group and comparable to control group values post IVIg induction.

Clinically stable and unstable patients with CIDP on IVIg treatment: pNfL was significantly greater in unstable patients than stable patients. A pNFL value above 16.6 pg/mL identified unstable treated CIDP from stable treated CIDP (sensitivity= 86.7%, specificity= 66.7%, area under ROC= 0.73).

Treatment withdrawal group: There was a strong and statistically significant correlation between pNfL concentration at time of IVIg withdrawal and the occurrence of relapse, suggesting an association of higher pNfL with active disease.

Conclusion pNfL concentrations may be a sensitive, clinically useful biomarker in assessing subclinical disease activity.

2278 AMA-VACC: CLINICAL TRIAL ASSESSING THE IMMUNE RESPONSE TO SARS-COV-2 MRNA VACCINES IN SIPONIMOD TREATED PATIENTS WITH SECONDARY PROGRESSIVE MULTIPLE SCLEROSIS

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10.1136/bmjno-2022-ANZAN.75

Objective To understand the longitudinal cellular and humoral immune responses to SARS-CoV-2 mRNA vaccines depending on the timing of vaccination and siponimod treatment.

Methods AMA-VACC is an open-label, three-cohort, prospective study in Germany with 41 multiple sclerosis patients currently treated with siponimod, any first-line DMT or without treatment at all. Cohort 1 received SARS-CoV-2 mRNA vaccination while continuing siponimod treatment, cohort 2 interrupted siponimod treatment for a full vaccination cycle and cohort 3 received vaccination during continuous treatment with first-line DMTs (glatiramer acetate, interferons, teriflunomide) or no current treatment. Primary endpoint is the rate of patients achieving seroconversion assessed by detection of serum neutralizing antibodies one week after SARS-CoV-2 mRNA vaccination. Furthermore, development and maintenance of SARS-CoV-2 specific T-cells is evaluated in all patients. Both parameters are analyzed in week one and month one and six after initial vaccination cycle and one month after a potential booster vaccination.

Results After a positive first interim analysis showing both SARS-CoV-2 neutralizing antibodies and T-cell responses one week after complete vaccination in siponimod patients data will be available in early 2022 for all patients at week one and later time points including first booster vaccinations.

Conclusions This analysis will provide first longitudinal data on the immune response after SARS-CoV-2 mRNA vaccination in siponimod treated SPMS patients and enable physicians and patients to make an informed decision on the coordination of SARS-CoV-2 mRNA vaccination and SPMS treatment.

2279 SEVERE REFRACTORY ANTIBODY-POSITIVE AUTOIMMUNE PANDYSAUTONOMIA POST-COVID19 VACCINATON: A CASE REPORT

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10.1136/bmjno-2022-ANZAN.76

Background COVID-19 vaccine-associated peripheral and central neuroimmunological disorders have been well described. We present the case of a 57 year old male who developed α3-ganglionic AChR antibody positive Autoimmune Autonomic Ganglionopathy (AAG) after completion of a two-dose course of mRNA (Cominarty) vaccination for COVID19.

Results A previously hypertensive 57 year old Vietnamese male presented with the subacute sequential onset of severe constipation, urinary retention, erectile dysfunction, sudomotor failure, sicca symptoms, non-reactive pupils and severe orthostatic hypotension shortly after receiving the second dose of an mRNA vaccine against COVID19. Autonomic testing revealed severe cardiovagal, adrenergic and sudomotor impairment, and tonic 'half-mast' pupils with evidence of sympathetic and parasympathetic denervation. Nerve conduction studies were normal. Investigations for common causes of autonomic failure were non-contributory to a diagnosis. Pathological α3-ganglionic ACHR antibodies were positive in serumas detected by a new flow cytometric immunomodulation assay. Malignancy was excluded. The patient was diagnosed with severe, treatment resistant acute pandysautonomia (AAG).

Conclusions While autonomic dysfunction has been previously reported post-COVID19 vaccination, to our knowledge this is the first reported case of antibody-positive AAG in this setting. The severity of this case is in marked contrast to the existing literature on idiopathic antibody-positive autoimmune pandysautonomia.

2282 SUSPECTING RASMUSSEN'S ENCEPHALITIS IN ADULTS: A CASE REPORT AND LITERATURE REVIEW

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10.1136/bmjno-2022-ANZAN.77

Objectives Adult-onset Rasmussen's encephalitis (A-RE) continues to present significant diagnostic and therapeutic challenges to clinicians, resulting in extensive investigations and an invariably long gap from presentation to diagnosis, which may subsequently correlate with poorer clinical outcomes. An increased awareness of the condition and a low threshold for suspicion are paramount to bridging this gap.

