Opsoclonus-myoclonus ataxia (OMA)

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Condition for which IVIg has an emerging therapeutic role.

Specific Conditions	• Opsoclonus myoclonus ataxia
Indication for IVIg Use	• Child with OMA. • Long-term maintenance therapy of OMA in association with other tumour therapies.
Level of Evidence	Insufficient data (Category 4a)
Description and Diagnostic Criteria	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 2–3% 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 (Kornberg 2004) consensus statement summarises several case reports suggesting that intravenous immunoglobulin (IVIg) is useful in idiopathic OMA and childhood paraneoplastic OMA associated with neuroblastoma.
Diagnosis Requirements	A diagnosis must be made by a Neurologist.
Qualifying Criteria for IVIg Therapy	Child with OMA. Child less than 18 years • Significant disability as measured by activities of daily living (ADL) or cerebellar functional scores. Long-term maintenance therapy of OMA in association with other tumour therapies. • Significant disability, as measured by activities of daily living (ADL) or cerebellar functional scores. AND • No response to a standard course of adrenocorticotrophic hormone (ACTH) therapy. OR • No response to a standard course of corticosteroid therapy. OR • Corticosteroid therapy is contraindicated.

Review Criteria for Assessing the Effectiveness of IVIg Use

Child with OMA.

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.

On review of the initial authorisation period

Clinical effectiveness of Ig therapy may be demonstrated by:

- Improvement in disability or no further deterioration compared to the qualifying assessment as measured by any formal assessment method

On review of a continuing authorisation period

Clinical effectiveness of Ig therapy may be demonstrated by:

- Improvement in disability or no further deterioration compared to the previous assessment as measured by any formal assessment method

Long-term maintenance therapy of OMA in association with other tumour therapies.

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Dose	
	Child with OMA. • **Induction Dose** - 1–2 g/kg in 2 to 5 divided doses. • **Maintenance Dose** - 0.4–1 g/kg, 4 to 6 weekly. Aim for the minimum dose to maintain optimal functional status. **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.**
	Long-term maintenance therapy of OMA in association with other tumour therapies. • **Induction Dose** - 1–2 g/kg in 2 to 5 divided doses. • **Maintenance Dose** - 0.4–1 g/kg, 4 to 6 weekly. Aim for the minimum dose to maintain optimal functional status. **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.**

Bibliography

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

Kornberg, AJ 2004, *Bringing consensus to the use of IVIg in neurology. Expert consensus statements on the use of IVIg in neurology*, 1st edn, for the Asia–Pacific IVIg Advisory Board, Melbourne, pp. 80–82.

National Institute of Neurological Disorders 2006, 'NINDS opsoclonus myoclonus information page', January. Available from: http://www.ninds.nih.gov/disorders/opsoclonus_myoclonus/opsoclonus_myoclonus.htm [cited 7 Dec 2007]

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