

identified cramp potentials in abductor digiti minimi, abductor pollicis brevis and flexor carpi ulnaris following application of a pressure cuff to the upper limb. Serological investigations for peripheral nerve hyperexcitability syndromes including voltage-gated potassium channel complex and GAD antibodies were negative. Treatment in the form of calcium, magnesium and Vitamin D replacement provided temporary symptomatic relief for the patient. Symptomatic therapy with carbamazepine and/or recombinant parathyroid hormone are being entertained. Normocalcaemic, normomagnesemic tetany following parathyroidectomy is a rarely reported phenomenon that is hypothesised to occur due to neuromuscular irritability resulting from relative hypocalcaemia following muscle membrane adaptation to long-standing hypercalcaemia

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PERAMPANEL MONOTHERAPY FOR FOCAL-ONSET SEIZURES (FOS): POST HOC ANALYSIS OF TREATMENT-EMERGENT ADVERSE EVENTS (TEAEs) BY TREATMENT PERIOD DURING FREEDOM STUDY 342

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10.1136/bmjno-2022-ANZAN.58

Objectives This post hoc analysis examines TEAE rates by Treatment Period in patients aged ≥ 12 years with newly diagnosed/currently untreated recurrent FOS, with/without focal to bilateral tonic-clonic seizures, who received perampanel monotherapy during Study 342 (FREEDOM; NCT03201900).

Methods During the Core Study, patients received perampanel 4 mg/day (4-week Pretreatment; 32-week Treatment [6-week Titration; 26-week Maintenance]) with the possibility to up-titrate to 8 mg/day. Patients could enter an Extension Phase for an additional 26 weeks (total: 52 weeks). TEAE rates were analysed by Treatment Period (Titration: Weeks 1–3; Steady State: Weeks 4–6; Maintenance: Week 7-end of 4/8-mg/day Maintenance). TEAE rates were calculated as number of events divided by total exposure, multiplied by 100.

Results Eighty-nine treated patients were included in the 4-mg/day group; of these, 21 patients were up-titrated and included in the 8-mg/day group. The rate of TEAEs/100 patient-months was highest during Titration (4 mg/day, 55.0; 8 mg/day, 86.4), lower during Steady State (4 mg/day, 25.2; 8 mg/day, 51.3) and lowest during Maintenance (4 mg/day, 12.4; 8 mg/day, 21.7). Treatment-related TEAEs and serious TEAEs with 4 mg/day exhibited the same pattern; rates with 8 mg/day were more variable. The most common TEAE/100 patient-months during Titration was dizziness (4 mg/day, 18.3; 8 mg/day, 57.6); lower rates of dizziness were reported during Steady State and Maintenance vs Titration.

Conclusions Perampanel monotherapy was generally well tolerated in patients aged ≥ 12 years with newly diagnosed/currently untreated recurrent FOS; TEAE rates generally decreased over time with treatment.

Funding Eisai Co., Ltd.

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FIRST-DOSE CHADOX1 VACCINATION AND ARTERIAL THROMBOSIS RISK

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10.1136/bmjno-2022-ANZAN.59

We describe three patients diagnosed with vaccine-induced immune thrombocytopenia and thrombosis syndrome (VITTs) who presented over a three-month period to a tertiary hospital, all of whom had both arterial and venous thrombosis. Case 1: A 53 year-old female had an acute left internal carotid artery (ICA) thrombus requiring intravenous alteplase and endovascular clot retrieval ten days following her first ChAdOx1 vaccination. Her admission was complicated by lower limb arterial thrombosis and pulmonary emboli. Case 2: A 67 year-old female presented with severe headaches 17 days following her first vaccination, and was found to have extensive cerebral venous sinus thrombosis (CVST) and intracerebral haemorrhage requiring decompressive craniectomy and drainage, and also developed multiple peripheral limb arterial thromboses. Case 3: A 57 year-old female who presented with convulsive status epilepticus after her first ChAdOx1 nCoV-19 vaccination ten days prior. She was found to have extensive clot burden with CVST complicated by haemorrhagic transformation of a venous infarct in addition to a complete left ICA occlusion needing thrombectomy. Similarly, she was found to have pulmonary emboli and arterial and venous limb thromboses. All patients received some combination of intravenous immunoglobulin, methylprednisolone, argatroban and ongoing apixaban or fondaparinux.

Conclusions Whilst venous thrombosis is well recognised in VITTs, we describe that the clinical spectrum can also commonly include arterial thrombosis, in the cerebrovascular and peripheral arterial tree. Furthermore, the presentation of this complication with arterial cerebral ischaemia acutely poses special difficulties in acute management given the degree of thrombocytopenia as a contraindication for thrombolysis.

