

THE 11th NATIONAL CONFERENCE

Multidisciplinary care in Parkinson's disease and parkinsonism

from science to practice

Conference Report



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multidisciplinary
conference
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at the
Royal College
of Physicians,
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Introduction

Parkinson's disease (PD) is increasingly on the agenda of clinicians, healthcare commissioners and the media. However, some less common parkinsonian syndromes – such as progressive supranuclear palsy (PSP), multiple system atrophy (MSA) and corticobasal degeneration (CBD) – do not always receive the attention they deserve. To raise awareness, the British Geriatrics Society Parkinson's Disease Special Interest Group's 11th national conference on multidisciplinary care in Parkinson's disease and Parkinsonism from science to practice focused predominantly on these devastating diseases.

Delegates to the meeting, held on 11 July 2006 at the Royal College of Physicians in London, discussed the genetic and neuropathological basis of the less common parkinsonian syndromes as well as the clinical pointers that aid differential diagnosis. The multidisciplinary panel of eminent speakers also considered management issues, such as:

- Treating autonomic dysfunction
- The challenging cognitive, behavioural and psychological changes
- Speech and swallowing problems.

While the meeting focused on the less common parkinsonian syndromes, much of the discussion was relevant to PD.

PSP, MSA and the other less common parkinsonian syndromes have a devastating impact on quality of life for patients and their families. Indeed, carers can find themselves enduring a 'living bereavement' as the disease ravages the patient's personality, cognition and bodily functions. Fortunately, delegates heard that a multidisciplinary approach and technological advances can help more patients to live in the community for longer. A nurse specialist also discussed how a dedicated 24-hour helpline, run by the PSP-Europe Association advises and supports patients, carers and healthcare professionals, many of whom know relatively little about these disabling conditions.

PD services usually manage patients with PSP, MSA and the other less common parkinsonian syndromes. The National Service Framework (NSF) for long-term conditions approached its first birthday and the National Institute for Health and Clinical Excellence (NICE) published its recommendations for PD around the time of the conference. To mark the anniversary, the Department of Health's National Director for Primary Care discussed some ways in which the NHS can translate these recommendations into improved patient care, which may mean questioning current practice. This report brings you some of the meeting's highlights.

Clinical and pathological diversity in the rarer parkinsonian syndromes

Dr David Williams, Clinical Research Assistant, Queen Square Brain Bank for Neurological Diseases, Institute of Neurology, University College London, London.

“You don’t need to try hard to get the diagnosis of PD right most of the time,” said **Dr David Williams**. “Ninety percent of bradykinetic, rigid patients have PD.” The remainder usually suffer from one of the less common parkinsonian syndromes. These syndromes often show markedly different prognoses from, and require different management strategies to, PD. Dr Williams therefore emphasised the importance of making the correct diagnosis.

James Parkinson described the disease that bears his name in 1817. PSP was not characterised as a separate disease to PD until 1963, when Drs Richardson, Steele and Olszewski described a condition they called ‘heterogeneous system degeneration’. Dr Williams commented that the first case had all the hallmarks of the disease, including: unsteady gait, slow thinking and difficulty with vision. The patient died six years after diagnosis, compared to a mean duration of 15 years with PD.

Since then, neurologists have characterised the hallmarks of PSP (Box 1). Falls among PSP patients, in particular, pose a problem for the multidisciplinary team, the patient and the NHS more generally. Dr Williams noted that 98% of patients with PSP fall at some point and 29% sustain a fracture. In contrast, 73% of PD patients fall at some point and 17% sustain a fracture. In MSA, the rates are 78% and 11% respectively. Fifty-seven percent of patients with PSP fall within two years of diagnosis, compared to 6% of those with PD. The pattern of fractures also differs between PSP and PD. Multiple fractures, especially down the midline, are more common in people with PSP than PD or controls. “PSP patients seem to lack the reflex to put their hand forward to break their fall. As a result, broken wrists are less common in PSP patients than in those with PD or controls,” Dr Williams told delegates.

Supranuclear gaze palsy is another hallmark of PSP that is uncommon among people with PD. The patient’s gaze becomes hypometric (the patient ‘undershoots’ when following a target) and then slows further. PSP initially hinders down gaze, although later upward gaze becomes impaired. As the disease progresses, patients show:

- Increased square wave jerks – they take their eye off a moving target, and then following a brief interval, return the gaze to the target. Healthy people also show square wave jerks but frequency is greater in PSP patients compared to controls
- Failure of convergence. Convergence refers to the normal ability to turn the lines of sight inwards and towards each other.

Box 1: Hallmarks of progressive supranuclear palsy

- Gradually progressive disease with age of onset >40 years
- Supranuclear gaze palsy – especially with vertical saccades (quick movements of both eyes in the same direction) and, later, vertical gaze and a fixed stare
- Dystonia of muscles in the neck and trunk
- Postural instability and frequent falls beginning in the first year after symptom onset
- Pseudobulbar palsy – a group of symptoms that includes difficulty with chewing, swallowing, and speech; inappropriate emotional outbursts
- Symmetrical akinesia
- Varying degree of dementia (especially later in the disease)
- Tau protein deposition in the subthalamus, thalamus and basal ganglion
- Little or no response to levodopa (750-1000mg daily for one month) or apomorphine.

“The ocular signs are subtle, but they help differentiate PSP from PD,” Dr Williams said. “Nevertheless, around 8% of PSP patients never show eye movement abnormalities.”

Research at Queen Square Brain Bank (QSBB) revealed several other differences between PSP and PD that potentially aid diagnosis. For example, decreased sense of smell is usual in PD but uncommon in PSP. Sustained visual hallucinations occur in only about 3% of PSP patients compared to 49% of those with PD. “If you have visual hallucinations, it argues for PD,” Dr Williams suggested.

