

Selected abstracts from the RCPE Neurology Symposium 2009

Held on 27 November 2009 at the Royal College of Physicians of Edinburgh

EFFICIENT NEUROLOGICAL ASSESSMENT

Dr Geraint Fuller, Consultant Neurologist, Gloucestershire Royal NHS Trust

Why should we be concerned about the efficiency of neurological assessment? The phrase hints at the worst type of management, all clipboards and stopwatches. However, with a limited number of neurologists in the UK, limited time and a vast number of patients with neurological disease, anything that increases efficiency is attractive.

The clinical encounter, the consultation, is the core element of clinical neurology. How can we improve the efficiency of the consultation? There are two elements within efficiency: what you do and how quickly you do it. What are we trying to do in the clinical consultation? This has been the subject of considerable research and thought within general practice. A significant part of the GP consultation is directed at determining the patient's agenda – recognising that unless you know what issue the patient wants addressed you will not be able to address it. The neurologist, however, must consider not only the patient's agenda but that of the GP, as well as providing their own perspective on a clinical problem. If you are addressing the wrong agenda, then no matter how well you do it your consultation will fail.

While there is no compelling evidence to support one approach to clinical assessment as the most efficient, there are a number of strategies to make the consultation last longer. Efficiency would improve if these strategies were avoided.

How best to elicit a history? The traditional order takes the current history out of context with the past medical history, which may lead to misinterpretation. To make the consultation last longer you can subject the patient to intense cross-questioning, seek constant clarification or seek second-hand medical opinions from the patient. Why not avoid doing this? Think about how you use examination, the smallest part of the consultation. Use the examination to assess clinical hypotheses. You are more likely to see things if you are looking for them.

When you enter the final phase of the consultation there are other opportunities to spin it out, such as telling the patient your plans for investigation and treatment before you have thought them through so they are incoherent and

illogical, or taking additional history as different treatment options cross your mind. Alternatively you could think through the issues, address the patient's and GP's agendas and then aim to answer the questions you would ask if you were the patient – you will save the patient from having to think of them and save yourself time.

COULD IT BE MITOCHONDRIAL?

Professor Patrick Chinnery, Professor of Neurogenetics, Newcastle University

The first pathogenic mitochondrial DNA (mtDNA) mutations were identified in patients with 'classical' mitochondrial disease syndromes more than 20 years ago, including mitochondrial encephalomyopathy with lactic acidosis and stroke-like episodes (MELAS), chronic progressive external ophthalmoplegia (CPEO) and Leber's hereditary optic neuropathy (LHON). However, since that time the number of different genetic defects found in patients with mitochondrial disorders has grown exponentially. This list includes not only primary mtDNA mutations, but also a wide range of nuclear gene defects which cause a secondary defect of mtDNA. The clinical spectrum of these disorders has expanded in parallel, and even in 2009 the phenotype has grown to include features not previously considered to be 'mitochondrial'. With ~1 in 200 of the population carrying a mtDNA defect, these are diseases that cannot be ignored in routine clinical practice, but how can the neurologist keep abreast of this rapidly evolving clinical sub-specialty? This presentation will provide an update on the clinical neurology of mitochondrial disorders, illustrated with practical examples of the approach to investigation, and will conclude with a 'rule of thumb' helpful in everyday neurological practice.

TAKING NEUROLOGY TO THE PEOPLE

Dr Christopher Allen, Consultant Neurologist, Addenbrooke's Hospital, Cambridge

Modern neurologists are not only interested in rare diseases but also in delivering an effective service for the solution of neurological clinical problems, both in hospital and in the wider community. In his talk Dr Allen describes the advantages of seeing neurology outpatients closer to their home in less intimidating circumstances than the hustle and bustle of the hospital clinic, while keeping the activity within the hospital's control and not distorting

the case mix of the clinic. Twenty-five per cent of patients seen by neurologists have unusual conditions that require advanced neurological expertise to identify them. Seventy-five per cent of outpatient consultations concern common problems that require confident and effective management, which is often best done outside the hospital environment. The quality of the consultation as perceived by the patient is vital in effectively managing the 30% of patients whose presenting symptoms are not caused by disease. Dr Allen presents some brief aphorisms of 'common neurosense' used to streamline the management of neurological problems, and briefly describes what happens in an effective neurological consultation. He also discusses the difficulties of squaring local innovations, such as consultants seeing patient nearer their home, with central NHS initiatives about patient care.

