research has shown that this enables patients to increase their physical activity[2]



Maintain a good posture:

- Keeping your back bent and shoulders slumped increases muscle strain causing breathing to be affected
- Avoid bending and lifting

Ask for help:
See page 12 in this booklet for further information about support available for you

If you require assistance to adapt your home or obtain equipment please contact your local authority to request an Occupational Therapy assessment.

PF CLINICAL TRIALS

- Clinical trials are research studies during which possible treatments for PF can be investigated through 3 phases
- These studies aim to confirm whether the treatment is safe and effective

PHASE 1

- Evaluation of safety
- Safe dosage determination
- Side effect identification

PHASE 2

- Testing whether the drug is effective
- Further evaluation of safety

PHASE 3

- Effectiveness of the drug is confirmed
- Side effects are monitored

Over **1 million** people participate in research every year in the UK [3]

How do I get involved?

- Search for clinical trials at **w:** www.bepartofresearch.nihr.ac.uk
- Speak to your lung specialist doctor or nurse to find out more information

WHAT ALLOWANCES CAN I RECEIVE?

Personal Independence Payment (PIP)

- £23.70 - £89.60 per week if you have not reached state pension age
- The amount received depends on how PF affects you
- You are assessed by a health professional
- To claim call 0800 917 2222 or write a letter to the following address to request a form: Personal Independence Payment New Claims, Post Handling Site B, Wolverhampton, WV99 1AH

Attendance Allowance

- £60.00 - £89.60 a week if you are over the state pension age
- To help with personal support
- To claim fill out the form available at w: www.gov.uk/attendance-allowance

Statutory Sick Pay (SSP)

- £96.35 a week if you are too ill to work
- Paid by your employer for up to 28 weeks
- To claim give your employer a doctor's 'fit note'

Carers Allowance

- £67.60 a week for your carer if you are being cared for at least 35 hours per week
- Claim online at w: www.gov.uk/carers-allowance/how-to-claim

Blue Badge

- Allows you to park closer to your destination
- For a list of requirements access w: www.gov.uk/transport/blue-badges
- Apply online at w: www.gov.uk/apply-blue-badge

Self Employed

- You cannot claim SPP but can apply for PIP and Universal Credit
- Apply for Universal Credit online at w: www.gov.uk/apply-universal-credit

Access an online benefits calculator to receive information about which benefits you are entitled to and how to claim at w: www.gov.uk/benefits-calculators.

Please note that the benefits and website links may change before this booklet is next reviewed.

SUPPORT FOR YOU AND YOUR FAMILY

Below are national and local support groups and charities that you can contact if you are struggling or you would like to speak to somebody.

Your care and management will be managed by your specialist respiratory doctors and nurses. Dr Hart and Dr Crooks are supported by specialist nurses Mark Major and Mandy Bell at Castle Hill Hospital in Hull.

NATIONAL

Pulmonary Fibrosis Trust

T: 01543 442191
E: info@pftrust.org
W: www.pulmonaryfibrosistrust.org

Action for Pulmonary Fibrosis

T: 01223 785725
E: info@actionpf.org
W: www.actionpf.org

ASTHMA+ LUNG UK

T: 03000 030555
E: info@asthmaandlung.org.uk
W: www.asthmaandlung.org.uk

SAMARITANS

If you are struggling to cope with anxiety or depression:

T: 116 123 **E:** jo@samaritans.org **W:** www.samaritans.org

LOCAL

NHS Hull University Teaching Hospitals NHS Trust

dovehouse hospice
caring for people with a life limiting illness

Hull and East Yorkshire PF Support Group

Every 3 months, usually on a Monday afternoon 14:00 - 16:00 at Castle Hill Hospital, Castle Hill Road, Cottingham, HU16 5JQ

T: 07769 742092

E: markmajor@nhs.net

Breathlessness clinic

Learn different strategies to manage breathlessness with a physiotherapist. You will be expected to put into practice what you have learned and progress will be reviewed at follow up visits.

T: 01482 784343

Is it an emergency or an urgent medical need?

Call 999 for emergencies or 111 for urgent medical problems

NOTES AND QUESTIONS

APPOINTMENT RECORD

DATE & TIME	PLACE	TO SEE	REASON

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The Pulmonary Fibrosis Trust was founded 10 years ago with the aim of providing personal support to people affected by PF. Their aim is to help patients with PF live their lives as independently and as comfortably as possible. Patients requiring advice or practical support can reach the Pulmonary Fibrosis Trust via the contact information detailed in this booklet.

The charity's work is funded solely by donations and it is thanks to these donations that they are able to continue providing the vital support that they do.

The authors declare that they have no conflict of interest with respect to the publication of this booklet.

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[1] Bloomer, A., 2020. Pulmonary fibrosis on the rise due to Covid-19. [online] Geriatric Medicine Journal. Available at: <https://www.gmjournal.co.uk/more-people-will-require-pulmonary-rehabilitation-due-to-covid-19> [Accessed 16 December]

[2] Swan, F., English, A., Allgar, V., Hart, S. and Johnson, M., 2019. The hand-held fan and the calming hand for people with chronic breathlessness: A feasibility trial. *Journal of Pain and Symptom Management*, 57(6), pp.1051-1061.e1.

[3] NIHR. 2020. Record number of patients take part in clinical research. [online] Available at: https://www.nihr.ac.uk/news/record-number-of-patients-take-part-in-clinical-research/11746?diaryentryid=44785 [Accessed 1 December 2021].

ACCESSIBILITY INFORMATION

If you or your carer requires this information in a different format, such as large print, braille or audio, due to disability, impairment or sensory loss, please advise a member of staff and this can be arranged. Alternatively access this information online by scanning the QR code below using a mobile device.

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