Dr Williams noted that around 20-25% of patients who show PSP on autopsy do not develop these classical features, partly because there appears to be more than one subtype of the condition. The QSBB team examined clinical case notes of patients with a pathological diagnosis of PSP on autopsy. Based on the clinical symptoms, they identified three distinct subtypes: Richardson’s syndrome (RS), PSP-Parkinsonism (PSP-P) and pure akinesia with gait freezing.

RS, which accounted for 54% of cases, is associated with an early onset of postural instability and falls, gaze difficulties and dementia. PSP-P occurred in 32% of cases. These patients showed non-symmetrical symptom onset, tremor and a moderate initial response to levodopa. Clinicians frequently misdiagnosed PD in these patients. Indeed, the initial diagnosis proved correct in 90% of RS patients, but in only 30% of those with PSP-P. The analysis revealed several other differences between PSP-P and RS:

- On average, people with RS showed a shorter duration of disease than those with PSP-P: 5.9 and 9.2 years respectively
- RS patients also tended to experience their first fall earlier in the disease than those with PSP-P
- RS patients died, on average, at a younger age than those with PSP-P: 72.1 and 75.5 years respectively.

The basal pons of people with PSP-P showed greater amounts of insoluble 3R-tau compared to RS. Adults express six versions (isoforms) of tau, a protein that forms part of the microtubules inside neurones. (Microtubules ensure that the nerve retains its correct shape and aid the flow of chemicals along the neurone.) Half of the isoforms have three sequences of amino acids repeated at their carboxyl-terminal (3R-tau). The other three isoforms have four repeat sequences (4R-tau). “The biochemical differences raise the question of the relationship between the conditions,” Dr Williams remarked.

Pure akinesia with gait freezing, the third form of PSP, accounted for 4% of cases examined by the QSBB team. These patients showed early-onset gait freezing, micrographia or stuttering, and hypophonia (weak, quiet voice) without other neurological signs. Patients with pure akinesia with gait freezing had longer disease duration (average 12.8 years) and a later onset of gaze palsy and dementia compared to RS patients.

Corticobasal degeneration (CBD) also seems to arise from the deposition of tau in the brain. However, the pattern of symptoms characteristic of CBD differs from PSP and PD:

- The motor symptoms of CBD tend to emerge asymmetrically
- Patients show apraxia – they cannot make familiar, purposeful movements
- Patients may exhibit alien limb syndrome, where a limb performs “meaningful” acts without the patient’s intention
- Patients with CBD may show frontotemporal dementia (FTD), which manifests as a progressive deterioration of linguistic ability and behaviour
- In common with PSP, levodopa usually produces little clinical improvement in CBD patients.

Dr Williams ended his presentation by saying that there is no cure for the atypical parkinsonian syndromes. Nevertheless, he suggested that as the prognoses differ so markedly, making the differential diagnosis is worthwhile.

The genetics of the atypical parkinsonian syndromes

Dr Huw Morris, Senior Lecturer in Neurology, Cardiff University and University Hospital of Wales, Cardiff.

Over the last few years, genetic studies have transformed our understanding of diseases as diverse as cystic fibrosis, cancer and coronary heart disease. According to **Dr Huw Morris**, genetic investigations are now helping researchers unravel the mechanisms behind atypical parkinsonian conditions, such as PSP and CBD. In the future, our growing understanding of their genetic basis may lead to disease-modifying therapies for these debilitating diseases.

Determining the genetic contribution to a disease can prove difficult. Dr Morris suggested that healthcare professionals could be reasonably certain that some diseases, such as Huntington's disease, are genetic, based on their clinical features. Other diseases, such as PD can occur in a familial, or much more commonly, non-familial (sporadic) form. A careful family history is important. If no-one else in the family has been affected, it is relatively unlikely that the disease is autosomal dominant, passed down from generation to generation. The risk of most Parkinsonian disorders increases with age and the age of onset offers a valuable clue as to whether the condition is sporadic or genetic. Dr Morris said, "It's unusual for a patient to develop PD in their 30s and such cases probably have a genetic basis".

Dr Morris explained that genetic tests currently fall into two groups:

- The validated, quality-controlled genetic tests funded by the NHS
- Tests used for research that have not yet made it into the clinic.

While researchers regularly discover genes that contribute to diseases, these need rigorous evaluation and validation before they become part of routine clinical practice. Once approved by the NHS, genetic testing has two main roles:

- Confirming the diagnosis. For example, genetic testing can aid the diagnosis of Huntington's disease and early onset dementia
- Predicting the risk in unaffected family members of developing disease.

These genetic tests can help in advising on appropriate clinical management and in providing appropriate genetic counselling for families. Predictive genetic testing requires specialist genetic counselling within medical genetics clinics. The take-up of predictive genetic tests is low, only around 10% of people at risk of developing Huntington's disease request the test.

Genetic testing as part of a research study can offer important insights into the pathology of a disease. For example, studies of a rare autosomal dominant form of fronto-temporal dementia (FTD) helped uncover the cause of least some cases of the atypical parkinsonian syndromes. PSP and CBD tend to be sporadic. However, around a third of patients with FTD have a family history of the condition. Genetic analysis showed that the gene responsible for one familial form of FTD with parkinsonism (FTDP-17) was tau. Dr Morris said, "For a long time, researchers thought that tau was a bystander protein. In other words, the protein wasn't directly pathogenic. The findings from the genetic studies suggest that abnormalities of tau deposition can be a primary problem that leads directly to disease." While autosomal dominant FTD is

uncommon, the studies helped researchers understand the pathogenesis of FTD, PSP and CBD. There are close pathological and clinical similarities between FTDP-17, PSP and CBD. Although PSP and CBD are sporadic, some affected family members with FTDP-17 have PSP as well as CBD-like syndromes and can present with parkinsonism.

