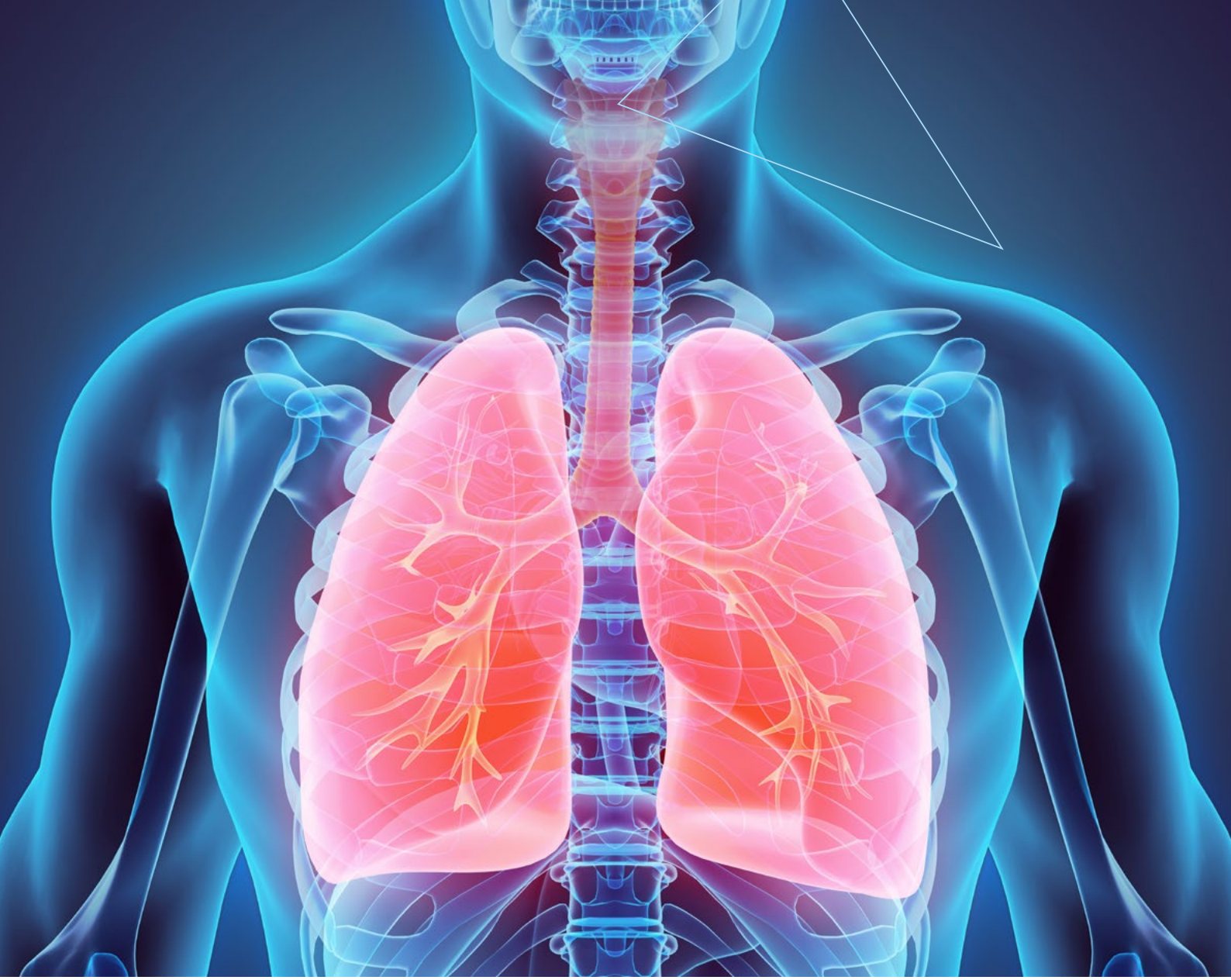


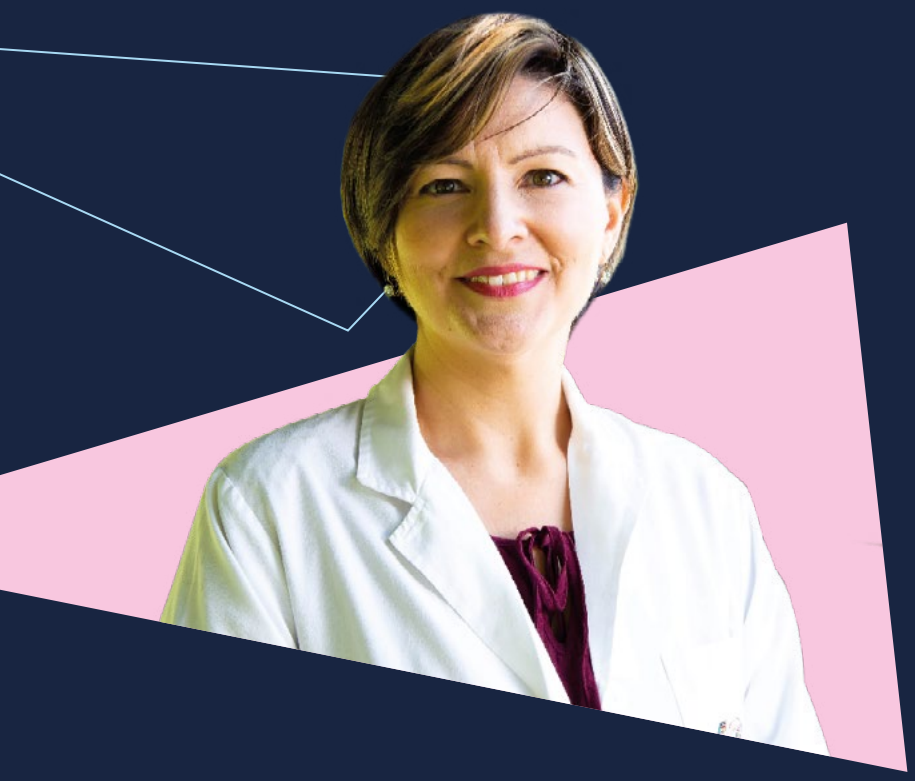
Therapy Guide

Idiopathic Pulmonary Fibrosis (IPF)

GKA.

The GKA Difference

IPF is a rare condition and can be a very tricky area for medical fieldwork – in fact, even the word “idiopathic” actually means that the cause is unknown. At GKA we have successfully completed medical fieldwork studies on IPF using our panel of respiratory specialists, respiratory nurses and patients; methodologies have included online, telephone interviews, web-assisted telephone interviews, face to face and in office interviews with both physicians and patients.



The Lowdown

Idiopathic pulmonary fibrosis is a poorly understood lung condition that causes scarring of the lungs. Over time, the build up of scar tissue (fibrosis) causes the lungs to become thick and hard, which ultimately results in the lungs losing their elasticity and an irreversible loss of the tissue's ability to transfer oxygen to the blood stream via the alveoli (air sacs). Unfortunately, IPF is a chronic and ultimately fatal disease that sees lung function progressively declining over time before failing completely.

Some facts about IPF

4 in 100,000
People in England are newly diagnosed each year[1]

1.7 Men
Are affected compared to every 1 woman[3]

People aged 70-75
are most commonly affected[2]

1 in 20
People with IPF have another family member with the condition[4]

Little is known about what triggers the disease, which is one of the many reasons that medical fieldwork is so important in this area; a number of factors have been suggested as potential triggers, including smoking, viral infections such as Hepatitis C or the Epstein-Barr virus, breathing in wood and/or metal dust and gastrooesophageal reflux disease (GORD), a condition where the acid from the stomach rises back into the throat and is accidentally inhaled into the lungs. There is also some evidence that IPF can be genetic, but as it is such a rare condition it remains hard to estimate how much of an impact each of these potential risk factors has on a person's chance of developing the disease.

Understanding IPF

Symptoms

One of the most common symptoms of idiopathic pulmonary fibrosis is shortness of breath when physically active, which gradually worsens over time. In fact, people suffering from IPF can feel breathless even from simple everyday activities such as walking and many even ignore their breathlessness at first, putting it down to getting old or being out of shape. Eventually however, even light activity such as getting dressed can be difficult. Other symptoms included a persistent dry cough, fatigue and weakness, a feeling of discomfort in the chest, loss of appetite and rapid weight loss.

