Rasmussen encephalitis

Condition for which Ig use is in exceptional circumstances only

Specific Conditions	• Rasmussen encephalitis
Indication for Ig Use	• Rasmussen encephalitis with concurrent steroid therapy unless contraindicated
Level of Evidence	Evidence of probable benefit – more research needed (Category 2a)
Description and Diagnostic Criteria	Rasmussen encephalitis is a chronic, progressive, focal encephalitis that is commonly accompanied by focal seizures, hemiparesis and cognitive decline. It is generally considered to be a disease of childhood, with most cases occurring in children younger than 10 years, although adult-onset cases do occur. Conventional anticonvulsant therapy is usually ineffective and hemispherectomy may be helpful in the correct setting. Generally, there is an active progressive phase of the disease, often lasting some years, followed by quiescence and no further progression of disease (Varadkar et al, 2014). Although historical data has generally reported seizure freedom only after functional hemispherectomy, some centres practise more aggressive immune therapies and report improved outcomes. Functional hemispherectomy provides the best opportunity of seizure freedom, however there will inevitably be post-surgical motor deficits, and often cognitive impairments post-hemispherectomy. Therefore, it can be argued that aggressive early immune therapy may reduce the surgical sequelae of motor and cognitive deficits (the decision regarding when and if to perform hemispherectomy is challenging and discussed in Figure 4 of Varadkar et al, 2014).
Justification for Evidence Category	A number of retrospective case series, open label studies and one randomized controlled study (Bien et al, 2012) were performed between 1996 and 2013. Hart et al (1996), Granata et al (2003) and Takahashi et al (2013) have reported benefit from immunomodulatory treatments including pulse steroids, intravenous immunoglobulin (IVIg), plasma exchange (PLEX) and tacrolimus. Benefit was seen in regard to reduced tissue and function loss and reduced chance of intractable epilepsy. A single case report described a good outcome with natalizumab (Bittner et al, 2013). Early therapies may yield the best outcomes. Varadkar et al (2014) reported that immunomodulatory treatments seem to slow rather than halt disease progression without changing the eventual outcome. Patients are often left with intractable epilepsy for which functional hemispherectomy remains the only effective cure.
Diagnosis Requirements	A diagnosis must be made by a Neurologist.

Qualifying Criteria for Ig Therapy	• Clinical features, EEG and MRI findings consistent with a diagnosis of Rasmussen encephalitis as per the European Consensus Statement (Bien, 2005) OR • Two of typical clinical features, MRI changes or histopathology findings consistent with a diagnosis of Rasmussen encephalitis as per the European Consensus Statement (Bien, 2005) AND • Significant level of disability as measured by an adapted Modified Rankin Scale (MRS) score of at least 3 points AND • Concurrent corticosteroid therapy OR • Corticosteroid therapy is contraindicated Review by a neurologist is required within 6 months of treatment and annually thereafter. It is recognised that the acute phase of Rasmussen encephalitis can last for at least 12 months followed by stabilisation of symptoms and residual disease. The aim of Ig therapy is to reduce the trajectory of deterioration in a progressive disease (Varadkar et al, 2014). Some patients will have aggressive and refractory disease, and in these cases hemispherectomy may be the preferred option (discussed in figure 4, Varadkar et al, 2014). For patients on maintenance treatment, review by a neurologist is required at least annually. It is recognised that patients will typically reach a stabilisation phase 18 months to 2 years from onset, at which time, a trial of weaning should be considered. Documentation of clinical effectiveness is necessary for continuation of IVIg therapy.
Review Criteria for Assessing the Effectiveness of Ig Use	Review by a neurologist is required within 6 months of treatment and annually thereafter. It is recognised that the acute phase of Rasmussen encephalitis can last for at least 12 months followed by stabilisation of symptoms and residual disease. The aim of Ig therapy is to reduce the trajectory of deterioration in a progressive disease (Varadkar et al, 2014). Some patients will have aggressive and refractory disease, and in these cases hemispherectomy may be the preferred option (discussed in figure 4, Varadkar et al, 2014). For patients on maintenance treatment, review by a neurologist is required at least annually. It is recognised that patients will typically reach a stabilisation phase 18 months to 2 years from onset, at which time, a trial of weaning should be considered. Documentation of clinical effectiveness is necessary for continuation of IVIg therapy. **Clinical effectiveness can be demonstrated by:** **On review of the initial authorisation period** • Monitoring of the rate of deterioration of symptoms and disability compared to the qualifying assessment, including as measured by the adapted Modified Rankin Scale (MRS) score **On review of a continuing authorisation period** • Monitoring of the rate of deterioration of symptoms and disability compared to the qualifying assessment , including as measured by the adapted Modified Rankin Scale (MRS) score AND • A trial of Ig weaning is planned for patients who have entered the residual stage of disease (permanent and stable neurologic deficits and continuing seizures) or a valid reason provided as to why a trial is not being planned or is contraindicated at this time

Dose	
	• **Induction Dose (IVIg)** - 2 g/kg in 2 to 5 divided doses. • **Maintenance Dose (IVIg)** - 1 g/kg monthly as a single dose. 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**.**

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