Original article

ALK Positive Lung Cancer (UK)

# Do Not Resuscitate (DNR) Decisions.

A Guide for Doctors, Patients and Families.

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ALK POSITIVE UK

**If you have ALK-positive lung cancer, you may wish to share this booklet with any doctor or other healthcare professional who may not be familiar with ALK-positive Lung Cancer.**

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# Contents



Contact details available on the back cover

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**The content of this ALK Positive Lung Cancer (UK) booklet is for information only and should not be used for the diagnosis or treatment of medical conditions. We are unable to offer specific advice and, if you are worried about any symptoms, please consult your doctor.**

ALK Positive Lung Cancer (UK)

# Do Not Resuscitate (DNR) Decisions.

## 1. Introduction

ALK-positive Lung Cancer (LC) is a rare form of lung cancer first identified in 2007. Many doctors will know little or nothing about it and will not have looked after a patient with it. Their knowledge will be based on poor survival rates for most other types of lung cancer diagnosed at Stage 4.

However, many ALK-positive LC patients have become well informed about:

- the nature of the disease
- the available treatments
- how effective treatments can be despite late-stage diagnosis
- the way it can progress
- further treatment options
- the fact that many ALK-positive LC patients are now living active, healthy lives for years.

We have produced this booklet for healthcare professionals, ALK-positive patients and their families to ensure:

- Everyone has up-to-date information on ALK-positive LC, treatments and life expectancy if a Do Not Resuscitate (DNR) Order becomes an issue.

- Key information is provided about DNR decisions, such as:
  - the framework for DNR decisions
  - the issues that need to be considered
  - the options if there is disagreement between healthcare professionals and patients or families.

**Sections 2 and 3** provide key facts about ALK-positive LC, current treatment options and survival rates. There is a glossary at the end of the booklet to explain some of the medical terms used.

**Sections 4 and 5** explain what is meant by Cardiopulmonary Resuscitation (CPR) and the decision-making process for DNR Orders.

**Section 6** explains the obligations of doctors to patients in their care in relation to a DNR decision.

**Section 7** explains how patients and families can challenge a doctor about their DNR decision.

Copies of this booklet can be obtained by:

- downloading from the charity's website at www.alkpositive.org.uk
- emailing hello@alkpositive.org.uk
- writing to the charity at the address at the end of the booklet
- telephoning 07975 632515.

## 2. Key facts about ALK-positive lung cancer[1]

ALK-positive LC is rare. It affects only about 4 in 100 patients with non-small cell lung cancer (NSCLC).

ALK-positive LC develops when the anaplastic lymphoma kinase (ALK) gene combines with another gene in an abnormal way to produce a hybrid ('mutant') protein. This protein allows cells to divide and multiply uncontrollably resulting in cancer and its spread.

Doctors do not yet know what causes the abnormal gene fusion which triggers ALK-positive LC.

It is not inherited from parents.

Nearly 7 in 10 patients have never smoked or smoked very little.

Half of patients are under 53 years of age at diagnosis.

Just over half of patients are female.

Nearly 9 in 10 patients are Stage 4 at diagnosis with spread of the cancer to another body organ (metastasis).

3-4 in 10 patients have LC which has spread to the brain (brain metastases or brain mets) at diagnosis[2].

Other parts of the body that the cancer may spread to are:

- bone
- organs in the abdomen, such as the liver, kidney or adrenal glands (intra-abdominal)
- between the lung lining and the lung, leading to a build-up of fluid (malignant pleural effusion)
- lymph nodes.

Half of patients with Stage 4 ALK-positive LC survive for over 6.2 years after treatment starts (median survival)[3].

## 3. Treatment

Current treatment options are effective for many ALK-positive LC patients, even though most patients are diagnosed at stage 4 with spread of the cancer (metastatic disease)[4].

Tyrosine Kinase Inhibitors (TKIs) are medicines taken daily by mouth that are used to target the cancer cells. This is known as 'oral targeted therapy'.

