Clinical Focus

Continuing education and moving points in medicine

Continuing Education Module 21: Brain disease

The challenges of progressive illness

The daily challenges of living with a deteriorating neurological disorder can be aided by planning and routine, writes **Alison Cashell**

Parkinson's disease (PD), first documented by Dr James Parkinson in 1817, is the second most common chronic neurodegenerative disease.[1] Progressive and disabling in its development, it affects approximately 9,000 people in Ireland, 1.2 million people in Europe, and 6.3 million worldwide.[2] It is estimated that PD affects 1-2:1,000 of the general population, affecting more men than women. This can be further broken down to 2:100 of the elderly. However, approximately 1:7 of those diagnosed will be under the age of 50.

According to the WHO, Parkinson's is just one of many neurodegenerative disorders that will surpass cancer as the most common group of medical conditions by 2040. In a recent HSE performance report,[3] neurology is one of six specialities accounting for high numbers of patients waiting longer than 12 months for an out patients department appointment.

PD is characterised by the degeneration of neurones in the basal ganglia which synthesise and store the neurotransmitter dopamine. It is thought at diagnosis, the person with PD will have already lost between 60-80% of their dopamine neurons. Dopamine is thought to be responsible for the initiation, planning and execution of movement, as well as motivation and thinking.

The cause is generally unknown, but there have been links identified to genetic disposition, environmental toxins, drugs, viruses or head injury. PD is slow and insidious in its progression and it always presents asymmetrically, but both sides will be affected as the condition advances.

There are two aspects to PD: motor symptoms and non-motor symptoms. Motor symptoms are the cardinal features of PD. For a diagnosis (as defined by the UK Brain Bank Criteria), bradykinesia must be present, along with at least one of the following: muscle rigidity; resting tremor (4-6Hz); and/or postural instability.

Approximately 70% of people with PD will have a tremor. Non-motor symptoms often dominate the clinical picture, and include constipation, fatigue, anxiety, depression, sweating, and insomnia to name a few. Studies suggest that non-motor symptoms, such as constipation, REM sleep behaviour disorder (RBD) and anosmia, can be present up to 20 years before diagnosis.[4]

NICE guidelines state that a person with suspected PD should be referred to a neurologist or geriatrician for a confirmed diagnosis, as according to some studies (UK Brain Bank), incorrect diagnosis occurs in up to 25% of cases, so review by a specialist should be arranged if possible.

Non-motor symptoms

Non-motor symptoms (NMS), such as anxiety and communication difficulties, can have a serious impact on the quality of life for people with PD. NMS can be a contributing factor for admission to hospitals and nursing homes, particularly later in the disease, as the symptoms such as postural hypotension worsen, which can lead to falls, and neuropsychiatric symptoms appear.

The addition of dopaminergic medications usually benefit the motor symptoms, but a recent study showed that very few NMS are positively affected by these therapies.[5]

Constipation is extremely common in PD and is often one of the symptoms present before diagnosis. Constipation is a product of a decrease in gut motility and a decrease in overall activity, which result in a lack of stimulation. As levodopa is absorbed in the bowel, constipation can cause delay or failure of medication, which can lead to unpredictable 'offs', a worsening of symptoms, both motor and non-motor.

Constipation may be manageable through diet and exercise, but the majority of people will require a laxative to ensure they go regularly. One of the most effective laxatives for the treatment of constipation associated with PD is Movicol (polyethylene glycol 3350 + electrolytes).[6]

Table 1: NICE guidelines for the clinical diagnosis of PD

Definition and differential diagnosis	Parkinson's disease should be suspected in patients with tremor, stiffness, slowness, balance problems and/or gait disorders
Expert vs non-expert diagnosis	Patients with suspected PD should be referred quickly and untreated to a specialist
Clinical diagnosis	PD should be diagnosed clinically and based on the UK Parkinson's Disease Society Brain Bank Criteria
Review of diagnosis	The diagnosis of PD should be reviewed regularly and reconsidered if atypical clinical features develop
Single photon emission computed tomography (SPECT)	123I-FP-CIT SPECT should be considered for patients with tremor where tremor cannot be clinically differentiated from parkinsonism