Methods/Results Here we present a case of a 36-year-old man with a prodrome of right sided sensory symptoms and subsequent right focal motor seizures, myoclonus/

polyminimyoclonus and neurological deficits. Paraneoplastic, autoimmune and neuroinflammatory screen were all unrevealing. The initial electroencephalogram (EEG) was normal; however, the repeat EEG showed focal slowing. Magnetic resonance imaging showed progressive T2 hyperintensity and volume loss predominantly over the left frontoparietal lobes, but also involved the right frontal lobe. Brain biopsy showed features of a T-cell predominant encephalitis. Rasmussen's encephalitis was suspected and immunotherapy with steroids and intravenous immunoglobulin were initiated, but he continued to deteriorate. He is now started on cyclophosphamide.

Conclusions Our case adds to the accumulating number of cases of A-RE reported in the literature. The rarity of the condition and overlap in clinical phenotype with other central nervous system diseases lead to delayed diagnosis in almost all reported cases. In our patient, the bilateral MRI changes and delayed recognition of seizures compounded to the diagnostic uncertainty. The main therapeutic options in adults are antiepileptic and immunomodulatory agents, and whilst there is no cure, early and aggressive therapy may result in improved patient outcomes.

2284 A CASE OF NEURODEGENERATION WITH BRAIN IRON ACCUMULATION (NBIA) PREVIOUSLY DIAGNOSED AS SPASTIC DIPLEGIC CEREBRAL PALSY

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10.1136/bmjno-2022-ANZAN.78

Objectives To present an 'ultrarare' case of neurodegeneration with brain iron accumulation (NBIA) that was previously diagnosed as spastic diplegic cerebral palsy.

Background NBIA is a very rare condition and affects both children and adults. It is clinically and genetically heterogenous and is characterized by pathological brain iron accumulation in the basal ganglia. Neurological manifestation include progressive extrapyramidal syndrome, pyramidal signs, cognitive dysfunction or ocular abnormalities. Neuroimaging shows T2 hypointense lesions bilaterally in the globus pallidi and substantia nigra on T2 weighted images

Case This is a case of a 24year old male who was diagnosed by paediatricians as 'cerebral palsy spastic diplegia' with intellectual disability. He was born at term and met all developmental milestones till age 3. He then had difficulties with gait, incoordination and delayed speech. He was diagnosed as a child as spastic diplegic cerebral palsy. His mobility declined further and he was assessed in adult neurology clinic for seizures. MRI brain, previously normal as a child, showed changes consistent with neurodegeneration with brain iron accumulation in the substantia nigra and atrophy of corpus collosum and cerebellum. Genetic testing confirmed heterozygous pathogenic mutations for PLA2G6 gene, securing diagnosis of phospholipase A2-associated neurodegeneration (PLAN).

Conclusion This case offers a reminder to re-evaluate diagnoses when clinical course of disorder does not fit the known pattern. Distinction between static and progressive clinical course is crucial. Regression in development should alert clinicians to metabolic and neurodegenerative disorders. NBIA, although 'ultrarare', should be considered as a differential if phenotypically appropriate.

2285 EFFICACY AND SAFETY OF AVALGLUCOSIDASE ALFA IN PARTICIPANTS WITH LATE-ONSET POMPE DISEASE AFTER 97 WEEKS OF TREATMENT DURING THE COMET TRIAL

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10.1136/bmjno-2022-ANZAN.79

Objectives Report efficacy/safety of avalglucosidase alfa (AVAL) in participants with late-onset Pompe disease in the extended treatment period (ETP) of Phase 3 COMET(NCT02782741) after a 49-week primary analysis period (PAP).

Methods At PAP enrollment, participants were treatment-naïve (n=100; age 16–78 years). All 51 participants receiving AVAL 20mg/kg every 2 weeks (qow) in the PAP continued this in the ETP (AVAL-arm). Of 49 participants receiving alglucosidase alfa (ALGLU) 20mg/kg qow in the PAP, 44 entered the ETP switching to AVAL 20mg/kg qow (Switch-arm).

Results Trends for improvement or stabilization from Baseline to Week 97 were observed for the primary and secondary outcomes of respiratory and motor function. Changes (LSmean[SE]) in forced vital capacity%predicted: AVAL-arm, +2.65[1.05]; Switch-arm, +0.36[1.12] and 6-minute walk test distance: AVAL-arm, +18.60[12.01]m; Switch-arm, +4.56[12.44]m. Similar trends occurred for muscle strength and quality of life measures. Treatment-emergent adverse events (AEs) in the ETP occurred in 49(96%) AVAL-arm and 42(95%) Switch-arm participants. Five discontinued during the ETP due to 6 treatment-emergent AEs; 4 were treatment-related (ocular hyperemia, erythema, urticaria, respiratory distress) and 2 were non-treatment-related (acute myocardial infarction, pancreatic adenocarcinoma). In the ETP, 12 AVAL-arm and 10 Switch-arm participants had treatment-emergent serious AEs (SAEs); 3 and 2 of them, respectively, had treatment-related SAEs. Switch-arm participants showed no safety- or immunogenicity-related concerns.