TKIs work by blocking the abnormal ALK fusion protein. This stops cells dividing uncontrollably and the cancer from growing and spreading.

2nd generation TKIs called Alectinib and Brigatinib are currently used to treat ALK-positive LC to start with. They have good brain penetration and, for many patients, are effective at treating patients who already have brain mets. In patients who do not already have brain mets, they protect the brain from spread of the cancer.

TKIs are generally well tolerated by patients. They have fewer side effects than conventional chemotherapy.

Often, there is no evidence of primary or metastatic disease on CT or MRI scans (called 'radiological resolution') after taking TKIs and this may last for years.

Over time, further gene mutations can lead to TKI resistance and 'progression' of cancer[4].

Regular monitoring of patients is needed. This includes CT scans every 3-months, and in many cases, MRI scans of the brain. This will ensure any progression is detected as early as possible.

The next step is usually a change of targeted therapy to the 3rd generation TKI called Lorlatinib.

After treatment with TKIs, progression in many patients resolves and they have further years where the cancer is controlled.

'Localised progression' may be treated with radiotherapy and the patient's current TKI.

Clinical trials of a 4th generation TKI are already running in the UK.

If there is further progression of the cancer or TKI intolerance, patients may be treated with chemotherapy and immunotherapy.

In 2019, median survival in the UK was recorded as 6.2 years. This is likely to increase significantly over time for patients treated with 2nd and 3rd generation TKIs.

TKIs allow many patients to live normal active, progression-free lives, without serious side effects, for many years.

## 4. CPR and resuscitation

Cardiopulmonary Resuscitation (CPR) is a treatment that can be given when a person's breathing stops (respiratory arrest) or heart stops beating (cardiac arrest).

CPR is a term which covers all procedures from basic first aid to more advanced techniques, like defibrillation[5]. The aim is to restart breathing and/or the heart when they have stopped.

Actions used in CPR, such as chest compressions, can cause bruising, break ribs and puncture lungs. Some people who are given resuscitation are left with permanent heart and/or brain damage.

CPR restarts the heart and/or breathing of between 10 and 20 people in every 100.

CPR is more likely to restart the heart and/or breathing of people with heart, lungs and other organs that are healthy and working well before a cardiac or respiratory arrest.

CPR is less likely to work in people who:

- already have previous organ damage, such as heart disease

- have a current severe illness, such as sepsis with multi-organ failure
- are frail
- are approaching the end of life.

## 5. Do not resuscitate (DNR)[6] orders

Do Not Resuscitate (DNR) may also be referred to as DNAR (Do Not Attempt Resuscitation) or DNACPR (Do Not Attempt Cardiopulmonary Resuscitation). In this booklet, we use the term 'DNR'.

DNR Orders:

- only state whether or not a person will be given CPR. Patients will still be given appropriate care, treatment, symptom control and support.
- allow the healthcare team not to try restarting a patient's heart or breathing.
- are recorded on a special form. Doctors and hospitals may use different forms. Examples include a DNACPR form, a treatment escalation plan or a recommended summary plan for emergency care and treatment (ReSPECT[7] form).
- can be reviewed and withdrawn if a patient's circumstances change. For example, if they recover from a severe illness.
- are signed to allow a patient to die with dignity and peace.

DNR is a medical decision made by the doctor. Doctors must:

- tell patients and families if a DNR order will be or has been completed
- explain the DNR order and why they think resuscitation would not be in the best interests of the patient.

Doctors should give patients the opportunity to talk about their wishes and preferences for the use of CPR.

However, the doctor makes the final decision about the use of CPR, even if the patient does not agree.

Since DNR orders apply only to CPR, having an Order should not affect other treatments a patient is given. However, research suggests this has been an area of confusion for some healthcare professionals.

