

2017 (v3.0) Proposed changes to v2.1 of the Criteria for the clinical use of intravenous immunoglobulin in Australia

CONDITION NAME IN v2.1	Potassium channel antibody-associated encephalopathy
PROPOSED APPROACH: To remove Potassium channel antibody-associated encephalopathy as a stand-alone condition in <i>Exceptional circumstances only</i> and allow access for eligible patients under Autoimmune encephalitis mediated by antibodies targeting cell surface antigens (AMAE) in <i>Emerging therapeutic role</i>	SUMMARY OF RATIONALE: Potassium channel antibody-associated encephalopathy has previously been endorsed by NIGAC and JBC with the revised name of Autoimmune encephalitis mediated by antibodies targeting cell-surface antigens (AMAE) by NIGAC and JBC as a condition for which Ig has an emerging therapeutic role. While the information relating to potassium channel antibody associated encephalopathy is unchanged from that previously approved by JBC in 2015, it is being re-submitted for completeness as two further indications are now also proposed to be included under AMAE. Re-submission of this condition also allows presentation of data specifically relevant to Potassium channel antibody-associated encephalopathy.
Role of Ig therapy, if appropriate: See AMAE –ATTACHMENT B for full details	
Access Information in v2.1	
Condition Category	Condition for which Ig use is in exceptional circumstances only (Chapter 7)
Level of Evidence	Insufficient data (Category 4a)
Description	Potassium channel antibody-associated neurologic syndromes include limbic encephalitis/subacute amnesic encephalopathy, Morvan syndrome, peripheral nerve hyperexcitability and autonomic ganglionopathy.
Qualifying Criteria	Potassium channel antibody-associated encephalopathy is considered to be an autoimmune, nonparaneoplastic, potentially treatable syndrome, but may respond to a variety of immunomodulatory agents, including intravenous immunoglobulin (IVIg).

References

Hudson LA, et al (2008) Reduplicative paramnesia in Morvan's syndrome. *Journal of the Neurological Sciences*, 267(1–2):154–7.

[http://www.jns-journal.com/article/S0022-510X\(07\)00656-9/abstract](http://www.jns-journal.com/article/S0022-510X(07)00656-9/abstract)

Vincent A, Buckley C, Schott JM, et al (2004) Potassium channel antibody-associated encephalopathy: a potentially immunotherapy-responsive form of limbic encephalitis. *Brain*, 127(3):701–12.

<https://www.ncbi.nlm.nih.gov/pubmed/14960497>

POTENTIAL OPERATIONAL IMPACT

The development of specific qualifying and review criteria will represent a change for this condition with the revised criteria providing significantly greater guidance for appropriate access to Ig therapy and patient management expectations. A neurologist will be required to diagnose and manage patient treatment, although it is recognised that this may be as part of a broader clinical team. There will be increased data entry required (e.g. compliance with the international consensus diagnostic criteria) compared to the previous Ig request process. Patients will be required to be assessed at qualifying and review using a Modified Rankin Scale, however, the scale will be presented as ‘drop-down’ menu options within BloodSTAR v3.0 and will be simple to apply. It is likely that as a result of greater awareness of the condition, diagnostic criteria will develop and requests for antibody testing may increase. There is limited testing currently available within Australia, however, availability is likely to improve over time.

POTENTIAL IMPACT ON PATIENTS, DEMAND AND EXPENDITURE

Potential impact on patients:

Under the changes communicated in 2015, patients with Potassium channel antibody-associated encephalopathy will be eligible to receive Ig therapy under the criteria for the condition ‘Antibody mediated autoimmune encephalitis’ (AMAE) as the condition has been renamed and broadened to include other disease groups. The formal access criteria proposed for this condition require that a neurologist, who may be part of a wider clinical team including clinical immunologists, makes the diagnosis and manages the treatment. This is because the condition is uncommon, can be misdiagnosed and it is important that the correct, early treatment is given to patients. Ig therapy is given at the same time as other treatments. These criteria provide greater guidance for prescribers by applying specific diagnostic criteria and ensuring that second line treatment is started very early when patients do not respond to first line treatment because this approach has been shown to result in improved patient outcomes.

For existing patients on Ig maintenance therapy, six monthly reviews are required to assess the effectiveness of the treatment to improve or stabilise symptoms and the degree of disability. Given that patients will already require very regular review by their neurologist, this requirement will not place an added burden on patients. If an existing patient on ongoing treatment is not being managed by a neurologist, referral to a neurologist will be required. If Ig therapy has not been effective in stabilising symptoms, it will be ceased as a different treatment should be used. A trial of reducing dose and then stopping Ig therapy will be considered by doctors when patients are well and symptoms are stable.

New patients authorised to receive Ig therapy will require an initial check after the first three months of treatment to confirm that Ig therapy has been effective in improving the severity of symptoms and that the level of disability has not worsened. If improvement has been demonstrated after four months treatment, Ig maintenance therapy will be continued. If a response has not been demonstrated, a second medication must have been started and a further six months Ig therapy will be given. Further six monthly reviews will assess how effective the combination therapy has been to improve or stabilise the symptoms and ensure that no further worsening has occurred in disability. If the combination therapy has not been effective, Ig therapy

		will be stopped and a different treatment approach will be required. The ongoing arrangements for maintenance therapy are as outlined above for existing patients.				
Impact on demand:		Potassium channel antibody-associated encephalopathy is one subgroup of conditions now recognised under the diagnosis AMAE, which has become increasingly recognised as a condition requiring Ig therapy as first line treatment. For this specific indication, Ig use has been relatively stable over the last two years, with higher levels in the preceding two years. Demand for Ig for all conditions included under AMAE is expected to continue to increase in line with the trend of previous years, although this may be partially offset by the requirement to meet formal diagnostic and review criteria, and the introduction of trials of weaning.				
	2011-12	2012-13	2013-14	2014-15	2015-16	The Specialist Working Group estimated magnitude of effect: No impact against projected demand
Patient number	75	45	34	29	25	
Total Grams issued	20,068	13,218	9,909	9,396	9,421	
% Total Grams issued	0.61%	0.37%	0.25%	0.21%	0.19%	
Specialist Working Group knowledge development opportunities and recommendations relevant to the transition to v3.0						
None identified at this stage						

END OF PUBLIC CONSULTATION DOCUMENT

Next review: Review will be undertaken as part of AMAE