



Progressive
Supranuclear Palsy

PSP

A Guide to PSP and CBD for General Practitioners and Community Nurses

Working for a world free of PSP and CBD

Contents

A Guide to PSP and CBD for GPs and Community Nurses

Introduction.....	3
The PSP Association.....	5
Progressive Supranuclear Palsy.....	6
Definition.....	6
Causes.....	6
Prevalence.....	7
Epidemiology.....	8
Familial.....	8
Symptoms.....	9
Cortico Basal Degeneration.....	11
Diagnosis.....	12
Treatment.....	12
Management of PSP.....	13
Medication.....	15
Difficult Conversations.....	18
Personal Choice.....	18
Cognition and Behaviour.....	19
End of Life Care.....	21
Support for Carers.....	22
Brain Donation.....	24
Future Treatments.....	25
Research.....	25
Useful Resources.....	26
References.....	28
Contact Details.....	31

Introduction

This booklet is part of a series of new publications for health and social care professionals, to help them support people who have Progressive Supranuclear Palsy (PSP) and Cortico Basal Degeneration (CBD).

The PSP Association actively supports both patients with PSP and CBD. References to PSP in this guide should be read, where appropriate, as PSP and CBD. Issues will be similar for both conditions, unless otherwise stated.

The purpose of this booklet is to provide General Practitioners with information about PSP and CBD and to assist them to improve the quality of life for those living with these conditions.

Decisions regarding treatment should be made by the patient, with guidance and support from their neurologist or movement disorder specialist, GP, PSP Nurse Specialist, (PSPNS), Parkinson's Disease Nurse Specialist (PDNS) and other members of their multi-disciplinary team members.

This booklet is intended as a guide only and cannot be substituted for the advice and experience of the consultant caring for the person living with PSP or CBD.

Thanks and gratitude go to the following for their advice and professional input into producing this booklet:

Maggie Rose and **Jill Lyons**, PSP Nurse Specialists,

Dr Gethin Prior, General Practitioner, South Wales,

Dr Mark Bayliss, Consultant in Older Adult Medicine, Brighton and Sussex University Hospitals Trust,

Dr Luke Massey, Clinical Research Fellow, Sara Koe PSP Research Centre,

Dr James Rowe Consultant Neurologist at Addenbrooke's Hospital and Wellcome Trust Senior Research Fellow,

Dr Boyd Ghosh, Clinical Research Associate and Honorary Clinical Fellow,

Department of Neurology Addenbrooke's Hospital,
Professor Andrew Lees, Director of the Reta Lila Weston Institute of Neurological Studies at University College London, Professor of Neurology at the National Hospital for Neurology and Neurosurgery, Queen Square, London and Chairman of the Progressive Supranuclear Palsy Association's Medical Advisory Panel,

Additional thanks go to **Hilary Kerby**, Community Matron, Warwickshire PCT.

This guide will be reviewed annually and we would welcome your feedback for improvement, which should be sent to the author, Samantha Pavey, at:

samantha.pavey@pspeur.org

Tel: 01747 841461

Or write to:

Samantha Pavey, PSP Nurse Specialist (South East)

The PSP Association,

167, Watling Street West,

Towcester,

Northants.

NN12 6BX.

The PSP Association

The PSP Association

Our mission is the conquest of PSP and CBD and the objectives of The PSP Association are to:

- Promote and sponsor research worldwide into PSP and CBD.
- Support affected patients, their families and carers.
- Provide information and engender awareness of PSP and CBD amongst the medical profession and the public at large.

Support for direct carers and people living with PSP or CBD who join the Association includes:

- Telephone advice and support by the PSP Nurse Specialists.
- A comprehensive information pack for direct carers, written in everyday language.
- Invitations to local support groups across the UK and Ireland.
- Subscription to *PSP Matters*, The PSP Association magazine, issued three times a year.
- Reduced rates for the Annual PSP Symposium, held at different locations across the UK.
- Access to the PSP website and web forum.

Support for professionals includes:

- Access to the Nurse Specialist telephone advice service.
- Training and PSP/CBD awareness sessions (available on request).

