1. Supernormality in Tibialis Anterior: Validation of a Novel Technique. C.E.G. Moore[1], R. Arunachalam[2] and D.C. Allen[2]. (Departments of Clinical Neurophysiology, Portsmouth NHS Trust[1], Southampton University Hospital NHS Trust[2], UK).

Traditional methods of studying muscle conduction velocity recovery cycles (VRCs) using paired stimuli are technically demanding and time consuming. We have used a recently described computerised method [Ref] to investigate VRCs in tibialis anterior in 7 normal subjects.

Muscle fibre conduction velocities were measured by direct muscle stimulation and recording. VRCs were recorded with 1, 2 and 5 conditioning stimuli (10ms apart) delivered at intervals from 2 to 1000 ms prior to the test stimulus.

The maximum velocity increase (i.e. supernormality) of ~7% was seen at an interstimulus interval of 10ms with 1 and 2 conditioning stimuli. A second phase of supernormality was seen at ~100ms, which increased in proportion to the number of conditioning stimuli, and exceeded the early supernormality with 5 conditioning stimuli.

There was no correlation between the muscle fibre conduction velocity and the degree of supernormality. The maximal supernormality is less in tibialis anterior than brachioradialis, but the coefficient of variation is similar (0.43 vs 0.37).

This method is relatively quick to perform and should give new insights into the pathophysiology of muscle disorders.

[Ref] Z'Graggen WJ, Bostock H. *Muscle & Nerve* **39**: 616-626, 2009

2. The Effects of Age, Sex, and BMI on Nerve Excitability Testing. J. McHugh[1], R. Reilly[2] and S. Connolly[1]. (Department of Clinical Neurophysiology, St Vincent's University Hospital, Dublin[1] and Department of Neural Engineering, Trinity College Dublin[2], Ireland).

Background: Nerve excitability testing evaluates properties of the axonal membrane at the point of testing. Subtle effects of age and gender have previously been reported and different explanations proposed including age-related changes in channel expression, and other "non-neural" effects.

Aims: To examine the effects of age, gender and body mass index on median motor nerve excitability testing in a large cohort of normal subjects.

Methods: 105 subjects aged 19 to 81 years (54 male; 51 female) were evaluated by history, physical examination, and neurophysiological assessment to qualify as normal controls. Median nerve excitability testing was performed at the wrist using an automated TROND protocol.

Results: The threshold of motor fibres increases and supernormality decreases with age. Threshold electrotonus (TE), the current-threshold relationship and strength duration constant are not substantially affected by age. Males have significantly higher thresholds than females although the difference is attenuated following correction for body mass index (BMI), which independently increases rheobase and unconditioned threshold.

Discussion: The small but significant effects of age and gender are consistent with those reported previously in two smaller reports. The effect of BMI has not been reported on previously and seems to exert a "non-neural" effect that limits the access of an applied stimulus to the node of Ranvier.

3. The Difference between Eccentric and Concentric Calf Muscle Loading: An *in vivo* Study of Force and EMG. S. Chaudhry[1], H.R.C. Screen[1], R.C. Woledge[2] and D. Morrissey[2]. (School of Engineering and Material Science[1] and Centre for Sports and Exercise Medicine[2], Queen Mary University of London, London, UK)

Achilles tendinopathy is common in sports people. The main conservative treatment is heavy load eccentric calf muscle training, but it is unclear why eccentric but not concentric exercise has been shown to be effective. This study compared the electromyography and tendon force during these exercises.

12 healthy volunteers (6 male and 6 female, age 27.8 ± 1.9 years) performed eccentric and concentric loading exercises for the Achilles tendon. Tendon force and electromyography were measured using motion analysis, force plate, ultrasound imaging and EMG recordings.

During concentric exercise, both the rate of change and the maximal muscle activation were significantly higher ($p < 0.01$) than during eccentric. Conversely, the rate of change and maximal tendon force were significantly higher ($p < 0.01$) during eccentric than concentric exercise. This higher force fluctuation may partially explain the difference in therapeutic effect, as tenocytes repair tendon in response to mechanical stimuli.

