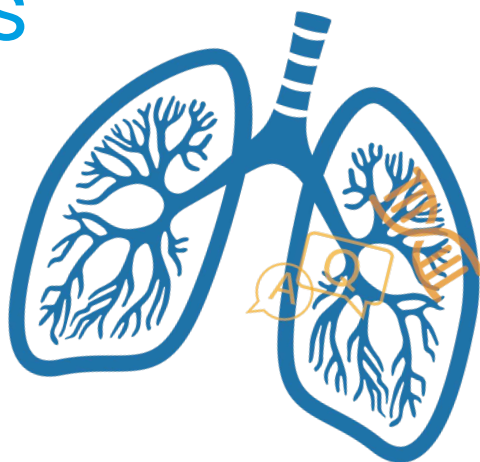


ALK+

Understanding your diagnosis

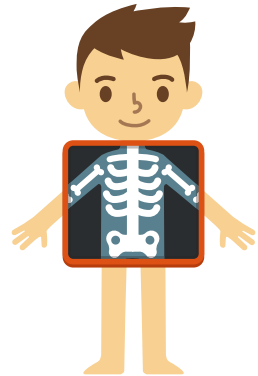


This booklet is for anyone diagnosed with ALK positive lung cancer, their family and friends. We hope it gives you an understanding of what ALK positive lung cancer is, treatment options available and how to get support if you want or need it.

Your journey so far...

Although procedure may differ slightly from hospital to hospital, you will have undergone various tests in order to receive your diagnosis.

This may have included an x-ray or CT scan of your chest, a bronchoscopy and a biopsy of your lung tissue. Your biopsy would have shown whether your cancer is associated with any genetic changes, such as changes to the anaplastic lymphoma kinase (ALK) genes, epidermal growth factor receptor (EGFR) genes or receptor tyrosine kinase (ROS1) genes.



This would have confirmed your ALK positive lung cancer diagnosis.

Remember that lung cancer can affect anyone.

- ALK positive lung cancer has no relation to smoking
- Two thirds of ALK positive lung cancer patients are women
- Over half of ALK positive lung cancer patients are younger than 50 when diagnosed
- ALK positive lung cancer accounts for 4% of NSCLC diagnosis'

What is ALK+ lung cancer?

ALK positive lung cancer is a type of Non-Small Cell Lung Cancer (NSCLC) where a mutation/ rearrangement of the Anaplastic Lymphoma Kinase (ALK) gene is present.

Our DNA is made up of chromosomes and genes, which are responsible for building new cells and instructing the cells how to behave. There can be abnormal changes to these genes, otherwise known as mutations. These mutations can instruct the cell to grow or behave abnormally.

The ALK gene provides instructions for making ALK receptor tyrosine kinase, which is a protein that is part of the signalling pathway that controls cell growth and division. ALK positive lung cancer occurs when part of the ALK gene is broken and attaches to another gene, which creates a fusion between ALK and another gene. This gene change is sometimes called 'rearrangement' or 'translocation'.

This type of mutation blocks messages which tell cells to stop growing. This means cells start to grow abnormally and lead to the growth of tumors.

Genetic changes can be considered as either somatic (changes occurring over a lifetime) or hereditary (changes inherited through families). The ALK gene mutation is considered to be *somatic*. This means that there is often no specific reason why the ALK gene undergoes a mutation or rearrangement.



Cancer stages explained

Your cancer stage describes the extent of lung cancer in your body and it is used to assess the prognosis of your cancer and treatment plan. Cancer stages consist of letters and numbers.

The letter T, N and M describe different areas of cancer growth.

T describes the growth of the main tumor

TX	Severity ↓
T0	
T1	
T2	
T3	
T4	

N describes how far the cancer has spread within nearby lymph nodes

NX	Severity ↓
N0	
N1	
N2	
N3	

M describes if cancer has spread to areas far from the lungs

M0	has spread
M1	hasn't spread

The number stages are **0, I, II, III and IV** and depend on how much the cancer has grown and how far the cancer has spread



Your medical team will be able to break down and explain your cancer stage to you.

Your next steps

It is completely normal to feel a wide variety of emotions after your diagnosis. You may feel angry, shocked, upset or worried. It's important to give yourself time to accept and understand your diagnosis and how this will affect your life. ALK positive lung cancer is a relatively unknown form of lung cancer, so you may have lots of questions. You can ask your medical team and support team any questions, at any time.

