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The Ig Criteria are changing.

Criteria for the clinical use of immunoglobulin in Australia (the *Criteria*)

Neurology conditions - Version 3 summary

The *Criteria for Immunoglobulin Use in Australia* (the *Criteria*) is changing to Version 3 from 22 October 2018. The *Criteria* has been extensively reviewed by expert neurologists via specialist working groups (SWG) under the auspices of the National Immunoglobulin Governance Advisory Committee (NIGAC) and the National Blood Authority. All changes to the *Criteria* have also been subject to public consultation. Following public consultation, the proposed changes were considered by SWG's and endorsed by NIGAC prior to approval by the Jurisdictional Blood Committee (JBC). Through this process the *Criteria* have been significantly strengthened in a number of important respects. Changes to each of the neurological medical conditions within the *Criteria* are outlined below.

NEW CONDITION NAME FOR VERSION 3 *CRITERIA*	SUMMARY OF PROPOSED CHANGES TO NEUROLOGY CONDITIONS FOR VERSION 3 *CRITERIA*	NEW INDICATIONS FOR USE FOR VERSION 3 *CRITERIA*
Chronic inflammatory demyelinating polyneuropathy (CIDP), (including IgG and IgA paraproteinaemic demyelinating neuropathies) (formerly, Chronic inflammatory demyelinating polyneuropathy [CIDP] [including IgG and IgA paraproteinaemic neuropathies])	• Existing patients will remain on the current arrangements until the next due review. At that time prescribers will need to select the appropriate indication and specific condition from a dropdown list. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and the initial review are limited to neurologists. Once a response has been demonstrated, subsequent reviews may also be performed by a general medicine specialist. • Objective measure of disability and response to treatment is required. The Overall Neuropathy Limitations Scale (ONLS) (and the Medical Research Council [MRC] sum score as a baseline for review) in adults and children greater than 10 years of age; or the Six Minute Walk Test and/or the Modified Rankin Scale (MRS) for children less than 10 years of age are required to qualify. This provides a consistent baseline that can be compared for clinical response to Ig therapy at review. • Demonstration of clinical benefit in relation to disability and walking is required after an initial treatment period of four months, and annually thereafter, in order to access further treatment. • Cessation of Ig therapy should be considered for all patients after 12 months of treatment unless contraindicated. If a patient has relapsed in the first six months of a trial off therapy, an option to recommence treatment is available. A further trial off might be considered after at least two years. • Dosing is set as 2 g/kg for induction and for maintenance therapy as 0.4–1 g/kg, two to six weekly. A maximum dose of 2 g/Kg may be given in any four week period.	• Treatment of chronic inflammatory demyelinating polyneuropathy (CIDP) for patients in whom walking is compromised or there is significant disability • Relapse of chronic inflammatory demyelinating polyneuropathy (CIDP) patients within six months of commencement of trial off Ig therapy

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Guillain-Barré syndrome including variants (GBS) (formerly Guillain–Barré syndrome [GBS])	• Existing patients will remain on current arrangements until authorisation expiry. • Diagnosis is limited to neurologists, general medicine physicians and paediatricians. • A description of the patient's symptoms is required alongside confirmation that the weakness is progressive and indicates a trajectory to significant disability. • Objective measure of disability is required. The severity of disability as measured by the Guillain–Barré syndrome (GBS) disability score for initial Ig therapy; and the Medical Research Council (MRC) sum score for relapsing patients is required to be reported. Where relevant, this will provide a consistent baseline that can be compared for clinical response to Ig therapy at review or subsequent relapse. • For patients who relapse after initial Ig therapy, access to a second dose of Ig can be requested using a separate indication on the advice of, and after assessment by, a neurologist. Prescribers must confirm an initial clinical response to Ig therapy and subsequent worsening weakness. • Dosing is set at 2g/kg.	• Initial therapy for GBS with significant disability and progression • Relapse in GBS - treatment related fluctuation with initial improvement and subsequent deterioration post IVIg treatment