CHRONIC DAILY HEADACHE

Dr Manjit Matharu, Senior Lecturer and Honorary Consultant Neurologist, Institute of Neurology and the National Hospital for Neurology and Neurosurgery, London

Chronic daily headache (CDH) refers to very frequent headaches (>15 days per month) for more than three months. An estimated 4–5% adults worldwide experience CDH. The incessant nature of CDH makes them among the most disabling headaches. The CDH group can be categorised into primary and secondary varieties. Secondary CDH has an identifiable underlying cause, such as abortive headache medication overuse, disorders of intracranial pressure, head trauma and vascular disorders. The major causes of primary CDH include chronic migraine, chronic tension-type headache, new daily persistent headache and hemicrania continua. These headache syndromes are often complicated by the overuse of analgesic leading to superimposed medication overuse headache. Various factors can increase the risk of developing CDH, including anxiety and depression, sleep disorders and obesity.

Primary headache syndromes were previously considered to be vascular in origin, but this hypothesis is no longer tenable as it is not supported by blood flow studies. The current concept is that primary headache disorders have a neurovascular basis. This hypothesis posits that primary headaches occur as a result of a disorder of the endogenous pain-modulating systems. Several supraspinal structures such as the periaqueductal gray and hypothalamus are responsible for the inhibition and facilitation of pain pathways. Some of these structures, besides participating in nociceptive control, modulate other sensory modalities and can alter brain blood flow. The abnormal function of these supraspinal structures – either by activation of descending systems that facilitate processing of pain signals by trigeminocervical

neurons, or suppression of descending pathways that inhibit such processing of pain signals or a combination of systems' dysfunctions – results in central hyperalgesia and an augmentation of the central perception of pain.

The management of CDH involves performing investigations such as neuroimaging and lumbar puncture when appropriate to exclude symptomatic causes. For the primary forms of CDH, it is important to diagnose the CDH subtype to enable appropriate treatment. Patients with strictly unilateral headaches warrant a trial of indometacin to exclude hemicrania continua. If a patient is overusing analgesics, it is imperative that this is addressed to exclude medication overuse headache (MOH). Most patients need trials of headache preventative medications; the options include tricyclic antidepressants, beta-blockers, serotonergic antagonists (pizotifen, nethersergide), neuro-modulators (sodium valproate, topiramate, gabapentin) and calcium channel antagonists. Recent data suggest that botulinum toxin and occipital nerve stimulation may be effective in the management of chronic migraine.

ISCHAEMIC STROKE – PREVENTION IS BETTER THAN CURE

Professor Graeme J Hankey, Consultant Neurologist and Head of Stroke Unit, Royal Perth Hospital, and Clinical Professor, School of Medicine and Pharmacology, University of Western Australia

Each year in southeast Scotland, where there are about one million residents, 2,000 new (incident) cases of ischaemic stroke are added to a pool of about 12,000 prevalent survivors of ischaemic stroke.

Without treatment, about 1,300 (65%) of the 2,000 new cases would ultimately die or remain dependent on someone for help in everyday activities within the next year. Appropriate and optimal acute treatment of all 2,000 incident cases of ischaemic stroke could save up to 115 (9%) of the 1,300 from death and dependency; thrombolysis could save up to 12, aspirin 20, organised care in a stroke unit 69 and decompressive surgery 13. Without treatment, about 600 (5%) of the 12,000 prevalent cases of ischaemic stroke would experience a recurrent stroke in the next year. Appropriate and optimal prophylactic treatment of all 12,000 prevalent cases of ischaemic stroke could save a substantial number of recurrent strokes; early carotid endarterectomy could save about 9, aspirin 60, aspirin and extended-release dipyridamole an additional 61, oral anticoagulation 77, blood pressure lowering 184, cholesterol lowering 58 and smoking cessation about 60.

Without treatment, about 1,400 of the 988,000 stroke-free population would experience a first-ever stroke. Appropriate and optimal screening, detection and prophylactic treatment of all high-risk individuals could

save a substantial number of first-ever strokes; blood pressure-lowering about 266, cholesterol-lowering 99 and smoking cessation about 236.

'An ounce of prevention is worth a pound of cure' (Benjamin Franklin).

IS THIS A NEUROPATHY, AND WHAT CAUSED IT?

