### Scleromyxedema

**Version:** 3.0  
**Published:** 20 October 2018

**Condition for which IVIg use is in exceptional circumstances only**

<table>
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<tr>
<th>Specific Conditions</th>
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<tr>
<td>- Scleromyxedema – skin involvement only</td>
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<td>- Scleromyxedema – skin and systemic disease</td>
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<table>
<thead>
<tr>
<th>Indication for IVIg Use</th>
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<tbody>
<tr>
<td>- Scleromyxedema (skin involvement only) unresponsive to steroids and other immunosuppressant agents or where contraindicated</td>
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<tr>
<td>- Scleromyxedema - systemic involvement as first line therapy</td>
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<tr>
<th>Level of Evidence</th>
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<tr>
<td>Insufficient data (Category 4a)</td>
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<tr>
<th>Description and Diagnostic Criteria</th>
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<td>Scleromyxedema is a chronic, idiopathic disorder characterised by excessive deposition of mucin in the skin in association with increased dermal collagen and absence of thyroid disease, and usually associated with a monoclonal gammopathy. Extracutaneous systemic manifestations also occur with neurologic, rheumatologic, pulmonary, renal, muscular and/or cardiac involvement.</td>
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This is a rare condition with proven difficulty in establishing specific or definitive treatment. Successful treatment of skin disease has been reported with melphalan, dexamethasone, thalidomide, lenalidomide and IVIg. In the three largest case series, the central nervous system (CNS) was predominantly involved in 10–15 percent of cases associated with poor clinical outcome, often with global CNS dysfunction with encephalopathy, seizures and coma. The pathophysiology of this fatal complication of scleromyxedema remains unknown. Evidence supporting effective treatment may suggest that targeting the paraprotein may be of benefit. Most of the reports of successful treatment of CNS manifestations are with IVIg.

<table>
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<th>Justification for Evidence Category</th>
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<td>Prognostic outcomes of therapy in this condition are poorly understood as the literature is limited to case reports or small series. A 2008 case series demonstrated full or partial remission in eight out of ten patients treated with IVIg, although maintenance therapy was required to maintain ongoing control of disease. Three of six patients treated with IVIg achieved full remission in a mainly retrospective multi-centre study of 30 patients in 2013, with three of the remaining patients achieving partial responses. In the European case series, IVIg was used in 13 out of 25 patients, all showing either complete or partial response. IVIg responders have included where patients were treated with Ig as first line therapy or after failure to respond to steroids and other immunosuppressant agents.</td>
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<th>Diagnosis Requirements</th>
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<td>A diagnosis must be made by an Immunologist or a Dermatologist.</td>
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Qualifying Criteria for IVIg Therapy

Scleromyxedema (skin involvement only) unresponsive to steroids and other immunosuppressant agents or where contraindicated

- Moderate to severe scleromyxedema proven by skin biopsy and confirmed absence of thyroid disease

AND

- Unresponsive to standard corticosteroid therapy and at least one other immunosuppressant
  OR
  - Immunosuppressant medication resulted in unacceptable side effects or significant toxicity
  OR
  - Corticosteroid and/or immunosuppressant medication are contraindicated

Review is required every six months by a dermatologist or immunologist. Response must be demonstrated at the initial review at six months and improvement must be demonstrated for continuation of supply.

Consideration should be given to a trial-off Ig therapy once the patient has achieved stabilised disease or clinical remission. The minimal effective dose should be prescribed.

Documentation of clinical effectiveness is necessary for continuation of IVIg therapy.

Scleromyxedema - systemic involvement as first line therapy

- Diagnosis of scleromyxedema proven by skin biopsy and confirmed absence of thyroid disease
  AND
  - Systemic manifestations of disease are present

Review is required every six months by a dermatologist or immunologist. Response must be demonstrated at the initial review at six months and improvement must be demonstrated for continuation of supply.

Consideration should be given to a trial-off Ig therapy once the patient has achieved stabilised disease or clinical remission. The minimal effective dose should be prescribed.

Documentation of clinical effectiveness is necessary for continuation of IVIg therapy.
Scleromyxedema (skin involvement only) unresponsive to steroids and other immunosuppressant agents or where contraindicated

Review is required every six months by a dermatologist or immunologist. Response must be demonstrated at the initial review at six months and improvement must be demonstrated for continuation of supply.

Consideration should be given to a trial-off immunoglobulin (Ig) therapy once the patient has achieved stabilised disease or clinical remission. The minimal effective dose should be prescribed.

Documentation of clinical effectiveness is necessary for continuation of IVIg therapy.

**Clinical effectiveness of Ig therapy can be assessed by:**

**On review of the initial authorisation period**
- A reduction in the number of lesions and severity of disease compared to the last assessment

**On review of a continuing authorisation period**
- Improvement in or stabilisation of disease compared to the previous assessment
  AND
- A trial of weaning and/or a trial-off Ig therapy is planned or if not planned, a reason is provided

Scleromyxedema - systemic involvement as first line therapy

Review is required every six months by a dermatologist or immunologist. Response must be demonstrated at the initial review at six months and improvement must be demonstrated for continuation of supply.

Consideration should be given to a trial-off Ig therapy once the patient has achieved stabilised disease or clinical remission. The minimal effective dose should be prescribed.

Documentation of clinical effectiveness is necessary for continuation of IVIg therapy.

**Clinical effectiveness of Ig therapy, can be assessed by:**

**On review of the initial authorisation period**
- A reduction in the number of lesions and severity of disease including systemic symptoms

**On review of a continuing authorisation period**
- Improvement in or stabilisation of disease compared to the previous review assessment
  AND
- A trial of weaning and/or a trial-off Ig therapy is planned or if not planned, a reason is provided
Dose

Scleromyxedema (skin involvement only) unresponsive to steroids and other immunosuppressant agents or where contraindicated

- **Induction dose** - 2 g/kg over 5 days
- **Maintenance dose** - 0.5 to 2 g/kg over 2 to 5 days, four to six weekly, or longer.
  
  A maximum dose of 2 g/Kg may be given in any 4 week period.

The aim should be to use the lowest dose possible that achieves the appropriate clinical outcome for each patient.

Refer to the current product information sheet for further information on dose, administration and contraindications.

Scleromyxedema - systemic involvement as first line therapy

- **Induction dose** - 2 g/kg over 5 days
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Bibliography


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