Progressive Supranuclear Palsy (PSP) and Cortico Basal Degeneration (CBD) are neurodegenerative diseases caused by the progressive selective death of nerve cells in the brain. This causes a disturbance of balance, voluntary movement, vision, (especially in PSP), speech and the swallowing mechanism. PSP and CBD are closely related, and in the early stages they may masquerade as Parkinson’s disease, Alzheimer’s disease or Motor Neurone Disease.

**Progressive Supranuclear Palsy (PSP)**

**What is PSP?**

Progressive Supranuclear Palsy (PSP) is a progressive neurodegenerative condition characterised by uncontrollable falls backwards, an unsteady gait, a difficulty looking up or down due to limitation of eye movement and slurred speech. Some patients also have behavioral and cognitive changes.

**Why is it called PSP?**

PSP is an acronym which stands for Progressive Supranuclear Palsy.

‘P’ Progressive – the disorder gets steadily worse over time

‘S’ Supranuclear means that the difficulties with movement occur at a high level in the brain above the final common pathway of the nuclei which process the messages to move the eyes and the limbs.

‘P’ ‘Palsy’ means a form of weakness.

**What causes PSP?**

PSP is associated with an over-production of a protein called tau in certain areas of the brain. In PSP it aggregates into clumps (called ‘neurofibrillary tangles’), which are believed to damage nerve cells. The mechanisms that underlie this process are unknown at present but it is of interest that severe head injury and some viruses and neurotoxins may also cause tau accumulation.

**So, is PSP Inherited?**

There is no evidence that PSP is an inherited disease like colour blindness or Huntington’s chorea, but there is research interest in the possibility that there may be some genetic susceptibility or risk factors that make some individuals constitutionally more at risk than others. These are the sort of genetic associations that are also being explored for common disorders like diabetes and high blood pressure.

**How many people in the UK have PSP?**

According to the most recent study there are at least 3,500 diagnosed cases of PSP in the UK, but the true figure could be much higher. It is now clear that PSP may present in many different guises making early and accurate diagnosis difficult even for neurologists.

**What are the main symptoms of PSP?**

PSP usually starts between the ages of 60 and 70 but may begin earlier, sometimes in people as young as 40. Early symptoms usually include loss of balance and unexpected falls (usually backwards), stiffness and problems with vision such as difficulty focussing or double vision. Depression, apathy, dislike of bright lights, personality changes, slurred speech and clumsiness are other common symptoms. Tunnel vision and tiny cramped handwriting are often reported by carers and there can be incontinence.
As the disease progresses, symptoms increase leading to choking bouts, recurrent falls leading to bruises and fractures, irritability and apathy, slowness of response and severe difficulties walking. Sometimes the eyelids close involuntarily and it becomes difficult to look up or down. Emotional lability is another common feature and some patients develop a tendency to repeat themselves.

Although the average life expectancy, statistically, for a patient with PSP is some five to seven years from onset of the disease, there is considerable variation in the symptoms and rate of progression of the disease in individual.

**What is the difference between PSP and Parkinson's disease (PD)?**

There is one type of PSP that can mimic Parkinson’s disease. It presents with slowness of movement, stiffness and sometimes a tremble and does not have the characteristic clinical picture of an upright stiff posture, eye movement problems, growling husky speech and poor balance which characterises PSP. This partly explains why PSP was in the past sometimes described as atypical Parkinson’s or Parkinson’s plus syndrome, terms which are no longer recommended.

**How Can I be certain of a Correct Diagnosis of PSP?**

There are no blood tests or brain scans which can diagnose PSP, although neurologists may use these investigations to exclude other conditions from the diagnosis. Accurate diagnosis of PSP depends on a characteristic constellation of symptoms and physical signs but absolute confirmation of the correct diagnosis requires a post mortem examination of the brain.

**What treatment for PSP is available?**

There are no disease modifying treatments for PSP but it is hoped that translational research will lead to trials of potential neuroprotective agents very soon. There are however a number of symptomatic therapies which may be used with benefit and patients with PSP should be seen regularly by a neurologist and a palliative care team.

**Cortico Basal Degeneration (CBD)**

**What is CBD?**

Cortico Basal Degeneration (CBD) is another degenerative tau protein brain disease that usually begins in the seventh decade of life. The cause of CBD is unknown and there is, yet, no effective treatment and no known cure. Although there are biological similarities to PSP, the clinical picture in the classical form is quite distinct.

**How many people in the UK have CBD?**

CBD is ten times less common than PSP and is considered a rare disorder. However there are a number of other pathologies that may present a clinical picture that closely resembles CBD and need to be distinguished from it, and it is also now evident that many patients found to have CBD at autopsy, present with cognitive and language disorders rather than with a movement disorder.

**What are the main symptoms of CBD?**

CBD presents classically with progressive numbness and then loss of use of one hand. Other early symptoms include jerking of the fingers, slowness and awkwardness for dexterous acts and an ‘alien limb’ (the inability to recognise the affected limb as one’s own). There is a slow, but relentless, progression of the disorder to involve firstly the arm and/or leg on one side and then the arm and/or leg on the other. There may also be disturbances of eye movements but these are normally less debilitating and prominent than in PSP. Patients with CBD also commonly complain of difficulty in controlling or using one hand, although the strength and sensation in the hand are normal. This appears to result from breakdown of the semi automatic
'motor routines' that we use in everyday life to put many complex individual muscle movements together into a highly skilled movement, for example, writing or tying up a shoelace.

In CBD the type of nerve cell damage in the brain is very similar to that seen in PSP.

**What treatment for CBD is available?**

As with PSP, there is currently no effective treatment to halt the progression of CBD. However, many symptoms of CBD can be treated and individual symptoms are usually managed in a similar way to those for PSP.

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**THE PSP ASSOCIATION**

The Mission of The PSP Association is to conquer PSP and CBD. It works in the interests of current and future sufferers of Progressive Supranuclear Palsy (PSP) and the associated condition, Cortico Basal Degeneration (CBD).

The objectives of the Association are to:

- **Promote and sponsor research worldwide into the cause, treatment and eventual cure of PSP and CBD**
- **Support affected families across Europe**
- **Provide information and raise awareness of this disease, particularly among health and welfare professionals.**

The PSP Association was formed in 1994 by Sara and Michael Koe, who were appalled by the lack of awareness and support for the disease after Sara was diagnosed with PSP. Since then, the Association has grown considerably, with over 20 staff (many part-time) and an annual income of more than £1 million.

Income is spent on research, awareness and support for patients and carers.

The Association currently allocates some £300,000 annually to research into the cause, diagnosis, treatment and eventual cure of PSP and CBD and leverages many times that amount from other institutions and public funds.

The Association was pivotal in setting up and funding the world’s first PSP Research Centre - the Sara Koe Research Centre - at the Institute of Neurology in London, which it continues to part fund.

The Medical Advisory Panel helps to coordinate world-wide research into PSP, CBD and related neurodegenerative diseases and the Association organises a biennial International Medical Workshop where research can be coordinated, progressed and presented by leading neurologists from across the world.

Support for carers and patients includes a 24/7 telephone helpline, linking subscribers to one of three nurse specialists; a carer’s information pack, full of practical guidance; local support groups and networks across the UK; the website, including the discussion forum, and an invitation to an annual Symposium, held in different locations throughout the UK, with briefings on research and support.

Meanwhile, a growing team of development officers are busily raising awareness of PSP/CBD amongst the relevant UK’s health and welfare services through briefings, presentations and
useful literature, to ensure that people affected by PSP and CBD have access to good support, when they need it.

For more information on PSP and The PSP Association, please contact:

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