

# GLILD

Granulomatous-Lymphocytic Interstitial  
Lung Disease

Information for patients,  
carers and families

This booklet has been written for people who have been diagnosed with, or are being tested for, a lung disease called GLILD. We hope that it answers most of your questions. There is space at the back for you to note down anything you want to ask your healthcare team. If you would like further information or have any particular worries please do not hesitate to ask your nurse or doctor.

### **What is GLILD?**

GLILD stands for granulomatous-lymphocytic interstitial lung disease. It occurs in some people living with common variable immunodeficiency disorders (CVID). We think between one in five and one in 10 people with CVID may be affected by GLILD, so although generally considered rare, it is not that unusual if you have CVID. CVID is associated with low levels of antibodies in the blood, so people living with CVID are prone to developing infections. The commonest lung problems in CVID are chest infections (such as pneumonia) or damage arising from such infections (which is called bronchiectasis). GLILD is different, however, because it arises not through an underactive immune system, but because parts of the immune system become over active. This over activity can damage the lung if it persists. These over-active immune cells may be present in other places too, such as the wall of the bowel causing diarrhoea, or the spleen causing it to become enlarged. GLILD is therefore best considered

as the lung manifestation of a problem that may be affecting many different parts of the body at the same time.

### **What are the symptoms of GLILD?**

Common symptoms of GLILD include having a cough, breathlessness, chest pain or fatigue. Some people don't show any symptoms, and this is known as being asymptomatic. In this situation GLILD is picked up by routine tests. It is important that you tell your healthcare team if you notice a change in your symptoms.

### **Why do some people with CVID get GLILD and others don't?**

The short answer to this is that we don't really know. It may be to do with the way the body recognises things that are 'foreign', and lots of research is going on to understand this more. Some people have a problem with a specific gene that is linked to an increased chance of developing GLILD.

### **How is GLILD diagnosed?**

GLILD is usually suspected because of changes found on a chest scan (usually a CT scan), or because of changes in the results of lung function tests. We recommend annual lung function tests for people with CVID and, if there were no problems on the previous scan, a CT scan every five years. The lung function tests are particularly helpful in assessing the severity of

GLILD and whether it is changing over time (or with treatment). Unfortunately there isn't a simple blood test that can detect GLILD (though of course you may have blood testing for other aspects of your COVID care).

The tests we use are:

A **chest CT scan** takes just a few minutes and typically involves lying on a table which moves through a scanner shaped like a doughnut (see picture below).



Your head will not be inside the scanner and therefore most people do not feel claustrophobic. Sometimes we advise an injection of dye (also known as contrast) into your veins at the time of the scan which helps us get more information from the images. The dye can be associated with a flushing sensation. A CT scan does

involve some exposure to radiation. However, it is a very important test in making or excluding the diagnosis, and assessing change over time.

**Lung function or breathing tests** involve blowing hard into a machine to measure lung capacity (spirometry), breathing a mixture of gases to look at how well the lungs can get oxygen into and out of the blood (gas transfer) and/or sitting in a glass box and breathing helium to assess the total amount of gas in the lungs (lung volumes). Each test gives different information. The gas transfer is most important in GLILD. We want to know what your best lung function is so we will need you to blow hard for the spirometry test. However, most people do not find the tests unpleasant and they are not usually painful.

Unfortunately, the diagnosis of GLILD cannot be made with certainty on a CT scan and lung function tests alone. Other problems can look similar, including common conditions such as infections, and rarer but potentially more serious complications, for instance lymphoma (a cancer of the blood cells). Therefore, further tests are usually recommended. These include:

**Bronchoscopy** is a visual inspection of the airways and a way to collect samples from deep inside the lung. This is done by passing a thin, flexible tube about the width of a pencil through the nose or mouth and down into the

airways. This allows us to take samples to rule-out infection, though the samples cannot reliably confirm GLILD. The test is usually done with sedation (not a full anaesthetic) and as a day case. It is safe, and most people do not find it too unpleasant. The commonest after effect is coughing, but you may also experience some degree of chest pain and fever because of the sampling. However, these effects generally settle over a few days.

**Lung biopsy.** The only way to diagnose GLILD with certainty (and to rule out other conditions including lymphoma) is to take a small piece of lung tissue away for analysis – this is called a biopsy. It is usually achieved with keyhole surgery called VATS (video-assisted thoracic surgery). Although generally safe, it is not a minor procedure and it usually requires a hospital stay. Most people will have a tube draining from the chest for a day or so after the operation. It is quite common to have some chest pain afterwards and this can sometimes be difficult to control and/or long-lasting. Because of all this, you should discuss the benefits and risks of a biopsy with your healthcare team.

### **Why is making the diagnosis of GLILD important?**

It is possible that without treatment, damage in the lung caused by GLILD could progress and become permanent. Therefore, it is important to diagnose and treat the condition as soon as possible.

## How is GLILD treated?

Because GLILD is so rare, medical experts do not always agree on the correct course of treatment. However, experts do agree about the importance of getting the basics of **CVID care** right if you also have GLILD. This includes, for example making sure that antibody trough levels were within the desired range, and ensuring infections are being prevented and treated properly.

Experts also agree that if treatment for GLILD is required then this should usually start with **steroids** (usually tablets of prednisolone/prednisone). Although steroids are natural hormones and essential for life, using them at these higher doses especially for a long time can result in side effects. Steroids can affect mood and sleep, interfere with blood sugar, cause change in appetite, weight gain and alteration in body fat distribution, result in increased susceptibility to infection (they work by damping down the immune response) and, when used longer term, lead to thinning of the skin and the bones (osteoporosis) among other problems. Of course, not everyone gets all these side effects.

Your doctor will speak to you about which treatment options are best for you. You should expect to be at the centre of the decisions made about your treatment and our specialist nurses and psychologists can offer you additional support.



## Advice from patients with GLILD

Don't panic

Provide family and friends with accurate information

Be positive

Use trusted sources of information

Talk to others – about both the positive and negative

Putting a slope on your bed helps with coughing at night

Don't focus on it



## References

Hurst JR, Verma N, Lowe D *et al.* BLF/UKPIN Consensus Statement on the Definition, Diagnosis and Management of Granulomatous-Lymphocytic Interstitial Lung Disease (GLILD) in Common Variable Immunodeficiency Disorders (CVID). *J Allergy Clin Immunol: In Practice*: 2017 in press

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