Importantly, there are a range of treatment options (otherwise known as targeted therapies) available for ALK positive lung cancer which can slow or stop the spread of cancer. Which route you choose depends on the results of your biopsy, your physical wellbeing, whether your cancer has spread and your personal preferences.

You can learn more about your treatment options in this booklet. It's a good idea to take time to consider your treatment plan and discuss this with your medical team, family and friends.

You may wish to consider your own preferences and priorities as you begin your journey and as you live with ALK positive lung cancer. As a starting point, you could think about:

- Do I understand what ALK positive lung cancer is – and what it means for me?
- Do I know what treatments are available to me?
- Do I know about the possible symptoms and side effects of these treatments?
- What kind of treatment do I want?
- Do I want a treatment with as few side effects as possible?
- Do I want the longest possible life regardless of the effects of treatment?
- Do I know what help I might need during my ALK positive lung cancer journey?
- How do I feel about my diagnosis?
- What can I ask my carer to do for me?
- Who else can I ask for help if I need it?
- What can I do to help myself?
- How might my diagnosis affect my relationships?
- Do I want to carry on working?
- Do I want to carry on supporting my family?
- Is there anything related to my ALK positive lung cancer I would like to know more about?
- Do I know where to go for more information?

Your medical team

You will meet and consult with many different healthcare professionals throughout your ALK positive lung cancer journey. It can feel overwhelming or be confusing to remember their different roles, so make sure you ask if you are unsure at any time.

In general, you will have an Oncologist (a doctor specialising in cancer) and an Oncology Nurse Specialist (a nurse specialising in cancer). You will also have your General Practitioner (local GP). If you want to, you can also have access to a Nutritionist (a specialist in nutrition and diet), a Counsellor or Psychologist (specialists in mental health and support) or an Occupational Therapist (specialist in general lifestyle support).



Your treatment options

Several treatments have been specifically developed to slow or stop the spread of ALK positive lung cancer. These can all be discussed with your medical team.

Traditional cancer treatments include radiotherapy (the use of radiation to treat cancer cells) and chemotherapy (the use of anti-cancer drugs to destroy cancer cells).

Generally, traditional cancer treatments are less effective for treating ALK positive lung cancer when used alone. Targeted therapies are often the recommended choice.



ALK inhibitors, also known as tyrosine kinase inhibitors (TKIs), have been developed to precisely target the mutation causing your cancer. These are referred to as targeted therapies.

Tyrosine kinase is an enzyme which instructs a cell to multiply and grow by binding to it via a receptor (protein). Tyrosine kinase inhibitors (TKIs) work by blocking the receptor (protein) to stop the growth and multiplication of the cell.

Crizotinib

Brigatinib

Ceritinib

Alectinib

Lorlatinib



These are all different types of tyrosine kinase inhibitors (TKIs). These are taken orally in tablet form. Your doctor will advise you on how to take your TKI, for example, how many times a day, with or without food or if there are any foods or medicines to avoid.

If your cancer starts to change or grow whilst you are receiving TKI treatment, you may need to change the drug you are currently taking. This is known as moving to a new 'line of treatment'. There are three eras of TKI development at present, with new treatment options being explored in clinical trials.

Please speak to your doctor about which targeted therapies are available. Each TKI is licensed by the National Institute for Health and Care Excellence (NICE) and may not be available to every patient in all circumstances.

You may also receive Stereotactic Radiosurgery (SRS) to targeted areas whilst receiving an ALK inhibitor drug. SRS (a non-surgical radiation therapy) can extend the length of treatment line should there be small areas of cancer progression.



Treatment side effects

It is common for ALK positive patients to experience symptoms of lung cancer alongside symptoms of their ALK positive lung cancer treatment. You should always discuss any symptoms or side effects with your medical team, especially if you think they are getting worse.

Side effects of targeted therapy, such treatment with TKIs, can include:

- Tiredness (fatigue)
- Nausea and vomiting
- Diarrhoea or constipation
- Changes in vision



Other possible side effects include: low white blood cell count, swelling / inflammation, liver problems, nerve damage (also known as neuropathy, which might feel like tingling or burning in the hands or feet) and problems with heart rhythm. Always consult your medical team if you are worried about any symptoms or side effects.

Having cancer might mean that you have a higher risk of developing a blood clot. A blood clot can be very serious. Most clots when diagnosed can be successfully treated so it's important to report any symptoms to your medical team immediately. Treatment for blood clots is usually blood thinners (anticoagulants).