4. The Diagnostic Yield of Two Sleep Activation Protocols in Paediatric Electroencephalography. A. Lee. (Department of Clinical Neurophysiology, Barts and The London NHS Trust, London, UK).

Routine surface EEG recording has a 30-50% chance of detecting interictal epileptiform discharges (IED) and sleep activation procedures are known to enhance irregularities that would otherwise remain undetected. At Barts and The London Trust, sleep-deprived (SD) activation was utilised exclusively in paediatric sleep studies until 2002, when the protocol was modified to a drug-induced (DI) procedure using trimeprazine. This has allowed comparison of the efficacy of

the two protocols in the 3 years preceding and in the 3 years following the changeover.

Electrographic data of 86 (48 boys, 38 girls) SD and 146 (87 boys, 59 girls) DI paediatric patients were evaluated. In addition to IED yield, localisation of evoked IED and patient-parent compliance were investigated.

The DI group yielded a significantly greater positivity (55.1%) for the presence of IED compared with the SD group (39.5%) ($P<0.017$). No significant differences were evident for localisation of IED ($P<0.312$) but DI sleep did show a much higher degree of patient-parent compliance ($P<0.000$). No gender differences were detected in either group.

Our study has demonstrated that higher yields of IEDs can be achieved with pharmaco-induced sleep studies. The findings differ in certain respects from some previous studies, which might reflect case selection bias, and study of the relevant factors may help further refine investigation of suspected epilepsy in children.

5. Phenotypic Assessment of Autosomal Recessive Congenital Stationary Night Blindness (CSNB) with and without Mutations in *GRM6*. **A.G. Robson[1,2], P. Sergouniotis[2], Z. Li[2], A.T. Moore[1,2], G.E. Holder,[1,2] A.R. Webster[1,2] (Moorfields Eye Hospital, London[1] and UCL Institute Of Ophthalmology, London[2], UK).**

Purpose: To describe cases of CSNB with and without mutations in *GRM6*.

Methods: Eight patients from 6 pedigrees were ascertained with autosomal recessive CSNB. International-standard full-field and pattern electroretinography (ERG; PERG), ON-OFF and S-cone ERGs were performed in 7 cases. Infant testing utilised skin electrodes. The coding region and intron-exon boundaries of *GRM6* were sequenced. Fundus images were reviewed.

Results: Seven of 8 individuals had mild to severe myopia. The scotopic rod ERG was undetectable and the bright flash ERG was electronegative in all. Photopic 30Hz ERGs showed borderline/marginal delay in 6 cases and were delayed in two elderly siblings with myopic degeneration. The transient cone ERGs had a bifid a-wave and sharply rising b-wave without oscillatory potentials. ON-OFF ERGs revealed reduced ON b-waves. The flash ERGs in the infant were qualitatively identical to those in her mother, in keeping with pseudo-dominant inheritance. The PERG was preserved in one case. DNA sequencing revealed mutations in *GRM6* in 4 of 8 cases.

Conclusions: A form of autosomal recessive complete CSNB caused by mutation in *GRM6* is described. Autosomal recessive complete CSNB is a heterogeneous condition.

6. An Evaluation of Neurophysiological Criteria Used in The Diagnosis of Motor Neurone Disease. **C.P. Douglass, R.H. Kandler, P.J. Shaw and C.J. McDermott. (Royal Hallamshire Hospital, Sheffield, UK).**

New criteria for the neurophysiological diagnosis of Amyotrophic Lateral Sclerosis/ Motor Neurone Disease (ALS/MND) were proposed in 2006 at an international symposium in Awaji-shima, Japan. They differ from the accepted revised El-Escorial criteria by considering fasciculation potentials to be evidence of acute denervation. In addition when assessing diagnostic certainty, the Awaji-shima criteria equate electrodiagnostic evidence of lower motor neurone dysfunction with clinical examination findings.

