BSCN abstracts October 2010

1. **Serial Imaging and Structure-Function Correlates of Abnormal Fundus Autofluorescence in Retinitis Pigmentosa. A. G. Robson,[1,2] A. Tufail,[1,2] F. Fitzke,[2] A. C. Bird,[2] A. T. Moore,[1,2] G. E. Holder[1,2] and A. R. Webster.[1,2] ([1]Moorfields Eye Hospital, London, UK and [2] UCL Institute of Ophthalmology, University College, London, UK).**

Purpose: To assess the functional and structural significance of parafoveal rings of high density fundus autofluorescence (AF) in patients with retinitis pigmentosa (RP) and to monitor changes over time.

Methods: Fifty two patients with RP were ascertained who had a parafoveal ring of high density AF and a visual acuity of 6/9 or better. International-standard pattern ERGs (PERG) were performed. AF imaging was repeated after 1-9 years (N=30). Optical coherence tomography (OCT) was used to measure the lateral extent of preserved outer retina according to the photoreceptor IS/OS band (N=35). Fine matrix mapping (FMM) was used to assess rod and cone sensitivity.

Results: Progressive ring radius reduction was detected in 57% and varied between 3 and 40% at a mean rate of between 0.8 and 15.8% per year. The lateral extent of the IS/OS lamina correlated with the ring width along the same OCT scan plane (R= 0.97, $p<0.005$, N=35) and with PERG P50 (R=0.72, $P<0.005$, N=34). FMM revealed progressive photopic visual field loss associated with ring constriction; scotopic losses were more severe and widespread.

Conclusions: Rings of increased AF surround areas of preserved outer retinal structure. The rate of ring constriction varies and is associated with progressive visual field loss.

2. **Modafinil for MS-Fatigue: A Pupillographic Study. G. Niepel[1], C.Constantinou[2] and E. Szabadi[2]. ([1]St George's Hospital, London, UK and [2]Queens Medical Centre, Nottingham, UK).**

Fatigue is a common and major symptom in multiple sclerosis (MS). It is generally agreed that multidisciplinary approaches to MS-fatigue management are most appropriate and also that convincing evidence for specific pharmacological agents does not exist.

Studies in MS-fatigue traditionally rely on self-reported measures of fatigue severity. A drug that has recently been evaluated is the wakefulness-promoting drug, modafinil. We assessed the effectiveness of modafinil, in MS patients with and without fatigue by assessing its effect on a number of objective measures of alertness and vigilance, including the Pupillographic Sleepiness Test (PST) and Critical Flicker Fusion Frequency (CFFF).

Compared to placebo, a significant response to modafinil was seen in MS fatigue sufferers in two measures of alertness on pupillography. Significant improvements were also noted in non-pupillary measures of alertness including; CFFF threshold, and the pause time on speech analysis.

3. **Healthcare Workers' Perceptions of Non-Epileptic Seizures Compared to Epilepsy. K. Whitehead, C. Worsley. M. Reuber and R. Kandler. (Sheffield Teaching Hospitals Foundation Trust, Sheffield, UK).**

Background : Many patients present to neurophysiology departments with epilepsy or non-epileptic seizures (NES). Anecdotal reports suggest that some patients with NES perceive healthcare staff as hostile or unsympathetic. We have therefore (i) investigated the attitudes of Neurosciences staff to patients with epilepsy and NES and (ii) compared the attitudes of neurosciences staff to the attitudes of A&E staff

Methods : 31 Neurosciences and 30 A&E staff were recruited. Participants were given two copies of an adapted version of the Illness Perception Questionnaire revised (IPQ-R) to complete separately for epilepsy & NES.

Results : Both Neurosciences and A&E healthcare workers thought epilepsy was more chronic than NES ($p<0.005$; Wilcoxon) and reported a greater understanding of epilepsy than NES ($p<0.001$; Wilcoxon). Neurosciences healthcare staff believed that patients with NES had greater personal control over their condition than patients with epilepsy ($p<0.001$; Wilcoxon).

Neurosciences staff self-reported a greater understanding of both conditions than A&E staff ($p<0.01$; Mann-Whitney). NES was thought to be more chronic by neurosciences than A&E staff ($p<0.005$; Mann-Whitney).

Conclusion : Our adaptation of the IPQ-R revealed some of the reasons why patients with NES may experience healthcare staff as unsympathetic. Qualitative methodologies may be more suitable to reveal the reasons for the differences in perception of epilepsy and NES.

4. **Using Muscle Fibre Conduction Velocity to Investigate Fatigue in Mitochondrial Disease. M. Bolland[1], M. Baker[1], E. Williams[1], S. Baker[1], D.M.Turnbull[2] and R.G. Whittaker[1,2]. (Institutes of [1]Neuroscience and [2]Ageing and Health, Newcastle University, Newcastle upon Tyne, UK).** Mitochondrial diseases are a variable group of genetic disorders that result from impairment in the ability of mitochondria to produce ATP through aerobic respiration. Fatigue is a major symptom common to all types. The origin of this fatigue remains unknown and its clinical evaluation is based upon subjective assessment of the patient's experience. A

technique to reliably assess fatigue on a physiological basis would expand our understanding and facilitate the development of treatments for this highly disabling symptom.

Surface-Electromyography (EMG) can be used to measure the Muscle Fibre Conduction Velocity (MFCV) - a marker of physiological fatigue. In controls this declines with fatiguing exercise. We used measurements of MFCV to investigate the mechanism of fatigue in mitochondrial disease.