In addition, access to all of the above carer benefits is available with a subscription to The PSP Association.

For access to the PSP Nurse Specialists, to arrange a training or awareness session or to obtain a subscription form, please see our contact details on the inside back cover.

Progressive Supranuclear Palsy

Progressive Supranuclear Palsy

Progressive Supranuclear Palsy was formerly known as Steele-Richardson-Olszewski Syndrome after the three doctors who first identified it in 1964. It is now generally referred to as PSP.

PSP is a neurodegenerative disease, classified as a movement disorder. Characteristics include the inability to look up or down (known technically as a supranuclear, vertical, gaze palsy) accompanied by symptoms resembling Parkinson's disease ('extrapyramidal symptoms' of slowness and stiffness), falls and cognitive dysfunction.

Definition

Pathologically, PSP is defined by the accumulation of a special 'junk' protein called tau, which forms neurofibrillary tangles in the brain. The principal areas of the brain affected are:

- the basal ganglia (particularly the subthalamic nucleus, substantia nigra and globus pallidus),
- the brainstem,
- the cerebral cortex,
- the dentate nucleus of the cerebellum.

In PSP there is premature death of neurons in the brain, as a result of a build up of the excess tau protein¹.

Causes

Despite the recent advances in the understanding of the biology of PSP, the

1 The tau protein is the major component of the fibrillary tangles seen in several neurodegenerative disorders, known as tauopathies. The accumulation of hyperphosphorylated tau isoforms comprise the abnormal neuronal and glial inclusions.

cause of the disease is still unknown.

Almost all cases appear to be sporadic, that is to say without a family background of PSP. It has been suggested that both environmental and genetic influences may be involved. However, the latest thinking is that there is a complex genetic component to PSP. Around twenty percent of the UK population carry a gene which provides a weak susceptibility to PSP (though with a very low level of risk), but the disease itself appears then to be triggered environmentally and selectively².

Prevalence

Research shows a prevalence of 6.4 per 100,000 of the population with PSP, suggesting that there are at least 4,000 people living with PSP in the UK.

Recent epidemiological studies suggest that the disorder is more common than previously thought, is frequently misdiagnosed, and that the majority of cases are not initially referred to a neurologist.

There has been no formal research into the prevalence of CBD, but it is thought by experts to be half as common as PSP.

"In the early stages PSP can mimic Parkinson's disease and be misdiagnosed as such. The two disorders have historically been linked."

Professor Andrew Lees

2 Golbe and coworkers performed a questionnaire survey of 75 patients with PSP and matched controls. Surveyed exposures included hydrocarbons, pesticides and herbicides, urban/rural living, occupation, trauma, education level, maternal age and family history of neurological diseases. Patients with PSP were less likely than controls to have completed 12 years of education.

The most common misdiagnoses were Parkinson's disease and cerebrovascular disease.

Uma Nath's study *Epidemiology of Progressive Supranuclear Palsy in the UK* in 2002 also highlighted the large number of cases of PSP where the diagnosis is missed or delayed and how the early manifestations of PSP lead to potential referral to a wide variety of specialists.

Epidemiology

- Average age of onset is 62 years, but it can affect people over 40.
- Median interval between onset and diagnosis is 3 years.
- Slight male predominance in most studies.

Familial

Fewer than 1% of those with PSP have a family member with the same disorder. A variant in the gene for tau protein called the H1 haplotype, located on chromosome 17, has been linked to PSP, but this genetic variation is common and is not enough to cause PSP on its own.

The role of heredity in PSP is currently under investigation, but the likelihood of transmitting PSP to one's children through genetic mutations is very small. If there is a family history of dementia, Parkinson's disease or PSP, the risks increase by a small degree, but are still not high. To date worldwide there have only been seven reported families with autopsy-proven PSP spanning more than one generation.

It is now accepted that there are several subvariants of PSP. Subdivisions were initially recognized from clinical symptoms, but it is now shown that different

areas of the brain are involved.

Symptoms

There is a large variation in symptoms with each patient, each of which can occur at any point in the progression of the disease. However, not every person gets every symptom.

