



SCLERODERMA IN CHILDREN

Jayne MacMahon, MD

Childhood Myositis Fellow, Paediatric Rheumatology
PGY6, University of Toronto
Division of Rheumatology | The Hospital for Sick Children



WHAT IS SCLERODERMA?

Scleroderma is a rare condition in children. It often affects the skin. It can cause the skin to become hard and tight. In some children, it can affect other organs (e.g., lungs, kidneys, joints, stomach, and intestines).

Scleroderma is an autoimmune disease. It happens when the body attacks its own skin, and sometimes organs, by mistake. In autoimmune diseases, our immune system (the chemicals and blood cells that are supposed to fight off germs) is overactive. Extra chemicals and blood cells get trapped in the blood vessels of the skin (and sometimes organs). This causes inflammation and damage there. It is a chronic (ongoing) condition. It may get worse over time.

SCLERODERMA IN CHILDREN



THERE ARE TWO TYPES OF SCLERODERMA IN CHILDREN.

The first is called 'localized scleroderma'. This is the most common in children. It affects the skin in one area of the body only.

The other type is called 'systemic scleroderma'. This can affect many areas of the skin, as well as the organs of the body. It is rare in children.

WHAT ARE THE SYMPTOMS?

It is important to remember that scleroderma can affect every child differently. The most common things that happen are:

- ▶ Tightness and swelling of the skin, sometimes the skin changes colour.
- ▶ Pain or swelling in the joints.
- ▶ Pale, tingling or numb fingers, often in cold weather or when stressed. (This is called Raynaud's Phenomenon).
- ▶ Hard bumps (calcium) under the skin.
- ▶ Sores on the fingertips or knuckles.
- ▶ Spider veins.
- ▶ Heartburn and trouble swallowing.
- ▶ Shortness of breath.

HOW IS IT DIAGNOSED?

If you or your doctor thinks your child has scleroderma, the first step is to be seen by a specialist doctor (called a 'pediatric rheumatologist'). The specialist will ask about your child's illness and give your child a check-up. They may ask for extra blood tests or X-rays. Depending on how your child is feeling, this may include:

- ▶ Blood and urine (pee) tests to look at blood counts, antibodies (chemicals found in the blood that may contribute to causing scleroderma), how well the liver is working, and how well the kidney is working.
- ▶ Imaging tests (such as X-rays, CTs, and MRIs). These look for any changes in the body's organs.
- ▶ Breathing tests (called Pulmonary Function Tests) to look at how well the lungs are working.
- ▶ Ultrasound of the heart (called an echocardiogram) to look at how well the heart and blood vessels are working.
- ▶ Biopsy of the skin to look at the skin more closely under a microscope.

SCLERODERMA IN CHILDREN

HOW IS IT MANAGED?



Scleroderma cannot be cured. The goal is to improve the skin and stop the organs from becoming damaged. Every child's treatment will be different.

IT MAY INCLUDE:

- ▶ Medicines (such as ibuprofen) to help with pain and reduce inflammation.
- ▶ Skin creams to work directly on softening the skin.
- ▶ Medicines to reduce the strength of the overactive immune system and stop inflammation. These are called 'immunosuppressive'. They can be given by mouth, by needle under the skin or through the vein.
- ▶ Treating symptoms such as heartburn or Raynaud's Phenomenon.
- ▶ Physical therapy and exercise to keep muscles strong, and the joints from tightening up.
- ▶ Regular visits with your child's rheumatology specialist.



Localised scleroderma treatment often must last for several years. It is very rare for localised scleroderma to change and become systemic scleroderma.

Children with systemic scleroderma are at a higher risk of getting damage to the skin and organs. With the proper treatment, patients may have little or no symptoms (i.e., remission) for years at a time.



REFERENCES

- Zulian F, Woo P, Athreya BH, et al. The Pediatric Rheumatology European Society/American College of Rheumatology/European League against Rheumatism provisional classification criteria for juvenile systemic sclerosis. *Arthritis Rheum.* 2007 Mar 15;57(2):203-12. doi: 10.1002/art.22551. PMID: 17330294.
- Foeldvari I, Culpo R, Sperotto F, et al. Consensus-based recommendations for the management of juvenile systemic sclerosis. *Rheumatology (Oxford)*. 2021 Apr 6;60(4):1651-1658. doi: 10.1093/rheumatology/keaa584. PMID: 33147624.
- Zulian F, Culpo R, Sperotto F, et al. Consensus-based recommendations for the management of juvenile localised scleroderma. *Annals of the Rheumatic Diseases* 2019;78:1019-1024.