

## Anaesthesia recommendations for patients suffering from

### **Systemic sclerosis**

**Disease name:** Systemic sclerosis

**ICD 10:** M34.0

**Synonyms:** Progressive systemic sclerosis, Scleroderma, CREST syndrome

Systemic sclerosis (SSc), also called scleroderma, is a multisystem connective tissue disease characterized by the excessive production of collagen, glycosaminoglycans and fibrinonectins within connective tissue. This results in the hardening and fibrosis of skin, mucus membranes, vasculature and internal organs. Clinical features include tightening and thickening of skin (skin sclerosis), Raynaud's phenomenon and involvement of various internal organs (particularly in the lungs). There are two major SSc phenotypes, limited cutaneous and diffuse cutaneous form, based on the extension of skin involvement.

Prevalence of scleroderma ranges from 4 – 489 cases per million worldwide, with an annual incidence of 0.6 – 122 million. Greater prevalence is seen in the US and Australia than in Europe and Japan. The ratio of women to men affected is 3:1 and it has a peak incidence in the fifth decade of life.

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Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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Find more information on the disease, its centres of reference and patient organisations on Orphanet: [www.orpha.net](http://www.orpha.net)

## **Typical surgery**

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Patients affected by systemic sclerosis may require surgery for any type of procedure, but will typically present for repeated esophageal procedures, dental treatment and surgical management of vascular insufficiency including cervical, lumbar and digital sympathectomy in addition to amputation. In severe forms, lung transplantation may be considered due to severe interstitial disease or pulmonary arterial hypertension.

## **Type of anaesthesia**

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There is no definite recommendation for either general anesthesia or regional anesthesia and choice of anesthetic technique will depend on the type of surgery, an understanding of the pathophysiology of the disease and careful preoperative assessment of the patient.

General anesthesia may be complicated by difficult intubation, high incidence of aspiration, due to gastro-esophageal reflux disease (GERD) and significant respiratory disease.

Regional anesthesia is a safe alternative to general anesthesia and a useful adjunct in the treatment of post operative pain and prevention of vasospastic crisis. Technical challenges exist in performing regional anesthesia due to difficulties in positioning the patient and altered anatomy. There is the potential for prolonged sensory blockade with peripheral nerve blocks.

## **Necessary additional diagnostic procedures (preoperative)**

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There is a five to eight fold increase in mortality associated with SSc, particularly in patients with pulmonary hypertension and cardiac involvement.

To assess the extent of pulmonary disease, such as pulmonary fibrosis, patients should have a chest radiograph and pulmonary function tests to demonstrate any reductions in compliance, vital capacity and diffusion capacity. A risk factor for increased mortality is a forced vital capacity less than 50% predicted. Pulse oximetry in air and arterial blood gases can be carried out to assess degree of hypoxaemia and consider cardiopulmonary exercise testing if available to assess functional ability.

Cardiac disease may present as pericarditis, pulmonary hypertension, congestive heart failure, cardiomegaly, systolic dysfunction, myocardial fibrosis, dilated or restrictive cardiomyopathy, conduction defects and arrhythmias. At-risk patients should have a base line ECG (although only 19% of patients will exhibit abnormal rhythm on continuous 24 ECG monitoring) and an echocardiogram.

Gastrointestinal disease may result in malnutrition, impaired absorption of vitamin K and electrolyte disturbance. All patients therefore require a full blood count, urea and electrolytes, liver function tests, bone screen and coagulation screen in addition to a group and hold or cross match depending on procedure.

### **Particular preparation for airway management**

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Dermal fibrosis will lead to up to 70% of patients having a pinched face, atrophied nasal alae and restricted mouth opening, compounded by temporomandibular joint fibrosis. Limited neck extension may occur along with blunting of the angle of the mandible. Difficulties with intubation and mask ventilation should therefore be expected and access to difficult intubation equipment including jet ventilation should be made immediately available.

Fiber-optic, blind oral or retrograde intubation techniques may be considered. Patients are prone to mucosal telangiectasiae, which may bleed profusely and therefore it is important to exercise careful airway manipulation techniques.