A common variant of the tau gene, known as the H1 haplotype, predisposes to both PSP and CBD. (The term haplotype refers to a raft of genetic variation along a chromosome, which is inherited together.) Almost all PSP patients have two copies (homozygous) of the H1 gene. However, around 50% of the general population – most of whom never develop PSP – are also H1 homozygotes. The H1 genetic variation increases the risk of developing PSP but is not sufficient to cause it. This suggests, Dr Morris said, that environmental and other genetic factors are also important in the pathogenesis of the atypical parkinsonian syndromes. Furthermore, the high prevalence of the H1/H1 genotype in the general population means that genetic testing would not be useful. On the other hand, another variant (H2) seems to protect against PSP. H2 seems to be especially common among people of Caucasian origin. “Understanding why H1 increases the risk of PSP should allow the development of new disease-modifying treatments,” Dr Morris concluded.

Cognition and neuropsychiatric aspects of parkinsonian syndromes

*Professor John R Hodges, MRC Professor of Behavioural Neurology, University of Cambridge
Department of Clinical Neurosciences and Medical Research Council – Cognition and Brain Sciences Unit, Cambridge.*

Richardson, Steele and Olszewski described the mental deterioration and the personality changes associated with PSP in their landmark 1963 paper. **Professor John Hodges** reported that the cognitive and neuropsychiatric aspects had remained relatively neglected for almost a decade, despite being highly influential in determining the clinical profile. Increasingly, clinicians recognise the value of assessing these outcomes.

In 1974, Albert and colleagues described the pattern of cognitive and behavioural symptoms characteristic of PSP: prominent apathy, mood changes, and frontal executive dysfunction such as impaired decision-making, planning and mental flexibility. Patients with PSP also tend to be forgetful, although this is a problem predominantly with information retrieval rather than ‘classic’ amnesia. “Patients perform relatively well when they receive cues or reminders,” Professor Hodges said.

Disruption to the normal function of the cortex’s frontostriatal structures, in particular the dorsolateral and medial frontal lobe, contributes to the cognitive and neuropsychiatric abnormalities in PSP. Although the frontostriatal structures show some damage from tau deposition in PSP, the disruption arises mainly from the marked changes in the basal ganglion. Nerves from the basal ganglion (the part of the brain that selects and initiates coordinated voluntary movements) project to the cortex. As a result, damage to the basal ganglion “switches off” the frontostriatal structures.

Clearly, clinicians should evaluate cognitive and neuropsychiatric function in patients suffering from the atypical parkinsonian syndromes. Unfortunately, Professor Hodges said that the Mini-Mental State Examination (MMSE), the main instrument used in many hospitals to assess cognitive function, is “almost useless” in patients with the atypical parkinsonian syndromes. “If the MMSE shows cognitive changes in these patients, the syndrome is either very, very advanced or they don’t have the disease.” He pointed out

that the maximum score on the MMSE is 30. A score of <20 indicates dementia and >25 is 'normal'. In a study that enrolled seven patients within a year of death, the MMSE scores ranged from 23 to 29. Only two patients scored less than 25. In other words, most of the patients were 'normal' on the MMSE.

Professor Hodges argued that the Addenbrooke's Cognitive Examination (ACE-R), which encompasses but adds a number of questions to the MMSE, is a better choice for the assessment of patients with the atypical parkinsonian syndromes. Indeed, the Addenbrooke's team developed the questionnaire to detect early Alzheimer's disease (AD), FTD, PSP and CBD. In the study mentioned above, all the patients scored below the threshold for probable dementia on ACE-R. Despite being more comprehensive than the MMSE, the ACE-R remains suitable for clinical use: administering ACE-R takes around 10 minutes in a healthy person and around 20 minutes in someone with PSP.

Professor Hodges described some of the ways in which ACE-R differs from the MMSE and how patients with various syndromes perform on different parts of the test:

- In common with MMSE, ACE-R ensures that the person is oriented in time (date etc) and space (country, town etc). Typically, patients with the parkinsonian syndromes perform accurately on this assessment, although their responses may be slower than those from age-matched controls. In contrast, people suffering from dementia with Lewy bodies or AD tend to show poor temporal and spatial orientation
- The MMSE asks patients to recall three words. The ACE-R adds a further test. The person repeats a name and address three times, and then tries to recall the address at the end of the test. Patients with AD or CBD perform poorly. In contrast, most patients with PSP or MSA are reasonably accurate on this test, especially after prompts. For example, most patients with PSP or MSA recognise the correct response when the tester provides three alternatives
- Professor Hodges commented that the test, asking people to name as many words as they can, beginning with a particular letter – which assesses working memory, executive function and phonological ability – is especially valuable in the assessment of patients with parkinsonian syndromes. During a related task, the tester asks people to name as many animals as they can. Typically, 'normal' people name more animals (14-20) than words beginning with a particular letter (12-18), although their performance depends on age and educational attainment. Patients with AD showed a marked decrement on this test, especially in animal fluency. In contrast, people with MSA tend to do well on fluency tests
- In another part of the MMSE, the tester asks people to name a pencil and a watch. Since this test is within the ability of many two-year-old children, Professor Hodges noted that, "This tells you that the level of brain damage reduced their cognitive abilities to that of a two year old," Professor Hodges noted. ACE-R poses a greater challenge by asking patients to name, *inter alia*, a giraffe, kite and crown.

Overall, Professor Hodges said:

- MSA patients show relatively little impairment on ACE-R compared to controls
- People with PSP tend to show marked deficits in verbal fluency, language and drawing. Pronounced apathy and sleep disturbances are also common among people with PSP
- People suffering from CBD tend to perform badly on tests assessing verbal fluency, language, visuospatial perception and memory.