- Poor balance, unsteady gait with reduced arm swing. Later a curving of the spine may develop, especially the neck.
- Falls, which are often backwards, and always without warning. Falls often occur within the first year of symptoms developing.
- Nuchal rigidity (stiffness in the nape of the neck, often accompanied by pain and spasm on attempts to move the head).
- Restricted eye movement with slowing of the up and down gaze, making mobility difficult. Later, restriction of horizontal saccades can make reading, among other things, difficult. Blurred vision, tunnel vision, photophobia can also occur, while eye closure (blepharospasm and apraxia of eye opening) can render the patient essentially blind. Corneal problems may also occur due to reduced blinking.
- Difficulty with swallowing, causing choking, weight loss, requiring possible Percutaneous Endoscopic Gastroscopy (PEG) feeding. People with PSP have a tendency to overfill their mouths and forget to swallow.
- A constant risk of aspirating, which can lead to aspirational pneumonia. Sialorrhea and drooling also occur due to poor control of saliva and poor lip seal.
- Difficulty communicating as speech becomes slurred, strained and unintelligible.

- Behavioural changes including apathy and disinhibition often occur. Depression, frustration, anger and difficulty coming to terms with the diagnosis are also features that arise frequently.
- A change in the level of cognition with problems understanding reasoning, loss of short-term memory and apathy. On the whole, the capacity to make decisions remains intact, even when communication is difficult. Often the person with PSP will lose the ability to appreciate danger, i.e. will still attempt to walk alone despite frequent falls. Reckless and impulsive behaviour is commonplace.
- Later, bladder control is affected and will eventually necessitate long term management. People with PSP will experience severe constipation and will require regular laxatives.

Cortico Basal Degeneration

Cortico Basal Degeneration

CBD is a related progressive neurological disorder characterised by nerve cell loss or deterioration and atrophy of multiple areas of the brain. The progression of the disease may be slower than the progression of PSP.

CBD usually starts on one side of the body first, for example, the loss of use of one hand, eventually spreading to the other side. It can cause jerky, awkward movements (myoclonus and apraxia) and sometimes uncontrolled actions made by the hand (alien limb³). This can make it difficult to do everyday tasks such as cleaning teeth, dressing or combing hair.

In common with PSP, there may be a disturbance of eye movements, although it is less striking than in PSP and problems with up gaze and down gaze are less common.

CBD patients may have difficulties in problem solving and other behavioural changes. Memory, language and vision can be affected, but not in many patients. Like PSP, CBD is a very individual disease and the rate of progression of symptoms varies considerably from person to person.

CBD mainly affects people in their 60s and 70s and symptom management has much in common with that recommended for PSP.

CBD – similar to PSP except:-

- Numbness, jerking fingers, loss of use of one hand.
- Asymmetric; progressively affecting arm and leg.
- Alien limb.
- Less common disturbance of eye movement.
- Increased behavioural changes (frontal lobe deficit).
- Slower progression generally.

3 Complex unintentional movements of one limb interfering with normal tasks.

PSP and CBD are so closely related that we often refer to them interchangeably (usually as 'PSP'), and The PSP Association supports people living with both.

Diagnosis

To date, definitive diagnosis of PSP can only be made by post mortem examination of the brain. Specialists can make the diagnosis with over 90% accuracy. A means to achieve earlier and better diagnosis would be of huge benefit to patients and would enable any new treatments to be administered at the earliest opportunity. Patients may wait 2-3 years before the diagnosis is reached.

Initially many of the symptoms present as idiopathic Parkinson's Disease (IPD) and it may be some time until their symptoms develop leading the specialist to suspect PSP. The neurologist may allow further time to elapse before being able to give a probable diagnosis.

Imaging techniques, including positron emission tomography, single photo emission computed tomography and magnetic resonance spectrometry, have improved the accuracy of diagnosis of parkinsonian syndromes. Magnetic resonance imaging has shown atrophy of midbrain structures in PSP and also of the pons. There may be some demyelination and gliosis of the superior cerebellar peduncle in PSP. At present, however, an MRI scan will usually be used to rule out other possible diagnoses.

Treatment

There is currently no treatment to stop or slow down the course of PSP, or to cure PSP. Management will be based around symptom control and quality of life.