A retrospective review of 205 consecutive sets of notes was performed, from patients who underwent neurophysiological assessment for suspected MND. The clinical signs and neurophysiological findings were combined according to the two sets of criteria (revised El-Escorial and Awaji-shima) and the diagnoses reached were compared to the interval diagnosis, to establish the sensitivities and specificities of each protocol.

An interval diagnosis of MND was recorded in 107 patients. The sensitivity of the Awaji-shima criteria in reaching a diagnosis of MND was 60.7% and the revised El-Escorial 28%, with a specificity of 95.9% for both criteria. The Awaji-shima criteria increased the sensitivity of diagnosis without affecting the specificity. Accepting EMG evidence of fasciculations as evidence of acute denervation increases the diagnostic certainty of MND and the new criteria allow earlier diagnosis of MND without increasing the false positive rate.

7. Fasciculations in Amyotrophic Lateral Sclerosis: For How Long Should I Listen? **K.R. Mills. (King's College Hospital, London, UK).**

Electromyographers carry a heavy responsibility in the diagnosis of ALS. A practical issue is how long must the EMG of a muscle be observed before it can safely declared free of fasciculation. This is particularly important when the muscle is uninvolved clinically or when seeking evidence of EMG abnormality in a cranial nerve innervated muscle. To answer this question, the discharge times of fasciculation potentials (FPs) were logged in 53 muscles (biceps 8, tibilalis anterior 27, first dorsal interosseous 17, trapezius 1) from 19 patients with definite ALS according to the modified El Escorial criteria. At least 50 FPs were recorded from each muscle giving total recording times of from 51.5 to 776.8 s and overall FP rates of 4.03 to 116.6 FP/min. From each dataset, the intervals to the nearest ms between 1 … 5 FPs were computed and the cumulative frequency distribution constructed for each set of intervals. Thus the probability of

observing up to 5 FPs with respect to the time elapsed from needle insertion could be calculated. To observe 1 FP with a probability approaching unity, then observation time should be 90 s. To be confident, most electromyographers would wait until more fasciculations were detected; to record 5 FPs with certainty, 180 s would be required. Of course in most cases, FPs would be detected much more quickly, the above times being the longest needed before deciding that fasciculations were almost certainly absent.

8. The Uncertainty of Nerve Conduction Measurements. J. Simmons and N.J. Smith. (University Hospital, Nottingham, UK).

The uncertainty of a measurement result is the interval within which the true value is expected to lie with stated probability, usually 95%. Uncertainty is assessed from statistics applied to repeated measurements (type A evaluation), and other factors (type B evaluation).

Subjects (n=37) underwent a standardised nerve conduction study protocol performed twice by the same physiologist, and once by a different physiologist. In the ulnar, tibial, peroneal, sural and medial plantar nerves, Type A evaluation of uncertainty was calculated from the within-subject standard deviation of pairs of values obtained by the same physiologist (repeatability) and different physiologists (reproducibility).

Sensory nerve action potential amplitude variability (measured peak-to-trough) was related to amplitude, and variability in latency was related to distance. The greatest uncertainties were sural and medial plantar nerve amplitudes, and ulnar sensory velocity. Reproducibility uncertainties were higher than repeatability, the greatest difference being in medial plantar sensory amplitude and in peroneal motor latency from ankle stimulation.

Uncertainty defines the range within which the true value could be zero, or on the opposite side of a reference value; uncertainty also establishes a critical difference for repeated results.

9. Modulation of Corticomuscular Coherence by Afferent Stimulation varies with Stimulus Intensity. V.M. McClelland and K.R. Mills. (King's College Hospital, London, UK)

The correlation within the frequency domain between rhythmic activity in the electro-encephalogram (EEG) and electromyogram (EMG) is known as corticomuscular coherence (CMC). CMC is task dependent and therefore likely to be physiologically important. CMC is reduced in studies where afferent feedback is blocked. However, the role of afferent inputs in modulating CMC remains uncertain. In most studies investigating CMC modulation, subjects perform a dynamic motor task. It is difficult to differentiate effects of a changing voluntary command from those of the changing afferent input accompanying these tasks. We have investigated the effect of a transient peripheral stimulus on the level of CMC during a maintained motor task.

