British Society for Clinical Neurophysiology

Scientific Meeting
Historic Dockyard, Portsmouth
13 July 2012

Abstracts (not peer reviewed)

1. Prognostic Value of Somatosensory Evoked Potentials, After Cooling, Following an Out-of-Hospital Cardiac Arrest. B.J. Haynes, C.E.G. Moore and S.P. Veal. (Department of Clinical Neurophysiology, Queen Alexandra Hospital, Portsmouth, UK).

Patients who experience an out-of-hospital (OOH) cardiac arrest commonly sustain some level of anoxic brain injury due to poor or delayed resuscitation. Somatosensory Evoked Potentials (SSEPs) have been shown to provide an early indicator of outcome in such patients. We studied 13 patients who were admitted to the Critical Care Unit (CCU) following OOH arrest and subsequently cooled. After re-warming, upper limb median SSEPs were recorded from the ipsilateral Erb's point and from the contralateral cortex (C3' or C4'), with concurrent EEG recording also obtained and graded I-V as per the Hockaday scale. Other physiological scales were also recorded. The cortical N^{20} was absent in 5 patients, none of whom survived, providing a 100% positive predictive value. Of the 8 patients who had a preserved N^{20}, 3 survived giving a negative predictive value of 37.5%; with 5 who did not survive having grade IV EEGs. The results of this study support the continued investigation into and use of SSEPs in this patient group, and highlights the value of simultaneous EEG recording.

2. Susac Syndrome: A Rare But Potentially Treatable Early Onset Dementia. R. Jain, G. Payne, B. Thomas and J. Heath. (Dept. of Clinical Neurophysiology, University Hospital of Wales, Cardiff, UK).

Background: Susac syndrome is a rare autoimmune disease characterised by the clinical triad of encephalopathy, branch retinal artery occlusion and sensory neural hearing loss, occurring mostly in young females.

Case report: A 52 year old woman presented with sub-acute onset headaches and deafness followed within weeks by progressive imbalance, memory impairment and confusional state. MRI of the brain revealed multiple sub-cortical white matter hyperintensities including the corpus callosum. The electroencephalogram (EEG) revealed theta range background slowing, bitemporal sharp activity and frontal intermittent rhythmic delta activity (FIRDA). A diagnosis of Susac syndrome was made based on clinical and MRI findings and treatment by IV Methyl prednisolone followed by oral steroids was instituted. At 3 months follow up, there was some improvement in the cognition and balance, while no significant changes were seen on the repeat MRI brain and the EEG.

Conclusion: Although rare, Susac syndrome is a potentially treatable disease, and if undiagnosed can lead to severe dementia, vision and hearing loss. The EEG shows non-specific changes including diffuse background slowing and FIRDA.

3. Carbamate Toxicity and The Neuromuscular Junction Shown By Single Fibre EMG. E.M. Sedgwick, V. Weerasinghe, N. Senaniyake, K. Naser and L. Karalliedde. (University of Peradenya, Sri Lanka).

Carbamates are widely used as insecticides. Their action is as inhibitors of acetylcholinesterase but unlike organophsophates (OP) the inhibition is not as long lasting and carbamate does not permanently bind with acetylcholinesterase. Signs of toxicity are those of cholinergic crisis and include muscle weakness, predominantly of the facial and respiratory muscles with fasciculations, ptosis and respiratory paralysis.

Treatment is as for OP poisoning and requires rapid atropinisation raise the heart rate and begin to dilate the pupils. Oxygen, fluids and respiratory support must be provided. Oximes are not used. Recovery is quicker than for OP poisoning. The sequelae of intermediate syndrome and late onset neuropathy have not been reported.

Here we report concentric needle single fibre EMG findings from orbicularis oculi on 6 patients who self-ingested carbamate insecticides. The study had the approval of the University of Perideniya Ethical Committee.

Patients were seen 18-120 hours post ingestion. Jitter values were raised at 18 hours and by 120 hours were returning to normal. One patient, seen at 3 and 5 days post ingestion was on respiratory support at 3 days with a median jitter of 35 μsec (Upper limit of normal 26 μsec) with 43% of fibres being abnormal but at day 5 was respiring spontaneously with a median jitter of 15 μsec and only 13% abnormal fibres.

Single fibre EMG shows the dynamics of neuromuscular dysfunction in carbamate poisoning which correlates with the clinical picture and is different from that seen in organophosporus poisoning.

