

**The British Society for Clinical Neurophysiology
(formerly the EEG Society)**

Abstracts (not peer reviewed) of the presentations to be made at the
Scientific Meeting, Bristol
2 July 2010

**1. The Neurophysiology of Myoclonus.
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Of all the involuntary movement disorders, myoclonus shows the greatest challenge to diagnosis and classification. The brief, shock-like muscle contractions (positive myoclonus) or inhibitions (negative myoclonus) take place in a fraction of a second. Hence, specification of the order of activation of different muscle groups and the duration of individual muscle activation is practically impossible by simple inspection of the motor phenomena. Surface EMG polygraphy provides information about the precise distribution of the muscle jerks, recruitment order of different muscles involved and the duration the EMG discharges; only extremely short duration bursts of EMG activity should be expected in epileptic myoclonus. It is often the case that standard EEG recordings fail to reveal the cortical accompaniments that precede the myoclonic jerks, in patients with epileptic myoclonus. The technique of jerk-locked EMG/EEG back-averaging allows identification of cortical transients occurring a few milliseconds before the onset of the EMG discharges. Finally, somatosensory evoked potentials and other more specialised techniques have a role to play in demonstrating cortical hyperexcitability (giant cortical evoked potentials and long loop reflexes).

In conclusion, neurophysiological examination of myoclonus can help confirm clinical diagnosis and provide important clues for the determination of the generator.

2. 'Sarcolemmal Electrotonus' – Effects of Membrane Polarisation on the Excitability of Human Muscle Fibres In-Vivo. R. Arunachalam, D. Allen and C.E.G. Moore. (Departments of Clinical Neurophysiology, Southampton and Portsmouth, UK).

Techniques capable of measuring components of muscle fibre membrane excitability, comparable to nerve excitability testing, show promise in understanding muscle pathophysiology [1,2]. Measurement of muscle fibre velocity recovery cycles (VRCs) after direct muscle stimulation has recently been developed. The early supernormality (Sn10) reflects membrane capacitance; the late supernormality (Sn100) reflects T-tubule function.

The effect of sub-threshold conditioning currents on muscle membrane function has not

been studied before. We investigated the effects of depolarising or hyperpolarising currents of 100ms duration presented immediately prior to the test stimulus, ranging in intensity from 0.5-5% of the test stimulus. Experiments were carried out in the tibialis anterior of four normal subjects, at rest and during the VRC.

The depolarising currents increase the relative refractory period (RRP) of the following test response. They also cause a reduction in the Sn10 seen in the VRC. In contrast, hyperpolarising currents decrease the RRP and increase the Sn10 in the VRC. This is similar to the effects reported in ischaemia and hyperkalaemia and therefore supports the hypothesis that the muscle sarcolemma is depolarised in these conditions [3].

The early supernormality is thought to be due to membrane capacitance change and is related to the depolarising after-potential (DAP) that follows a muscle action potential. We have demonstrated that changes in sarcolemmal excitability can be induced by sub-threshold currents and it is not essential to have an action potential. Our results suggest that the term DAP is misleading as the effects on the muscle membrane seems to be equivalent to a hyperpolarising event.

Refs: [1] Kiernan MC, Bostock. Brain 2000; 123: 2542-51. [2] Z'Graggen WJ, Bostock H. Muscle and Nerve 2009; 39: 616-26. [3] Z'Graggen WJ, Bostock H. Clin Neurophysiol 2010; 121: 874-81.

3. Clapham's Sign; Stretch Related Facial Muscle Contraction After Complete Denervation. L. Clapham, D. Allen, R Arunachalam and J. Cole. (Wessex Neurological Centre, Southampton and Poole Hospital, UK).

In complete facial nerve palsy, physiotherapists inspect the inside of the mouth for complications. During such examinations, it was noticed that, if the cheek was stretched, there followed ipsilateral contraction of the facial muscles of expression.

We report four cases; two with complete facial nerve section during surgical procedures, the others after trauma and infection. Facial muscle contraction could be elicited 7-8 weeks after nerve injury and persisted for months. It occurred a second or so after stretch, lasted for up to 60s and fatigued with repeated stretch. EMG (n=3) showed complete denervation. Stretch

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induced contraction was associated with an increase in fibrillation.

We postulate that this mass contraction is a direct effect of stretch on the preserved muscle. Though myocardium contracts in response to stretch without membrane depolarisation [1], here it was associated with increased fibrillations. Stretch activated channels have been described on somatic muscle membranes, with increased sensitivity after denervation, though their role is unclear [2]. Clapham's sign may reflect this increased sensitivity.

This novel sign suggests at least partial, continuing, function in the contractile apparatus of the denervated muscle.

