# Susac syndrome

## Condition for which Ig use is in exceptional circumstances only

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| **Specific Conditions** | • Susac syndrome |
| **Indication for Ig Use** | • Probable or definite Susac syndrome in concurrence with high dose corticosteroids |
| **Level of Evidence** | Insufficient data (Category 4a) |
| **Description and Diagnostic Criteria** | Susac syndrome is a rare, microangiopathic disorder characterised by encephalopathy, hearing loss and retinal artery branch occlusions. Case reports show benefit of intravenous immunoglobulin (IVIg) or plasma exchange in combination with corticosteroids, generally with, or in mild cases without, other immunosuppressive agents. |
| **Justification for Evidence Category** | There are no randomised controlled trials (RCTs) nor prospective series in Susac syndrome. However, there is a very poor natural history and a clear response to multi-agent immunosuppressive therapy including intravenous immunoglobulin (IVIg) in case series (Mateen, 2012). A recent series has suggested equal or possibly greater efficacy from plasma exchange over IVIg, and plasma exchange should be considered where available (Vodopivec, 2016). |
| **Diagnosis Requirements** | A diagnosis must be made by an Immunologist, Neurologist, Rheumatologist or an Ophthalmologist. |
| **Qualifying Criteria for Ig Therapy** | • Probable or definite diagnosis of Susac syndrome has been made by the presence of at least two of the following:<br>  ◦ Encephalopathy with typical MRI brain changes including corpus callosum lesions or characteristic diffusion weighted imaging (DWI) hyperintense lesions<br>  ◦ New sensorineural hearing loss or tinnitus<br>  ◦ Branch retinal artery occlusions (BRAOs) or ischaemia or arterial wall hyperfluorescence (AWH) on fluorescein angiography<br>AND<br>• A baseline assessment of disability is conducted as measured by the Modified Rankin Scale (MRS)<br>AND<br>• Corticosteroid therapy is being given concurrently<br>OR<br>• Steroid therapy is absolutely contraindicated<br>IVIg should be used for four months (induction plus three maintenance cycles) before determining whether the patient has responded. If there is no benefit after this treatment, IVIg therapy should be abandoned.<br>Review by a neurologist, immunologist rheumatologist or ophthalmologist is required within four months of treatment to determine whether the patient has responded, and annually thereafter.<br>For stable patients on maintenance treatment, review by a neurologist, immunologist, ophthalmologist or rheumatologist is required at least annually. A trial off IVIg should be attempted after a year of therapy, unless there is a contraindication to doing so, or the patient has previously relapsed after an earlier trial of withdrawal of IVIg.<br>Documentation of clinical effectiveness is necessary for continuation of IVIg therapy. |

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| **Review Criteria for Assessing the Effectiveness of Ig Use** | IVIg should be used for four months (induction plus three maintenance cycles) before determining whether the patient has responded. If there is no benefit after this treatment, IVIg therapy should be abandoned.<br><br>Review by an neurologist, immunologist rheumatologist or ophthalmologist is required within four months of treatment to determine whether the patient has responded, and annually thereafter.<br><br>For stable patients on maintenance treatment, review by a neurologist, immunologist, ophthalmologist or rheumatologist is required at least annually. A trial off IVIg should be attempted after a year of therapy, unless there is a contraindication to doing so, or the patient has previously relapsed after an earlier trial of withdrawal of IVIg.<br><br>Documentation of clinical effectiveness is necessary for continuation of IVIg therapy.<br><br>**Clinical effectiveness of Ig therapy can be demonstrated by:**<br><br>**On review of the initial authorisation period**<br><br>• Stabilisation of, or improvement in the severity of symptoms<br><br>AND<br><br>• Stabilisation of, or improvement in disability as measured by the Modified Rankin Scale (MRS) score compared to the qualifying assessment<br><br>**On review of a continuing authorisation period**<br><br>• Improvement in or stabilisation of symptoms compared to the previous review assessment<br><br>AND<br><br>• Stabilisation or improvement in disability as measured by the Modified Rankin Scale (MRS) compared to the previous review assessment<br><br>AND<br><br>• A trial of weaning towards cessation of Ig therapy is planned once the patient is stable or a reason is provided as to why a trial is not planned<br><br>A trial off IVIg should be attempted after a year of therapy, unless there is a contraindication to doing so, or the patient has previously relapsed after an earlier trial of withdrawal of IVIg. |
| **Dose** | • **Induction Dose (IVIg)** - Up to 2 g/kg over 2 to 5 days.<br>• **Maintenance Dose (IVIg)** - 0.5 - 1 g/kg every 2 to 6 weeks.<br><br>The aim should be to use the lowest dose possible that achieves the appropriate clinical outcome for each patient.<br><br>Refer to the current product information sheet for further information on dose, administration and contraindications. |

**Bibliography**


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