Detecting these differences may allow the multidisciplinary team to tailor management to each patient's cognitive and neuropsychiatric profile.

The burden of autonomic dysfunction in MSA

Professor Christopher Mathias, Professor of Neurovascular Medicine, Imperial College School of Medicine and the Institute of Neurology, University College London, London.

Catherine Best, Multiple System Atrophy Specialist Nurse, Sarah Matheson Trust and the National Hospital for Neurology and Neurosurgery, London.

Dysfunction of the autonomic nervous system is the hallmark of MSA. However, **Professor Christopher Mathias** observed, it is increasingly clear that autonomic dysfunction contributes to other parkinsonian syndromes. Studies that enrolled PD patients report prevalences for orthostatic hypotension of between ‘rare’ and 58%. In the latter study, 39% of the PD patients reported symptomatic orthostatic hypotension. Professor Mathias suggested that patients’ ages, Hoehn and Yahr (a scale that measures severity of PD), stage as well as concurrent diseases and treatments could all influence the prevalence of orthostatic hypotension among people with PD and, therefore, account for the differences between the studies.

Professor Mathias reminded delegates that the autonomic nervous system comprises parasympathetic and sympathetic nerves. These supply every organ in the body and act in an integrated manner to control key bodily functions, e.g. blood pressure and body temperature. Furthermore, humans have a number of cerebral centres and pathways, which directly and indirectly are concerned with autonomic function – as is being increasingly recognised by neuroimaging studies. As the system supplies every organ, autonomic dysfunction causes widespread signs and symptoms including:

- Genitourinary problems, including incontinence, erectile problems and ejaculatory failure
- Gastrointestinal disorders, including constipation and aspiration – the latter can prove fatal
- Respiratory problems
- Sleep disorders, which clinicians increasingly recognise as problematic for many MSA patients
- Sweating disorders. PD patients can show excessive sweating over their entire body surface area. In contrast, among MSA patients, anhidrosis and heat intolerance can be important clinical problems.

Nevertheless, orthostatic hypotension is the cardinal feature of MSA, Professor Mathias commented. Technological advances, such as the Portapress used with a 60° head tilt on a table, allow the non-invasive assessment of blood pressure response. “This procedure should be offered to all patients,” he said.

In healthy people, blood pressure remains within normal limits when the table raises the subject from horizontal to 60°. By contrast, in a person with MSA, blood pressure falls for a protracted time and there is no compensatory increase in heart rate as seen in healthy people. Numerous factors influence the severity of the orthostatic hypotension, including:

- The ambient temperature – because of the lack of compensation, heat-related vasodilatation can lead to orthostatic hypotension in people with MSA
- The normal circadian variation in blood pressure, which means that orthostatic hypotension tends to be more marked in the morning
- The time of the patient’s last meal. Blood vessels in the gastrointestinal tract dilate after a meal. Again, because patients lack compensatory mechanisms, postprandial orthostatic hypotension can develop in people with MSA
- Exercise, which in healthy people tends to increase blood pressure. In people with MSA, in contrast, exercise tends to reduce blood pressure.

Professor Mathias emphasised the importance of considering these factors when assessing whether a patient shows orthostatic hypotension. In some cases, the orthostatic hypotension may be asymptomatic. However, the low blood pressure can contribute to a range of symptoms, including:

- Dizziness, visual disturbances, transitory cognitive defects etc. These arise from cerebral hypoperfusion
- Muscle hypoperfusion, which may lead to discomfort, especially in the neck. (This is the so-called ‘coat-hanger ache’.) In some cases, orthostatic hypotension can produce severe pain
- Non-specific symptoms, including weakness, lethargy and falls.

Against this background **Catherine Best** discussed the management of autonomic dysfunction. “The first step is to discuss when and why the symptoms occur,” she told delegates. “For example, some people are fine when they are moving around, but they develop symptoms when they stand still, because they lack the calf and thigh responses that prevent pooling.” In other cases, a worsening of the orthostatic hypotension may be the first symptom of an illness or infection. “This is one exception to the rule of not using antibiotics as soon as possible if patients present with ‘cold’ like symptoms or a sore throat,” Ms Best observed.

Management of orthostatic hypotension aims to prevent syncope and falls, improve patients’ day-to-day function and optimise their health-related quality of life. Achieving this, Ms Best said, means taking a tripartite approach to management – one that encompasses education, non-pharmacological treatments and drugs. “Education and non-pharmacological approaches are important. They give control back to the patient.” For example, Ms Best suggests that patients with MSA:

- Avoid changing position quickly
- Avoid too much activity in the early morning
- Avoid performing too many activities in a short time
- Avoid warm environments
- Avoid high carbohydrate diets, which trigger insulin release. (Insulin is a vasodilator.)
MSA patients should eat small meals – these limit the increase in insulin levels, and reduce the risk of postprandial orthostatic hypotension, thereby helping patients remain alert
- Avoid lying flat in bed. Raising the head of the bed during sleep seems to stimulate the renin angiotensin system, which promotes fluid retention. Nocturia can pose a problem for MSA patients
- Avoid straining on the toilet. “Bladder and bowel management is an important element in the management of MSA,” Ms Best commented
- Use calf pump exercises and suggest that patients cross their legs when standing or sitting. These support blood pressure
- Ensure adequate intake of fluid and salt. “Drinking 500ml of fluid in five minutes increases blood pressure for about 15 minutes. The increase in blood pressure usually lasts around an hour,” Ms Best said. This approach is especially valuable for people who

Table 1: Examples of drugs used to manage complications associated with MSA

Indication	Drug
Blood pressure problems	Fludrocortisone, ephedrine, midodrine
Bladder problems	Desmopressin (DDAVP), oxybutinin
Reduce postprandial hypotension	Octreotide

don't want to take medications for the orthostatic hypotension." Nevertheless, many MSA patients benefit from taking one or more drugs, tailored to their particular problems (Table 1).