Patients are at risk of aspiration due to GERD, which can be severe. Rapid sequence induction should be undertaken with caution due to the risk of failed/difficult intubation. Sellick's maneuver may also be ineffective due to fibrosis or the oesophagus and impair view at laryngoscopy further.

In particularly difficult situations it may be necessary to consider awake tracheotomy with local anesthesia.

### **Particular preparation for transfusion or administration of blood products**

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There is no definite recommendation for transfusion; administration of blood products will depend on type of surgery, patient symptoms and advice from senior hematology clinicians.

### **Particular preparation for anticoagulation**

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In rare cases, scleroderma patients have antiphospholipid antibodies and are at higher risk of vascular thrombosis, however there is no definite recommendation for anticoagulation; administration will depend on type of surgery, patient symptoms and senior clinical advice.

### **Particular precautions for positioning, transport or mobilisation**

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Due to flexion contractures, positioning should ideally be guided by the awake and co-operative patient. Due to vascular insufficiency, pressure areas should be carefully padded and checked regularly. A vacuum mattress should be considered for patient transportation.

During the surgical procedure, Trendelenburg position may favour pulmonary aspiration and should therefore be avoided, unless the airway is secure.

The patient's temperature should be maintained at all times to prevent vascular crisis and digital ischemia. Sweating is hindered and therefore care should also be taken not to overheat the patient, which may present as malignant hypertension.

Patients are prone to developing dry eyes which may be compounded by scarring of the eyelids preventing complete closure. Eyes should be carefully lubricated and padded to avoid corneal abrasions.

## **Probable interaction between anaesthetic agents and patient's long-term medication**

Immunosuppressants are the mainstay of treatment increasing the risk of post operative infective complications. Additional steroid cover should be provided in patients on glucocorticoids.

Angiotensin–converting enzyme inhibitors are the first line antihypertensive agents in patients with Systemic Sclerosis and may produce refractory hypertension post induction of anaesthesia.

## **Anaesthesiologic procedure**

In cases where general anaesthesia is required, endotracheal intubation is advocated to decrease the risk of aspiration.

Ventilation may be challenging due to reduced lung compliance and protective lung strategies should be employed to prevent barotrauma.

Induction agents, volatiles, depolarising and non depolarising muscle relaxants, and reversal agents are all safe in Systemic Sclerosis.

Use of vasopressive amines can worsen Raynaud's phenomenon and is associated with an increased risk of digital ischemia, treatment with iloprost can be proposed in accordance with the patient's hemodynamic status and before necrotic lesions occur.

There is a strong association of renal and gastrointestinal disease and therefore, non steroidal anti-inflammatories should be avoided. Patients can also be sensitive to opioids. Where possible opiate sparing techniques should be used including the use of regional anaesthesia, which may also be considered as a safe alternative to general anaesthesia in high risk cases.

Regional anaesthesia may be challenging due to patient positioning, altered fascial planes and prolonged sensory blockade. The use of ultrasound to identify structures and spread of local anaesthesia is therefore recommended. The spine is frequently spared in systemic sclerosis and many of the challenges and complications associated with regional anaesthesia are therefore reduced by neuroaxial approaches. Marked hypotension can occur secondary to anaesthesia induced vasodilatation and may be refractory to inotropes. Excessive fluid administration may result in pulmonary oedema once vascular tone is restored. Techniques that enable gradual or incremental adjustment of block height, such as epidural or combined epidural spinal anaesthesia, are therefore preferable.

## **Particular or additional monitoring**

Routine monitoring (as per Association of Anaesthetists Great Britain and Ireland guidelines) is advocated in all SSc patients. Dermal thickening, flexion contractures and vasoconstriction may make it difficult to obtain intravenous access and non-invasive blood pressure readings. This may necessitate the use of invasive monitoring and central venous access.

Radial arterial cannulation can precipitate Raynaud's phenomenon and even subsequent necrosis. Moreover, some patients have a macroangiopathy with radial artery thrombosis. It is important to alternate pulse oximeter probes between digits during surgery, as failure to do

so can result in precipitation of ischaemic damage. Patients with severe cardiac disease and pulmonary hypertension may benefit from cardiac output monitoring although the presence of esophageal fibrosis, aortic disease and altered vascular performance may effect the accuracy of newer cardiac output monitors.