Management of PSP

The multi-disciplinary team offers the best approach to management and works towards improving quality of life. For most patients it helps if the GP refers to the following team members once a possible diagnosis has been made by the neurologist:

- If the neurologist has not already referred the patient to the local Parkinson's Disease Nurse Specialist (PDNS), the GP should do this. The PDNS will usually cover PSP and CBD within their role. They will have knowledge of medication usage, be able to advise and monitor any changes and also know which services are available to the person with PSP/CBD locally. They can liaise with the PSPNS if required.
Some people will also benefit from support at local PD groups.
- Referral to The PSP Association which will allow the person access to the PSP Nurse Specialists and the telephone helpline and gain an awareness of their illness. Joining the Association will also allow their details to be stored on the database for future research/drug trials. They will also receive invitations to their nearest PSP support group.
- Early referral to a Physiotherapist (PT) (neuro physiotherapist if available) will allow gait, balance and falls education, as well as advice regarding bed/chair transfers, walking aids and wheelchair services.
- Referral to an Occupational Therapist (OT) as required for assessment of functional ability, adaptations to the home and garden, aids for eating, bathing, etc.
Stairs are a particular hazard when eye movements are affected.
- Early referral to a Speech & Language Therapist (SLT) who will be able to advise on communication needs and also assess swallowing difficulties.
As silent aspiration can become an issue the team should be guided by the

regular input and advice from the SLT.

- Referral to a Dietician who can advise on calorific intake (weight loss is common) and types of foods to avoid/eat safely as dysphagia worsens. The Dietician will liaise with the SLT if PEG feeding becomes necessary and the person with PSP chooses this option.

- Referral to the Community Matron (CM) who will often case manage the complex issues of the condition, with support from the MDT.

Where there is no CM referral should be made to the district nurse for support and case management.

- Referral to an Orthoptist or eye specialist. As the ocular function itself is unaffected, it is not always necessary to refer to an optician. However, other ocular problems (cataracts, glaucoma) must not be ignored. The orthoptist can advise on the effectiveness of prism glasses.

Some patients, especially those with CBD, may qualify for a certificate of sight impairment, allowing access to talking books, etc.

The GP can treat any infections of the conjunctiva and the use of artificial tears is helpful with the reduced blinking in PSP.

- As the illness progresses, referral to the Continence Nurse Advisor will be beneficial to allow the person with PSP support to manage their bladder dysfunction (urgency/frequency/nocturia/retention) and constipation which can be severe.

N.B. Early bladder involvement may indicate a diagnosis of Multiple System Atrophy (MSA⁴) rather than PSP.

- The Social Worker can assist with a needs assessment and carer assessment

⁴ MSA is a similar movement disorder characterised by parkinsonism, autonomic dysfunction and cerebellar problems. www.msaweb.co.uk

and then help to provide an appropriate package of care. As the illness progresses they can also advise on regular respite periods or suitable long term nursing care homes if this becomes necessary. The PSP nurse specialists are available to give teaching sessions to care homes, hospices and care agencies if required.

- Referral to a Community Psychiatric Nurse (CPN) may be required if the person with PSP develops behavioural problems or signs of early dementia. Hallucinations and psychotic episodes can also occur, although often in association with medications or infection. Unresponsive depression may require a referral to a psychiatrist or a counsellor.
- Palliative / Hospice Outreach Team. As the illness progresses, respite for the carer becomes necessary. As many care homes will be unfamiliar with the person with PSP/CBD's needs, respite periods can vary in success. Fortunately, hospices now cater for long term neurological conditions, and regular respite periods can thus ensure an excellent standard of care for the person with PSP, and offer essential support for the carer. When end of life care is then required, the person with PSP and their families will be better supported by staff they already have a rapport with and who understand the complexities of a neurodegenerative illness. They are also skilled in broaching end of life issues and assisting people to consider their end of life wishes, so early referral to the palliative team can be very helpful.

Medication

There is no specific treatment for PSP, but the medications used for Parkinson's disease are often prescribed by the specialist in movement disorders. For around one third of people there will be limited symptom improvement. In general,

they do not provide dramatic or long lasting improvements and can cause unwanted side effects.

