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## Review Article

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## Mixed Connective Tissue Disease (MCTD)' in the World

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### Abstract

Mixed connective tissue disease is a term used by some doctors to describe a disorder characterized by features of systemic lupus erythematosus, systemic sclerosis, and polymyositis.

The diagnosis is based on symptoms and the results of blood tests to detect levels of characteristic antibodies.

Treatment varies depending on the severity of symptoms and may include nonsteroidal anti-inflammatory drugs, hydroxychloroquine, corticosteroids, immunosuppressive drugs, or a combination. Raynaud phenomenon, joint pains, various skin abnormalities, muscle weakness, and problems with internal organs can develop.

**Keywords:** Mixed connective tissue disease, blood tests, drugs.

### Introduction

**Mixed connective tissue disease** is a term used by some doctors to describe a **disorder** characterized by features of systemic lupus erythematosus, systemic sclerosis, and polymyositis. Raynaud phenomenon, joint pains, various skin abnormalities, muscle weakness, and problems with internal organs can develop.

### Overview:

**Overview of Autoimmune Disorders of Connective Tissue.** In an autoimmune disorder, antibodies or cells produced by the body attack the body's own tissues. Many autoimmune disorders affect connective tissue and a variety of organs. Connective tissue is the structural tissue that gives strength to joints, tendons, ligaments, and blood vessels.

Autoimmune rheumatic disorders include

Autoimmune myositis

Eosinophilic fasciitis

Mixed connective tissue disease

Relapsing polychondritis

Sjögren syndrome

Systemic lupus erythematosus (SLE)

Systemic sclerosis

Rheumatoid arthritis and spondyloarthritis are examples of other autoimmune disorders that affect connective tissue.

- Autoimmune disorders can affect other tissues in the body beside connective tissue, and some people with autoimmune disorders of connective tissue have other kinds of autoimmune disorders, such as Hashimoto thyroiditis (an autoimmune thyroid disorder that can lead to overactivity or underactivity of the thyroid gland). Diagnosis: A doctor's evaluation
  - Laboratory tests
  - Sometimes established criteria

An autoimmune disorder of connective tissue (also called an autoimmune rheumatic disorder or autoimmune collagen-vascular disorder) is diagnosed

on the basis of its particular symptom pattern, the findings during a physical examination, and the results of laboratory tests (such as blood tests and biopsies). For some of these disorders, doctors can also base the diagnosis on an established set of criteria.

Sometimes the symptoms of one disease overlap with those of another so much that doctors cannot make a distinction. In this case, the disorder may be called undifferentiated connective tissue disease or an overlap disease.

### Symptoms:

In autoimmune disorders, inflammation and the immune response may result in connective tissue damage, not only in and around joints but also in other tissues, including vital organs, such as the kidneys and organs in the gastrointestinal tract. The sac that surrounds the heart (pericardium), the membrane that covers the lungs (pleura), and even the brain can be affected. The type and severity of symptoms depend on which organs are affected.

Most autoimmune rheumatic disorders increase the risk of developing cholesterol deposits (plaques) in arteries, resulting in hardening of the arteries (atherosclerosis).

### Treatment:

Corticosteroids and/or other drugs that suppress the immune system.

Many autoimmune disorders of connective tissue are treated with corticosteroids, other drugs that suppress the immune system (immunosuppressive drugs), or both.

People who take corticosteroids, such as prednisone, are at risk of fractures related to osteoporosis. To prevent osteoporosis, these people may be given the drugs used to treat osteoporosis, such as bisphosphonates, denosumab, teriparatide, and supplemental vitamin D and calcium. People whose immune system is suppressed by corticosteroids and other drugs and by autoimmune disease itself are often given drugs to prevent infections such as by the fungus *Pneumocystis jirovecii* (see prevention of pneumonia in immunocompromised people).

In people who have overlap diseases, doctors treat symptoms and organ dysfunction as they develop.

Because people with autoimmune disorders have an increased risk of infection, it is important for them to get recommended vaccinations, such as the influenza vaccine and the pneumococcal vaccine.

Although many people who have autoimmune disorders of connective tissue have tried changing their diet to reduce the inflammation caused by these disorders, no "anti-inflammatory" diet has been shown to alleviate symptoms or change the course of the disorder.



Is mixed connective tissue disease fatal? Despite **treatment, mixed connective tissue disease** worsens in about 13% of the people, causing potentially **fatal** complications. Causes of death include pulmonary hypertension (mainly) and heart **disease**. The prognosis is worse for people who have mainly features of systemic sclerosis or polymyositis.

### What are the symptoms of mixed connective tissue disease (MCTD)?

Fatigue.  
Muscle pain with no apparent cause.  
Joint pain.  
Low-grade fever.

Raynaud phenomenon (reduced blood flow to the fingers, toes, ears, and nose, causing sensitivity, numbness, and loss of color in these areas).  
What is the treatment for mixed connective tissue disease? Other ways to control symptoms of mixed connective tissue disease include: **Nonsteroidal** anti-inflammatory drugs. These medications, such as ibuprofen (Advil, Motrin IB, others) or naproxen sodium (Aleve), may help relieve the pain and inflammation if your condition is mild. Is Mctd curable? **mixed connective tissue disease (MCTD)** is

a rare autoimmune disorder. ... Some cases of **MCTD** also share symptoms with rheumatoid arthritis. There's no **cure** for **MCTD**, but it can usually be managed with medication and lifestyle changes. It shouldn't have much of an impact on your life expectancy. Is mixed connective tissue disease a disability?