Cancer cells release substances that damage your bones, making them weak and more likely to break. If you experience bone pain or weakness, please consult your medical team to discuss bone strengthening treatments.

ALK positive lung cancer and the brain

Sometimes cancer can spread from one part of the body to another, for example lung cancer may spread to the brain. This is still lung cancer, but is now in the brain, so it is called brain metastases. Although any type of cancer can spread to the brain, some types are more likely to do so, including non-small cell lung cancer (NSCLC). In fact, brain metastases may affect around half of patients with ALK positive lung cancer.

Some people with brain metastases may show symptoms, while others may not. If your cancer has spread to the brain, you may experience headaches, weakness, sickness, mood swings / behaviour changes, fits or seizures, coordination problems, confusion, lethargy / tiredness, problems reading or talking.

Let your medical team know straight away if you experience any of these symptoms so that they can give you an MRI scan to test for brain metastases. It's a good idea to have regular brain MRIs to detect brain metastases as early as possible.



Living with ALK positive lung cancer

Once you have come to terms with your diagnosis and considered your options for treatment, you may like to think about your lifestyle moving forward as a lung cancer patient. It's beneficial to consider the following:

- Eating a variety of nutritious food will provide you with all the essential nutrients your body needs to stay as healthy as possible.
- Although you may suffer from fatigue, trying to remain as physically active as possible as this will improve your physical function and promote your mental health. Just 5 minutes of activity a day (walking, gardening, cycling etc.) is a great start.
- Maintaining relationships and social interaction is really important during your ALK positive lung cancer journey. It may also be worth reviewing any sources of stress.
- Although ALK positive lung cancer is not associated with smoking, it's advisable to avoid smoking and second-hand smoke.



Advice for carers

It can be a very difficult, confusing and upsetting time when a family member or friend is diagnosed with ALK positive lung cancer. It's important to ensure that you have dealt with your own emotions and feelings before you begin to support someone with cancer.

Providing both emotional and practical support can be hugely beneficial to patients, whether this is simply being a friendly face to talk to, helping drive to and from appointments or helping with daily chores.

If you are a carer and feel in need of support yourself, get in touch with a local support group (for example ALK Positive UK at www.alkpositive.org.uk) or speak to your loved one's medical team. It's very important that your mental and physical health is looked after in order for you to be able to care for someone with cancer.

Did you know?

- You can speak to your doctor about clinical trials and Compassionate Use Programs if certain medicines are not licensed.
- You can apply for a medical exemption certificate to help with the costs of prescriptions. Visit www.nhs.uk for more information.
- You may be eligible for Personal Independence Payment (PIP) – a benefit that helps with the extra costs of a long-term health condition. Visit www.gov.uk for more information.
- If you drive, you may need to inform the DVLA of certain medical or health conditions if they affect your driving. Visit www.gov.uk for more information.
- You may be entitled to a free flu jab. Speak to your GP about this or visit www.nhs.uk for more information.
- If you own a smart phone, there are lots of apps available to help schedule and remind you of appointments and medication. If not, a dedicated note book or diary works just as well. It's also a good idea to note down any questions that you think of or side effects you experience between appointments.
- You should have a 24/7 emergency number to contact out-of-hours or at weekends if you need advice or support outside of office hours.
- If you don't have any metal in your clothing or body, you don't need to wear a hospital gown for scans.
- Removing ECG electrode pads can be uncomfortable if you have hair on your chest. It may be worth considering shaving or removing hair in this area prior to ECG tests.
- You are entitled to have a say in your treatment plan. Don't be afraid to research, ask questions or request information relating to your diagnosis and treatment.
- You will be required to take regular blood tests during your diagnosis and treatment. There are lots of tips for making this process easier, including drinking plenty of water before tests, running your hands under warm water or requesting a nurse who is trained with a butterfly needle.
- You are always entitled to have a friend, family member or chaperone with you during any consultation or meeting.

Glossary of terms

ALK: A gene that provides instructions to make an enzyme (made of protein) in the body called ALK receptor tyrosine kinase.

ALK mutation: An ALK mutation occurs when the ALK gene is broken and attaches to another gene.

ALK inhibitor: A type of anti-cancer drug that specifically targets and blocks the growth (or inhibits) cancer cells caused by the ALK mutation.

Biopsy: A procedure that involves taking a sample of suspicious tissue from the body to test for certain changes or patterns of growth.

Bone metastases: Bone metastasis or 'bone mets' occurs when cancer cells from the primary tumour relocate to the bone. Prostate, breast, and lung cancers are most likely to spread to the bone.