Refs: [1] Stelzer and Moss, *J Gen Phys*, 2006, 128, 461-471 [2] Squecco, et al. *Neurol Res*. 2008, 30, 155-9.

4. Cross Correlation Analysis in Stimulated Single Fibre EMG. T. Tidswell¹, and B. Packham². (¹Royal Free Hospital and ²University College, London, UK).

Introduction: Stimulated single fibre EMG (SSFEMG) can rapidly assess neuromuscular junction (NMJ) instability, however data analysis is complex, time consuming, requires manual input thus raising possible operator bias.

Method: SSFEMG data, from a concentric facial EMG needle in orbicularis oculi with 10Hz monopolar needle stimulation of the facial nerve, was analysed by cross-correlation in two consecutive patient cohorts referred for possible myasthenia; 42 patients were followed up for clinical outcome, either definite (antibody positive), probable (treatment responders, antibody negative) or unlikely (no response, other diagnosis) myasthenia; in the next 57 patients cross-correlation was compared with MCD analysis.

Results: Outcome data was recorded in 36/42 of the first cohort: 17/17 with probable or definite myasthenia had abnormal correlation, 17/19 non-myasthenics had normal correlation. In the second cohort 38/38 with normal MCD had normal correlations, 18/19 with abnormal MCD had abnormal correlations.

Conclusion: Cross-correlation analysis of SSFEMG data is accurate in the detection of NMJ instability or stability when compared with either independent clinical outcome data or with existing

MCD analysis, with the advantage that this is an automated and rapid analysis technique.

5. EEG And VEP Abnormalities in a Patient With Heidenhain's Variant of Creutzfeldt-Jacob's Disease. T. Yermakova and A.A.A. Bajalan. (Department of Clinical Neurophysiology, Hull Royal Infirmary, Hull, UK).

The Heidenhain's variant is a rare variant of sporadic Creutzfeldt-Jacob's disease (sCJD). It initially affects the occipital cortex and presents with early, prominent visual manifestations. The affected patients often present to ophthalmologists.

We describe a new case with characteristic clinical features of Heidenhain's variant of sCJD before very obvious cognitive changes. The neurophysiological abnormalities were as follows:

The PRERGs and PRVEPs were not recordable. FVEPs showed normal early subcortical and abnormal cortical responses.

The EEG showed repetitive focal sharp theta 1.0 – 1.5 per second in the posterior regions with right sided emphasis.

The findings are consistent with previously published data from a small number of patients where visual symptoms corresponding to focal periodic sharp waves on EEG were documented.

We emphasize the importance of awareness of the condition amongst medical professionals and the role of Neurophysiological assessment in the diagnosis early in the course of the disease.

6. Is It Safe or Useful to Hyperventilate Patients With Mitochondrial Disease? L. Romaniuk¹, D.M. Turnbull² and R.G. Whittaker.^{1,2}. (¹Department of Clinical Neurophysiology, Royal Victoria Infirmary, Queen Victoria Road, Newcastle upon Tyne, UK and ²Mitochondrial Research Group, Institute of Ageing and Health, Newcastle University, Newcastle upon Tyne, UK).

Background: Mitochondrial diseases are a group of phenotypically variable conditions in which central nervous system disorders are common. Patients can develop encephalopathy, epilepsy and cognitive decline and are commonly referred for EEG. A sub-group of patients are also at risk of stroke-like episodes (SLE). These are transient focal neurological deficits, the

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pathophysiology of which is unclear. Hyperventilation (HV) carries an increased risk of stroke in patients with cerebrovascular disease. We wanted to assess whether this risk also applies to patients with mitochondrial disease.

Methods: 282 EEGs from 139 patients with mitochondrial disease between 1994 and 2009 were reviewed. The number of patients undergoing hyperventilation, changes in the EEG during HV, and risk of subsequent stroke-like episode was assessed.

Results: 51 of 139 patients performed HV. In 31 patients, no EEG changes were observed, in the remaining 20 an accentuation of previously seen abnormalities was observed. In no cases did diagnostically useful changes appear. 4 of the patients had a history of SLE; in three this preceded the HV by >1 year, and in one postdated HV by five years.

Conclusion: HV is probably safe in mitochondrial disease, but adds no useful information to the EEG.