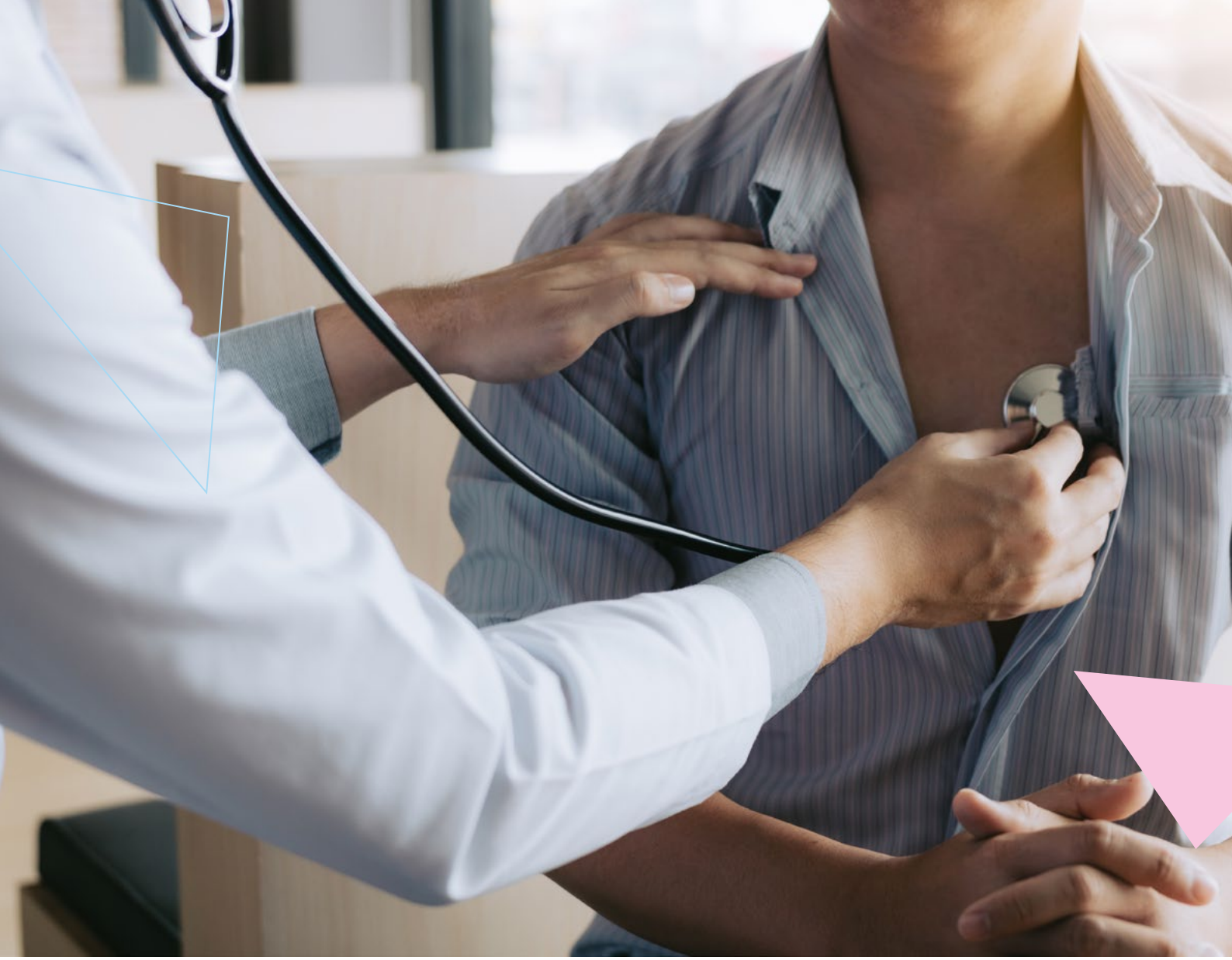
Another change could be clubbing that can affect some people's finger and toe nails. Nails change shape to become 'drumstick ilke'; the tips of the fingers get bigger and nails curve around the finger tips with the base of the nail feeling spongy.

Diagnosis

Identifying IPF is hard as it can often be confused with other lung conditions such as chronic obstructive pulmonary disease (COPD) due to similar symptoms. Successful diagnosis will be a joint effort by a specialist multi-disciplinary team including several doctors who are experts in lung conditions.

Typically they will need to carry out a number of tests and examinations including:

- **Chest x-ray and CT scan**: Although it doesn't show the lungs in much detail, a chest x-ray can help to identify more obvious problems that could be causing symptoms such as cancer or a build-up of fluid. If IPF is suspected, a CT scan will create a more detailed image of the lungs to identify scarring. On the CT Scan IPF often shows up as a distinctive pattern described as **honeycomb lung** - the image showing lots of empty pockets or bubbles where more solid-looking lung tissue should be.



- **Breathing and blood tests**: Lung function or pulmonary function tests are used to see how quickly air moves in and out of the lungs and how much air the lungs can hold. A blood test will also be used to identify how well the lungs transfer oxygen into and carbon dioxide from the blood.
- **Lung biopsy**: If all the above tests aren't conclusive, a lung biopsy will be carried out via keyhole surgery under general anaesthetic to remove and analyse a small sample of lung tissue for signs of scarring.
- **Bronchoscopy**: If doctors still aren't sure what the problems is, they will insert a narrow tube with a camera into the airways to look out for anything abnormal and take small tissue samples for testing. A local anaesthetic is used to numb the throat so the procedure doesn't hurt.