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Inflammatory myopathies: polymyositis (PM), dermatomyositis (DM) and necrotising autoimmune myopathy (NAM) (formerly Inflammatory myopathies: polymyositis [PM], dermatomyositis [DM] and inclusion body myositis [IBM])	• Existing patients will remain on the current arrangements until the next due review. At that time prescribers will need to select the appropriate specific condition from a dropdown list. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and review is limited to immunologists, neurologists and rheumatologists. • Diagnosis proven by muscle biopsy in adult patients, and either muscle biopsy or typical MRI and muscle enzyme changes with rash in children, is required. • Objective measure of muscle weakness and response to treatment is required. The use of the Medical Research Council (MRC) sum score in an adult, or the Childhood Myositis Assessment Scale (CMAS) in a child, provides a consistent baseline that can be compared for clinical response to Ig therapy at review. • For patients with significant dysphagia limiting dietary intake, a description of the degree of disability is required. Where possible video-fluoroscopy should be used to demonstrate pharyngeal involvement, unless video-fluoroscopy presents an unacceptable risk. • Ig therapy is reserved for patients who have failed to respond to corticosteroid treatment, unless such treatment is contraindicated or has resulted in unacceptable side effects. • Demonstration of clinical benefit in relation to muscle weakness or dysphagia is required after an initial treatment period of four months, and annually thereafter in order to access further treatment. • Cessation of Ig therapy should be considered for all patients after 12 months of treatment unless contraindicated. • Dosing is set as 2 g/kg for induction and for maintenance therapy as 1 g/kg four weekly. Existing patients may require transitioning to the dose levels permitted under this condition, if these are being exceeded.	• Treatment of significant muscle weakness or dysphagia unresponsive to corticosteroids and other immunosuppressant agents in adults with biopsy-proven PM or DM or NAM or children with clinical, biochemical and imaging abnormalities consistent with definite PM or DM or NAM

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Inflammatory myopathies – inclusion body myositis (IBM) (formerly Inflammatory myopathies: polymyositis [PM], dermatomyositis [DM] and inclusion body myositis [IBM])	• Existing patients will transition automatically to the new criteria. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and review is limited to immunologists, neurologists and rheumatologists. • Confirmatory biopsy is required unless absolutely contraindicated. Where possible video-fluoroscopy should be used to demonstrate pharyngeal involvement, unless video-fluoroscopy presents an unacceptable risk. A description of the degree of disability, including intolerance for solid dietary textures or aspiration episodes is required. • Demonstration of clinical benefit in relation to dysphagia, dietary intake and/or aspiration events is required after an initial treatment period of four months, and annually thereafter, in order to access further treatment. • Cessation of Ig therapy should be considered for all patients after 12 months of treatment unless contraindicated. • Dosing is set as 2 g/kg for induction and for maintenance therapy as 0.4-1 g/kg four to six weekly. A maximum total dose of 1g/kg may be given in any four week period. Existing patients may require transitioning to the dose levels permitted under this condition, if these are being exceeded.	• Patients with inclusion body myositis (IBM) who have dysphagia limiting dietary intake

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Lambert-Eaton myasthenic syndrome (LEMS)	• Existing patients will transition automatically to the new criteria. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and review is limited to neurologists. • Objective measure of disability and response to treatment is required. The use of the Medical Research Council (MRC) sum score to assess all patients is required; unless autonomic symptoms predominate. This provides a consistent baseline that can be compared for clinical response to Ig therapy at review. • Demonstration of clinical benefit in relation to disability or weakness is required after an initial treatment period of four months, and annually thereafter, in order to access further treatment. • For patients with severe disease who may slowly deteriorate between infusions and therefore not demonstrate response on the day of the annual review assessment, evidence that additional immunosuppressant agents have been prescribed is required when requesting ongoing Ig therapy. • Dosing should be reduced progressively and consideration should be given annually to a trial off Ig therapy once the patient has achieved stabilised disease or clinical remission. A maximum total dose of 2 g/kg may be given in any four week period. Existing patients may require transitioning to the dose levels permitted under this condition, if these are being exceeded.	• Additional therapy for LEMS patients with disability where symptomatic therapy is insufficient

