Opsoclonus-myoclonus ataxia (OMA)

Version: 3.0
Published: 20 October 2018

Condition for which IVIg has an emerging therapeutic role.

Specific Conditions	• Idiopathic opsoclonus-myoclonus ataxia • Paraneoplastic associated neuroblastoma • Paraneoplastic associated small cell lung cancer • Paraneoplastic associated breast cancer • Paraneoplastic associated other tumour type
Indication for IVIg Use	• Treatment of OMA initially diagnosed in a child • Second-line treatment of OMA in adults following the use of corticosteroids
Level of Evidence	Insufficient data (Category 4a)
Description and Diagnostic Criteria	Opsoclonus-myoclonus ataxia (OMA) is an immune-mediated monophasic or multiphasic disorder consisting of opsoclonus (conjugate chaotic eye movements), cerebellar ataxia, and arrhythmic myoclonus affecting the trunk, the head and the extremities. OMA may be either paraneoplastic or idiopathic, presumably para-infectious (e.g. post-viral). In children, OMA complicates about two to three percent of neuroblastomas. In adults, it may occur in association with several cancers, most commonly small-cell lung cancer and breast cancer.
Justification for Evidence Category	The Asia–Pacific IVIg Advisory Board (2004) consensus statement summarises several case reports suggesting that IVIg is useful in idiopathic OMA and childhood paraneoplastic opsoclonus-myoclonus ataxia (OMA) associated with neuroblastoma.
Diagnosis Requirements	A diagnosis must be made by a Neurologist.
Qualifying Criteria for IVIg Therapy	Treatment of OMA initially diagnosed in a child • Diagnosis of OMA in a patient younger than 18 years of age AND • Significant disability as measured by the Cerebellar Functional System Score with a value of at least two points IVIg should be used for six months before determining whether the patient has responded. If there has been no benefit after six months treatment, IVIg therapy should be abandoned. Review by a neurologist is required after the first six months of treatment. For stable patients on maintenance treatment, review by a neurologist is required at least annually. Documentation of clinical effectiveness is necessary for continuation of IVIg therapy.

Second-line treatment of OMA in adults following the use of corticosteroids

- Adult with OMA unresponsive to a standard course of corticosteriod therapy

  OR

- Adult with OMA and corticosteroid therapy has resulted in unacceptable side effects or significant toxicity

  OR

- Adult with OMA and corticosteroid therapy is contraindicated

AND

- Significant disability as measured by the Cerebellar Functional System Score with a value of at least two points

IVIg should be used for six months before determining whether the patient has responded. If there has been no benefit after six months of treatment, IVIg therapy should be abandoned.

Review by neurologist is required after the first six months of treatment. For stable patients on maintenance treatment, review by a neurologist is required at least annually.

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

Review Criteria for Assessing the Effectiveness of IVIg Use	Treatment of OMA initially diagnosed in a child
	IVIg should be used for six months before determining whether the patient has responded. If there has been no benefit after six months treatment, IVIg therapy should be abandoned. Review by a Neurologist is required after the first six months of treatment. For stable patients on maintenance treatment, review by a Neurologist is required at least annually. Documentation of clinical effectiveness is necessary for continuation of IVIg therapy. **Clinical effectiveness of Ig may be assessed by:** **On review of the initial authorisation period** • Clinical improvement in, or stabilisation of, opsoclonus symptoms after six months treatment compared to the qualifying assessment AND • Improvement or stabilisation in the degree of **disability** compared to the qualifying assessment as measured by the Cerebellar Functional System Score **On review of a continuing authorisation period** • Clinical improvement in or stability of opsoclonus symptoms compared to the previous review assessment AND • Improvement, or no further deterioration in **disability** compared to the previous review assessment as measured by the Cerebellar Functional System Score

Second-line treatment of OMA in adults following the use of corticosteroids

IVIg should be used for six months before determining whether the patient has responded. If there has been no benefit after six months of treatment, IVIg therapy should be abandoned.

Review by neurologist is required after the first six months of treatment. For stable patients on maintenance treatment, review by a neurologist is required at least annually.

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

Clinical effectiveness of Ig therapy may be demonstrated by:

On review of the initial authorisation period

- Clinical improvement in. or stabilisation of, opsoclonus symptoms compared to the qualifying assessment

  AND

- Improvement or stabilisation in disability compared to the qualifying assessment as measured by the Cerebellar Functional System Score

On review of a continuing authorisation period

- Improvement in or stabilisation of, opsoclonus symptoms compared to the previous review assessment

  AND

- Improvement or no further deterioration in the degree of disability compared to the previous review assessment as measured by the Cerebellar Functional System Score

Dose

Treatment of OMA initially diagnosed in a child

- **Induction Dose** - 1–2 g/kg in 2 to 5 divided doses.
- **Maintenance Dose** - 0.4–1 g/kg, 4 to 6 weekly.

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, administartion and contraindications.

Second-line treatment of OMA in adults following the use of corticosteroids

- **Induction Dose** - 1–2 g/kg in 2 to 5 divided doses.
- **Maintenance Dose** - 0.4–1 g/kg, 4 to 6 weekly.

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.

Bibliography

Bibliography

Glatz, K, Meinck, HM & Wildemann, B 2003, 'Parainfectious opsoclonus-myoclonus syndrome: high dose intravenous immunoglobulins are effective', *Journal of Neurology, Neurosurgery and Psychiatry*, vol. 74, no. 2, pp. 279–80. https://jnnp.bmj.com/content/74/2/279

Kornberg, AJ, for the Asia–Pacific IVIg Advisory Board 2004, '*Bringing consensus to the use of IVIg in neurology. Expert consensus statements on the use of IVIg in neurology*', 1st edn, Asia–Pacific IVIg Advisory Board, Melbourne, pp. 80–82. http://www.indaps.org/files/1/Consensus%20Statements_0511.pdf%20for%20website.pdf

Kurtzke, JF 1983, 'Rating neurologic impairment in multiple sclerosis: An expanded disability status scale (EDSS)', *Neurology*, vol. 33, no. 11, pp. 1444–1452. http://n.neurology.org/content/neurology/33/11/1444.full.pdf

National Institute of Neurological Disorders and Stroke 2006, ' Opsoclonus Myoclonus Information Page'. Available from: https://www.ninds.nih.gov/Disorders/All-Disorders/Opsoclonus-Myoclonus-Information-Page

Generated on: 2 April 2019