Ms Best concluded by stressing the importance of a team approach to the management of patients with MSA. For example:

- Physiotherapists can help reduce the risk of falls and improve posture during activities
- Occupational therapists can suggest useful pieces of equipment, such as bed-raisers and perching stools
- Dietitians can design diets that limit carbohydrate intake, while maintaining weight
- Social workers can help patients access the benefits, care packages and other entitlements.

Together, these approaches can help maximise patients' quality of life.

Speech, swallowing and breathing difficulties in MSA and PSP

Tricia Gilpin, Senior Speech and Language Therapist, National Hospital for Neurology and Neurosurgery, London.

Tricia Gilpin introduced her presentation on MSA and PSP with an outline of the differences between these two conditions and PD. Severe speech and swallowing problems can occur early in the course of MSA and PSP and are often among the presenting symptoms.

There are three different aspects to MSA:

- Extrapyrarnidal or Parkinsonian signs (MSA-P)
- Cerebellar signs (MSA-C)
- Autonomic impairment.

The type of dysarthria (speech disorder due to disturbances in muscular control of the speech mechanism) depends on the disease subtype:

- Patients with MSA-P will have hypokinetic dysarthria. The speech will have "typical Parkinsonian monotony and low volume". It may be quiet, slow, hesitant, and monopitch
- Patients suffering from MSA-C may exhibit ataxic dysarthria. Speech may be staccato and explosive, or slow and slurred with imprecise consonants
- Orthostatic hypotension can reduce speech volume and promote profound exhaustion. "Patients may simply not want to talk," said Mrs Gilpin.

Other presenting symptoms in MSA may be sleep and breathing problems. In some patients an increase in 'sleep talking' may indicate the onset of the disease. In others, the muscles that open the vocal cords (the abductors) are affected leading to excessive snoring and stridor. "Abductor paralysis, of a degree which endangers life, is fairly common in this disease," according to Williams *et al*¹. Of the 12 patients in this study, 75% had excessive snoring, 50% experienced stridor, and 33% required tracheostomy. "The multidisciplinary team should discuss the possibility of non-invasive night-time ventilation and possible tracheostomy with patients and their family to improve day-time quality of life," she commented.

Patients with PSP present with a different pattern of dysarthria to MSA. In 1993, a study by Kluin *et al*² found all 44 patients had mixed dysarthria with spastic characteristics, (slow, harsh, unintelligible speech) hypokinetic and sometimes ataxic components. They also exhibited palilalia (repetition of syllables or words) and echolalia. Several other aspects of the condition could hinder communication, such as masked face, facial dystonia, language dysfunction and difficulty initiating conversation. In addition, “dementia may be present in up to 100% of PSP patients³.”

People with PSP or MSA also commonly experience swallowing problems (dysphagia). Mrs Gilpin showed a videofluoroscopy clip showing the normal swallow and then discussed the problems that may occur in each disease. In MSA patients may experience difficulties at the oral, pharyngeal and oesophageal stages. In a study of 10 MSA patients⁴, 90% showed poor control of the bolus at the oral stage with delayed initiation of the swallow in 80% and pooling of residue in the pharynx in 100% of the patients. Additionally it has been found anecdotally at Queen Square that many MSA patients suffer from dysmotility at the oesophageal stage. Video clips were shown to illustrate the problems at all three stages. In 1997, a study by Litvan *et al*⁵ of 27 patients with PSP, 96% had abnormal swallows and those with cognitive problems had significantly more dysphagia. Symptoms included poor tongue motility, delayed initiation of the swallow and pooling in the pharynx. Neumann *et al*⁶ in 1996 described ‘mouth stuffing and rapid drinking’ in at least 50% of patients with PSP. Many of the patients with MSA and PSP had difficulty chewing, excessive or insufficient saliva, coughing or choking, a wet or ‘gurgly’ voice and a weak cough.

Mrs Gilpin discussed the assessment of patients with speech and swallowing problems. She suggested beginning with a full history of the disease and the speech and swallowing problems, from the patient and the carer, followed by a cranial nerve assessment and trials of different consistencies of food and drink. If appropriate, a videofluoroscopy would be arranged. A formal or informal dysarthria (speech) assessment would be carried out.

Advice for swallowing difficulties would include encouraging the patient to sit upright at 90°, and in the case of PSP, to discourage overloading the mouth. Placing the plate on a pile of telephone directories can be helpful for those who have supranuclear gaze palsy affecting downward vision. In PSP supervision may be required due to the cognitive difficulties. The speech and language therapist may advise modification of the consistencies of the diet, possibly thickening drinks or trying a ‘chin tuck’ when swallowing. In addition they may discuss the placement of a percutaneous endoscopic gastrostomy (PEG).

Speech and language therapists can help with communication problems by encouraging facial expression and eye contact. Good breathing techniques can help speech, while relaxation can reduce the anxiety that can further impair communication. Early in the disease process work on articulation and volume control may be helpful. Mrs Gilpin said, “The aim should be to preserve speech for as long as possible.”

Communication aids may be appropriate for some patients. Lightwriters can be helpful in MSA, but tend to be less suitable for PSP patients due to visual and cognitive problems. Patients may find ABC and picture charts helpful or even typed sheets with key words and phrases printed in a large font. “Patients can use these to express their needs,” she observed.