Dr James Overell, Consultant Neurologist, Institute of Neurological Sciences, Southern General Hospital, Glasgow

Long lists of causes of peripheral neuropathy make peripheral nerve disease a dry and uninspiring subject. A simple clinical classification scheme based on the answers to six questions (What systems are involved? What is the distribution of weakness? What is the nature of the sensory involvement? Is there any evidence of upper motor neurone involvement? What is the temporal evolution? Is there any evidence for a hereditary neuropathy?) enables the clinician to recognise characteristic patterns and investigate relevant subgroups appropriately.

Length-dependant mixed sensori-motor axonal neuropathies are common, and usually only mildly disabling. They are investigated with a standard series of tests including glucose studies, vitamin B12 (with metabolites) and electrophoresis. Chronic inflammatory demyelinating neuropathies usually cause proximal or upper limb weakness, and are often associated with raised cerebrospinal fluid protein levels. They generally respond to immunotherapies such as intravenous immunoglobulin or steroids. A specific subtype known as distal-acquired demyelinating symmetric neuropathy causes distal weakness and sensory ataxia due to distal demyelination, and is often associated with IgM paraproteinaemia and antibodies to myelin-associated glycoprotein. Recent studies suggest worthwhile clinical responses to rituximab. Vasculitic neuropathy usually presents as a subacute, painful multifocal mononeuropathy, and is treated with immunosuppressants. Subacute sensory neuronopathy (or dorsal root ganglionopathy) leads to disabling sensory ataxia, and is a characteristic paraneoplastic syndrome, often secondary to small cell lung cancer. It normally precedes a diagnosis of cancer.

The use of the simple clinical classification scheme presented leads to early recognition of disorders requiring further investigation and the identification of treatable subgroups.

DOES THIS PATIENT HAVE DEMENTIA?

Professor Adam Zeman, Professor of Cognitive and Behavioural Neurology, Peninsula Medical School, Exeter

The diagnosis of dementia is a demanding task, which can fall either to physicians or psychiatrists. Psychiatrists would not attempt this in 30 minutes as a rule – physicians should also try to avoid doing so (but sometimes must)! I will first define dementia, emphasising that it is a broad category, calling for further specification; outline an approach to history-taking based on an understanding of the subcomponents of cognition, the importance of screening for depression, and the value of obtaining history from an informant; discuss the Addenbrooke's Cognitive Examination as a relatively succinct but highly informative instrument; touch on the importance, in some cases, of general medical, neurological and mental state examination; and run through the standard, and some more advanced, approaches to investigation. Given that the consultation will conclude with a few minutes of explanation and discussion, the assessment process generally requires 45 minutes to one hour. Try not to rush.

IMAGING THE SPINE

Dr David Summers, Consultant Neuroradiologist, Western General Hospital, Edinburgh

Since the development of clinical computed tomography in the 1970s and subsequently magnetic resonance (MR) imaging in the early 1980s, imaging of the brain and spine has been one of the major areas of MR practice. Despite the considerable technological developments since then, the cervical spine remains a challenging area to image, as a consequence of the physical characteristics of the neck and the small size of the structures concerned. There is also relatively poor correlation between the near ubiquitous imaging features of ageing, which include disc signal changes, facet joint arthropathy, chronic disc protrusions and osteophyte formation, and clinical features of cervical radiculopathy or myelopathy. Other incidental findings at the skull base, deep facial spaces or thyroid may also result in unnecessary further investigation. Examples of relevant imaging findings and appropriate strategies for access and use of cervical imaging will be discussed.

SURGICAL MANAGEMENT OF CERVICAL DEGENERATIVE DISORDERS: EVIDENCE-BASED PRACTICE

Mr Nicholas Todd MD FRCS, Consultant Neurosurgeon and Spinal Surgeon, Newcastle

There are three clinical syndromes:

- Mechanical neck pain with pain referred into the paraspinal muscles, shoulders and/or upper arms;
- Cervical radiculopathy, typically caused by compression of a single cervical nerve root. There is usually radicular pain. There may be dermatomal numbness, myotomal weakness and/or change of a reflex;
- Cervical myelopathy with numb clumsy hands, gait and balance problems.

Surgical decision-making

The history and signs need to fit very closely with the imaging. The indication for surgery is almost always the degree of functional impairment that the patient has. We need to consider the approach (anterior, posterior or lateral, or a combination). We need to consider whether the spine needs to be decompressed and/or stabilised. If a fusion is to be performed, we need to consider whether to use autologous bone or allografts. Disc replacement is an option.

