A.04

The classification of autosomal recessive cerebellar ataxias: a consensus statement from the society for research on the cerebellum and ataxias task force

M Beaudin (Québec)* A Matilla-Dueñas (Badalona) B Soong (Taipei) J Pedroso (São Paulo) OG Barsottini (São Paulo) H Mitoma (Tokyo) S Tsuji (Tokyo) JD Schmahmann (Boston) M Manto (Charleroi) GA Rouleau (Montreal) CJ Klein (Rochester) N Dupre (Québec)

Background: There is currently no accepted classification of recessive cerebellar ataxias, a group of disorders characterized by important genetic heterogeneity and complex phenotypes. The objective of this task force was to build a consensus and develop a clinical and pathophysiological classification for recessive ataxias. **Methods:** The work of this task force was based on a scoping systematic review of the literature that identified recessive disorders characterized primarily by a cerebellar motor syndrome and cerebellar degeneration. The task force regrouped 12 international ataxia experts who decided on general orientation and specific issues. **Results:** We identified 59 disorders that are classified as primary recessive ataxias. For each of these disorders, we present geographical and ethnical specificities along with distinctive clinical and imagery features. The primary recessive ataxias were organized in a clinical and a pathophysiological classification, and we present a general clinical approach to the patient presenting with ataxia. We also identified a list of 48 complex multisystem disorders in which ataxia is a secondary feature. **Conclusions:** This classification is based on a scoping systematic review of the literature and results from a sconsensus among a panel of international experts. It promotes a unified understanding of recessive cerebellar disorders for clinicians and researchers.

A.05

Five-year clinical and health economic outcomes in patients with late functional improvement post-stroke: A population-based cohort study

A Ganesh (Calgary)* R Luengo-Fernandez (Oxford) PM Rothwell (Oxford)

Background: We recently demonstrated that late functional improvement between 3-12 months post-stroke occurs in about one-fourth of patients with ischaemic stroke. It is unknown whether this improvement is associated with better long-term clinical or health-economic outcomes. **Methods:** In a prospective, population-based cohort of 1-year ischaemic stroke survivors (Oxford Vascular Study;2002-2014), we determined changes in functional status (modified Rankin Scale[mRS], Rivermead Mobility Index[RMI], Barthel Index[BI]) from 3-12 months post-stroke. We examined the association of late improvement (by ≥ 1 mRS grades, ≥ 1 RMI points, and/or ≥ 2 BI points) with 5-year mortality, institutionalization (Cox regressions), and health/social-care costs (generalized linear models), adjusted for age/sex/3-month disability/stroke subtype. **Results:** Among 1,288 1-year survivors, 1,135 had 3-month mRS>0, with 319(28.1%) demonstrating late improvement. 1-year survivors with late mRS improvement had lower 5-year mortality (aHR:0.68,95%CI 0.51-0.91,p=0.009), institutionalization

(aHR:0.48,0.33-0.72,p<0.001), and costs (margin -$17,369,-25,271 to -9,469,p<0.001). These associations remained on excluding patients with recurrent strokes during follow-up (e.g. 5-year death/institutionalization aHR:0.59,0.44-0.79,p<0.001) and on examining late improvement per RMI and/or BI (e.g. 5-year death/institutionalization aHR:0.67,0.53-0.84,p=0.001). **Conclusions:** Late functional improvement post-stroke is associated with lower 5-year mortality, institutionalization, and costs. These findings should motivate patients and clinicians to maximize late recovery and encourage payers to consider access to rehabilitative services for at least 1-year post-stroke.

A.06

Assessing inter-rater reliability in localizing sleep-related hypermotor seizures: a video-based survey

PM Lobbezoo (Utrecht) L Nobili (Genoa) S Gibbs (Montreal)*

Background: Sleep-related hypermotor epilepsy (SHE) is a focal epilepsy characterized by abrupt sleep-related hypermotor seizures (SRHS) with complex semiology. Although difficult to localize within the frontal lobe recent studies using intracerebral EEG recordings have suggested the existence of four distinct semiology patterns (SP) organized in a rostro-caudal manner. It remains unclear however if these SP are clinically useful. **Methods:** We aimed to estimate the inter-rater reliability (IR) of classifying SP in SHE amongst epilepsy and sleep medicine experts. Following a short training session, ten experts were asked to review and classify 40 videos of SRHS in patients with confirmed SHE. IR was calculated using Kappa statistics. **Results:** SP1 and SP4, who are at the opposite ends of the SHE semiology spectrum, had substantial IR (0.82 and 0.67, respectively). Meanwhile, SP2 and SP3 showed fair agreement (0.25 and 0.35, respectively) and represented the major source of variance, with a small difference favouring epilepsy experts. **Conclusions:** Amongst epilepsy and sleep medicine experts, IR of classifying SRHS into four SP was only mildly satisfactory. SP1 and SP4 were shown to be easily recognizable while SP2 and SP3 were frequently confounded. Improvements in SP recognition are needed before widespread clinical use.

A.07

Efficacy, safety, and tolerability of ubrogepant for the acute treatment of migraine: a single-attack phase 3 study, ACHIEVE I

DW Dodick (Scottsdale) RB Lipton (Bronx) J Ailani (Washington) K Lu (Madison) H Lakkis (Madison) M Finnegan (Madison) JM Trugman (Madison) A Szegedi (Madison) G Davidovic (Marham)*

Background: To evaluate efficacy, safety, and tolerability of ubrogepant, an oral CGRP receptor antagonist, for acute treatment of a single migraine attack. **Methods:** Multicenter, randomized, double-blind, placebo-controlled, parallel-group, single-attack, phase 3 study (NCT02828020). Patients randomized 1:1:1 to placebo, ubrogepant 50mg, or ubrogepant 100mg had 60 days to treat one migraine attack (moderate/severe pain intensity). Co-primary efficacy endpoints: pain freedom 2 hours post initial dose and absence