One particular overlap syndrome is characterized by features of scleroderma, lupus, and polymyositis and is referred to as **mixed connective tissue disease (MCTD)**, also known as Sharp's syndrome. ... First, Social Security has a **disability** listing for **connective tissue diseases** in its **disability** evaluation handbook. What causes connective tissue disorder?

These can affect blood flow to the organs and other body tissues. Vasculitis can involve any of the blood vessels. Mixed connective tissue disease. People with **MCTD** have some features characteristic of several diseases, including **lupus, scleroderma, polymyositis** or **dermatomyositis**, and **rheumatoid arthritis**. Can mixed connective tissue disease be fatal? These can affect blood flow to the organs and other body tissues. Vasculitis can involve any of the blood vessels. Mixed connective tissue disease. People with **MCTD** have some features characteristic of several diseases, including **lupus, scleroderma, polymyositis** or **dermatomyositis**, and **rheumatoid arthritis**.



What are the signs and symptoms of polymyositis?  
**The common symptoms of polymyositis include:**

- Muscle pain and stiffness.
- Muscle weakness, particularly in the belly (abdomen), shoulders, upper arms, and hips.
- Joint pain and stiffness.
- Trouble catching your breath.
- Problems with swallowing.
- Irregular heart rhythms, if the heart muscle becomes inflamed.

**How does connective tissue disease affect the body? In this systemic disorder, immune cells attack and inflame the membrane around joints. It also can affect the heart, lungs, and eyes. ... Systemic lupus erythematosus (SLE):**

**A disease that can cause inflammation of the connective tissue in every organ of the body, from the brain, skin, blood, to the lungs. What causes connective tissue disorder? These can affect blood flow to the organs and other body tissues. Vasculitis can involve any of the blood vessels. Mixed connective tissue disease. People with MCTD have some features characteristic of several diseases, including lupus, scleroderma, polymyositis or dermatomyositis, and rheumatoid arthritis. Is connective tissue disease hereditary? Some of these disorders have no clear cause, and some are inherited. Certain hereditary disorders cause**

connective tissue **throughout the body to form abnormally. In general,** hereditary connective tissue disorders **develop in childhood but last throughout life.** Overview of Connective Tissue Disorders in Children

Connective tissue is the tough, often fibrous tissue that binds the body's structures together and provides support and elasticity. Muscles, bones, cartilage, ligaments, and tendons are built mostly of connective tissue. Connective tissue is also present in other parts of the body, such as the skin and internal organs. The characteristics of connective tissue and the types of cells it contains vary, depending on where it is found in the body. Connective tissue is strong and thus able to support weight and tension.

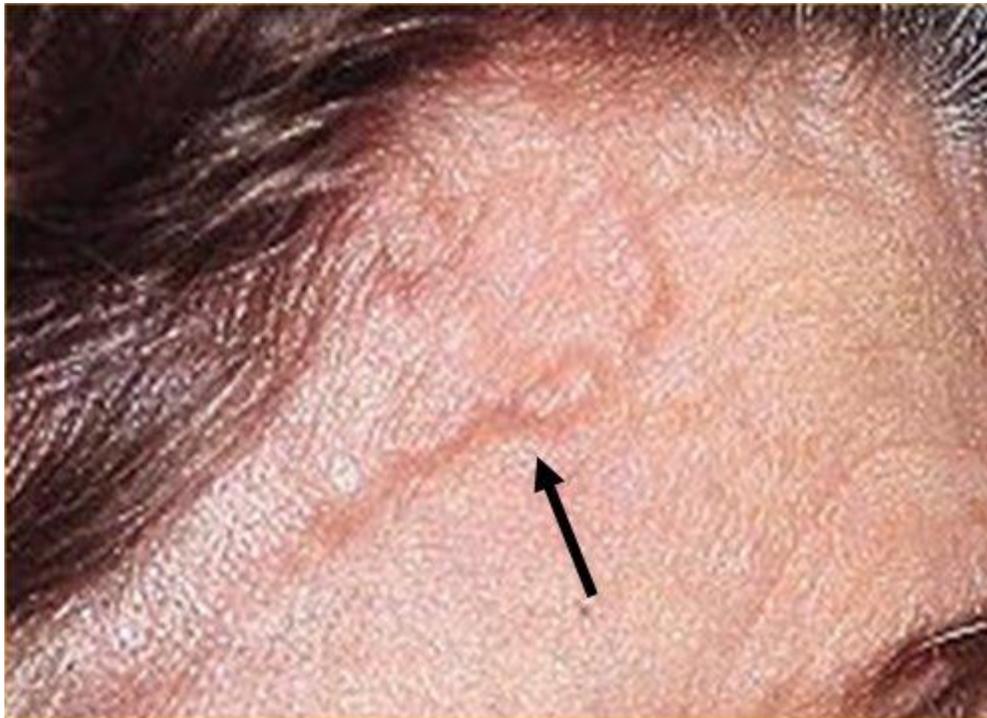


There are over 200 disorders that involve connective tissue. Specific disorders discussed here include