Brain metastases: When cancer that started in another part of the body – such as the lungs – spreads to the brain.

Cancer: A disease where cells grow and reproduce uncontrollably.

Central nervous system: Part of the nervous system that consists of the brain and spinal cord.

CT scan: A computerised tomography (CT) scan uses X-rays and a computer to create detailed images of the inside of the body. CT scans are sometimes referred to as CAT scans or computed tomography scans.

Gene: The building blocks of your DNA (genetic material) that determine your characteristics, e.g. hair or eye colour.

Genetic mutation: A fault (or change) in the DNA sequence that makes up a gene. A 'somatic' mutation is a change that is not inherited from a parent. These are usually prompted by environmental factors or may occur randomly. A 'germline' mutation occurs in the germ cells (which eventually develop into eggs or sperm) and is passed from parent to child. Gene mutations have varying effects on health, depending on where they occur.

Hereditary: Characteristics (or diseases) that are passed on from parents to their children in their genes.

Lines of treatment (i.e. first-line, second-line): The first line of treatment is the first drug given to a patient to treat a specific disease (usually accepted as the best treatment for that disease). Second-line or third-line treatments might be given if the previous treatments stop working, and so on.

Metastases: When cancer spreads from one part of the body to another part of the body.

Molecular testing: Checks for certain changes in a gene or chromosome that may cause the development of a specific disease or disorder.

MRI scan: Magnetic resonance imaging (MRI) is a type of scan that uses strong magnetic fields and radio waves to produce detailed images of the inside of the body. An MRI scanner is a large tube that contains powerful magnets. You lie inside the tube during the scan. You cannot wear anything metal if you have an MRI scan.

Neuropathy: Nerve damage that can be caused by medication, tumour growth or surgery. Symptoms vary depending on the nerves affected. You may have pain, sensitivity, numbness or weakness. This is often in the hands, feet or lower legs. The nerves controlling digestion and blood pressure can also be affected, so you may have constipation, dizziness or other symptoms.

NSCLC: Non-small cell lung cancer – about 85% of lung cancers are NSCLC. ALK+ lung cancer is a form of NSCLC, which accounts for 4% of NSCLC cases. Oncologist: A doctor who treats cancer and provides medical care for a person diagnosed with cancer.

PET scan: Positron emission tomography (PET) scans are used to produce detailed 3-dimensional images of the inside of the body. The images can clearly show the part of the body being investigated, including any abnormal areas, and can highlight how well certain functions of the body are working.

Pneumologist: A doctor specialising in lung diseases and conditions.

Targeted therapy: Targeted therapy drugs interfere with the way specific cancer cells signal or interact with each other. This can stop them from growing and dividing.

Translocation: The movement of something from one place to another. In the case of ALK positive gene translocation, refers to the rearrangement of the ALK gene.

Tumour: A lump / swelling caused by the abnormal growth of tissue. Can be benign or malignant (i.e. cancerous)

Tyrosine kinase inhibitors (TKIs): Block chemical messengers (enzymes) called tyrosine kinases. Tyrosine kinases help to send growth signals in cells, so blocking them stops the cell growing and dividing.



About ALK Positive Lung Cancer (UK)

ALK Positive Lung Cancer (UK) is a registered charity established by patients, their families and friends. We are a community from different backgrounds and walks of life - each with our own individual story. Established in 2018, our purposes are to provide support and advocacy and to improve the overall survival and quality of life of ALK positive lung cancer patients across the United Kingdom.

We are recognised by the National Institute for Health and Care Excellence (NICE) as an organisation to be consulted on new ALK positive treatments.

We do not offer medical advice, but we have an active Facebook Group for patients, family and carers where experiences can be shared. We hold regular meetings in the UK and also share information on social media.

If you are living in the UK with ALK positive lung cancer, or if a member of your family or a close friend is, please come and join us.



hello@alkpositive.org.uk
www.alkpositive.org.uk
@UKALK1





Supporting patients of ALK Positive Lung Cancer

This booklet has been put together by the ALK Positive Lung Cancer (UK) charity, which is run by patients of ALK positive, their family and friends.

For answers to any questions you may have about ALK Positive Lung Cancer (UK), or to join our online community, contact us at hello@alkpositive.org.uk

To find out more about ALK Positive Lung Cancer (UK), visit www.alkpositive.org.uk

Please remember, you are not alone and support is always available.

*Please note we do not offer medical advice.
Charity no. 1181171*