Graded electrical stimuli (1.0–2.5 times perceptual threshold (T)) were applied to the dominant index finger in 5 healthy adults performing a simple key grip task. CMC between sensorimotor cortex EEG and intrinsic hand muscle EMG was detected between 14-36Hz. Stimuli above 2.0T produced a reduction in the background 14-36Hz CMC, followed by a rebound increase of CMC in this frequency range. This increase outlasted any reflex evoked cortical activity and continued up to 2.5 seconds post-stimulus. The depth of CMC modulation varied with stimulus intensity, providing further evidence that CMC is influenced directly by afferent information.

10. Rhythmical EEG Changes and Stereotyped Clinical Features in Stretching Syncope: A Challenging Differential Diagnosis with Seizures. P.G. Sarrigiannis[1], M. Randall[2], R.H. Kandler[1], M. Reuber[2], R.A. Grunewald[2] and K. Harkness[2]. (Department of Clinical Neurophysiology[1] and Neurology[2], Royal Hallamshire Hospital, Sheffield, UK).

Episodes of stereotyped simple motor manifestations, like stretching, followed by loss of consciousness can be easily misdiagnosed as seizures. We describe 3 male patients (aged 21, 26 and 20) with video-EEG evidence of stretch syncope. In all patients, the attacks were characterised by an initial fairly stereotyped phase of stretching and holding of breath. Generalised rhythmical EEG slow wave abnormalities soon emerged. Clinically, there was a variable degree of loss of consciousness with asymmetric jerks in the more prolonged attacks. A significant period of tachycardia (captured on ECG) coincided with the phase of stretching/holding of breath and the generalised slow wave abnormalities on the EEG. The aforementioned clinical EEG and ECG changes define this distinctive clinical syndrome: stretch syncope. There was an element of obsessive compulsive disorder in all three cases.

Simultaneous video-EEG recordings, transcranial doppler of the middle cerebral arteries and continuous (finger) arterial pressure measurements were performed in all three patients during their stretching syncopal attacks. At the time the patients lost consciousness, a dramatic drop in the cerebral blood flow and in the systemic arterial pulse pressure took place. Simultaneously, slow wave abnormalities emerged on the EEG. To our knowledge, such complete recordings (including a detailed hemodynamic analysis) have not previously been reported.

11. **Accuracy of Stimulated Single Fibre EMG in the Diagnosis of Myasthenia.** **B. Packham[1] and T. Tidswell[2]. (University College London[1] and Royal Free Hospital, London[2], UK).**

Introduction: Stimulated single fibre EMG (SSFEMG) is a rapid test of neuromuscular junction instability but there is a paucity of literature about the clinical application of the technique. The aim of this study was to compare SSFEMG abnormalities with clinical outcome in patients referred to Neurophysiology with suspected myasthenia.

Methods: 42 consecutive patients (2008-2009) had SSFEMG of orbicularis oculi, as part of their assessment, with a concentric "facial" EMG needle and 10Hz monopolar stimulation of the facial nerve; data was analysed on a Keypoint EMG machine. SSFEMG was abnormal if ≥10% of motor fibre potentials had jitter >40micros. Clinical outcome was assessed by independent, blinded case note review and defined as either definite myasthenia (antibody positive), probable myasthenia (an antibody negative treatment responder) or unlikely myasthenia (no response to treatment or alternative diagnosis made).

Results: 36 patients had follow-up information: 17 had definite or probable myasthenia (17/17 abnormal SSFEMG) and 19 unlikely myasthenia (3/19 had abnormal SSFEMG, all three had alternative neuromuscular disease).

Conclusions: The study confirms that SSFEMG has a high sensitivity for neuromuscular junction instability.