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Multifocal motor neuropathy (MMN)	• Existing patients will remain on the current arrangements until the next due review. At that time prescribers will need to select the appropriate indication from a dropdown list. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and review is limited to neurologists. • Clinicians are required to provide information about the clinical phenotype and motor weakness when requesting Ig therapy for new patients. • Objective measure of disability and response to treatment is required. The use of the Overall Neuropathy Limitations Scale (ONLS) to assess all patients provides a consistent baseline that can be compared for clinical response to Ig therapy at review. • Demonstration of clinical benefit in relation to disability and weakness is required after an initial treatment period of four months, and annually thereafter, in order to access further treatment. • Cessation of Ig therapy should be considered for all patients after 12 months of treatment unless contraindicated. In previously responding patients who relapse within six months, further treatment can be requested using a separate indication. A further trial off might be considered after an additional two years therapy. • Dosing is set as 2 g/kg for induction and for maintenance therapy as 0.4–1 g/kg, two to six weekly.	• First-line and maintenance therapy for multifocal motor neuropathy (MMN) • Relapse of multifocal motor neuropathy (MMN) patients within six months of commencement of trial off immunoglobulin therapy

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Myasthenia gravis (MG)	• Existing patients will be managed in one of two ways depending on their current indication for Ig use: » Existing shorter-term authorisations that do not have continuing therapy will remain on current arrangements until authorisation expiry. » Existing authorisations that allow continuing treatment will transition automatically to the new criteria. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and review is limited to neurologists. • Where clinicians can confirm that the patient is in myasthenic crisis or at risk of crisis with symptoms of respiratory insufficiency, Ig therapy is supported as an alternative to plasma exchange. • For patients with planned surgery, clinicians are required to confirm the patient has advanced MG disease with either severe symptoms or bulbar symptoms or respiratory involvement. • In myasthenic crisis, or where access to maintenance therapy is required in patients with advanced MG disease, objective measure of disability and response to treatment is required. The use of the Myasthenia Gravis Composite (MGC) score to assess all patients provides a consistent baseline that can be compared for clinical response to Ig therapy at review. • Ig maintenance therapy is reserved for patients who have failed to respond to at least two alternative therapies concurrently, unless a reason is provided as to why these therapies cannot be prescribed. IVIg should be regarded as a stopgap treatment while using short-term drugs such as pyridostigmine and while introducing effective immunotherapy. • Demonstration of clinical benefit in relation to symptoms and disability is required after an initial treatment period of four months, and annually thereafter, in order to access further Ig maintenance therapy. • Cessation of Ig therapy should be considered for all patients after 12 months of treatment unless contraindicated. • Dosing is set as 1-2 g/kg for induction and for maintenance therapy as 0.4-1 g/kg four to six weekly. A maximum total dose of 1g/kg may be given in any four week period.	• Myasthenic crisis as an alternative treatment to plasma exchange • MG prior to surgery and/or thymectomy in patients with advanced disease, bulbar symptoms or respiratory involvement, as an alternative treatment to plasma exchange • As maintenance therapy for moderate to severe MG when other treatments have been ineffective or caused intolerable side effects

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Stiff person syndrome (formerly Stiff person syndrome [Moersch–Woltmann syndrome])	• Existing patients will transition automatically to the new criteria. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and review is limited to neurologists. • Objective measure of disability and response to treatment is required. The use of the Modified Rankin Scale (MRS) and the Distribution of Stiffness Index scores provide a consistent baseline that can be compared for clinical response to Ig therapy at review. • Demonstration of clinical benefit in relation to stiffness and disability is required after an initial treatment period of six months, and annually thereafter, in order to access further treatment. • Dosing is described as up to 2 g/kg for induction and for maintenance therapy as 0.4-1g/kg, two to six weekly, titrated to the individual's response. A maximum dose of 2 g/kg may be given in any four week period.	• Stiff person syndrome or variants with significant disability

