Infantile Spasms and Treatment. J. Martinez, S. Penfold, Z. Agirre-Arrizubieta. (Department of Clinical Neurophysiology, East Kent Hospitals University NHS Foundation Trust, UK).

We assessed treatment options in children with infantile spasms (IS) during a three-year-period (2007-2010) at East Kent Hospitals.

Medical histories were reviewed to obtain aetiology, neurological findings, MRI, video-EEG studies, treatment and follow-up.

A total of 10 patients with IS were studied. Aetiology: symptomatic in 9 patients and unknown in 1. Most common IS: flexion in 43.6% and mixed in 36.4%. Therapeutic response was broadly satisfactory: 60% seizure free; 20% partial response, 10% with more than 50% decrease and 10% with no IS but other seizures. Polytherapy: noted in 70% of patients. At follow-up, development was normal in 1 patient and delayed in 9 (2 mild, 3 moderate and 2 profound).

Optimal treatment of IS remains challenging. In East Kent Hospitals, the commonest current therapy is a combination of 2 of the following: ACTH, steroids, vigabatrin and topiramate. The outcome and prognosis of IS appeared similar or mildly improved compared with previous experience. However, safer and more effective therapies are needed to improve long-term outcome of these patients.

Velocity Recovery Cycles in Biceps Brachii: New Insights Into The Origin of The Early Supernormality. C.E.G. Moore1,3, R. Arunachalam2 and D.C. Allen2,4 (Departments of Clinical Neurophysiology Portsmouth NHS Trust1 and Southampton University Hospital NHS Trust2 and Universities of Portsmouth3 and Southampton4, UK).

Traditional methods of studying muscle conduction velocity recovery cycles (VRCs) using paired stimuli are technically demanding and time consuming. We have previously reported the use of an automated computer driven methods to investigate VRCs in tibialis anterior (TA). This has shown promise in the investigation of muscle disease, in particular critical illness myopathy.

We now present results from the investigation of biceps. Muscle fibre potentials were recorded using standard technique for direct muscle stimulation. VRCs were recorded with 1, 2 and 5 conditioning stimuli (10ms apart) prior to the test stimulus. The interstimulus intervals (ISI) between the last conditioning stimulus and the test stimulus were varied from 2 to 1000ms.

The early supernormality (ISI~10ms) in biceps was greater than in TA. In addition there were distinct differences in the amount of early supernormality after 1, 2 or 5 conditioning stimuli. The early supernormality has hitherto been explained purely in terms of membrane capacitance and should be independent of the number of conditioning pulses. Our findings of increased supernormality with multiple conditioning stimuli, however, indicate that in some groups of fibres accumulation of potassium may also play a significant role. This may have implications when interpreting results in muscle diseases.


Idiopathic small fibre neuropathy (ISFN) is associated with intraepidermal nerve fibre loss and an increased prevalence of impaired glucose tolerance (IGT). It has been suggested that the dysglycaemia of IGT and additional metabolic risk factors contribute to small nerve fibre damage in these patients.

25 patients with ISFN and 12 aged-matched control subjects underwent detailed evaluation of neuropathic symptoms, neurological deficits (Neuropathy deficit score (NDS); Nerve Conduction Studies (NCS); Quantitative Sensory Testing (QST) and Corneal Confocal Microscopy (CCM)) to quantify small nerve fibre pathology. 8 (32%) of patients had IGT. Whilst all patients with ISFN had significant neuropathic symptoms, NDS, NCS and QST except for warm thresholds were normal. Corneal sensitivity was reduced and CCM demonstrated significant reductions in corneal nerve fibre density (P<0.0001), nerve branch density (P<0.0001), nerve fibre length (P<0.0001) and an increase in nerve fibre tortuosity (P<0.0001). These parameters did not differ between ISFN patients with and without IGT or correlate with BMI, lipids and blood pressure.

CCM provides a sensitive non-invasive means to detect early small nerve fibre damage in patients with ISFN and metabolic abnormalities do not relate to nerve damage.

Are There Differences Between Han Chinese and Caucasians in Transcranial Magnetic Stimulation (TMS) Parameters? Xiang Yi1, K. Fisher2, K. Mansoor3, Ming Lai5, S. Baker2. (Mechanical and System Engineering4, Institute of Neuroscience5, Newcastle University, Royal Victoria Infirmary5, Newcastle, UK).

An experiment has been performed studying a range of TMS parameters in Chinese and Caucasian subjects. Sixteen subjects were studied in each group. A circular coil at the vertex was used for stimulation, whilst recording surface electromyograms (EMG) from right first dorsal interosseous. In the passive state we measured motor evoked potential (MEP) threshold, MEP
recruitment, short interval intracortical inhibition (SICI) and intracortical facilitation (ICF). The MEP threshold, recruitment and silent period were measured in active muscles.

Chinese subjects showed significantly higher passive thresholds (p<0.01), less inhibition in of the motor response (SICI p< 0.01 ) and the silent period was shorter (p< 0.05 ).