7. Mechanisms of Double Fasciculations in ALS. K.R. Mills. (King's College, London, UK).

Fasciculation potentials (FPs) were recorded from 63 muscles of 28 patients with definite amyotrophic lateral sclerosis; at a single needle site, up to 15 separable FPs could be recognized. Double FPs were defined as any second discharge occurring in the 100ms following the triggering FP and were classified as DSFPs where the second FP was the same as the first and DDFPs where the second discharge was different. DDFPs occurred in 48.1% of the 451 FPs identified, whereas DSFPs occurred in 15.4%. The incidence of DSFPs was significantly higher in weak muscles showing neurogenic change on EMG than in strong muscles with no other EMG abnormality. Pooled time interval histograms of DDFPs and DSFPs were constructed. Two peaks of discharge interval were found with DSFPs; histograms of DDFP intervals were flat implying no interaction between different FPs. DSFPs occurred in all muscles at intervals of 4-10ms; in tibialis anterior, a peak at 30-50ms was also seen. Of the 48 DSFPs, the waveforms of the two FPs in this early peak were identical in 38 (79.1%). In the peak at 30-50ms, it was noted that while the triggering FP could have a variable waveform, the second discharge was identical on each occasion. Axonal excitability studies have shown a period of super-excitability

in the 3-20ms following an action potential; it is therefore suggested that DSFPs with short intervals arise during the phase and furthermore, they arise from the same patch of abnormal membrane. DSFPs occurring at longer latency probably arise as F-responses; the triggering FP can arise from multiple distal triggering sites, but since the second FP is identical and has therefore been propagated through the axonal arborisation leading to fibre activation in the same order, it must have arisen proximal to branch points.

8. Evaluation of Motor Evoked Potentials (By Transcranial Magnetic Stimulation-TMS) As Marker of Disease Severity And Progression in Primary Progressive Multiple Sclerosis. B. Patil, N. Kane, D. Cottrell, P. Walsh and S. Butler (Frenchay Hospital, Bristol. UK).

Introduction: Primary Progressive Multiple Sclerosis (PPMS) is relentlessly progressive producing significant disability. Currently there is no effective disease modifying agent in PPMS. Various clinical scores have been used to evaluate the disease severity and progression. These scales have limitations and there is need for more objective markers of disease progression that may help evaluate various impending therapeutic interventions.

Methods: Patients with a diagnosis of PPMS, without contraindication for TMS, were recruited from a larger ongoing PPMS trial at Frenchay Hospital. Patients were assessed with clinical scores and multi-modal evoked potentials on recruitment, at 6 months, 1 and 2 years thereafter. TMS was done using a circular coil (Magstim 200 & Synergy EMG machine). Central Motor Conduction Times (CMCT) was calculated using the F-wave method.

Results: Motor Evoked Potentials (MEP) data of 22 patients with PPMS and 5 controls were analysed. At a particular given point, the clinical scores and MEP scores did not have strong correlation [(EDSS vs. Total MEP scores with Spearman's rho (0.34, p=0.11), and Average 9 Hole Peg test with Upper Limb MEP scores-Spearman's rho (0.31, p= 0.18)]. When the Extended Disability Scale (EDSS) and total MEP scores were followed over time, the MEP scores showed significant deterioration (Wilcoxon signed rank test W=55, p=0.0574) but the EDSS did not. There is also some suggestion that initial high MEP scores may be predictive of worsening of clinical scores (9 Hole Peg test and 25 ft Walk

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test) over a period of time but the numbers are too small for this to be statistically significant.

Conclusion: The MEP measurement does not correlate with disease severity of PPMS as measured by clinical scales. However, MEP's may be a useful tool in monitoring the disease progression over time as there are indications that they are probably more sensitive to change than clinical scales.

9. A Technical Evaluation of The Neurometrix 'Advance' Handheld EMG Machine. A. Kumar, J. Martinez and J. D.P. Bland. (Department of Clinical Neurophysiology, East Kent Hospitals NHS Trust, UK).

The 'Advance' EMG machine, is a battery powered, handheld device which relies upon internet access and a remote fileserver for its data storage and reporting facilities and which is markedly more compact than portable versions of conventional EMG machines. This study reports the ability of this device to produce comparable nerve conduction recordings to those obtained from a full-size EMG machine. A variety of sensory and motor nerve action potentials were recorded from staff members using the same input signals by transferring the electrode connections between the Advance and a Dantec Keypoint, without disturbing the electrode placements. Recordings were made in a random order, with the same stimulus intensity set on each machine. Automatic cursor placements made by the machines were accepted except when obviously disturbed by artefact. For sensory and mixed nerve potentials the differences between the two machines were (mean/SD) 0.04/0.1 msec for onset latency, 0.01/0.07 msec for negative peak latency and 1.54/2.78 microV for negative peak/subsequent positive peak amplitude. The Advance recorded consistently lower SNAP amplitudes, averaging 5% smaller than those obtained from the Keypoint. For motor potentials the difference in onset latency was 0.035/0.14 msec and the difference in baseline to peak MAP amplitude was 0.44/0.41mV.