References

1 - https://www.healthexchange.org.uk/body-parts/introduction-728/
2 - http://www.nhs.uk/Conditions/pulmonary-fibrosis/Pages/Introduction.aspx
3 - http://pmj.bmj.com/content/76/900/618.full
4 - http://www.nhs.uk/Conditions/pulmonary-fibrosis/Pages/Introduction.aspx

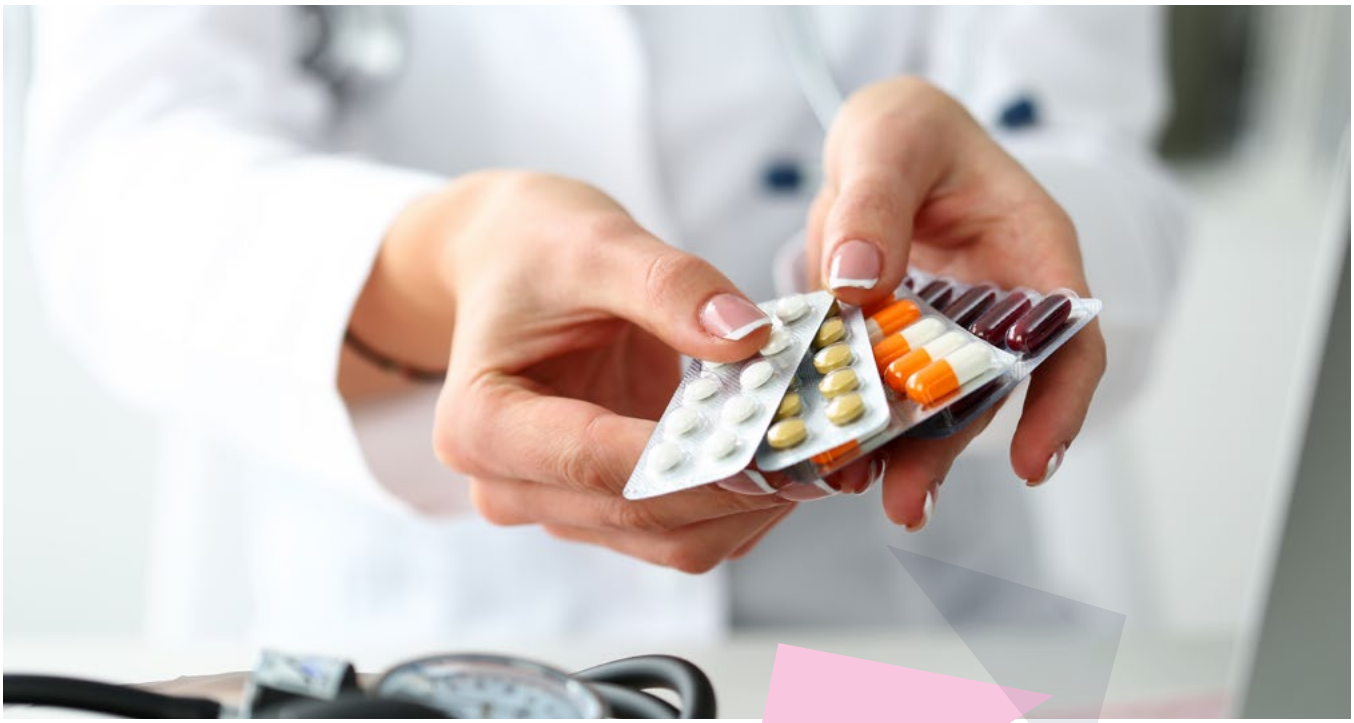
Treating IPF

Whilst there's currently no cure for IPF, there are several treatments available that can help to relieve symptoms and slow down progression, including self-care measures such as stopping smoking and exercising regularly, taking medication to reduce that rate at which scarring worsens, breathing oxygen through a mask and pulmonary rehabilitation exercises to help you breath more easily. In extreme cases, a lung transplant might be the only option, however donor lungs are very rare.

Medications

Some patients may take steroid tablets to help to ease breathing, and there is some evidence that combining a corticosteroid with an antioxidant can slow the progression of IPF in the short-term, although its long-term effectiveness is uncertain. More commonly, patients tend to take medications which ease symptoms of breathlessness, and a steroid to reduce the rate that the scar tissue hardens.

Sildenafil is a medication that can help to widen the blood vessels in the lungs, improving blood flow whilst reducing blood pressure. Another treatment option is pirfenidone, an antifibrotic medication that is commonly used for treating adults with mild to moderate IPF. Whilst more studies are needed on its long-term effects, current research shows that pirfenidone is able to slow the progression of IPF by reducing the rate at which the scarring worsens.



We are GKA.

If you have been asked to carry out a healthcare market research study surrounding IPF or one of the related conditions, why not give us a call today?

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