4. Auditing Referrals of EEGs in Elderly Patients and Adherence to Guidelines. O. Lee, and A. Kurian. (Neurophysiology Dept., Southampton General Hospital, Southampton, UK).

EEGs are often requested for elderly patients for various indications. This audit analysed EEG referrals in elderly patients and adherence to NICE guidelines. The purpose was to ascertain percentage of inappropriate referrals, and initiate strategies to tackle this.

Referral data for first-time EEGs in patients aged over 65 were collected retrospectively over one year (2011). EEG referral requests were assessed for compliance with NICE guidelines for epilepsy 2012 (CG137), which states that EEG should be performed to support a diagnosis of epilepsy in adults in whom clinical history suggests a seizure is likely to be epileptic in origin. EEGs should not be performed in probable syncope.

191 EEG referrals from various sources were made during the study-period. 122(64%) referrals adhered to guidelines for seizures. Of the remaining, 27(14%) were for syncope and 42(22%) for other indications. Only 6(3%) EEGs were concluded to have epileptiform activity. Interestingly, EEG findings suggested encephalitis/encephalopathy in 35(18%) patients.

Excluding EEGs for other indications, 18% of referrals did not adhere to guidelines. It is proposed that this information is disseminated among referring clinicians, and re-audited.

5. Improving Specificity of Stimulated Single Fibre EMG in The Diagnosis of Myasthenia in Children. J. Deeb[1,2] and M. Pitt[1]. ([1]Dept of Clinical Neurophysiology, Great Ormond Street Hospital for Children Foundation Trust, London, UK and [2]Dept of Clinical Neurophysiology, Queen's Hospital, Romford, Essex, UK).

The diagnosis of myasthenic syndromes in children presents considerable difficulty to the neurophysiologist. Stimulation single fibre EMG (StimSFEMG) most likely represents the most useful technique with a high sensitivity but low specificity.

Whereas in adults it is possible to screen out those cases likely to enter the differential diagnosis of myasthenia before testing, this is not possible in children. The integration of StimSFEMG into an investigative protocol may represent the best way forward to improve the specificity of the study.

The analysis includes EMG results of 303 children who had had StimSFEMG, needle EMG study and /or repetitive nerve stimulation. All included patients had a final clinical diagnosis. No child with a neurogenic EMG was later found to have myasthenia. EMG findings of a myopathy and a decrement on repetitive nerve stimulation were strongly associated with a diagnosis of myasthenia. An investigative protocol is described commencing with StimSFEMG, followed by needle EMG assessment of bulbar muscles, repetitive nerve stimulation and finally EMG of the limb muscles.

6. Differences in Excitability Properties of Low and High Threshold Motor Nerves: The Importance of The HCN Channel Current (Ih). J. Jeevananthan and C.E.G Moore. (Dept. of Neurophysiology, Queen Alexandra Hospital, Portsmouth and University of Portsmouth, UK).

The introduction of nerve excitability studies of peripheral nerves has increased our understanding of axonal function and questioned previously held beliefs. It was always assumed that the functions of myelinated motor and sensory nerves were identical. This was partly due to the similar properties demonstrated in supramaximal nerve conduction studies.

Giving a little thought to the matter, however, would suggest otherwise. Why do we get spontaneous sensory activity, in the way of parathesiae, without associated fasciculations after mild ischaemia. Previous studies have shown an increased strength duration time constant (persistent nodal sodium activation) in sensory nerve fibres. This was thought to be the mechanism to explain their lower threshold. The increased nerve excitation seen in neuromyotonia, however, was not accompanied by changes in SDTC.

Recent excitability studies have shown physiological differences between high and low threshold motor fibres and between motor and sensory fibres and suggest that the inwardly rectifying HCN channel current (Ih) has an important role in maintaining the resting membrane potential.

With ethics committee permission we studied 7 normal subjects using the Trond protocol targeting thresholds at 10, 20, 40 and 60%, in some cases utilising very strong hyperpolarising currents to further investigate the HCN current (1).

Our results confirm and expand on the findings of Trevellion (2) that inward rectification is greater in lower threshold motor fibres and that this remains true when using strong hyperpolarising currents.

These findings have important implications for the better understanding and treatment of diseases characterised by hyper and hypoexcitability of peripheral nerves with the HCN channel as a novel target for pharmaceutical intervention.

Refs : (1) Tomlinson et al : Muscle and Nerve 2009. (2) Trevellion : J Physiol.2010: 588