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### **Possible complications**

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Patients with systemic sclerosis are at increased risk of failed or difficult intubation and aspiration.

Patients with pulmonary disease will have reduced oxygen reserve and impaired pulmonary compliance. They may therefore desaturate suddenly, particularly during airway maneuvers and may be difficult to ventilate with risk of barotrauma. They can be sensitive to opiates and are of high risk of postoperative respiratory failure especially in the presence of severe disease (Vital Capacity of less than 1 Liter)

Patients may have severe cardiac disease resulting in systolic dysfunction, conduction defects and arrhythmias. They have a relatively reduced intravascular compartment and may become profoundly hypotensive as a result of anesthesia induced vasodilatation and tolerate dehydration and blood loss poorly. Rebound pulmonary oedema may occur on restoration of vascular tone.

Patients are at increased risk of cerebrovascular events. Uremia and malignant hypertension may also cause seizures.

Stress, pain, dehydration, hypothermia and vasoconstrictors therapy may induce vasospastic crisis leading to peripheral ischaemia and ulceration.

Sweating is hindered and patients are at risk of hyperthermia.

Wound healing can be impaired due to poor peripheral perfusion and patients may be prone to pressure sores.

Malnutrition and immunosuppressant therapy may increase patients' susceptibility to infection.

There is potential for prolonged sensory blockade with peripheral nerve blocks, although there is no evidence that patients are at increased risk of permanent nerve injury following regional anaesthesia, and full sensory function is usually returned within 24 hours.

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### **Postoperative care**

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Postoperative care will depend on type of surgery and disease severity. Postoperative ventilation may be required given the high risk of postoperative respiratory failure, and admission to High Dependency or the Intensive Care Unit admission may be indicated.

Continuous ECG monitoring or invasive monitoring is indicated in the postoperative period in those with cardiac manifestations of the disease. It is important to maintain euvoemia throughout the peri and post operative period to avoid renal crises or pulmonary oedema.

Patients are at significant risk of developing scleroderma renal crisis (SRC) in the post operative period. This may present with acute onset and or progressive worsening of arterial hypertension (>150/85 mmhg confirmed during at least 2 different measurements) and oligo/anuria. They may go on to develop thrombotic microangiopathy, thrombocytopenia, and haemolytic anaemia. Patients with diffuse cutaneous scleroderma lasting less than 5 years are of particular risk and if suspected the diagnosis may be confirmed by measuring serum haptoglobin and schizocyte levels following discussion with haematology.

Mobilization can prove difficult due to contractures, malnutrition and prolonged sensory blockade from regional anaesthesia and additional assistance may be required.

Thromboembolic stockings should be avoided due to peripheral vascular disease and risk of ischemia.

Postoperative analgesia should avoid non-steroidal anti-inflammatory drugs and opiates should be used with caution.

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### **Information about emergency-like situations / Differential diagnostics**

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*caused by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the disease*

Vasospastic crisis may be triggered by stress, pain, dehydration, exposure to cold and use of inotropic drugs. It can present with severe pain, hypertension, tachycardia and peripheral ischemia. Calcium channel blockers are usually the first line treatment although, iloprost and sildenafil may also be effective. Regional anesthesia can provide good pain relief in addition to vasodilatation. A sympathectomy of the affected area may be required.

All anaesthetists anaesthetizing patients with SSc should be familiar with difficult and failed intubation protocols.

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### **Ambulatory anaesthesia**

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Availability of ambulatory anesthesia will be guided by severity of disease, surgical procedure and local guidelines. It is unlikely to be appropriate in any but the mildly affected.

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### **Obstetrical anaesthesia**

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Systemic sclerosis does not usually affect fertility but there is a high incidence of miscarriage, still birth and premature labour. The disease is accelerated in 50% of cases and women with widespread multi-organ involvement may be counseled against continuation of pregnancy.

Renal scleroderma may present as pre-eclampsia, is differentiated by raised plasma renin, and treated with angiotensin-converting enzyme inhibitors.

Pregnant women with systemic sclerosis should have experience obstetric lead care and multidisciplinary team involvement. Early epidural anesthesia is recommended for labour as there is a high risk of obstructive labour and need for expedient operative delivery.

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