

Parkinson's Disease Information Sheet 1.1

Description and Incidence

Parkinson's Disease

Parkinson's disease (Pd) is a progressive neurological condition which is characterised by both motor (movement) and non-motor symptoms. Information Sheet 1.2 describes the major symptoms in detail.

The condition was first described in 1817 by Dr James Parkinson in *An Essay on the Shaking Palsy* in which he reported in detail the symptoms of six patients. His accurate description of the motor symptoms remains unchallenged to present times. Dr Parkinson highlighted the fact that 'the intellect remains intact'. Dementia is not seen early in the disease process and research suggests that dementia may develop in approximately 20% of advanced cases.

The cause of the disease remains unknown (hence the term Idiopathic Parkinson's disease) and this is the case for approximately 95% of people with Pd (PWP). However, in the early 1960s it was found that the symptoms are primarily related to a deficit of the neurotransmitter dopamine as a result of degeneration of dopamine producing neurons within the substantia nigra in the basal ganglia in the mid-brain.

These dopamine producing neurons slowly die resulting in a gradual loss of dopamine and a subsequent imbalance of the neurotransmitters dopamine and acetylcholine. It is widely accepted that more than 70% of the dopaminergic neurons have been lost before the cardinal signs and symptoms reach a point when diagnosis can be made. Hence, many people affected by Pd and their families can retrospectively describe a gradual onset of symptoms.

The rationale for the gradual cell death is a longstanding topic for research and many theories exist. The most commonly

explored theories are:

- Environmental toxins
- Oxidative stress
- Genetic factors
- Multi-factorial

Environmental toxins: Parkinsonian symptoms have been linked with exposure to a range of metals and chemicals including manganese, copper, mercury, carbon monoxide and cyanide. All may result in Parkinsonism but the mechanisms of causing damage vary and post mortem examination differs from that of Idiopathic Pd. Industrial pollutants, herbicides and pesticides have also been considered as possible causative factors. Most studies in this area, however, are small and the results inconclusive or not statistically significant.

Oxidative stress: Oxidative processes are part of natural aging and take place at the cellular level. Free radicals and hydrogen peroxide are produced as part of this process and both may be toxic to cells and the membranes of neurons. As many of the symptoms of Pd are similar to common signs of aging and Pd shows marked reduction of dopamine production it has been suggested that Pd may be an accelerated aging process.

Genetic factors: A positive family history of Pd is reported in approximately 15% of cases which equates to 85% having no genetic link. In 1997 genetic studies in USA saw the identification of the first genetic mutation linked with Pd in a gene that produces the protein α -synuclein. This was a monumental discovery for several reasons; it shows that genetic study of Pd is possible; it provides researchers with a window into the disease; and it brings closer together familial and sporadic Pd. The onset of genetic Pd is often at a younger age than sporadic Idiopathic Pd.

Multi-factorial: The most recent theory regarding the cause of Pd suggests that a genetic predisposition with an environmental trigger factor may be linked to the development of the condition. Studies in this area are ongoing.

Currently, a definitive diagnosis can only be made at post mortem when the loss of dopamine producing neurons within the basal ganglia is evident together with the presence of Lewy bodies within the basal ganglia and brain stem. Lewy bodies are abnormal protein deposits which are thought to disrupt the brain's normal functioning.

Two recent Australian advances are the development of a blood screening process to identify the genes implicated in Pd and another laboratory test to measure the levels of the protein alpha-synuclein in the blood. This protein is implicated in Pd as PWP have low levels while people without Pd have high levels. These tests are not yet available.

Incidence and Prevalence

Pd is a global phenomenon recognised in all cultures. It is estimated to affect approximately 6.3 million individuals worldwide. Exact figures are not available from any source in Australia. However, the prevalence of Pd in the United Kingdom is estimated at 1.6 per 1000 and in the USA it is estimated as 1.5 million. Pd is considered to be the second most common progressive neurological condition (dementia being the most common).

Increasing age is unequivocally associated with increasing risk for Pd. Incidence is documented as 1:100 for people over 65 and much lower prior to this age. There is some evidence that the incidence decreases after the age of 85. Although Pd is related to aging it has been well documented that the underlining process is distinct to natural aging.

While Pd is more common in older people it is accepted that the term 'young onset' Pd is attributed to those diagnosed under 40 years. Prior to the age of 21 the term 'juvenile onset' may be used.

Currently the average age range of diagnosis is 55 to 65. Pd is slightly more common in males than females (ratio 5:4) and there is a slightly higher incidence in Caucasian races than in Asian and African races.

Pd may affect anyone at any time. Well known identities with Pd include Muhammad Ali, Michael J Fox, Janet Reno, Billy Graham, Pope John Paul II, Don Chipp and Adolf Hitler.

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