Axial neck pain

One of the difficulties in selecting patients for surgery where there is only axial nerve pain is that there are a considerable number of potential pain generators. These include degenerate discs, facet joints and musculo-ligamentous complexes. A pain generator could be multifactorial at any one level and/or at multiple levels. The initial role of management is to exclude more serious alternative diagnoses such as infection, malignancy or an inflammatory disorder. Cervical epidurals have a high incidence of complications with no benefit. Facet joint blocks probably offer no long-term benefits. Radiofrequency lesioning offers short-term benefit only.

There is no evidence base for surgery for axial neck pain. If it is felt that a single level of degenerative disc disease is probably a pain generator, about 62% of patients improve in the short term.¹ Disc replacement has become a common procedure for axial neck pain. There is no evidence base for its use.

Radiculopathy

The natural history of a cervical radiculopathy is good, with pain settling in six to eight weeks in the majority of patients. There are few good controlled studies of treatments.² The indications for surgical decompression are persistent pain (some would say six to eight weeks, others up to 12 weeks of radicular pain associated with significant functional impairment).

Progressive motor weakness is an indication for urgent surgery. A number of procedures, including anterior discectomy alone, anterior discectomy with bone grafting and plates, or allograft and plates, or disc replacement, and/or posterior foramenotomy, are all associated with an 85–90% likelihood of improvement in radicular pain. There appears to be no great difference between the various techniques, but there are no randomised controlled trials.³

Myelopathy

The natural history of a cervical myelopathy is debated. It was initially thought that the natural history was relatively benign, but more recent studies have suggested that in the medium term there is usually progressive neurological deterioration.^{4,5}

Where there are modest symptoms with minor functional deficits and where the imaging demonstrates a modest degree of spinal cord compression with no myelomalacia, watching and waiting may be reasonable. The compressive abnormalities usually lie anteriorly and therefore anterior approaches are usually favoured. These include single or multilevel discectomies or corpectomy with anterior reconstruction.

Where the pathology lies posteriorly (for example, a synovial cyst or ligamentous hypertrophy), laminectomies can be performed to decompress from posteriorly. Laminectomies will also decompress where there is multilevel anterior compression provided there is a preserved lordosis which allows the spinal cord to 'come off' the anterior compressive osteophyte(s).

In most patients surgical decompression halts neurological deterioration (assuming there is no complication of surgery). There may be improvement in the patient's condition, but often this is not functional. For example, the patient may say their hands are less numb but they are not functionally better. Improvement is less likely where the myelopathy is either rapidly progressive or severe, or the compression is at multiple levels, or where there is myelomalacia prior to surgery.

References

- 1 White AA, Southwick WO, Deponte RJ et al. Relief of pain by anterior cervical-spine fusion for spondylosis. *J Bone Joint Surg Am* 1973; 55:525–34.
- 2 Wainner RS, Gill H. Diagnosis and nonoperative management of cervical radiculopathy. *J Orthop Sports Phys Ther* 2000; 30:728–44.
- 3 Herkowitz HN, Kurz LT, Overholt DP. Surgical management of cervical soft disc herniation. A comparison between the anterior and posterior approach. *Spine* 1990; 15:1026–30.
- 4 Symon L, Lavender P. The surgical treatment of cervical spondylotic myelopathy. *Neurology* 1967; 17:117–27.
- 5 Matsunaga S, Sakou T, Taketomi E et al. The natural course of myelopathy caused by ossification of the posterior longitudinal ligament in the cervical spine. *Clin Orthop Relat Res* 1994; 305:168–77.

SURGERY FOR DEGENERATIVE CERVICAL SPINE DISEASE: DON'T OPERATE

Dr Richard Davenport, Consultant Neurologist, Western General Hospital, Edinburgh

There are more than 4,000 operations performed on the cervical spine in the UK annually, and much debate among surgeons about which operation is the most suitable, and which expensive form of artificial disc/cage/insert should be used. Such enthusiasm will of course be underpinned by persuasive evidence from large, well-conducted randomised controlled trials comparing surgery with conservative management. I shall review what we know about the epidemiology of cervical spondylotic myeloradiculopathy, including the natural course of the condition, before presenting the wealth of evidence regarding surgical intervention, including the most up-to-date systematic review, which includes all the available data.