Mrs Gilpin discussed the quality of life issues surrounding the insertion of a PEG or tracheostomy. Litvan *et al*⁵ in 1997, looking at PSP patients, felt that indicators for PEG were:

- Videofluoroscopy shows aspiration on all textures
- The patient loses more than 10% of their body weight
- There is fever of unknown origin
- Patients take more than an hour over a meal.

Finally, Mrs Gilpin suggested that therapists and other members of the multidisciplinary team should have on-going discussions with the patient and their family about overall management and the placement of a PEG. In some cases it may be possible to continue small amounts of oral feeding with the majority of nutrition and hydration being provided by the PEG, “our aim should be to maximise the quality of life,” Mrs Gilpin concluded.

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Specialist equipment and technology

Ruth Groves, Clinical Specialist Occupational Therapist and Local Therapist Coordinator for the Regional Environmental Control Equipment Service (RECES) North Thames (North London, Hertfordshire and Essex).

“Environmental control systems enable the most severely disabled members of our society to remain in, or regain, control over their immediate home environment, thereby increasing their independence, autonomy and quality of life,” **Ruth Groves** said at the start of her presentation. “Environmental control systems allow disabled people to operate electrical appliances from a central device that looks rather like a large remote control – although it does far more than just operate the television.”

Ms Groves explained that RECES was devolved from the Department of Health in 1994, following the publication of a document by the British Society for Rehabilitation Medicine, called ‘Prescription for Independence’. This was later revised in 2000 as ‘Electronic Assistive Technology’. The services are regional and are set to develop further as the government implements the Telecare initiative and Integrating Community Equipment Schemes (ICES) evolve across England.

Environmental control equipment services differ in several ways in various regions, e.g. in their exact referral criteria. However, Ms Groves remarked that the main criteria are:

- An inability to call for assistance or contact their carer
- An inability to control their environment by any other means
- The person being left on their own for considerable periods.

Table 2: Funding of environmental control systems in North London, Hertfordshire and Essex

Funded	Not funded
Call for help	Door openers
Door release	Electrical work to install the system
Intercoms, with pre-recorded messages	BT sockets
Light and lamp controls	Motorised curtain tracks
Bed control	Communication aids
Riser and recliner chair control	Close circuit television
	Carpentry work to install the system

Such people will need to be able to control their door, raise the alarm if necessary and be able to operate a system that has an appropriate level of complexity to meet their needs. “In some cases, we may need a trial installation to ensure that patients can use the system,” she said.

Ms Groves added that RECES North Thames would not install environmental control systems solely to operate entertainment systems. (Table 2 shows some examples of what RECES will and will not fund.) An environmental control system costs between £3000 and £5000 and funds are limited. RECES North Thames installs environmental control systems only when there is no other more economical means to meet the patient’s needs. As a result, an occupational therapist’s report is useful with the referral letter. This also allows the service to decide if the referral is ‘urgent’, ‘soon’ or ‘routine’. With conditions such as MSA, Ms Groves stressed the importance of referring as early as possible. However, the atypical parkinsonian syndromes account for a relatively small proportion of the caseload in North Thames. Currently, multiple sclerosis, cerebral palsy and spinal cord injury account for the greatest proportion of cases: 30%, 21% and 12% respectively.

The initial assessment takes about 1 to 1.5 hours. “We compare what the patient needs to do with what the service can provide,” Ms Groves said. It is sometimes possible to make some changes almost immediately. The team reviews patients each year by phone and aims to visit the client at home every three years. Nevertheless, in some cases follow-up is more intensive: the team may see a client with motor neurone disease every three or four weeks. Increasingly, RECES North Thames can offer integrated technologies: a wheelchair with a console that controls its movement, a communication aid and environmental control systems. These technological advances offer disabled patients an unprecedented level of control over their environment.

Getting the care right

Maggie Rose, PSP Specialist Nurse, Progressive Supranuclear Palsy (PSP-Europe) Association, Towcester, Northamptonshire.

“Many people find that in the early days following diagnosis, they forget the information they were given in a busy clinic,” **Maggie Rose** said. Consequently, the PSP-Europe Association helpline can become an invaluable point of contact for the person with PSP or their carer, by talking through issues with them as they arise, answering questions truthfully and being available for support.”

Often the advice needed by patients or their carers is relatively simple, such as increasing fluid intake to avoid urinary tract infections or suggesting that the patient is propped up to avoid chest infections. However, in most cases, Ms Rose said, patients and their carers just want someone to listen. “We can act as a safety valve. Often patients are in shock after they get the diagnosis. They are afraid of the unknown. They want to know what it means, how progressive the symptoms are likely to be and whether their children develop PSP.”

Ms Rose stressed that the helpline provides a backup to regional services. “The helpline does not cut across, but adds to the advice and support given by professionals locally including the palliative care team, the PD nurse specialist, the neurology nurse and the neurologist,” she said. For example, patients can use the helpline to discuss the advantages and disadvantages of PEGs. “Often healthcare professionals don’t have the time to fully address these issues,” she said.

The PSP-Europe Association helpline is available 24 hours a day, 365 days a year. “Many worries arise in the evenings or at weekends,” Ms Rose noted. “They come when the carer is on his or her own in the evening after the patient has gone to bed, when the person they are caring for is in respite care or they have just visited a care home. I listen to their concerns and, to gain their confidence and trust, the conversation cannot be hurried. My time is my main gift to my clients.” She also encourages carers and patients to attend the support groups that the PSP-Europe Association runs in many parts of the countries – many benefit from sharing their experiences.

“PSP can lead to personality changes and some people say that it is like living with a stranger, it is a living bereavement,” Ms Rose said. “The person with PSP may be apathetic and aggressive. They may develop obsessions and express very little gratitude. This can be very distressing for the carer and hints of these emotions may emerge during the call. We help them express the emotions and let them know that it is justified to feel this way.”

