Cryptogenic Fibrosing Alveolitis / CFA

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What Is Cryptogenic Fibrosing Alveolitis (CFA)?

This disease has a difficult name. It may help by explaining what the words mean:

**Cryptogenic** - means of unknown cause.

**Fibrosing** - refers to scarring and in this disease it is scarring of the lungs.

**Alveolitis** - refers to inflammation of the tiny sac-like air spaces in the lung where carbon dioxide and oxygen are exchanged.

CFA is also known as Usual Interstitial Pneumonitis (UIP) or Interstitial Pulmonary Fibrosis (IPF).

The scarring or fibrosis of the lungs is the main feature of CFA. This progresses and causes the lungs to feel stiff and this makes breathing difficult leading to breathlessness, especially when doing activities that are more strenuous. The rate of progression is very variable from very slow to rapid.

CFA is a relatively rare condition although more people are being diagnosed with the disease. It is estimated that 5 people out of every 100,000 of the population have CFA. Equal numbers of men and women get CFA and most cases are diagnosed between the ages of 40 to 70.
What Causes CFA?

It is not known what causes CFA hence the word “cryptogenic”. There is some idea that it is an autoimmune disorder. This is a condition in which the body's immune system attacks its own tissues, so called “friendly fire”. Another theory is that it develops as an after effect of an infection, most likely a virus. In rare cases, it is an inherited condition, which is known as familial idiopathic pulmonary fibrosis. Additional research is being done to determine whether there is a genetic tendency to develop the disease.

There are some conditions, which develop similar scarring to CFA, where the cause is known. These include the diseases Rheumatoid Arthritis / Systemic Lupus Erythematosis (SLE) and other rheumatological diseases. Some drugs e.g. amiodarone can cause scarring, as well as some ‘dusts’ e.g. asbestos.

If you are diagnosed with the disease, you can be reassured that:

- CFA is not an infection and therefore, can not be caught by other people.
- CFA is not a form of cancer.

How Will CFA Make Me Feel?

The symptoms of CFA are given below:

- Shortness of breath. This is usually the first symptom noticed by patients. This may first be noticed after taking exercise. The scarring may be quite advanced before symptoms are first noticed.

- A ‘dry’ cough which means that no sputum or lung secretions are produced.

- Tiredness and weight loss.

- The tips of fingers can swell up. This is called clubbing.

- Chest infections may be more common.
How Is CFA Diagnosed?

The doctor may suspect the diagnosis if on examination they find crackles (like opening Velcro) when listening to the bottom of the lungs and clubbing of the fingers.

Investigations that are carried out may include:

- **Blood Tests**
  Regular blood tests will give the doctor information about your general well being.

- **Lung Function Test (LFTs)**
  Lung function tests require the patient to breathe into a mouthpiece. The mouthpiece, in turn, is connected to a machine that measures the amount of air the patient breathes in and out over a certain period. The results tell the doctor how well the lungs are working.

- **Chest X-ray**
  In a CFA patient, the X-ray usually shows scarring in the lower part of the lungs.

- **Computed Tomography (CT scan)**
  A CT scan provides a very detailed picture of the lungs and is often enough to confirm the diagnosis of CFA. It involves lying still inside a long, dome-shaped machine that allows x-rays to pass through the chest. It takes up to 20 minutes for the procedure to be completed.

Sometimes further tests need to be done to confirm the diagnosis. These include:

- **Bronchoscopy**
  In this procedure, a narrow, flexible, lighted tube is put down the windpipe and into the major tubes of the lung. This procedure is carried out under sedation and may be a little uncomfortable, but it is not painful. There are two types of samples that may be taken at the time of bronchoscopy.

  a) **A Transbronchial Lung Biopsy**
     This procedure allows a very small sample of lung to be taken to confirm the diagnosis.

  b) **Bronchoalveolar Lavage**
     A ‘washing’ test may be done. This involves using a small amount of sterile salty water to wash off some cells lining the lungs. This ‘wash’ helps to rule out other causes of lung disease such as infections. It also helps to assess the type of inflammation in the lungs.