Levodopa

Sinemet / Madopar is often tried in the early stages and may also be used to rule out Parkinson's disease. Small doses are tried and then titrated up gradually to prevent side effects; commonly nausea for the first few weeks, which can be improved with domperidone (other antiemetics may make the extra pyramidal symptoms worse).

Levodopa may help with initiation of movement, muscle rigidity, and bradykinesia. If it doesn't benefit the person with PSP, it can be reduced slowly and discontinued, although a trial of at least 3 months should be allowed.

Dopamine agonists

Drugs such as Mirapexin (Pramipexole) and Ropinirole (Requip) may be tried to improve the parkinsonian symptoms. This is very individual and it may take some time to find a working dose. These medications may be used in conjunction with Levodopa.

Rotigotine (neupro) patches may be useful when swallow is impaired.

Amantadine (Symmetrel)

Amantadine is considered by many neurologists to be one of the best treatments available for PSP. It can be helpful for improving balance and preventing falls. There is no research based evidence for Amantadine but anecdotal studies show it is beneficial in some cases. It can be used in conjunction with Levodopa or dopamine agonists. In some patients it can cause hallucinations which are dose dependant.

Amitriptyline

This can often be helpful for improving sleep, lifting mood and can also be

beneficial for pain from muscle rigidity. A small dose at night can be increased slowly as required. This may also help with drying up secretions. Higher doses may lead to confusion or cognitive dysfunction.

Atropine

In the form of eye drops used sublingually, this can be very helpful with sialorrhoea and is often the first line of treatment. **(N.B. it is not licensed for use in the mouth)**. Normal dose is Atropine 0.5% two drops t.d.s. If not effective, Glycopyrronium (Glycopyrrolate) can be used in tablet form (or in liquid for a PEG) to aid sialorrhoea. The use of hyoscine should usually be avoided as this can often cause confusion and hallucinations in PSP/CBD.

For severe sialorrhoea, the patient may consider Botulinum toxin injections to the salivary glands, but this is usually performed by an ENT surgeon and therefore may not be available in all areas.

Botulinum Toxin

Often offered by neurologists in a specific botox clinic, this may be beneficial for involuntary eyelid closure. It may last up to 4 months and therefore needs to be repeated regularly. It may not be effective in all cases.

Painful or disabling Dystonia in some neck muscles, hands and feet may also respond to botox injections.

Muscle relaxants

These may be helpful for muscle rigidity, the commonest used are Baclofen 10mg t.d.s. and Clonazepam 0.5mg nocte.

Clonazepam may also benefit sleep disorders.

Gabapentin may be of benefit for neuropathic pain. However, this is difficult to treat, and it may be necessary to refer back to the neurologist and to a pain clinic if required.

Laxatives

Will be required on a daily basis for the majority of people living with PSP. Despite a healthy high fibre diet, constipation will be a common symptom, often severe. Movicol would be used initially, with other laxatives added as required. Parkinson's disease medications will be ineffective if the person with PSP is constipated.

Pain Control

Although not common, pain relief may be required at end of life. Diamorphine will often be used by the palliative care team if anxiety is present.

Difficult Conversations

In many instances, difficult conversations about end of life issues are put off by families as being too difficult to address. The PSP Nurse Specialists frequently take calls from distressed family members who have not felt able to discuss end of life issues and who are now faced with advocacy for the person with PSP, often not knowing what their wishes are. Often the professional will need to broach these subjects whilst the person with PSP can still make their wishes clear as verbal communication can become increasingly difficult as the disease progresses.

Personal Choice

For many people with PSP the time will come when they lose the ability to swallow safely and following assessment by the SLT, the neurologist may suggest PEG feeding. The person with PSP should be supported to make an informed decision regarding this. For many, once artificial feeding is in place, they will no longer struggle to get adequate nutrition and hydration and may indeed gain weight. For others, they may decide their quality of life is so poor that they

no longer wish to prolong their life with artificial feeding. Patients should be supported in either decision. If a PEG is declined by the person with PSP, it is often the family members who have difficulty accepting this decision.