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Eculizumab for acute neuromyelitis optica spectrum disorder relapses

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10.1136/bmjno-2022-ANZAN.60

Eculizumab is an effective medication approved for the treatment of neuromyelitis optica spectrum disorder (NMOSD) in maintaining disease remission, in patients who are aquaporin-4 water channel autoantibody (AQP4-IgG) seropositive. The efficacy of eculizumab in an acute relapse of NMOSD however is still under review. We describe a 46 year-old female who presented with acute left monocular vision loss on a background of bilateral optic neuritis treated fifteen years prior as suspected NMOSD. She had very poor vision from the right eye

(6/60). On presentation she was not on any long-term immunosuppressive agents. Her serum was positive for AQP4-IgG and MRI brain and spine demonstrated areas of demyelination in the corpus callosum and thoracic spine. She underwent plasmapheresis for five consecutive days but continued to clinically deteriorate with ongoing blindness in her left eye (light perception only). She was subsequently administered eculizumab with weaning oral corticosteroids. Clinically her vision improved to counting fingers and she remains on maintenance eculizumab infusions in the community. At 3 months, there is a steady improvement but still significant loss of central vision from that eye.

Conclusions The utility of eculizumab in NMOSD may assist with treating acute episodes. This theoretically accords with the mode of action in inhibiting conversion of C5 to C5a/b, perhaps arresting the acute inflammatory process in this disease. Given that disease burden and mortality in NMOSD is almost entirely related to relapses, increased use of eculizumab acutely could potentially aid recovery from an attack and therefore, minimise accrual of disability.

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MGBASE: READY, SET, GO! THE LAUNCH OF AN INTERNATIONAL ELECTRONIC DATABASE FOR PATIENTS WITH MYASTHENIA GRAVIS

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10.1136/bmjno-2022-ANZAN.61

Objectives To develop and implement the first international observational database for patients with Myasthenia Gravis (MG) to advance collaborative outcome-based MG research and improve the quality of care for patients with MG.

Methods The MGBase was developed based on the highly successful Multiple Sclerosis registry, MSBase. This approach leverages the existing IT infrastructure and governance structures of the MSBase registry. Designed to be used during regular outpatient consultations, MGBase provides a longitudinal graphical display of the patient disease course, therapies and outcomes. The development of the MGBase data entry fields and minimum data set was guided by an MG special interest group comprising national and international MG experts. Members of this group have subsequently formed the MGBase scientific leadership group responsible for determining the overall direction and scope of the MGBase registry.

Results MGBase was launched in December 2021 with the first patients recruited at two Melbourne tertiary centers. It is anticipated that another four national centers and several international centers will start recruiting patients within the next 6 months. Data from the first 21 patients enrolled in MGBase demonstrates a mean age of 60.1 years (62% female) with mean disease duration of 4.74 years. Five patients had a recorded exacerbation in the last 12 months. Further clinical and demographic data will be presented

Conclusion MGBase is the first observation international registry launched for patients with MG. The MGBase registry is dedicated to evaluating outcomes data in MG through collaborative international research.

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AN UNEXPECTED DIAGNOSIS OF NEUROFIBROMATOSIS TYPE 2 IN A PATIENT WITH ACUTE VERTIGO AND SUDDEN HEARING LOSS

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10.1136/bmjno-2022-ANZAN.62

Objectives We report a case of an acute vestibular syndrome with hearing loss to highlight that hearing loss is a red flag for sinister causes, and to demonstrate the utility of quantitative vestibular testing.

Methods/Results A 29-year-old woman was seen in our neuro-otology clinic for residual imbalance following an episode four months earlier of acute spontaneous vertigo and sudden right-sided hearing loss. Our clinical examination showed post head-shaking nystagmus to the left, a positive head impulse test to the right and a rightward Unterberger test. These signs were consistent with right-sided peripheral vestibulopathy, such as seen after vestibular neuritis, but given the history of hearing loss we arranged vestibular testing and imaging. Video head impulse testing showed reduced gains in all 3 right semi-circular canals, but unexpectedly also in the left lateral and posterior canals. Vestibular-evoked myogenic potentials demonstrated impaired otolith function on the right. Air conduction audiometry showed mild high frequency hearing loss on the right. Auditory brainstem response suggested bilateral retro-cochlear pathology. MRI imaging subsequently revealed bilateral vestibular schwannomas, conferring a clinical diagnosis of neurofibromatosis Type 2; this was later confirmed by genetic testing. Progress imaging at 12 months demonstrated increased schwannoma size, so she was commenced on bevacizumab. This proved effective, as progress imaging showed schwannoma shrinkage and repeat vestibular testing also showed improved vestibular function.

Conclusions Hearing loss is not typical for vestibular neuritis and should always prompt imaging to exclude other causes. Vestibular tests can help determine the underlying aetiology.

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IDENTIFYING AND APPROPRIATELY TREATING ADULT ARACHNOID CYSTS

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10.1136/bmjno-2022-ANZAN.63

Objectives Arachnoid Cysts a common finding found incidentally on CT scans in outpatient or inpatient settings. Differentiating benign cysts with complex or more likely symptomatic lesions is a useful skill for all practitioners to aid in counselling and triage of what can be a distressing incidental finding.

Methods A summary table of benign features and differentiating simple vs complex cysts has been created through retrospective review on clinical experience and data obtained through literature searches.

Results A guideline for referral to specialist services and an educational resource about the condition has been created.