- Chondromalacia patellae
- Cutis laxa
- Ehlers-Danlos syndrome
- Infrapatellar tendinitis
- Marfan syndrome
- Nail-patella syndrome
- Osteochondrodysplasias

- Osteogenesis imperfecta
- Pseudoxanthoma elasticum

Some of these disorders have no clear cause, and some are inherited. Certain hereditary disorders cause connective tissue throughout the body to form abnormally. In general, hereditary connective tissue disorders develop in childhood but last throughout life.



### Diagnosis

- A doctor's evaluation
- X-rays
- Biopsy
- Analysis of genes

Most hereditary connective tissue disorders are diagnosed based on their symptoms and findings during a physical examination.

X-rays can reveal bone abnormalities that may be associated with a connective tissue disorder.

A biopsy (removal of a tissue sample for examination under a microscope) can also help. The tissue is usually removed using a local anesthetic, which numbs the area.

Analysis of genes, usually from a sample of blood, may help doctors diagnose some hereditary disorders.

**What's the outlook?** Despite its complex range of symptoms, MCTD generally isn't too serious. Most people are able to manage their symptoms with a combination of medication and lifestyle changes. Talk to your doctor about establishing a long-term treatment plan that works best for your symptoms.

### Article resources;

Medically reviewed by Nancy Carteron, MD, FACR on January 4, 2018 — Written by Corinne O'Keefe Osborn. **Are there any risk factors?** Some people with MCTD have a family history of it, but researchers haven't found a clear genetic link.

According to the Mayo Clinic, it seems to be most common among women under 30, but it can strike at any age. **Conclusion:** Medication can help manage the symptoms of MCTD. Some people only require treatment when their disease flares up, but others may need long-term treatment.

Medications used to treat MCTD include:

- **Nonsteroidal anti-inflammatory drugs (NSAIDs).** Over-the-counter NSAIDs, such as ibuprofen (Advil, Motrin) and naproxen (Aleve), can treat joint pain and inflammation.
- **Corticosteroids.** Steroid medications, such as prednisone, can treat inflammation and help to stop your immune system from attacking healthy tissues. Because they can result in many side effects, such as high blood pressure, cataracts, mood swings, and weight gain, they're usually used only for brief periods of time to avoid long-term risks.
- **Antimalarial drugs.** Hydroxychloroquine (Plaquenil) can help with mild MCTD and probably prevent flare-ups.

- **Calcium channel blockers.** Medications such as nifedipine (Procardia) and amlodipine (Norvasc) help manage Raynaud's phenomenon.
- **Immunosuppressants.** People with severe MCTD may require long-term treatment with immunosuppressants, which are drugs that suppress your immune system. Common examples include azathioprine (Imuran, Azasan) and mycophenolate mofetil (CellCept). These drugs may be limited during pregnancy due to the potential for fetal malformations or toxicity.
- **Pulmonary hypertension drugs.** Pulmonary hypertension is the leading cause of death among people with MCTD. Doctors may prescribe drugs like bosentan (Tracleer) or sildenafil (Revatio, Viagra) to prevent pulmonary hypertension from getting worse. In addition to medication, several lifestyle changes can also help:
  - **Get regular exercise.** A moderate level of physical activity four to five times per week will help improve muscle strength, reduce blood pressure, and decrease your risk of heart disease.
  - **Quit smoking.** Smoking causes your blood vessels to narrow, which can make the symptoms of Raynaud's phenomenon worse. It also raises blood pressure.
  - **Get enough iron.** About 75 percent of people with MCTD have iron deficiency anemia.
  - **Eat a high-fiber diet.** Eating a healthy diet rich in whole grains, fruits, and vegetables can help keep your digestive tract healthy.

Protect your hands. Protecting your hands from the cold can reduce your chances of having a Raynaud's phenomenon flare-up. Inheritance: The role of genetics in the onset of mixed connective tissue disease (MCTD) is still unclear. Some people with MCTD have family members who are also affected by the condition. This suggests that in some cases, an inherited predisposition may contribute to the development of MCTD. People with an inherited or genetic predisposition have an increased risk of developing a certain condition due to their genes.<sup>[3][5]</sup>

#### In-Depth Information

Medscape Reference provides information on this topic. You may need to register to view the medical textbook, but registration is free.

Mixed Connective-Tissue Disease

Pediatric Mixed Connective Tissue Disease

Dermatologic Manifestations of Mixed Connective Tissue Disease.

The Merck Manual for health care professionals provides information on Mixed connective tissue disease.

The Monarch Initiative brings together data about this condition from humans and other species to help physicians and biomedical researchers. Monarch's tools are designed to make it easier to compare the signs and symptoms (phenotypes) of different diseases and discover common features. This initiative is a collaboration between several academic institutions across the world and is funded by the National Institutes of Health. Visit the website to explore the biology of this condition.

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