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Acute disseminated encephalomyelitis (ADEM)	• Existing patients will be managed in one of two ways depending on their current indication for Ig use: » Existing shorter-term authorisations that do not have continuing therapy will remain on current arrangements until authorisation expiry. » Existing authorisations that allow continuing treatment will remain on the current arrangements until the next due review. Prescribers will need to select the appropriate indication and specific condition from a drop down list. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and review is limited to neurologists. • Confirmation that the diagnosis of ADEM is consistent with International Paediatric MS Study Group (IPMSSG) criteria is required. • Ig therapy is reserved for patients who have failed to respond to corticosteroids, unless there is a contraindication or intolerable side effects to such therapies. • For patients with monophasic ADEM maximum treatment time is three months. • For patients with recurrent or multiphasic ADEM, a total of 12 months treatment is permitted providing there is clinical benefit demonstrated after the first six months. Cessation of Ig therapy is required after a full 12 months therapy. • Previously responding patients with multiphasic ADEM, who relapse within six months following a trial off therapy, may be permitted further maintenance therapy under a separate indication. For these patients, clinical benefit must be demonstrated at review every six months, with a further trial of cessation considered for all patients after 12 months of treatment unless contraindicated. • Dosing is set as up to 2 g/kg for induction and for maintenance therapy as 1 g/kg four to six weekly. A maximum total dose of 1g/kg may be given in any four week period.	• Monophasic ADEM unresponsive to corticosteroid therapy or where corticosteroids are contraindicated • Recurrent or multiphasic ADEM unresponsive to steroid therapy or where corticosteroid therapy has become intolerable or is contraindicated • Relapse of patients with recurrent or multiphasic ADEM within six months of commencement of trial off immunoglobulin therapy

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IgM paraproteinaemic demyelinating neuropathy	• Existing patients will transition automatically to the new criteria. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and review is limited to neurologists. • Objective measure of disability and response to treatment is required. The use of the Overall Neuropathy Limitations Scale (ONLS) to assess all patients provides a consistent baseline that can be compared for clinical response to Ig therapy at review. • Ig therapy is reserved for patients who have failed to respond to at least two alternative therapies, unless there is a contraindication or intolerable side effects to such therapies. • Demonstration of clinical benefit in relation to symptoms and disability is required after an initial treatment period of four months, and annually thereafter in order to access further treatment. • Cessation of Ig therapy should be considered for all patients after 12 months of treatment unless contraindicated. • Previously responding patients, who relapse within six months following a trial off therapy, may be permitted further maintenance therapy under a separate indication, provided clinical benefit continues to be reported at review. For these patients a further trial of cessation should be considered after an additional two years of Ig therapy. • Dosing is set as 2 g/kg for induction and for maintenance therapy as 0.4-1 g/kg two to six weekly. A maximum total dose of 2g/kg may be given in any four week period.	• IgM paraproteinaemic neuropathy with significant progressive disability where other therapies have failed or are contraindicated • Relapse of patients with IgM paraproteinaemic neuropathy within six months of commencement of a trial off Ig therapy

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Multiple sclerosis - (MS) (relapsing remitting multiple sclerosis [RRMS]) (formerly Multiple sclerosis [MS])	• Existing patients will remain on current arrangements until authorisation expiry. Further treatment may be requested by submitting a new application. • Diagnosis and review is limited to neurologists. • Confirmation of clinically definite relapsing/remitting multiple sclerosis (RRMS) by brain or spinal cord MRI scan is required where the patient has had severe relapse at least twice during the previous two years. • Ig therapy is reserved for patients who have failed to respond to a course of high dose methylprednisolone, unless there is a contraindication or intolerable side effects to such therapy. • For prevention of further relapse confirmation is required that the patient has failed to respond to all other standard MS therapies, unless there is a contraindication to such therapies, or they are unavailable. • To access maintenance therapy, objective measure of disability and response to treatment is required. The use of the Expanded Disability Status Scale (EDSS) provides a consistent baseline that can be compared for clinical response to Ig therapy at review. • Demonstration of clinical benefit in relation to disability and walking is required after an initial treatment period of six months, and annually thereafter to access further treatment. After a maximum of 12 months treatment, patients should be re-assessed as to whether a more appropriate treatment is available. A new authorisation request will be required for any subsequent course (after 12 months) as appropriate. • Dosing is set as 1-2 g/kg for induction and for maintenance therapy as 0.4-1 g/kg, four to six weekly.	• Severe relapse of clinically definite relapsing remitting multiple sclerosis (RRMS) with no response to high dose methylprednisolone or where methylprednisolone is contraindicated • Prevention of relapse of clinically definite relapsing remitting multiple sclerosis (RRMS) with no response to high dose methylprednisolone or where methylprednisolone is contraindicated

