

Hospital Checklist

A Guide to Nursing Patients with Systemic Sclerosis (Scleroderma)

SYSTEMIC SCLEROSIS (an uncommon disease)

Scleroderma & Raynaud's UK (SRUK) has produced this leaflet because, due to the rarity of the disease, many health professionals will not be used to caring for people with systemic sclerosis (scleroderma). Whilst in hospital many patients will have different needs according to exactly how they are affected. If you have any thoughts on how to improve this information for future edits, please let us know.

Systemic sclerosis (often referred to as scleroderma) is a rare connective tissue disease affecting many different organs and systems of the body. Symptoms, disease course and outcome vary enormously between patients. There are two major subsets of the disease, depending on the extent of skin involvement – limited cutaneous systemic sclerosis (skin involvement only distal to elbows and knees) and diffuse cutaneous systemic sclerosis (both distal and proximal skin involvement). Face is normally affected in both subsets. Even though the disease mechanisms are still poorly understood, research has shown that three processes are involved in the pathogenesis of systemic sclerosis – autoimmune inflammation, vasculopathy and fibrosis.

Below is a list of the more common symptoms and signs of systemic sclerosis, although not all patients will experience every single one of them, and some patients experience only very few:

- thickening and tightness of the skin, which may be limited to fingers and toes or be very extensive, affecting the whole body;
- Raynaud's phenomenon (vasospasm causing colour changes in the hands and feet, usually triggered by temperature changes or emotional stress);
- pitting scars over the fingertips, ulcerations, critical ischemia and, in extreme cases, gangrene can develop over fingers and toes;
- calcinosis (depositions of calcium in the tissues, most often over fingers and bony prominences, but can develop anywhere);
- telangiectasia (dilated capillaries) can occur over different parts of the body, most often over the face and hands;
- dryness of the eyes, mouth and vagina;
- microstomia (small mouth);
- fixed flexion contractures (permanent bending) may occur as the tendons and joint linings become thickened and usually affects fingers, toes, elbows and knees;
- generalised stiffness in the muscles and joints is very common and can affect even people with mild disease;
- oesophageal dysfunction resulting in dysphagia (difficulties swallowing), reflux and heartburn;
- bowel dysfunction causing a variety of symptoms ranging from persistent diarrhoea to severe constipation and bowel pseudo obstruction;
- anorectal involvement can cause faecal incontinence;
- patients can often suffer with breathlessness on exertion caused by development of scar tissue in the lungs (lung fibrosis) or high blood pressure in the lung arteries (pulmonary arterial hypertension);
- clinically significant cardiac disease as a result of scleroderma is rare, although many patients can have abnormalities on ECG or echocardiography; and
- acute renal failure (hypertensive renal crisis) can occur especially in people with early diffuse cutaneous systemic sclerosis, which is why regular blood pressure monitoring is essential in patients with systemic sclerosis.

Nursing Care of Patients with Systemic Sclerosis

Because systemic sclerosis is a rare condition, often health professionals have little experience with the disease. This leaflet was developed in order to summarise disease-specific problems that patients may encounter whilst in hospital and provides nursing staff with additional information on how to help manage those problems. Below there is a checklist of some of the main problems a person with scleroderma may experience and how to manage these problems.

For you as a person with systemic sclerosis – below there is a checklist where you can tick all of the problems that apply to you. This will help nursing staff know how they should help you manage these problems as they may not have looked after someone with scleroderma before. Please tick the boxes which apply to you. Add anything extra in the “Notes” section at the end, and hand to the nursing staff on admission to a ward.

PROBLEM	MANAGEMENT	TICK
Raynaud's Phenomenon	Provide extra blankets. Avoid draughts. Maintain warmth, particularly pre & post-op.	
Ulceration & slow healing	Provide appropriate dressing. Assist with activities of daily living as necessary. Refer to occupational therapist if necessary.	
Tight skin	Extra care required with venepuncture.	
Dry skin	Assist with moisturising if required	
Painful hands & feet	Avoid injury, e.g. during transfer & ambulation.	
Painful joints	Assist with repositioning. Provide extra pillows. Massage & application of heat. Painkillers, anti-inflammatory medications. Physiotherapy assessment.	
Impaired mobility	Assistance may be required for feeding & oral hygiene. Possible difficulties with intubation.	
Sicca symptoms (dry mouth, dry eyes), Sjogrens Syndrome	Ensure drinking water readily accessible. Maintain good oral hygiene, especially when patient unable to drink. Assist with application of eye drops or ointment if patient unable to self-administer, particularly pre & post-op & prior to sleeping.	
Oesophageal involvement including difficulty swallowing	Elevate head of bed. Provide extra pillows. Sit upright when eating & after meals. Administer anti-acids. Discuss food preferences. Ensure adequate & appropriate dietary intake.	
Bowel involvement (diarrhoea &/or constipation; faecal incontinence)	Assess for dietary requirements & medication regime. May need to refer to dietician.	
Shortness of breath on exertion	Allow patient to set the pace during physical activity.	
Reduced capacity to cope	Create calm, supportive environment. Encourage stress reduction techniques. Refer for social work assessment.	
Increased sensitivity	Take extra care; patient has increased pain threshold.	
Pre-operative guide	Anaesthetic assessment prior to surgery. If patient has pulmonary arterial hypertension, the anaesthetist needs to know well in advance of operation as surgery & anaesthesia carry increased risks. Maintain warmth to allow optimum venous access.	

Scleroderma & Raynaud's UK

We are here for you, to support you through your diagnosis and to help you understand and manage your condition.

We know how important it is to join a community that understands your condition, provides expert information and support, works for you to improve access to treatment and care whilst driving research into more effective treatments as we work to find a cure.

Ways we can support you

Our informative website has up to date, specialist information on Scleroderma and Raynaud's, personal stories from the community on managing the conditions and details on services and events that are available in your local community. You can find information, support and friendship through our online community: www.sruk.co.uk

Our helpline operates 365 days a year providing support and information to everyone affected by or interested in Scleroderma and Raynaud's: **0800 311 2756**. Our experienced helpline volunteers understand the challenges and difficulties you face and provide a confidential space to talk through worries or concerns.

To order printed copies of SRUK publications, please call: **020 3893 5998** or email: info@sruk.co.uk

Find out more about the work of the charity and how we can support you by visiting our website: www.sruk.co.uk or calling our friendly team on: **020 3893 5998**.

Interested in joining our community?

We have a variety of ways that you can join the community. Our most popular is to become a member. For a small subscription fee you will receive 4 magazines per year, free attendance at annual conference, priority booking for educational events and member only discounts for our shop products.

Find out more about all the ways you can join the community by visiting our website: www.sruk.co.uk or calling our friendly team on: **020 3893 5998**.

We are here for you,
when you need us.

0800 311 2756

Our helpline operates
365 days a year

Visit us today

www.sruk.co.uk

Sources used

We rely on several sources to gather evidence for our information. All our information is in line with accepted national or international clinical guidelines where possible. Where no guidelines exist, we rely on systematic reviews, published clinical trials data or a consensus review of experts. We also use medical textbooks, journals, and government publications.

If you would like further information on the sources we use on a particular publication, please contact the Information and Support Services team at info@sruk.co.uk

Valuing your feedback

As someone who has received a copy of this booklet, we would very much value your opinion on whether it meets the needs of people affected by Scleroderma and Raynaud's. Please complete the survey online at sruk.co.uk/publications

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