



# Parkinson's Disease Information Sheet 1.6

## Parkinson's Plus Conditions

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**Idiopathic Parkinson's disease (Pd)** is typically a slowly progressive neurological condition which is related to a deficit of dopamine as a result of degeneration of dopamine producing neurons.

In addition to Idiopathic Pd there are several neurological conditions which are described as atypical Parkinsonism which may be grouped under the term Parkinson's Plus. These include;

- Progressive Supranuclear Palsy
- Multi-System Atrophy
- Cortical Basal Degeneration
- Lewy Body Disease

### **Progressive Supranuclear Palsy (PSP)**

PSP was originally described by Steele, Richardson and Olsewski in 1964 and originally bore their names. The usual age of onset is between 50-70. Published cases of PSP reveal a slightly higher male prevalence (2:1). The condition is rare with the prevalence being approximately 7:100,000 over the age of 55.

There is no known cause for PSP and association with toxic substances, geographic region, occupation has not been found. There is no familial association reported.

The progression of PSP is more rapid than in Pd. The average length of illness from diagnosis to death is 5-6 years with the range being 2-11 years. Ongoing support for both the person with the condition and the family is essential.

Early signs and symptoms are varied and include;

- Impaired downward gaze
- Unsteady gait
- Bradykinesia
- Rigidity
- Speech disturbances

The characteristic clinical sign of PSP is a supranuclear palsy which results in impaired voluntary eye movements however the eyes move normally in response to passive head movements. This finding implies that the neurological change is above the nuclei that produce head movements, hence the name. Downward gaze is first affected followed by upward gaze and later horizontal gaze.

Blink rate is markedly reduced (more so than in Pd) and there may be an ocular stare with the upper eyelids resting above the irises.

Those affected by PSP often exhibit neck extension rather than the stooped posture of Pd.

Falls, due to imbalance, occur relatively early in the disease process often within a year or two of symptom onset and commonly involve falling backwards.

Later symptoms include;

- Facial stiffness
- Swallowing difficulties
- Marked bradykinesia
- Rigid posture
- Lack of mobility

The introduction of levodopa therapy fails to achieve a positive response and symptoms are managed as they develop.

Referral to a Speech Pathologist is essential to assess swallowing and to supervise the introduction of modified diet and strategies to ensure a safe swallow in order to prevent aspiration pneumonia which is a major risk.

The involvement of a Physiotherapist will address mobility with emphasis on safety of gait and fall prevention.

## **Multi-System Atrophy (MSA)**

The term MSA was introduced in 1969 and prior to that time the following terms were used to describe various manifestations of the condition;

- Striatonigral degeneration
- Shy-drager syndrome
- Olivopontocerebellar atrophy

MSA is a sporadic progressive neurological condition which is demonstrated by Parkinsonian features of bradykinesia, rigidity, postural instability and autonomic nervous system involvement.

There is currently no reliable information concerning the incidence and prevalence of MSA. Clinical symptoms commonly begin in the 60's however occasionally may commence as early as the 40's with a slightly higher incidence in males.

Early signs and symptoms are the gradual onset of autonomic nervous dysfunction;

- Urinary dysfunction
- Impotence
- Postural hypotension
- Disturbed balance

In addition the early presentation of symmetric (bilateral) Parkinson's like symptoms (muscle rigidity and bradykinesia) may be indicative of MSA rather than Pd (unilateral presentation). Non-response to levodopa therapy is an additional indicator although many patients will experience a partial (but often short lived) response.

Diagnosis of MSA rests on the clinical history and neurological examination. The relentless progression of MSA is more rapid than Pd. However, there is a

considerable variation of disease progression.

The later symptoms include;

- Marked communication changes
- Swallowing difficulties
- Inspiratory stridor
- Urinary incontinence
- Increased muscle rigidity

Due to the frequent lack of efficacy of Pd medications in MSA, allied health involvement to manage the symptoms is essential;

- Physiotherapy to maintain mobility and balance
- Occupational therapy to maintain functional independence for as long as possible
- Speech therapy to address speech and swallowing problems. The provision of alternative communication aides may be of benefit in the latter stages.

Medical and pharmaceutical management may address such symptoms as postural hypotension, impotence and constipation.

## **Cortico Basal Degeneration (CBD)**

CBD, first described in 1968, is described as a late adult (60-80) onset progressive neurological condition for which the cause is unknown. It remains a rare disease of unknown incidence and prevalence. It may affect either gender.

Common indicators of CBD are:

- Asymmetric rigidity and bradykinesia
- An inability to perform purposeful tasks especially noted in one limb
- Clumsiness, jerking and sensory impairment affecting a limb (distinctive indicators)
- Postural and action tremor

Diagnosis of CBD rests on the clinical history and neurological examination. Progression is more rapid than Pd. Treatment is limited and management is symptomatic.

### **Dementia with Lewy Body (DLB)**

Dementia with Lewy bodies (DLB) is a progressive neurological condition which involves symptoms suggestive of Pd (tremor, muscle rigidity, bradykinesia) however, early onset memory loss is marked.

In the early 1900's Dr Friederich H Lewy discovered abnormal protein deposits in the brain. These proteins were found in the mid brain where they deplete dopamine hence causing Parkinsonian symptoms. In DLB these Lewy bodies are diffuse through other areas of the brain including the cerebral cortex. The neurotransmitter acetylcholine is depleted causing disruption of perception, thinking and behaviour.

DLB is a common cause of dementia second only to Alzheimer's disease (AD).

It has recently been accepted as a disease in its own right. DLB affects both genders. Aging is considered to be the greatest risk factor for DLB with onset typically between 50 and 85. Having a family member with DLB may increase a person's risk.

Early indications of DLB are;

- Tremor and muscle rigidity similar to that seen in Pd
- Difficulties with concentration and attention
- Fluctuating cognition
- Early onset visual hallucinations
- Difficulties judging distance

The symptoms of DLB vary from person to person and may mimic other diseases in the early years. As a result it is not uncommon for patients with DLB to be initially diagnosed with Pd or AD.

DLB is progressive, eventually leading to complete dependence. Currently the medical management for DLB is similar to that for AD and at times Pd medications may be used to address the motor symptoms.

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