NEW CONDITION NAME FOR VERSION 3 *CRITERIA*	SUMMARY OF PROPOSED CHANGES TO NEUROLOGY CONDITIONS FOR VERSION 3 *CRITERIA*	NEW INDICATIONS FOR USE FOR VERSION 3 *CRITERIA*
Opsoclonus-myoclonus ataxia (OMA)	• Existing patients will remain on the current arrangements until the next due review. At that time prescribers will need to select the appropriate indication and specific condition from a dropdown list. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and review is limited to neurologists. • Objective measure of disability and response to treatment is required. The use of the Cerebellar Functional System Score (CFSS) to assess all patients provides a consistent baseline that can be compared for clinical response to Ig therapy at review. • In adults, Ig therapy is reserved for patients who are concurrently treated with corticosteroids, unless there is a contraindication or intolerable side effects to such therapies. • Demonstration of clinical benefit in relation to disability and symptoms is required after an initial treatment period of six months, and annually thereafter, in order to access further treatment. • Dosing is set as 1–2 g/kg for induction and for maintenance therapy as 0.4–1 g/kg, four to six weekly.	• Treatment of OMA initially diagnosed in a child • Second-line treatment of OMA in adults following the use of corticosteroids

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Autoimmune encephalitis mediated by antibodies targeting cell–surface antigens (AMAE) (formerly included Hashimoto's encephalopathy, limbic encephalitis – non-paraneoplastic, limbic encephalitis – paraneoplastic and potassium channel antibody-associated encephalopathy)	• Existing patients being treated for Hashimoto's encephalopathy, limbic encephalitis – non-paraneoplastic, limbic encephalitis – paraneoplastic and potassium channel antibody-associated encephalopathy will remain on the current arrangements until the next due review. At that time prescribers will need to select the appropriate specific condition and indication from a dropdown list under the AMAE condition. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and review is limited to neurologists. • A description of clinical features and serology testing results is required. When serology cannot be used to confirm diagnosis, other diagnostic criteria will be applied under the indication for suspected AMAE using MRI, CSF analysis or EEG investigations to ensure that the clinical findings are consistent with encephalitis. • Objective measure of disability and response to treatment is required. The use of the Modified Rankin Scale (MRS) to assess all patients provides a consistent baseline that can be compared for clinical response to Ig therapy at review. • Demonstration of clinical benefit in relation to symptoms and disability post infusion is required after an initial treatment period of three months, and six monthly thereafter in order to access further treatment. • Cessation of Ig therapy should be considered for all patients after 12 months of treatment unless contraindicated. • Dosing is set as 2 g/kg for induction and for maintenance therapy as 0.4-1 g/kg four weekly. Existing patients may require transitioning to the dose levels permitted under this condition, if these are being exceeded.	• Confirmed antibody mediated autoimmune encephalitis (AMAE) or limbic encephalitis – cell surface antibody positive • Suspected antibody mediated autoimmune encephalitis (AMAE) – antibody results not available or seronegative AMAE or seronegative limbic encephalitis

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Neuromyelitis optica spectrum disorders (NMOSD) (formerly Devic disease [neuromyelitis optica])	• Existing patients will remain on the current arrangements until the next due review. At that time prescribers will need to select the appropriate specific condition from a dropdown list. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and review is limited to neurologists. • In patients with acute relapse of NMOSD, initial Ig treatment is permitted with confirmation that the diagnosis is consistent with criteria of the International Panel for NMO Diagnosis with or without the presence of AQP4 or MOG antibodies. • Ig therapy is reserved for patients who have failed to respond to a standard course of corticosteroids, immunosuppressant agents and plasma exchange, unless there is a contraindication or intolerable side effects to such therapies. • Objective measure of disability and response to treatment is required. The use of the Expanded Disability Status Scale (EDSS) provides a consistent baseline that can be compared for clinical response to Ig therapy at review. • Ig will be authorised for four weeks in the first instance. In responding patients, who have a further relapse, maintenance therapy may be provided, except where AQP4 antibodies have been confirmed. • Demonstration of clinical benefit in relation to symptoms is required every six months in order to access further treatment for patients who have relapsed after Ig therapy. • Cessation of Ig therapy should be considered for all patients after 12 months of treatment unless contraindicated. • Dosing is set as 2 g/kg for induction and for maintenance therapy as 1 g/kg, four to six weekly, in combination with an immunosuppressant agent. Patients on a higher dose may need to reduce dosing. A lower dose has been created to support maintenance and weaning off.	• Acute relapse of NMOSD with significant disability and corticosteroids and/or plasmapheresis have failed, are contraindicated or unavailable (one month treatment only) • Further significant relapse of NMOSD post Ig therapy with significant disability and resistant to corticosteroids and other immunosuppressant agents

