



[www.printo.it/pediatric-rheumatology/AU/intro](http://www.printo.it/pediatric-rheumatology/AU/intro)

# Scleroderma

Version of 2016

## 1. WHAT IS SCLERODERMA

### 1.1 What is it?

The name scleroderma is derived from Greek and can be translated as "hard skin". The skin when affected by this disease becomes shiny and hard. There are two different types of scleroderma: localised scleroderma and systemic sclerosis.

In localized scleroderma, the disease is limited to the skin and tissues located underneath but not the internal organs. It can involve the eyes and cause uveitis, it may also involve the joints and cause arthritis. The skin can be affected in oval patches (morphea) or occur as a tight band (linear scleroderma).

In systemic sclerosis, the process is more severe and widespread and involves not only the skin but also some of the internal organs of the body.

### 1.2 How common is it?

Scleroderma is a rare disease. There is no detailed registry of children in Australia, but international estimations of its frequency never exceed 3 new cases in 100,000 people every year. Localized scleroderma is the most common form in children and affects predominantly girls. Only about 10% or less of children with scleroderma are affected by systemic sclerosis.

### 1.3 What are the causes of the disease?

Scleroderma is an inflammatory disease but the reason for the

---

inflammation has not been discovered yet. It is probably an autoimmune disease, which means that the immune system of the child reacts against itself. The inflammation causes swelling, redness or colour change from the inflamed tissues and then an overproduction of thickened (scar) tissue.

#### **1.4 Is it inherited?**

No, there is no evidence of a genetic link for scleroderma to date, although there are a few reports of the disease running in families.

#### **1.5 Can it be prevented?**

There is no known prevention for this condition. This means that you as a parent or a patient could not have done anything to prevent the onset of the disease.

#### **1.6 Is it infectious?**

No. Some infections may trigger the disease onset but the condition itself is not infectious and affected children do not need to be isolated from the others.