It is also important to know the individual's wishes with regard to admission to hospital, treatment with antibiotics, and resuscitation measures.

As it is so difficult for a spouse or family member to act as an advocate without knowing the person's wishes, the PSP Nurse Specialists try to encourage discussions early on. This enables the person living with PSP to consider advance decisions and enduring power of attorney.

The PSP Association has information leaflets on advanced directives (living wills) and the PSP Nurse Specialists can discuss this with the patient and their family.

Cognition and Behaviour

Cognitive dysfunction and personality changes are common, but they are not considered to be a 'true dementia'. There is some debate regarding this.

Steele Richardson et al describe:

- forgetfulness,
- slowing of thought processes, emotional or personality changes (apathy or depression with occasional outbursts of irritability),
- impaired ability to manipulate acquired knowledge.

In many neurological disease states associated with subcortical pathology a similar pattern of dementia exists. The neurobehavioural changes of progressive supranuclear palsy thus typify a clinical pattern which may be referred to as subcortical dementia.

PSP patients can experience cognitive and behavioural changes suggesting

abnormal functions in the frontal lobes. Cognitive changes consist of a decline in frontal lobe functioning, such as slow information processing and retrieval, concrete thinking, impaired reasoning and difficulty in planning tasks. Behaviourally, patients often exhibit apathy, leading to decreased motivation.

Family members can find the personality changes very distressing and often state this as the most difficult symptom to deal with.

Key Issues:

- A delayed response
- Poor judgement
- Lack of self awareness
- Difficulty in organisational skills and sequencing
- Lack of motivation
- Lack of empathy.

These are generally more prominent later in the disease course.

Most people living with PSP express a sense of isolation when first diagnosed. As many professionals will not be familiar with PSP the person can feel very uncertain and alone. When coming to terms with the diagnosis and changes to the life they may have planned for themselves they can experience a sense of loss and grief. Antidepressants and anxiolytics can help ease these symptoms and may also help with the emotional lability.

As one's intellect remains largely intact, the person with PSP will be aware of their general deterioration and will naturally be very frightened, however they also have a lack of awareness regarding their abilities and will continue to attempt to walk unaided.

Loss of higher intellect, reckless behaviour and the added problem of not being able to look down, all contribute to the possibility of falls and injury. People

with moderately advanced PSP are at higher risk if left alone due to the risk of falling or choking.

End of Life Care

Although PSP has an individual rate of progression, deterioration is inevitable and the person living with PSP may eventually become immobile, with severe communication difficulty, poor swallow, restricted vision, incontinent, yet with their intellect largely intact. It is very difficult to give an accurate individual prognosis.

For most people end of life will occur within 5 to 9 years from the onset of symptoms, but there is very wide variation in life expectancy.

Frequent bouts of infection (UTI or pneumonia) with poor recovery in between, increased apathy, increased periods spent sleeping, weight loss, worsening bulbar symptoms (speech and swallowing), may all indicate progression of the disease. Often infections are difficult to diagnose as pyrexia or other symptoms may not always be present, but early treatment with antibiotics is crucial to be effective.

Many people living with PSP (and their families) fear that their end of life will be caused by a traumatic choking episode and may be painful. This is rarely the case and they require reassurance regarding this. The commonest cause of death is from aspiration pneumonia, which can be peaceful and pain free if managed well. Towards end of life, anxiolytics and analgesia may be required. Involvement of the palliative care team can ensure that those who remain alert can have appropriate medication to ease any anxiety and pain relief if necessary.

PSP should be included on the GP palliative care register and included as part of the Gold Standards Framework.

Support for Carers

Support for Carers

Being a carer for someone with a long term condition can be overwhelming. There can be many strains upon the relationship especially where communication and empathy from the patient is restricted or cognition issues are present. Carer stress is common and they may experience any of the following:

- guilt
- disbelief
- helplessness
- anger
- depression
- sleep disturbance
- inability to concentrate
- weight loss/gain
- inability to cope
- anxiety about loss of income
- embarrassment
- lack of knowledge
- exhaustion.