Surface-EMG signals were recorded from the biceps brachii using a custom-built electrode with 8 linearly arranged contacts. 5 patients with mitochondrial disease and 7 controls performed sustained isometric contractions at 50% Maximal Voluntary Contraction for 5 minutes. EMG data was analysed using cross-correlation and coherence analysis.

The mean resting MFCV was similar in both groups. The mean final MFCV was 5.4±0.4 m/s (mean±SEM) for patients and 3.6±0.7m/s for controls. The rate of decline was considerably less among patients with a mean drop of 9.5±1.9% compared to 35±9.2% in controls ($P \leq 0.05$). This suggests a different mechanism of fatigue in mitochondrial disease to that seen in controls, with changes in the excitability of the muscle fibre membrane playing a minor role. Further research is required to establish the biochemical basis of this finding.

5. Effect of Arterial Baroreflex Modulation on Postural Sway. A. Nicotra[1], M. Bissa[2], G. DeBarbieri[2], A. Bharadwaj[3] and L. Bernardi[2,4]. ([1]West London Neurosciences Centre, Charing Cross Hospital, London, UK, [2]Department of Motor Sciences, University of Pavia, Italy, [3]Defence Institute of Physiology and Allied Sciences (DIPAS), New Delhi, India, and [4]Department of Internal Medicine, University of Pavia and IRCCS S.Matteo, Pavia, Italy).

A direct effect of the autonomic nervous system (ANS) on posture has never been demonstrated. To test this hypothesis, we studied thirteen healthy subjects (age 26±5) on a force platform with and without stimulation of arterial (carotid) baroreceptors by sinusoidal neck suction (pure motion-independent autonomic stimulus) at different frequencies (0.05, 0.10, 0.125, 0.15, 0.175, 0.20, 0.30Hz) during controlled breathing (15br/min, 0.25Hz).

Spectral analysis showed that neck suction significantly ($p<0.05$) increased oscillations synchronous in RR interval (from 0.10 to 0.20Hz), systolic and diastolic blood pressure (from 0.05 to 0.15Hz), and postural sway (from 0.10 to 0.30Hz in both antero-posterior and medio-lateral planes). Changes were greater in the left than in the right foot.

Our results indicate that postural sway is modulated by the ANS and is influenced by phasic stimulation of the arterial baroreceptors.

6. Neurophysiological Assistance to Neurosurgeons in Conus Region Surgery. A. Forster, J. Lumsden , E.K. Labram and P. Bhatt. (Aberdeen Royal infirmary, Aberdeen, UK).

Conus tumours, spina bifida, and tethered cords can provide a severe challenge to the neurosurgeon, and a range of logically-applied neurophysiological techniques can prove of considerable value in identifying structures. Cord midline may need to be defined in distorted lower cord to guide the surgical incision (midline myelotomy to minimize damage) utilising cord stimulation with a bipolar probe and recording antidromic SAP and motor evoked responses (1) and roots localised either by mechanically evoked EMG or stimulation via the probe.

Monitoring can be performed for long tracts, via posterior tibial SEPs and transcranially evoked motor potentials, and multichannel free running EMG can be used for detecting root, nerve, and corticospinal tract irritation. Bulbocavernosus reflex type responses may also be visualised or intentionally monitored (2).

We have evolved our methods for this area of intraoperative neurophysiology over 10 cases. Athough numbers are small the techniques have proved very useful in minimizing complications, principally by guiding myelotomy localization and giving warning of impending root injury. Clear localization of roots is usually readily obtained, with stimulated and free running EMG emerging as the most useful toosl.

Refs: (1) Quinones-Hinojosa A, Gulati M, Lyon R, Gupta N, Yingling C. Neurosurgery. 2002 51(5):1199-1206 ; (2) Deletis V, Vodusek DB: Neurosurgery1997 40:88-93.

7. Water, Water Everywhere! S. Murphy[1], R. Lonergan[1], C. Albertyn[1], P. O'Sullivan[1], R.P. Murphy[1] and M.D. Alexander[2] (Departments of Clinical Neurophysiology[2] and Neurology[1], The Adelaide and Meath Hospital Dublin, Incorporating the National Children's Hospital, Dublin, Ireland).

Case History: A 29 year old woman, with previous symptomatic idiopathic hypokalaemia, presented in acute renal failure. She had two consecutive convulsions, preceded by visual disturbance. CT brain was non-diagnostic but EEG demonstrated a clear disturbance of occipital cortical function. On questioning, the patient admitted to the ingestion of up to18 non-steroidal anti-inflammatory drugs (NSAIDs) daily for eight years for a chronic daily headache. Based on history and EEG findings, a reversible posterior leucoencephalopathy was postulated.

The patient developed further seizures, hypertension and spastic paraparesis. EEG in ITU demonstrated an alpha coma with burst suppression pattern. An emergency MRI of brain (T2 and FLAIR sequences) demonstrated diffuse

high signal (vasogenic oedema) but with a parieto-occipital predominance. Prompt optimisation of blood pressure and intracranial pressure was undertaken and she improved clinically and radiologically over the next four weeks.

Discussion: This is the first report of a NSAID-induced reversible leucoencephalopathy in an adult. This case illustrates potential neurological complications of NSAID overuse, the importance of a detailed drug history and how EEG studies can assist diagnostic localisation before neuro-imaging. In addition, recovery after electrographic alpha coma is unusual and highlights the potential reversibility and importance of speedy identification of posterior (pan)leucoencephalopathy.