2267 THE MONASH STATUS EPILEPTICUS STUDY (MOSES) – AGE, INITIAL GCS AND INPATIENT ONSET, NOT TIME TO TREATMENT, IS ASSOCIATED WITH IN-HOSPITAL MORTALITY AND MORBIDITY

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Objectives Status epilepticus (SE) is a medical emergency with high mortality and morbidity. We aimed to determine the predictors of in-hospital mortality and increased disability in an Australian setting.
Methods We retrospectively reviewed medical records between Jan 2020 and Dec 2020 to identify patients diagnosed with status epilepticus. Data regarding in-hospital mortality, modified Rankin Score (mRS), medical history, management and outcomes were collected from the electronic medical records. Multivariable logistic regression was performed.
Results We identified 157 patients meeting the inclusion criteria. In-hospital mortality was 20.4%(32/157) and 40.8%(64/157) had an increase in their mRS. An aetiology was identified in 71.3%(112/157). Only 42.7%(67/157) received first-line benzodiazepine therapy.

After adjusting for confounders, age, presenting Glasgow Coma Score (GCS) and inpatient onset of SE were associated with in-hospital mortaltiy. For every 1 year increase in age, the odds of in-hospital mortality increased by 1.05 (95%CI 1.01–1.08). For every 1 point decrease in GCS, the odds of in-hospital mortality increased by 1.13 (95%CI 1.01–1.25). Inpatient onset had greater odds of in-hospital mortality (Odds ratio (OR) of 4.42, 95%CI 1.71–11.49). Age, GCS and inpatient onset of SE also independently predicted increase in mRS. Time to first, second and third-line therapy did not predict mortality or morbidity.
Conclusions Status epilepticus was associated with a high rate of mortality and morbidity. Less than one-quarter of patients had timely provision of first-line SE treatment. Age, initial GCS and inpatient onset of SE were the strongest predictors of in-hospital mortality and morbidity.

2268 ESTIMATING THE PROFILE OF RESPONDERS TO TREATMENT: DO DIFFERENT PATIENTS SHOW BENEFITS ON DIFFERENT OUTCOMES?

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Objective This study aims to identify the baseline profile characteristic of super-responders to siponimod in the Phase 3 EXPAND study on four domains of progression (Expanded Disability Status Scale [EDSS]; upper limb function using the 9-hole peg test [9HPT]; ambulation using the timed-25-fooot walk test [T25FWT]; and cognitive processing speed using the single digit modalities test [SDMT]) using and innovative statistical approach
Methods This is a post-hoc analysis of the phase III EXPAND trial comparing siponimod (n=1099) vs placebo (n=546) in SPMS. A response score was derived from baseline characteristics describing participants with a more pronounced treatment effect on each of the 4 clinical endpoints and evaluated optimal division into non-responders/responders according to Zhao L et al. 2013.
Results In the whole cohort, the effect of siponimod on time to confirmed progression for each of the 4 outcomes was:

- EDSS: HR=0.79, p=0.0103;
- 9HPT: HR=0.86, p=0.23;
- T25FW: HR=0.95, p=0.53;
- SDMT: HR=0.75, p=0.001.

Four different responder profiles (RSP) were obtained and validated, all showing a significant interaction with treatment, thus defining responders to each of the 4 outcomes. Overall, 1290/1645(78%) patients were pronounced siponimod-treatment responders in at least one of the 4 clinical outcomes.
Conclusions 78% of SPMS patients had a large treatment benefit with siponimod on at least one of the 4 clinical outcomes.

2270 A CASE OF TRANSIENT VERBAL AUDITORY AGNOSIA FROM SEIZURES

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Introduction Verbal auditory agnosia is a rare phenomenon predominantly caused by bilateral vascular lesions. Here we describe a case due to seizures to highlight the importance of comprehensive language assessments, and discuss implications on normal language processing and childhood epileptiform encephalopathy.
Case A 65-year-old man presented following two witnessed generalised tonic-clonic seizures. He had a significant background of small cell lung cancer which was managed with chemotherapy and local radiotherapy five months prior. On admission he was initially considered to be delirious. Detailed examination revealed a profound mixed aphasia for spoken language with preservation of reading and writing. In retrospect he was also able to appreciate music and environmental sounds. An MRI demonstrated a left temporal pole metastatic lesion with left temporal leptomeningeal disease. His EEG showed bilateral centrotemporal epileptiform discharges. Following treatment with levetiracetam and clobazam his language deficits improved within five days with a concomitant normalisation of his EEG. He was able to recount his experience of hearing speech 'flow together' making individual words indistinguishable.
Conclusions This case demonstrates an atypical aetiology of a rare condition. It emphasises the need for detailed language assessments, and that common conceptualisation of language dysfunction (e.g., receptive and expressive aphasia) do not completely describe spoken language processing; dual-stream models of language function may be more useful paradigms. Additionally, there are clinical and electrophysiological parallels

to the Landau-Keffler syndrome in children and thus may provide clues to the pathophysiological aetiology of this syndrome.