Introduction: As Motor Neuron Disease (MND) progresses there may be pathological or compensatory changes in cortical motor network activation outside the normal somatic representation, which may be due to excitotoxic damage of local inhibitory circuits leading to inappropriate activation of the motor pathway or due to increased peripheral motor effort leading to greater central drive.

Method: Functional magnetic resonance imaging of the blood oxygenation response (BOLD-fMRI) during a calibrated handgrip task of 5-30% maximum grip strength was performed in 12 healthy controls, 12 MND subjects and 12 subjects with multifocal motor neuropathy (MMN), under placebo (i.v. saline) or midazolam conditions and images analysed with statistical parametric mapping software (SPMv5).

Results: Statistically similar BOLD-fMRI signal increases were seen in all groups in contralateral primary sensorimotor cortex and ipsilateral cerebellum, and statistically similar decreases in BOLD activation of the cortical motor networks with midazolam.

Conclusion: There is no evidence for motor cortex reorganisation in patients with MND, using this calibrated motor paradigm; the comparable BOLD-fMRI signal reduction following midazolam in all groups suggests that neuronal inhibition via GABA potentiation is not impaired in MND.


Myoclonic status in non-progressive encephalopathies (MSNE) is an epileptic syndrome in development characterized by diffuse EEG abnormalities associated with positive and/or negative phenomena correlated with transient or recurring motor, cognitive or behavioural disturbances. MSNE is now included in the ILAE classification of electroclinical epilepsy syndromes in infancy.

The recognition of MSNE may guide diagnostic workup and suggest an underlying cause such as the presence of a genetic syndrome like Angelman syndrome, brain malformation or hypoxic ischaemic encephalopathy.

We describe the clinical and video-EEG features of two adult patients with MSNE and discuss diagnostic and management challenges in this difficult to treat epilepsy syndrome. The recognition of MSNE resulted in the correct identification of Angelman syndrome in one patient who was otherwise considered to have learning disability of undetermined aetiology.

7. Examining the Reproducibility of Median Motor Nerve Excitability Testing. J.C. McHugh1, R. Reilly2, S. Connolly1. (Department of Clinical Neurophysiology, St. Vincent’s Hospital Group, Dublin, Ireland1 and Department of Neural Engineering, Trinity College Dublin, Ireland2).

Introduction: Threshold tracking allows functional assessment of axonal membrane function in peripheral nerve, and the technique has potential for use in longitudinal studies.

Objective: To examine the test-retest reliability of nerve excitability in a healthy cohort.

Methods: Fifteen subjects had median motor excitability studies performed by the same operator on three occasions (twice on the same day, and once after one week). ANOVA was used to analyse and compare the within-subject and between-subject variances for different parameters.

Results: In general nerve excitability testing had excellent reproducibility. The parameters with the greatest reproducibility were superexcitability and minimum I/V (current threshold) slope. F-ratio of between-subject versus within-subject variance 7.30 (p < 5x10^-5) for superexcitability and 7.0 (p < 5x10^-3) for minimum I/V slope. The least reproducible parameters were strength-duration time constant, and threshold parameters such as rheobase, in which the between-subject and within-subject differences were not significantly different.

Conclusion: Nerve excitability testing is a reliable test over time, and should be suitable for use in longitudinal patient studies.

8. Measures of Peripheral Nerve Excitability Demonstrate Rapid And Sustained Improvement in Nerve Function Following Renal Transplantation. C.E.G. Moore1, M. Todd2, J. Mason2, M. Connolly2. (Departments of Clinical Neurophysiology1 and Renal Medicine2, Queen Alexandra Hospital, Portsmouth, UK and University of Portsmouth2, UK).

Neural dysfunction is a common and important feature of advanced chronic kidney disease. The pathophysiology of ‘uraemic neuropathy’ remains ill understood. Measures of axon excitability have implied that chronic depolarisation, secondary to hyperkalaemia, may be the major pathological factor. The ability to reduce potassium levels with dialysis is however,
short lived and a more prolonged normalisation of the associated depolarisation may be required (Ref).

We present results from a patient receiving a living-related renal transplant. Testing was performed 8 months, and 1 day pre-transplant and at 1 day, and 3 months post-transplantation. 8 months prior to transplantation peripheral nerve excitability was significantly diminished, with threshold changes to depolarising and hyperpolarising stimuli (‘fanning-in’) (p<0.005) and reduced sub- and superexcitability in the recovery cycle (p<0.02). These abnormalities worsened up until transplantation.

23 hours post-transplant there was a marked recovery towards normal. This was too rapid to be explained by changes in neuronal cytostructure or remyelination. By three months post-transplantation all measures of axonal excitability had normalised. The neuropathy symptom score had improved and there was no further deterioration in ‘routine’ nerve conduction studies.

These findings show ‘proof of principle’ that decreasing the amount of hyperkalaemia related depolarisation improves nerve membrane function with a potential reversal of neuropathy.

Ref: Krishnan et al. Brain 2005;128:2164-74