Table 2: UK PDS Brain Bank Criteria for the diagnosis of PD*

Step 1: Diagnosis of a parkinsonian syndrome	Bradykinesia and at least one of the following: • Muscular rigidity • Rest tremor (4–6 Hz) • Postural instability unrelated to primary visual, cerebellar, vestibular or proprioceptive dysfunction
Step 2: Exclusion criteria for PD	History of: • Repeated strokes with stepwise progression • Repeated head injury • Antipsychotic or dopamine-depleting drugs • Definite encephalitis and/or oculogyric crises on no drug treatment • More than one affected relative • Sustained remission • Negative response to large doses of levodopa (if malabsorption excluded) • Strictly unilateral features after three years • Other neurological features: supranuclear gaze palsy, cerebellar signs, early severe autonomic involvement, Babinski sign, early severe dementia with disturbances of language, memory or praxis • Exposure to known neurotoxin (MPTP) • Presence of cerebral tumour or communicating hydrocephalus on neuroimaging
Step 3: Supportive criteria for PD	Three or more required for diagnosis of definite PD: • Unilateral onset • Rest tremor present • Progressive disorder • Persistent asymmetry affecting the side of onset most • Excellent response to levodopa • Severe levodopa-induced chorea • Levodopa response for over five years • Clinical course of over 10 years

** From Hughes AJ et al. Accuracy of clinical diagnosis of idiopathic Parkinson's disease. J Neurol Neurosurg Psychiatry 1992 Mar; 55(3): 181-4*

Both written and spoken communications can be impaired in PD. Handwriting can become small and cramped; appear 'scribbly' and difficult to read. Speech and language problems are varied between dysarthria and dysphasia. People with PD will frequently have a soft voice, and they are likely to lack expression in their voice. They may also experience festination of speech or a hesitancy of initiating words. These issues may be worse during periods when the medication is not working or is 'wearing off'.

Speech and language difficulties can have a huge negative impact on a person's life, and may lead to social isolation. The therapy known as LSVT (Lee Silverman Voice Treatment) was specifically developed for people with PD to help them improve their speech by teaching them to 'think loud'. The four-week course of classes is conducted by qualified STLs and can be very successful.

There are many sleep disorders associated with PD, ranging from daytime problems, such as excessive daytime sleepiness (EDS) and fatigue, to night-time problems such as insomnia, vivid dreaming and RBD.[7] The first line of treatment should be education regarding good sleep hygiene.

EDS may be present due to a feature of the disease or as a side effect of the dopaminergic medications. Treatment is varied, depending on the severity of sleepiness. Some people may have a good response caffeine supplements, or some may need medication used to treat narcolepsy (eg. modafinil – off license). Fatigue is often reported as one of the most disabling symptoms of PD,[8] and can be difficult to manage as it can be due to the physical impact of the disease, or as a result of night-time issues.

As with all cases of insomnia, establishing whether the lack of sleep is due to being unable to fall asleep, or being unable to stay asleep is essential. Nocturia is a common urinary symptom and waking up during night due to tremor, stiffness or an inability to turn in bed due to 'wearing off' is frequently reported.

RBD involves people with PD acting out vivid and action-packed dreams, and may result in waking tired due to their movements during the night-time. It is often the bed partner who reports these problems, as the person themselves is unlikely to be aware of it unless they wake themselves during the night, maybe falling out of bed or thrashing out against something.

Nutrition

There is no specific diet that someone with PD should follow, or specific foods they should avoid. It is important to advise people taking levodopa therapy to avoid protein at tablet time, particularly at later stages of the disease, to improve efficacy of the dose. This is because the two compete for absorption at the blood brain barrier, and the medication loses, so if taken with diary or with meals, the dose may appear inadequate.[9]

It is important to ensure that levodopa is taken 30-40 minutes prior to, or an hour and a half after a meal with a full glass of water, juice or cordial (to promote transit to the jejunum, where it is absorbed from). If nausea occurs as a side-effect of taking it on an empty stomach, it is usually short lived, and may be treated with domperidone.[10]

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References available on request from nursing@medmedia.ie (Quote: Cashell, A. WIN 2014; 22 (2): 41-42)