Often the carer will require psychological support (and even sometimes antidepressants) in order to cope with the illness. Regular support and respite periods will be necessary in order for the carer to continue to deal with supporting the person living with PSP/CBD. Professional counselling may also be required.

Referral to support groups, carers groups and local organisations may be beneficial and may make the carer feel less isolated. Hospices do not treat a patient in isolation and a lot of the services they offer to the person living with

PSP/CBD are available to their carers also.

Social Services should carry out a carer's assessment to ensure their needs are being catered for.

Many of the carers will not ask for help until they reach crisis point; GP's and Community Matrons are in a position to prevent this.

Carers should be registered as carers on the practice register and be given priority for treatment. Most people living with PSP should be booked for double appointments due to slowness of speech and movement. Home visits will be necessary as the disease progresses.

N.B. Where the term 'carer' is used throughout the text the author is referring to the main family member caring for the person with PSP, rather than the paid carer/professional.

Brain Donation

Brain Donation

Brain Banks have been established to collect tissue samples and whole brains donated by people with PSP and others with neuro-degenerative disease.

Examples include the Queen Square Brain Bank in London and the Cambridge Brain Bank. These are an invaluable resource for research which will enable researchers to better understand what causes PSP/CBD and how the diseases progress. In the long term, this will enable us to develop a much needed diagnostic test and an effective treatment.

The Sara Koe PSP Research Centre (SKRC), the first dedicated PSP Research Centre in the world, was established in January 2002 at the Institute of Neurology in London. The SKRC's role is to co-ordinate PSP research within the UK, provide support to those involved in PSP research and to act as a link with PSP research worldwide.

The SKRC administers a PSP DNA and Brain Bank and is closely linked to the Queen Square Brain Bank which stores the brains of people who have died from various neurodegenerative diseases as well as the brains from disease free donors.

People living with PSP will often enquire about brain donation, as will their family members who wish to do something positive to help with research. In the first instance they can discuss this with their specialist, or call the telephone number below for information and support.

Queen Square Brain Bank Contact Details

Susan Stoneham Tel: 020 7679 4266 or 0207 837 8370.

Cambridge Brain Bank Contact Details

Beverley Haynes / Jenny Wilson Tel: 01223 217336

Other local brain banks can be located at:

www.hta.gov.uk/donations/howtodonateyourbody/donatingyourbrain.cfm

Future Treatments - Stem cell treatment

For many people with neurodegenerative diseases, including PSP, the potential of stem cell treatment lies in the possibility that it could lead to ways of producing new neurons to replace those lost in the brain through the PSP/CBD process. Whilst exciting, stem cell research is still in its infancy and there is a long way to go before treatments are available.

It is important that this is recognized as all too often stem cell research is overhyped and misrepresented in the media. This gives false hope to those who find themselves in very desperate situations.

Research

The PSP Association is the only UK organisation dedicated to funding research which aims to:

- develop a better understanding of the causes of PSP
- establish the best ways of caring for people with PSP
- develop an effective treatment for PSP
- find a cure for PSP.

Ongoing Research at The PSP Association

- Control of tau gene expression.
- High field quantitative Magnetic Resonance Imaging in PSP.
- Randomized placebo-controlled trial of valproic acid in patients with PSP.
- Cerebrospinal fluid biomarker discovery in PSP and related tauopathies.
- Haplotype regulation of alternative splicing at the MAPT locus.