By offering advice and support, the PSP-Europe Association aims to maximise quality of life for patients and their carers. “We aim to keep their lives as normal as possible for as long as possible,” Ms Rose explained. For example, the helpline often advises how patients and carers can travel, by offering practical advice about insurance, notifying airlines and so on. She remarked that one couple got around the problems of eating in a hot environment and keeping clothes clean by going to a nudist camp!

Finally, Ms Rose highlighted the fact that healthcare professionals also use the helpline. “GPs, therapists, specialist nurses or social care providers, whose experience of the illness may be limited, often ask about symptom control and prognosis,” she said. For example, the helpline may help nursing homes appreciate that incontinence could be due to a neurological bladder rather than attention seeking. “The helpline is at the heart of the PSP-Europe Association,” Ms Rose concluded: “In providing advice and support to anyone caring for someone with PSP, the organisation hopes it helps them get the care right.”

NSF and NICE guidelines for PD: from rhetoric to reality

David Colin-Thome OBE, National Clinical Director for Primary Care, Department of Health, London.

At the time of the conference in July, the National Service Framework (NSF) for Long-term Conditions was nearly a year old and the National Institute for Health and Clinical Excellence (NICE) PD guidelines were about to be published. **Dr David Colin-Thome** considered the challenges that healthcare professionals face when translating the recommendations made in these documents into improved services for patient care.

Dr Colin-Thome said that the NSF and NICE guidelines reflect a change of focus in the NHS more widely. This change in focus aims to give patients and the public more say about, and control over, the management of their conditions. “The emphasis is on health and independence,” he commented. This requires the delivery of an increasing proportion of health care in the community. However, until recently, community services were relatively under-funded compared to secondary care. Dr Colin-Thome suggested that the emphasis on secondary care was “outmoded compared to other countries”. Primary care led commissioning aims to restore the balance. Commissioning, he explained, is the process by which the primary care organisations identify health needs and prioritise services to secure the best care to meet these needs within the available budget.

Commissioners should help maximise patient choice, a key element in the new focus in the NHS. According to Dr Colin-Thome, patient choice embraces three elements:

- Patients should have the power to shape their treatment pathway and keep control of their management
- The treatment pathway should reflect patients' preferences for what services the NHS delivers, when, where and how
- Services should be personalised around the patient.

Nevertheless, the NSF and NICE guidelines have yet to realise their full potential. Dr Colin-Thome argued that translating the recommendations into improved services might mean making difficult choices, including an evidence-based disinvestment in some current services. For example, he noted that the NHS currently spends £800 million year on statins, a group of drugs used to lower abnormally raised levels of lipids in the blood. A wholesale switch from the most expensive statin to the cheapest could save £300 million annually without compromising clinical outcomes. He also highlighted the large volume of outpatient care and follow-up appointments, as well as the fact that a third of referrals are now between consultants. "Primary care could deal with much of this work," he remarked. "There is a huge variation in what we do across the country, without any clear justification."

Against this background, Dr Colin-Thome suggested that devolving budgets to primary care, which is responsible for 90% of healthcare, offers "a huge opportunity to improve quality and responsiveness". For example, community pharmacists and nurses could review patients with long-term conditions. Moreover, primary care-led commissioning should allow community, secondary and social care to work together more effectively and efficiently. "We need to challenge the way in which things are currently done," Dr Colin-Thome said. "In many cases we have the money; we choose to use it in 'interesting' ways. We need to begin to look at how we can use the money more effectively".

Poster presentation and prize session

Dr Peter Fletcher, Consultant Physician/Geriatrician, Gloucestershire Hospitals NHS Foundation Trust, introduced the poster presentation and prize session. The quality of the content and presentation was impressive. The following is a summary of the main messages of each poster displayed at the conference.

Medication prescribing and administration in Parkinson's

Rachel Bradley, Dorothy Robertson, Isobel Wyber, Royal United Hospital Bath, Bath.

This study retrospectively reviewed hospital records to examine the prescribing and administration of medications for PD. They also used a semi-structured interview to explore the knowledge and experiences of 79 patients with PD.

Prescribing was incorrect in 43% of patients on admission falling to 26% on discharge. Inaccurate timing emerged as the most common error. Medication administration was incorrect in 51% of patients on admission and 35% after 24 to 48 hours, with a median of five doses omitted per hospital stay, usually for 'no reason', 'drug not available' and 'nil by mouth'. Themes that emerged from the semi-structured interview included anxiety, loss of control, negative thoughts, decline in physical

function, concerns about staff knowledge, ward routines, inflexible drug trolley rounds and prompting staff about timings.

The authors concluded that the quality of PD drug prescribing and administration is sub-standard in many cases. This resulted in patients experiencing a variety of psychological problems. Based on their findings, they have developed specific guidance for ‘unavailable’ medication and advice on how to manage PD patients who experience swallowing difficulties.

Patients’ adherence and knowledge of prescribed Parkinson’s medication

Steffan Gwent, Brian Wood, Northumbria Healthcare NHS Trust, Wansbeck General Hospital, Ashington, Northumberland.

Researchers from Northumbria Healthcare NHS Trust audited adherence and knowledge of prescribed medication among 149 patients from PD clinics across Northumberland. The audit employed the following standards:

- 100% of patients should know which PD drugs they are taking; 67.1% met this standard
- 100% of patients should know the frequency of dosing; 90.6% met this standard, 0% of patients miss a dose, 55.0% stated that they had missed a dose of PD drug(s) while 12.1% missed a dose in the week before the audit.

“The deviation from the standards set indicates a shortfall in adherence and knowledge of medication used in PD,” the authors concluded. “Factors such as dementia may affect medicine-taking behaviour and knowledge of therapy in this patient population.”

