Lung Inflammation (ILD)	Regular pulmonary function tests to assess lung capacity, HR-CT scan required to assess extent and severity of damage. Dependent on severity, this may require steroids or immunosuppressants such as Azathioprine or Cyclophosphamide	Very rarely seen, can accompany scleroderma or lupus
Pulmonary hypertension (PH)	Regular echocardiogram to assess pressure inside pulmonary artery. Further investigations required according to specialist centre assessment. This is very rare and requires highly specialised management	

Drugs used in MCTD

Steroids, either oral, intramuscular or intravenous, are used when the disease is active in order to gain control. These are often used for those with internal organ problems and drugs that suppress the immune system, such as Methotrexate, Azathioprine or Cyclophosphamide, are used to help reduce the steroid dose over time and maintain remission. Whilst these drugs do carry some significant potential side effects, the risks of these are always outweighed by the risks of a very active disease such as MCTD and doses are always kept to a minimum to help reduce and minimise risk.

Take home messages

MCTD is a less commonly seen connective tissue disease that can cause a wide variety of different symptoms. Whilst some can develop internal organ complications, this is not always the case. There are three important messages for you to remember from this leaflet:

1. Learning to live with and finding ways to manage your symptoms are important parts of coming to terms with your illness.
2. You can improve symptoms such as fatigue by pacing and planning periods of activity and rest, listening to your body and setting realistic achievable goals (more information can be found about these in other LUPUS UK leaflets)
3. Pay attention to the variant or type of MCTD that you have and keep in touch with your rheumatology team if you develop any new symptoms or if you are at all concerned.

THE LUPUS UK RANGE OF FACT SHEETS

A range of fact sheets are available as follows:

1. LUPUS Incidence within the Community
2. LUPUS A Guide for Patients
3. LUPUS The Symptoms and Diagnosis
4. LUPUS The Joints and Muscles
5. LUPUS The Skin and Hair
6. LUPUS Fatigue and your Lifestyle
7. LUPUS and Pregnancy
8. LUPUS and Blood Disorders
9. LUPUS and Medication
10. LUPUS and the Kidneys
11. LUPUS and Associated Conditions
12. LUPUS and the Brain
13. LUPUS The Heart and Lungs
14. LUPUS The Mouth, Nose and Eyes
15. LUPUS and Light Sensitivity
16. LUPUS and the Feet
17. LUPUS and Men
18. LUPUS and Mixed Connective Tissue Disease

FACT SHEETS

LUPUS UK is the registered national charity caring for people with lupus and has over 6,000 members who are supported by the Regional Groups.

LUPUS UK acknowledges with gratitude the assistance of Sue Brown MSc RGN NIP EN (G) Consultant Nurse in Rheumatology (Connective Tissue Diseases) Royal National Hospital for Rheumatic Diseases NHS Foundation, Bath in the provision of clinical information towards the production of this fact sheet.

LUPUS UK also thanks the Wooler Walkers (Northumberland) for their valued fundraising towards the cost of producing the fact sheets.

Please contact National Office should you require further information about lupus. LUPUS UK will be pleased to provide a booklist and details of membership.

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PW 05/13 1K

LUPUS and Mixed Connective Tissue Disease

Published by LUPUS UK
Reg. Charity Nos 1051610, SC039682

LUPUS
and Mixed Connective Tissue Disease

What is Mixed Connective Tissue Disease?

Mixed connective tissue disease (MCTD) is an auto-immune disease that was first recognised in 1972 and is considered to be a cousin of lupus. We now understand that there can be an overlap in many auto-immune diseases that may involve any number of different variants of connective tissue diseases. MCTD is the commonest seen combination, and symptoms of MCTD usually either appear as a lupus or a scleroderma variant. Some people have a polymyositis variant so have problems with muscle inflammation as well. Often with time, MCTD may evolve into one of the conditions of lupus, scleroderma or polymyositis. Dermatomyositis can also be seen when the inflammation affects the skin as well as the muscles.