2271 SAFETY AND TOLERABILITY OF CONVERSION TO SIPONIMOD WITH AND WITHOUT TITRATION IN PATIENTS WITH RELAPSING MULTIPLE SCLEROSIS: INTERIM RESULTS OF THE PHASE 3B EXCHANGE STUDY

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Objectives EXCHANGE is a 6-month, open-label, single-arm trial of conversion to siponimod in patients with MS. Here we report interim analyses evaluating safety and tolerability of converting to siponimod from other disease-modifying therapies (DMTs).

Methods Analysis included patients aged 18–65 years with advancing forms of RMS and on continuous oral/injectable DMTs for ≥3 months at time of consent. Patients previously on teriflunomide required 11–14 days accelerated washout. Those converting from fingolimod immediately switched to siponimod 2mg, with no dose-titration. All other patients were titrated from 0.25 to 2mg over 6 days. Primary endpoint was incidence of adverse events (AEs) possibly related to siponimod treatment.

Results 163 patients (74.2% female; mean age 46.6 years; mean baseline EDSS score 3.9) were eligible for safety analysis. Most common prior DMTs were oral and injection therapies: 30.7% fingolimod, 27.6% glatiramer acetate/IFNβ, 20.9% dimethyl fumarate, and 17.2% teriflunomide. In safety analysis, 31.3% of patients reported ≥1 AE possibly related to siponimod treatment. Most common AEs by preferred term were headache(8.0%), dizziness(4.3%), nausea(3.7%), bradycardia(3.1%), and fatigue(3.1%). There was no decrease in heart rate at 6 hours post 1st dose from baseline in the overall or any of the prior DMT groups. In the subgroup of fingolimod patients (n=7) who were switched to siponimod without dose titration, mean heart rate (SD) was 73.1 bpm (18.1) at 6 hours post 1st dose vs 68.4 bpm (10.8) at baseline.

Conclusions Immediate conversion from other DMTs to siponimod had an acceptable safety and tolerability profile, with no unexpected findings.

2272 SARCOIDOSIS AND INCLUSION BODY MYOSITIS OVERLAP

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Sarcoidosis and inclusion body myositis (IBM) are inflammatory disorders of unknown aetiology that share a potentially causal association. We report on 2 patients with pulmonary sarcoidosis who developed inclusion body myositis. Case 1 is a 68 year-old-man who presented with a 2 year history of dysphagia, reduced grip strength and difficulty climbing stairs on a background of biopsy-proven pulmonary sarcoidosis. Examination revealed dysarthria, mild neck flexion weakness, symmetric distal upper limb weakness most pronounced in flexor digitorum profundus, and proximal lower limb weakness. Creatine kinase level was three times the upper limit of normal. Muscle biopsy showed an inflammatory myopathy with rimmed vacuoles, COX negative fibres, and MHC1 upregulation, consistent with IBM. His condition has slowly progressed over 8 years but he remains ambulant with a walking stick. Case 2 is a 57 year-old-woman who developed proximal leg weakness predating a biopsy-proven diagnosis of pulmonary sarcoidosis by 5–10 years. Weakness accelerated over a 12 month period with a switch from prednisone to methotrexate for her underlying sarcoidosis. Examination revealed symmetric distal upper limb and proximal lower limb weakness. Creatine kinase level was twice the upper limit of normal. Muscle biopsy findings were consistent with IBM. Inclusion body myositis is associated with various autoimmune conditions. The association with sarcoidosis is well-recognised and may relate to shared Th1-mediated immunopathology. Recognition of the characteristic clinical pattern of weakness and pursuit of muscle biopsy are essential in differentiating IBM from sarcoid myopathy and other causes of weakness in patients with sarcoidosis.

2276 PLASMA NEUROFILAMENT LIGHT CHAIN AND DISEASE ACTIVITY IN CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY

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Objectives Serum biomarkers of neuronal damage might improve reliability, accuracy, and objectivity of assessments of disease activity in chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) where differentiating patients in remission from those with active but stable disease on therapy is a challenge to the clinician and trial design. We explored associations between plasma neurofilament light chain (pNfL) and disease activity in CIDP patients and examined its usefulness in determining disease remission.

Methods We examined pNfL in untreated CIDP patients (n=10) before and after intravenous immunoglobulin (IVIg) induction treatment, differences in pNfL in patients on maintenance IVIg treatment (stable patients n=15, unstable patients n=9), and in clinically stable IVIg treated patients (n=10) in whom we stopped IVIg to determine their disease activity and need for maintenance IVIg.

Results Untreated patients with CIDP: pNfL was significantly higher in patients than an age-matched, healthy control