- **Surgical Lung Biopsy**
  Often a larger, but still very small piece of lung needs to be removed and examined to make the diagnosis of CFA. This is the ‘gold standard’- internationally agreed, way of making a diagnosis. A surgical biopsy can be taken during an ‘open’ procedure where
a 5-6 cm cut will be made between the ribs. In the newer technique of thoracoscopy, biopsies are taken via a fibreoptic telescope. This is a thin, tube-like instrument containing a tiny camera, which is inserted into the chest. This camera allows the surgeon to see the lungs on a video monitor. In a patient with no other major illness, recovery from a surgical lung biopsy is quite quick, taking on average 4 to 7 days.

Diagram of the Lungs

![Diagram of the Lungs](image)

What Is The Course Of The Disease?

The disease usually develops slowly, sometimes over years. The early stages are marked by alveolitis, an inflammation of the air sacs called alveoli, in the lungs. The job of the alveoli is to allow the transfer of oxygen from the lungs into the blood and the elimination of carbon dioxide from the lungs and out of the body.

As CFA progresses, the alveoli become damaged and scarred. This stiffening makes breathing difficult and brings on a feeling of breathlessness, especially when doing activities that are more strenuous.
How Is CFA Monitored?

Your doctor will wish to see you on a regular basis to monitor the disease. Your condition will be monitored through:

- Your symptoms
- Chest X-rays
- Breathing or Lung Function tests
- Blood tests

The results of these tests will help the doctors to decide how best to manage the treatment of your particular symptoms.

The Clinic will give you access to the specialists in this disease. In this way we hope to be able to offer you the best health care with the minimum amount of disruption to your life. The clinic can provide support and expert advice for your particular condition.

What Treatment Will I Be Given?

Treatment of CFA is difficult.

The first line treatment for CFA is steroids. These are usually started at a moderate to high dose and then reduced gradually to a low dose if there is a response to them. If there is no response, they will be stopped after a couple of months. Only about 20% of patients show an improvement with steroids.

If patients require a high dose of steroids to control their symptoms, other medication may be used in an attempt to reduce the amount of steroids needed. The main alternative or add-on drug is Azathioprine, see later.

Other treatments used are inhalers, which may ease your breathlessness.

You may also need oxygen therapy, in the later stages of the illness, which can be set up for you in your home. Oxygen can be obtained from compressed oxygen cylinders, which are supplied by a local pharmacy and delivered regularly to the home, or from an oxygen concentrator machine, which filters the room air to produce a greater concentration of oxygen. These are both available on NHS prescription.

Unfortunately, 50% of patients die within 5 years of diagnosis, even with treatment.
What Are The Side Effects Of Steroids?

It is understandable that you may be concerned about starting steroids. They, like all medicines, do have side effects, which is why the risks and benefits of treatment have been weighed up carefully by your doctor in discussion with you.

You will receive a steroid card that you should carry with you at all times and read carefully.

- **Risk Of Infection**
  Steroids at high doses do reduce your ability to fight infection. This means -
  a) You should promptly seek medical advice if you think you have an infection.
  b) Avoid contact with anyone with chicken pox or measles.

- **Risk Of Stopping Treatment Suddenly**
  If you have been taking steroids for more than three weeks, do not stop taking them suddenly without medical advice. This can make you feel very unwell and is potentially dangerous. Steroids must be reduced in stages.

- **Weight Gain**
  Steroids increase appetite, which can lead to weight gain. It is important that you watch your diet carefully.

- **Bone Thinning Or Osteoporosis**
  Long term steroids can cause bone thinning. You will be carefully monitored for this. In order to help reduce the risk of bone thinning, it is recommended that you:

  a) Do not smoke.
  b) Avoid drinking large amounts of alcohol.
  c) Take part in regular weight-bearing exercise such as walking or running.
  d) You may be offered calcium preventative treatment.
What Do I Need to Know About Azathioprine?

- **How Does the Drug Work?**
  Azathioprine reduces inflammation and suppresses the body’s immune system.

- **What Dosage of the Drug Will I Take?**
  It is usual to start on a low dose of azathioprine. It may need to be increased. The dose given is linked to your body weight. The drug can take 6-12 weeks before any benefits are seen.

- **What Are the Possible Side Effects?**
  Azathioprine can cause sickness, diarrhoea, skin rashes, and loss of appetite or hair loss. Another important side effect is an increased susceptibility to infections.
  Tell your doctor if you develop signs of infection such as:
  - a sore throat or fever
  - bruising or bleeding without an obvious reason
  - or jaundice.
  **Particular infections to avoid are chicken pox and shingles.** If you have not had chicken pox before and feel that you have been exposed to it, you should contact your doctor straight away, as you may need to start special treatment.