Useful Resources

Useful Resources

Carers UK Equal Partners

Tel: 0808 808 7777

Web: <http://www.carersuk.org>

Counsel + Care

Tel: 0845 300 7585

Web: <http://www.counselandcare.org.uk>

Crossroads Caring for Carers UK

Tel: 0845 450 0350

Web: <http://www.crossroads.org.uk>

Cure PSP

Web: <http://www.psp.org>

Department for Work and Pensions

Web: <http://www.dwp.gov.uk/directgov>

Disabled Holiday Directory

Tel: 01348 875592

Web: <http://www.disabledholidaydirectory.co.uk>

National Care Forum

Tel: 024 7624 3619

Web: <http://www.nationalcareforum.org.uk>

National Care Association

Tel: 020 7831 7090

Web: <http://www.nationalcareassociation.org.uk>

NHS Carers Blog

Web: <http://talk.nhs.uk/blogs/carers/default.aspx>

NHS Carers Direct

Tel: 0808 802 0202

Web: <http://www.nhs.uk/Carersdirect/Pages/CarersDirectHome.aspx>

Princess Royal Trust for Carers

Tel: 0844 800 4361

Web: <http://www.carers.org>

Remap Charity

Tel: 0845 130 0456

Web: <http://www.remap.org.uk>

Respite Care

Tel: 0845 644 4932

Web: <http://www.bettercaring.com>

Telecare Services Association

Tel: 01625 520320

Web: <http://www.telecare.org.uk>

References

References

Eric R Eggenberger, DO, MS, FAAN, Zeba F Vanek, MD, MBBS, DCN, David Clark, DO, (2010) **Progressive Supranuclear Palsy**, *Medscape emedicine*
<http://emedicine.medscape.com/article/1151430-overview>

Rodger J. Elble, MD, PhD, Professor and Chair of Neurology, Director, Neurology Residency Program, Southern Illinois University School of Medicine, (1987 - 2008) **Progressive Supranuclear Palsy**, The National Organization for Rare Disorders (NORD)

Steven Karceski, MD, (25/11/2008) **Patient page. Progressive supranuclear palsy**, *Neurology*, 71(22):e70-2.

Progressive Supranuclear Palsy, The Association for Frontotemporal Dementias (AFTD), <http://www.ftd-picks.org/frontotemporal-dementias/disorders/psp>

Steele J C, Richardson J C, Olszewski J, (Apr 1964) **Progressive Supranuclear Palsy. A heterogeneous degeneration involving the brain stem, basal ganglia and cerebellum with vertical gaze and pseudobulbar palsy, nuchal dystonia and dementia**. *A. M. A. Archives of Neurology*, 10:333-59

E. Tolosa, F. Valldeoriola, M.J. Marti, Neurology Department, Hospital Clinic, Faculty of Medicine, University of Barcelona, Spain, (1994) **Clinical diagnosis and diagnostic criteria of progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome)**, *Journal of neural transmission*.

Supplementum, 42:15-31

Williams D R, de Silva R, Paviour D C, Pittman A, Watt H C, Kilford L, Holton J L, Revesz T, Lees A J. The Queen Square Brain Bank for Neurological Disorders, University College London, UK, (Jun 2005) **Characteristics of two distinct clinical phenotypes in pathologically proven progressive supranuclear palsy: Richardson's syndrome and PSP-parkinsonism.** *Brain* 128(Pt 6):1247-58.

Williams D R, Holton J, Strand C, Pittman A, de Silva R, Lees A J, Revesz T. The Queen Square Brain Bank for Neurological Disorders, University College London, UK, (2007) **Pathological tau burden and distribution distinguishes progressive supranuclear palsy-parkinsonism from Richardson's syndrome.** *Brain* 130(6):1566-1576.

Further Reading

<http://www.brt.org.uk/dr-rohan-de-silva>

<http://www.brt.org.uk/prof-john-hardy>



Notes

Contact Details

The PSP Association

PSP House

167, Watling Street West

Towcester, Northants, NN12 6BX

Tel: 01327 322410

PSP Nurse Specialists:

Katie Rigg (North East England & Scotland) Direct Line: 01434 382564

katie.rigg@pspeur.org

Kat Haines (North West England, N. Wales & Ireland)

Direct Line: 01995 601533 kat.haines@pspeur.org

Samantha Pavey (South East England) Direct Line: 01747 841461

samantha.pavey@pspeur.org

Jill Lyons (South West England & S. Wales) Direct Line: 01934 842366

jill.lyons@pspeur.org

Fax: 01327 322412

E-Mail: psp@pspeur.org

Web: www.pspeur.org

The PSP Association is a Company limited by guarantee Registered number: 2920581

Registered Charity numbers: England and Wales 1037087 / Scotland SC041199

Registered Office: PSP House, 167 Watling Street West, Towcester, Northamptonshire NN12 6BX