Patients should discuss any concerns with their doctor. They should be clear about their treatment plan and the role of the DNR Order in that plan[8].

Patients are encouraged to have a discussion with their oncologist about 'advance care plans' and DNR decisions. Most hospitals have a form which records care plans in a patient's medical notes. This is referred to when there is any acute event, such as an emergency admission to hospital because of an accident or serious infection. Care plans can inform medical professionals that a patient should be given full active treatment,

including resuscitation, if that is what is agreed.

## 6. Obligations of doctors

Doctors in the UK are regulated by the General Medical Council (GMC)[9]. This means all doctors have a duty of care to:

- act in the best interest of the patient.
- offer treatment options based on up-to-date clinical evidence.
- be fully informed about the patient and their medical condition, the nature of the disease(s) affecting the patient and the risk of their heart or breathing stopping, and how likely it is that a patient will be revived if CPR is given.
- not consider their personal views of the patient's quality of life.
- take all reasonable steps to prolong a patient's life but are under no obligation to prolong life regardless of consequences.

Doctors also have an obligation[9] to seek advice or a second opinion from a colleague with relevant experience. This could be a healthcare professional from another specialty, such as oncology or palliative care. This should happen where:

- the doctor and the healthcare team have limited experience of the patient's condition.
- they are in doubt about the benefits, burdens and risks of a DNR decision for the patient.
- there is a major difference of opinion over a DNR decision between the doctor and patient.

## 7. Challenging a DNR decision

If patients disagree with a DNR decision they can ask the doctor the following questions:

- What knowledge and experience of ALK-positive LC do you have?
- Have you discussed the DNR decision with my oncologist or a medical colleague with more experience of ALK-positive LC?

Patients who want to challenge a DNR decision can talk to the hospital Patient Advice and Liaison Service (PALS). They can also ask a doctor who knows them well to be involved, such as their oncologist or possibly their GP.

## 8. Glossary

| Medical term | Explanation |
|---|---|
| Brain metastases or brain mets | Cancer that has spread to the brain. |
| Computerised tomography (CT) scan | A non-invasive medical imaging technique that uses x-rays to create 3D images of the inside of the body. |
| CPR | Cardiopulmonary Resuscitation can be given to try restart a person's breathing or if their heart stops beating. |
| Defibrillation | Patients are given a jolt of electricity to the heart. This can help restore the heart's rhythm and get it beating normally again. |
| DNR Order | A Do Not Resuscitate (DNR) Order instructs medical professionals not to use CPR if a patient's heart or breathing stops. |
| Inherited | A characteristic passed on from parents to children. |
| Localised progression | Means that the cancer has spread to a limited number of sites. |
| Malignant pleural effusion | Build-up of fluid in the chest between the lung lining (pleura) and the lung caused by cancer. |
| Magnetic resonance imaging (MRI) scan | A non-invasive medical imaging technique that uses radio waves and strong magnetic fields to create detailed images of the inside of the body. |
| Median age | The age at which half the people being considered are below and half are above. |
| Median survival | The length of time that half the people being considered live longer and half live less. |
| Metastatic spread or metastatic disease | Cancer that has spread from one part of the body (primary location) to another part of the body. |
| Oral targeted therapy | Example: Medicines such as Tyrosine Kinase Inhibitors (TKIs), which are taken daily by mouth. They are used to target the cancer cells. |

Glossary continued overleaf

## 8. Glossary continued

| Medical term | Explanation |
|---|---|
| Primary disease | The place where the disease begins growing. |
| Radiological resolution | No evidence of the cancer is seen on CT or MRI scans. |
| Stage 4 | Stage 4 is the most advanced stage of cancer. It occurs when cancer cells spread from their original location to other parts of the body. |
| 2nd, 3rd and 4th generation TKIs | Newer targeted drugs designed to improve on 1st generation drugs by improving patients' response to treatment. |

## 9. References


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For further information, please visit **www.alkpositive.org.uk**

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