- **What Checks Will I Have to Have?**
  When taking azathioprine it is very important to have regular blood tests. The drug causes fewer blood cells to be produced and can sometimes cause liver problems. **Azathioprine should not be taken if you are not having regular blood tests.**

- **Can I Take Other Medications?**
  Drugs can interact with one another. Before taking any ‘over the counter’ medicines check with your doctor whether they are likely to cause you side effects.

- **Can I Drink Alcohol?**
  There is no specific reason to avoid alcohol with azathioprine.

- **Is Azathioprine Safe in Pregnancy?**
  You should usually not take this drug whilst you are pregnant. If you are planning to have a family or have become pregnant whilst taking azathioprine, you should contact your doctor as soon as possible. This drug should not be taken if breast-feeding.

- **Is it OK to Have Immunisation Injections?**
  Injections that contain live vaccines such as Rubella (German Measles) or polio should not be taken. It is safe to have vaccines against flu.
How Can I Help Myself?

The best way to help yourself is to ensure that you eat a sensible healthy diet that is recommended for everyone. Maintaining your correct weight and taking regular exercise all helps, in addition to getting enough rest in the day.

It is particularly important that CFA patients do not smoke and avoid smoky atmospheres.

There is no specific diet for CFA. We have provided the outline of a healthy and balanced diet.

**Reduce These Foods:**
- Reduce your intake of saturated fats. This includes animal fats and dairy products.
- Oils and fats to avoid are vegetable (polyunsaturated) oils and margarines (partially hydrogenated fats and transfatty acids).

**Increase Intake of These Foods:**
- Use extra virgin olive oil for salad dressing and as a substitute in recipes using oil.
- Eat lots of fruit and vegetables – the present recommended government daily intake is 5 portions of fruit / vegetables a day.
- Eat more oily fish such as salmon, herring and sardines. These are good sources of protein.
- Other good sources of protein are chicken, fish in general, pulses and nuts.

CFA Research

At UCL, we have an active research program looking at what is the most effective drug treatment for CFA patients. We are always looking for volunteers to assist us with this research which only requires providing us with a blood sample and answering questions for our CFA database.

If you would like to take part in our research or just want to know more details about the studies we are doing please contact the Respiratory Research Nurse, in the first instance: Tel 0207 387 9300 ext 4135 or email: catherine.read@uclh.org

Consultants Involved in the Research:

Dr Helen Booth
Consultant Thoracic Physician
UCL Hospitals
London W1T 3AA
Tel 0207 380 9005
Useful Contacts

**The British Lung Foundation**
78 Hatton Garden
London
EC1N 8LD
Tel: 0207 831 5831
Website: www.lunguk.org
E-mail: blf@britishlungfoundation.com

Breathe Easy Club
The British Lung Foundation has a network of regional offices across Britain. Their dedicated teams run events, promote the work of the British Lung Foundation and support Breathe Easy groups. They are contactable through the British lung Foundation.

**Breathe Easy**
Breathe Easy is part of the British Lung Foundation and is a support network for patients with breathing problems.
E-mail: breatheasy@britishlungfoundation.com

**CFA & Interstitial Lung Association (SILA)**
The Secretary, SILA
Chest Clinic Office
Victoria & Albert Ward
King’s College Hospital
Denmark Hill
London SE5 9RS
Website: www.CFA.org.uk
E-mail: info@CFA.org.uk
Glossary

**Alveoli** - The tiny sac-like air spaces in the lung where carbon dioxide and oxygen are exchanged.

**Biopsy** - The removal of a small sample of tissue from the body for examination under a microscope. This helps in diagnosis.

**Bronchoscopy** - A procedure carried out under local anaesthetic to look in the lungs. A miniature video camera at the end of a flexible tube is put down into the airways.

**Clubbing** - The broadening or thickening of the fingertips and toes.

**Crackles** - The sound that the lung makes when listened to through a stethoscope that can indicate scarring in the lungs.

**Fibrosis** - Scarring

**Immune System** - The body’s ability to fight infection.