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Rasmussen encephalitis (formerly Rasmussen syndrome)	• Existing patients will transition automatically to the new criteria. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and review is limited to neurologists. • A description of symptoms and confirmation that the patient has clinical features along with test findings including by EEG, MRI or histopathology which are consistent with a diagnosis of Rasmussen encephalitis as per the European Consensus Statement is required. • Ig therapy is reserved for patients who are concurrently treated with corticosteroids, unless there is a contraindication or intolerable side effects to such therapies. • Objective measure of disability and response to treatment is required. The use of the Modified Rankin Scale (MRS) to assess all patients provides a consistent baseline that can be compared for clinical response to Ig therapy at review. • Demonstration of clinical benefit in relation to symptoms and disability is required after an initial treatment period of six months, and annually thereafter in order to access further treatment. • Cessation of Ig therapy should be considered for all patients after 12 months of treatment unless contraindicated. • Dosing is set as 2 g/kg for induction and for maintenance therapy as 1 g/kg four weekly. Existing patients may require transitioning to the dose levels permitted under this condition, if these are being exceeded.	• Rasmussen encephalitis with concurrent steroid therapy unless contraindicated

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Susac syndrome	• Existing patients will transition automatically to the new criteria. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • The diagnosing and reviewing specialists are limited to neurologists, immunologists, rheumatologists or ophthalmologists. • A description of symptoms is required and probable or definite diagnosis has been made by at least two of the following: encephalopathy with diagnostic MRI brain changes, new sensorineural hearing loss or tinnitus, and/or branch retinal artery occlusions/ischaemia or arterial wall hyperfluorescence on angiography. • Ig therapy is reserved for patients who are being concurrently treated with corticosteroids, unless there is a contraindication or intolerable side effects to such therapies. • Objective measure of disability and response to treatment is required. The use of the Modified Rankin Scale (MRS) in all patients provides a consistent baseline that can be compared for clinical response to Ig therapy at review. • Demonstration of clinical benefit in relation to symptoms and disability is required after an initial treatment period of four months, and annually thereafter in order to access further treatment. • Cessation of Ig therapy should be considered for all patients after 12 months of treatment unless contraindicated. • Dosing is set as 2 g/kg for induction and for maintenance therapy as 0.5-1 g/kg two to six weekly. Existing patients may require transitioning to the dose levels permitted under this condition, if these are being exceeded.	• Probable or definite Susac syndrome in concurrence with high dose corticosteroids

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Childhood epileptic encephalopathy (formerly Epilepsy)	• Existing patients will remain on the current arrangements until the next due review. At that time prescribers will need to select the appropriate specific condition and indication from a dropdown list. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and review is limited to neurologists. • Diagnosis by EEG is required, and patients must demonstrate refractory epilepsy (at least weekly) with neurodevelopmental or cognitive issues. The keeping of a seizure diary is encouraged in order to be able to assess the clinical response to Ig therapy. • Ig therapy is reserved for patients who have failed to respond to corticosteroids and more than two anticonvulsants or surgery, unless there is a contraindication or intolerable side effects to such therapies. • Demonstration of clinical benefit in relation to symptoms and severity and/or frequency of seizures are required after an initial treatment period of three months, and annually thereafter in order to access further treatment. • Cessation of Ig therapy should be considered for all patients after 12 months of treatment unless contraindicated. If patients relapse and seizures start again after Ig therapy has been stopped, a further request for ongoing Ig therapy can be made for those who have previously responded. • Dosing is set as 2 g/kg for induction and for maintenance therapy as 1 g/kg over two to five days monthly.	• Children with epileptic encephalopathy resistant to anti-epileptic medications and steroid therapy or steroid responsive but dependant • Relapse of epileptic encephalopathy following a trial of weaning from Ig therapy in a patient previously demonstrating response