Visuospatial bias in left hemiparkinson’s disease

Paul M Greenhouse, Alison C Lee, Dorothy Robertson, PD Research Group, School of Social Sciences, Bath Spa University, Bath.

The authors of this poster noted that in some patients PD distorts spatial perception. As a result, PD patients may experience difficulties walking through doorways or down narrow corridors, which may suggest a visual bias towards one side.

To investigate spatial perception in PD patients, the researchers used pairs of horizontal parallel lines of equal length (34cm) separated by (14cm) projected on the left or right side of a large screen. Marks bisected the centre of one line and at varying positions to the right or left of centre on the other. Participants verbally indicated which line showed the central bisection.

The poster presented preliminary findings from five patients with primarily left-sided motor symptoms and four that experienced right-sided symptoms. Patients with primarily left-sided motor symptoms showed a bias for leftward bisections, particularly for stimuli on the left of the screen. Patients with primarily right-sided motor symptoms showed a similar pattern of errors to seven age-matched controls. The leftward bias in patients with primarily left-sided motor symptoms was greatest for stimuli presented within the image of a doorway. This suggests that the study may be relevant to visuospatial problems encountered in the ‘real world’ and may inform rehabilitation strategies.

Causative factors of hospital admissions in Parkinson's patients

Zaki Ibrahim, Christopher Jones, Brian Wood, Northumbria Healthcare NHS Trust, Wansbeck General Hospital, Ashington, Northumberland.

Researchers from Northumbria Healthcare NHS Trust reviewed causes of hospital admissions among 20 people with PD. They aimed to determine if any of the admissions were potentially preventable. Consultant specialists reviewed the admissions and classified nine as being related to PD: falls (3 admissions); pneumonia (2 admissions); other respiratory (2 admissions); dysphagia (1 admission); PD medication related (1 admission). The authors considered that only the latter admission was potentially preventable and called for follow-up studies to confirm the findings.

Teaching emergency doctors and nurses about Parkinson's

Subramanian Manickam, Judith Graham, James George, Cumberland Infirmary, Carlisle.

Previous studies suggest that PD patients often do not receive their medication promptly when admitted to hospital, especially as an emergency. A group from Cumberland Infirmary, Carlisle, sent a questionnaire to 20 foundation doctors and 20 nurses to, firstly, assess their knowledge about and attitude to PD and, secondly, discover their preferred learning styles.

The doctors and nurses surveyed showed poor knowledge about the special medication requirements of PD patients and the emergencies that commonly arise for this group. They tended to regard PD as a low priority chronic disease, with little relevance to the emergency department. The preferred learning styles were activist and pragmatic. The doctors and nurses had little time for reflection.

The group developed a practical teaching programme based on the results of the questionnaire. A follow-up questionnaire demonstrated that doctors' and nurses' knowledge had improved. "Importantly, their attitude to PD also changed – they saw it as a chronic disease, in which acute complications were common and preventable," the authors commented. They added, however, that future studies need to determine whether the teaching programme improved the management and outcomes of PD patients admitted to hospital.

A goal-orientated activity and exercise group for people with Parkinson's

Gail Keith-Baker, Claire Berncastle, Liskeard Community Hospital, Liskeard, Cornwall.

This poster outlined the aims, activities and interim results of the PD therapy group, which has been running for patients in East Cornwall since January 2006. The group comprised seven patients at a similar stage of disease and who experienced similar practical problems.

Before the first session, patients completed the PDQ39 questionnaire to assess baseline quality of life. To meet goals identified during the baseline assessment, therapists developed activities and exercises that emphasised conductive education management strategies.

Interim results suggest that the approach improved patients' quality of life, timed walking and some memory abilities. More than 90% of the goals set showed positive outcomes and the therapists received several letters of commendation from patients and carers. The team plans to re-evaluate patients after three months to see if these improvements are maintained. Another group of eight patients will start the programme at the end of May 2006.

Driving in Parkinson's

Rajiv Singh, Brian Pentland, John Hunter, Frances Provan, Astley Ainslie Hospital, Edinburgh.

Researchers from Astley Ainslie Hospital, Edinburgh, reported the results of a study of 154 people with PD who were referred to the centre's driving assessment centre. The multidisciplinary team used a combination of clinical tests, reaction times on a test rig and an in-car driving test to assess whether PD patients were suitable to drive.

The team considered that 66% of the PD patients were fit to continue driving, although 46 required automatic cars and 10 needed car modifications. Several factors were correlated with unsuitability to drive: increasing disease severity; advancing age; other medical conditions, particularly dementia; disease duration; reaction time on test rig; and the score on driving test. Drug treatment and length of driving history did not predict whether a patient would be unsuitable to drive. Severe disease, slow reaction time, moderate disease associated with another medical condition and the score on car testing emerged as showing the strongest correlation with driving ability.

In conclusion, the authors noted that most PD patients are safe to drive, although many benefit from car modifications or using an automatic vehicle. "A combination of clinical tests and in-car driving assessment will establish safety to drive and a number of clinical correlates predict likely outcome," they added.

Poster Prizes

First prize was awarded to **Rajiv Singh, Brian Pentland, John Hunter and Frances Provan** for **Driving in Parkinson's**.

Second prize was awarded to **Gail Keith Baker and Claire Berncastle** for **A goal-orientated activity and exercise group for people with Parkinson's**.

Third prize was awarded to **Rachel Bradley, Dorothy Robertson and Isabel Wyber** for **Medication prescribing and administration in Parkinson's in an acute hospital**.

This report was written by Mark Greener, Freelance Medical Writer.

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Medical Education Partnership
53 Hargrave Road, London NW1 5SH.
Telephone: 020 7561 5400
Email: info@mepltd.co.uk
Web: www.mepltd.co.uk