Diagnosing MCTD

For many, to gain a diagnosis of any connective tissue disease is difficult and this is still the case for MCTD. As the immune system is over-active in these conditions, most people with MCTD will have positive Anti-Nuclear Antibodies (ANA) and also antibodies to ribonuceloprotein (RNP). The specific antibody for MCTD is called Anti-U1-RNP. Other diagnoses should always be excluded before coming to a diagnosis of MCTD, especially in people who do not carry any MCTD associated antibodies.

Who gets MCTD?

As is seen in many other rheumatological conditions, more women than men have MCTD and it can occur at any age. Children can be diagnosed with MCTD, where Raynaud's phenomenon (a circulatory disorder affecting mostly the hands and feet), fatigue and pain tend to be the commonest symptoms. As signs and symptoms can change over time, children with a diagnosis of MCTD should always be closely monitored and these should be acted upon if any other features develop.

Symptoms of MCTD

The most commonly seen symptoms in MCTD include:

- Joint pain and swelling
- Fatigue
- Skin rashes
- Hair loss
- Dry eyes and dry mouth
- Muscle inflammation
- Raynaud's phenomenon
- Tight skin and/or 'sausage-shaped' digits
- Dysphagia - Difficulty in swallowing
- Trigeminal neuralgia – inflammation of the nerves affecting the jaw line.

Complications of MCTD

Some people with MCTD can develop internal organ problems although this is rarer. All people with MCTD should pay attention to any new episodes of breathlessness and/or heart palpitations. Regular heart and lung investigations should always be requested in order to look out for possible signs of interstitial lung disease (ILD) or pulmonary hypertension. These conditions, although in some cases serious, are now much better diagnosed and treated in specialist centres that see people with MCTD on a regular basis. Investigations need to be balanced according to symptoms and for many, these complications may never occur.

Treatments for MCTD

Treatments for MCTD vary dependent on the types of symptoms that you may have. The following table describes some of the treatments used, but it is important to remember that your specialist team will always assess how active your disease is and suggest treatments according to your variant of MCTD. This table describes the symptoms and important points and in which variant you would expect these to occur.

Symptoms	Treatments and important points	Associated with
Raynaud's phenomenon	High dose vitamin regimes and certain drugs can help to improve circulation e.g. Nifedipine. Poor circulation can lead to fingertip ulceration that may require antibiotics or intravenous treatments	Lupus Scleroderma Polymyositis
Joint pain and swelling	Painkillers such as Paracetamol or anti-inflammatory drugs can help. In some, drugs like Hydroxychloroquine or Methotrexate can be used. If severe, steroids can be used (tablets or injections). X-rays will show any damage to joints, if this is the case then drugs such as Methotrexate can be used	Lupus Scleroderma Rheumatoid Arthritis
Skin rashes	Topical creams can be used, sometimes needing steroid based creams. Hydroxychloroquine can help especially in those with a lupus variant. Rashes need to be treated effectively in order to control disease activity and minimise a flare of symptoms	Lupus Dermatomyositis
Difficulty in swallowing	Medication can help to reduce acid secretion in the stomach if reflux is a problem. This needs to be investigated fully to find the best treatments	Scleroderma Polymyositis
Tight skin and/or 'sausage-shaped' digits	Cream and moisturise hands regularly. Monitor the extent of skin thickening and report any change to your rheumatology team. Be careful of the development of digital ulcers especially in those with severe Raynaud's phenomenon	Scleroderma Polymyositis
Dry eyes and dry mouth	Symptoms can be improved by replacement eye drops/ointments (Hypromellose, Lacrilube) and saliva replacements. See your optician and dentist regularly to assess any damage caused by the dryness	Sjögren's Syndrome Lupus Scleroderma Polymyositis
Fatigue	Drugs such as Hydroxychloroquine can have some benefit in the lupus variant of MCTD. See your nurse or occupational therapist for advice to help manage the fatigue	All connective tissue diseases
Hair loss	This can be related to active disease so make sure you mention this when attending your appointment. Some people can experience hair loss as a side effect of certain medications. This is very difficult to treat and may need referral to a dermatologist or tricologist	Lupus
Muscle inflammation	Dependent on severity, this may require steroids or immunosuppressants such as Azathioprine or Cyclophosphamide. Regular monitoring is required to assess active disease and to prevent side effects associated with medication	Polymyositis