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Paediatric autoimmune neuropsychiatric disorder associated with streptococcal infections (PANDAS) or paediatric acute neuropsychiatric disorders (PANS) (formerly Paediatric autoimmune neuropsychiatric disorder associated with streptococcal infection [PANDAS])	• Existing patients will transition automatically to the new criteria. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and the initial review are limited to neurologists and immunologists. • Confirmation of a sudden onset of obsessive-compulsive disorder or severely restricted food intake associated with infection is required, along with a description of additional neuropsychiatric symptoms. • Ig therapy is reserved for patients who have failed to respond to standard antibiotic therapy. • Objective measure of disability and response to treatment is required. The use of the Modified Rankin Scale (MRS) is required at qualifying, and one of the Tics-Yale, OCD-CY-BOCS or Anxiety–SPENCE scales is required to assess improvement at review. • Demonstration of clinical benefit in relation to symptoms and disability is required after an initial treatment period of one month, and every three months thereafter, to access further treatment. • In stable patients, a trial of weaning towards cessation should be considered at each review. • Dosing is set as up to 2 g/kg for induction and for maintenance therapy as 1–1.5 g/kg four to six weekly. Existing patients may require transitioning to the dose levels permitted under this condition, if these are being exceeded.	• Paediatric autoimmune neuropsychiatric disorder associated with streptococcal infections (PANDAS) or paediatric acute neuropsychiatric disorders (PANS) unresponsive to trial of antibiotic therapy, and significant impairment requiring intervention • Relapse of paediatric acute neuropsychiatric disorders (PANDAS) or paediatric acute neuropsychiatric disorders (PANS) symptoms within three months of commencement of trial off Ig therapy
Hashimoto encephalopathy (steroid-responsive encephalopathy associated with autoimmune thyroiditis) ! no longer stand-alone (see AMAE)	• This condition will no longer be listed separately as patients can qualify where they have autoimmune encephalitis under Autoimmune encephalitis mediated by antibodies targeting cell-surface antigens (AMAE) (see above). • Existing patients will remain on the current arrangements until the next due review. At that time prescribers will need to select the appropriate indication and specific condition from a dropdown list. For these patients, additional clinical information will be required, as a one-off during transition to the new condition, to ensure the patient meets the new criteria.	• See AMAE

NEW CONDITION NAME FOR VERSION 3 *CRITERIA*	SUMMARY OF PROPOSED CHANGES TO NEUROLOGY CONDITIONS FOR VERSION 3 *CRITERIA*	NEW INDICATIONS FOR USE FOR VERSION 3 *CRITERIA*
Limbic encephalitis - nonparaneoplastic ! no longer stand-alone (see AMAE)	• This condition will no longer be listed separately as patients can qualify where they have autoimmune encephalitis under Autoimmune encephalitis mediated by antibodies targeting cell-surface antigens (AMAE) (see above). • Existing patients will remain on the current arrangements until the next due review. At that time prescribers will need to select the appropriate indication and specific condition from a dropdown list. For these patients, additional clinical information will be required, as a one-off during transition to the new condition, to ensure the patient meets the new criteria.	• See AMAE
Potassium channel antibody-associated encephalopathy ! no longer stand-alone (see AMAE)	• This condition will no longer be listed separately as patients can qualify where they have autoimmune encephalitis under Autoimmune encephalitis mediated by antibodies targeting cell-surface antigens (AMAE) (see above). • Existing patients will remain on the current arrangements until the next due review. At that time Prescribers will need to select the appropriate indication and specific condition from a dropdown list. For these patients, additional clinical information will be required, as a one-off during transition to the new condition, to ensure the patient meets the new criteria.	• See AMAE
Diabetic amyotrophy (formerly Diabetic amyotrophy [diabetic proximal neuropathy or diabetic lumbosacral radiculoplexus neuropathy]) ! no longer supported	• Ig therapy is not supported for Diabetic amyotrophy (diabetic lumbosacral radiculoplexus neuropathy; DLRPN) as there is insufficient evidence to support the use of IVIg in this condition. There are no published controlled clinical trials, and one registered randomised clinical trial was terminated prematurely without publication. This approach is consistent with recommendations of the Canadian and North American IVIg criteria. A single prospective, randomised double-blind controlled trial of intravenous methylprednisolone, demonstrated significantly more improvement in neuropathic pain in the IVMP group, but no significant improvement in neuropathy impairment score (Class 1). • Existing patients will remain on current arrangements until authorisation expiry.	• N/A

NEW CONDITION NAME FOR VERSION 3 *CRITERIA*	SUMMARY OF PROPOSED CHANGES TO NEUROLOGY CONDITIONS FOR VERSION 3 *CRITERIA*	NEW INDICATIONS FOR USE FOR VERSION 3 *CRITERIA*
• Paraneoplastic cerebellar degeneration • Paraneoplastic Subacute Sensory Neuropathy (formerly Paraneoplastic neurological syndromes) ! no longer supported	• Ig therapy is not supported for paraneoplastic cerebellar degeneration, associated with onconeural antibodies directed against intracellular antigens (Yo, Ma2, Hu, Ri). • Ig therapy is not supported for paraneoplastic subacute sensory neuropathy, associated with onconeural antibodies directed against intracellular antigens (Hu, Ri, CV2/CRMP5). • Response to immune therapy is poor in these paraneoplastic disorders, likely due to cytotoxic T-cell mediated neuronal loss. Therefore, onconeural autoantibodies are considered biomarkers for the presence of tumours rather than pathogenic mediators of neurologic disease, and should motivate the search for an associated malignancy. • Tumour resection and/or oncological treatment are the most effective therapies for these paraneoplastic neurologic syndromes, with case series reporting variable roles for corticosteroids, cyclophosphamide and rituximab. • Existing patients will remain on current arrangements until authorisation expiry.	• N/A

Please don't Ignore this.

The Ig Criteria are changing.

NEW CONDITION NAME FOR VERSION 3 *CRITERIA*	SUMMARY OF PROPOSED CHANGES TO NEUROLOGY CONDITIONS FOR VERSION 3 *CRITERIA*	NEW INDICATIONS FOR USE FOR VERSION 3 *CRITERIA*
Sjögren's syndrome associated neuropathy (formerly Sjögren's syndrome)	• Existing patients will remain on the current arrangements until the next due review. At that time prescribers will need to select the appropriate specific condition from a dropdown list. For these patients, additional clinical information will be required, as a one-off during transition, to ensure the patient meets the new criteria. • Diagnosis and the initial review are limited to neurologists. Subsequent reviews can be managed by a neurologist, immunologist or rheumatologist. • Demonstration of significant disability in patients with primary Sjögren's syndrome associated neuropathy without necrotising vasculitis is required. • Objective measure of disability and response to treatment is required. The use of the adapted Modified Rankin Scale (MRS) in all patients provides a consistent baseline that can be compared for clinical response to Ig therapy at review. • Ig therapy is reserved for patients who have failed to respond to corticosteroids and immunosuppressant agents, unless there is a contraindication or intolerable side effects to such therapies. • Demonstration of clinical benefit in relation to symptoms and disability post infusion is required after an initial treatment period of four months, and six monthly thereafter, in order to access further treatment. • Cessation of Ig therapy should be considered for all patients after 12 months of treatment unless contraindicated. • For patients who relapsed within six months of a trial off Ig therapy, further therapy may be requested using a separate indication, and a further trial off should be considered each 12 months. • Dosing is set as 1-2 g/kg for induction and for maintenance therapy as 0.4-1 g/kg four to six weekly. A maximum total dose of 1g/kg may be given in any four week period.	• Severe, primary Sjögren's syndrome associated neuropathy that is unresponsive to corticosteroid and immunosuppressant therapy • Relapse of Sjögren's syndrome associated neuropathy within six months of trial off Ig therapy