

Progressive
Supranuclear Palsy

PSP

**A Guide to Progressive
Supranuclear Palsy (PSP)
and Cortico Basal
Degeneration (CBD)**

Working for a world free of PSP

A GUIDE TO PSP AND CBD

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A GUIDE TO PSP AND CBD

Overview

This Guide to Progressive Supranuclear Palsy (PSP) and Cortico Basal Degeneration (CBD) has been edited and approved by Professor Andrew Lees, Consultant Neurologist at the National Hospital for Neurology and Neurosurgery, Queen Square and Director of the Reta Lila Weston Institution of Neurological Studies, University College London. Professor Lees is also the Chairman of the Progressive Supranuclear Palsy Association's Medical Advisory Panel.

This Guide provides information to assist health and social care professionals in the support of people with PSP or CBD. It complements:

- 'A Physician's Guide to PSP' in DVD format, issued to assist accurate early diagnosis. This has been issued to all consultant Neurologists, Geriatricians and Ophthalmologists in the UK, as well as to all PD Nurses and Speech and Language Therapy centres.
- the Carers' Information Pack which gives practical and comprehensive advice, symptom management and support and financial advice to carers of people with PSP or CBD.

Copies of all three publications are available from The PSP Association.

About PSP and CBD

Progressive Supranuclear Palsy (PSP)

PSP is a neurodegenerative disease involving the progressive death of nerve cells in the brain stem and basal ganglia. In this area lies the substantia nigra, which is also affected in Parkinson's Disease (PD). Damage to the substantia nigra accounts for the slowness and awkwardness of movement that the two diseases have in common. However, several other important brain regions are affected in PSP that are normal in PD and some of the microscopical abnormalities in PSP more closely resemble those found in Alzheimer's Disease (AD) or the punch drunk syndrome of boxers.

The distribution of these lesions is, however, markedly different in AD. Symptoms generally appear first in the sixth or seventh decade of life, however, as diagnosis improves cases from the early 40's are becoming more common, too. PSP can present with a wide variety of symptoms, in different orders, that progress at different rates. Symptoms include:

- Balance and Movement. Frequent and unexpected falls are common. The falls are often backwards and can result in severe injury with fractures. Strategies to avoid falls and minimise their impact when they do occur should be recommended. A motor recklessness compounds the risk of falls with impulsive behaviour.
- Vision. Eye movement problems are common. Upgaze and, more specifically, downgaze are usually affected. The inability to look down can lead to falls, and the inability to see what is on a plate can lead to messy and distressing eating habits. Double or tunnel vision may occur, although the ability to focus directly ahead is usually retained. The inability to scan smoothly from side to side can lead to an inability to read. Involuntary closure of the eyelids can lead to functional blindness despite normal visual acuity, giving the inaccurate impression of sleep or disinterest. A dislike of bright lights may occur and the blink rate is also slowed, leading to soreness of the eyes.
- Speech. The speech is often slow and slurred. It loses its tone and has a growling quality. Repetition of words or phrases can occur.
- Swallowing. Choking especially on liquids is a common handicap and difficulty swallowing solids may lead to weight loss. Some people require assisted feeding through a gastrostomy (PEG) to avoid recurrent chest infections and restore adequate nutrition.
- Behavioural Changes. Apathy, irritability or truculent behaviour may present, although more rarely. Many people become quieter and quieter and communication becomes harder. Mood swings may occur.
- Bowel and Bladder Function. Constipation and difficulty in initiating urine flow can be common. A need to pass urine several times during the night due to an unstable bladder is common.

Throughout the management of these symptoms, it is important to appreciate that although communication may be restricted and there may be behavioural problems, intelligence is for the most part unaffected.

Cortico Basal Degeneration (CBD)

Cortico Basal Degeneration (CBD) is another neurodegenerative disease that causes disturbances of movement and behaviour and most commonly presents in the sixties and seventies. It is much less common than PSP, although it is pathologically similar, and symptom management has much in common with that recommended for PSP.

CBD causes the loss of use of one hand. A stroke or brain tumour may be initially suspected. Other early symptoms include jerking of the fingers, slowness and awkwardness for dextrous acts and a feeling that the affected limb no longer is part of the body. Patients may additionally have disturbances of higher sensory function and problems with the execution of complex motor tasks, leading to difficulties in everyday activities like cleaning their teeth or dressing and combing hair, despite reasonably normal strength. Unlike PSP, CBD often initially presents asymmetrically, but it progressively spreads to affect the other side of the body. In common with PSP, there may be a disturbance of eye movements, although it is less striking than in PSP and paralysis of vertical eye movements is uncommon.

CBD patients may also have difficulties in problem solving even though memory is relatively unaffected. Depression, apathy, irritability, agitation and disinhibition can occur.

Pathology

Although CBD and PSP share some clinical similarities, the abnormalities seen by the pathologist at post-mortem are quite characteristic, allowing the two related disorders to be distinguished. Sometimes patients who are considered to have CBD in life are found to have the pathological changes of PSP. The biological pathological processes which occur in the two disorders are, however, similar making it appropriate to link them together. *The PSP Association therefore embraces people with CBD and sponsors research into this disorder.*

Incidence and Prevalence

PSP is much more common than previously believed. Recent research gives an incidence of 5.3 per 100,000 of people over 50 years of age, and a prevalence of 6.4 per 100,000 of the whole population, which means that PSP is at least as common and as devastating as, for example, Motor Neurone Disease. There has been no formal research into the prevalence of CBD, but it is thought by experts to be half as common as PSP. Cases of both PSP and CBD are often labelled as Parkinson's plus or atypical parkinsonism when an element of doubt exists about the precise diagnosis.

Text Reference

References to PSP in the rest of this guide should be read, where appropriate, as PSP and CBD.

Health and Welfare Support for those with PSP

The GP and Neurologist

Best management of PSP requires an integrated multi-disciplinary approach with early palliative care intervention. Contact should be made with the relevant health professionals at an early stage. There are no current treatments to extend the course of the disease, but there are a range of drugs and treatments available to assist with symptom management.

As a patient, your GP is your 'gatekeeper' to the NHS and will have seen you and referred you to a neurologist for accurate diagnosis. They will be the lynchpin in putting together a multi-disciplinary team to oversee the care and support you may need. Your neurologist will advise on drugs and treatment.

Further visits may be arranged to monitor the progress of your disease and any changes in prescription. If you are concerned or unhappy about a diagnosis, or lack of diagnosis, you are entitled to ask for a second opinion. However, you should be aware that PSP is a particularly difficult disease to diagnose, especially in the early stage. The PSP Association Nurse Specialists attend clinics in London, Cambridge, Newcastle,

Manchester and Cardiff. Otherwise, you may see a Parkinson's Disease Nurse Specialist. In either case their role is to support your neurologist.

Drugs Several drugs may assist symptom management:

- Amantadine has gained a reputation as a symptomatic therapy to help PSP, although there is no convincing evidence base yet.
- L-dopa (Sinemet or Madopar) provides an effective treatment for PD, but unfortunately L-dopa only benefits some people with PSP. It can sometimes help the slowness, stiffness and balance problems of PSP, but usually not the speech, vision or swallowing difficulties. It usually loses any initial benefit after two or three years.
- Amitriptyline, although an anti-depressant drug, has been shown to have a mild effect in improving the movement problems in PSP. The dosage should be increased slowly to at least 100 mg per day (taken divided into at least two doses) for an adequate trial in PSP. It is also a good sleep medication for elderly people and may benefit an unstable bladder.
- Botulinum Toxin can be an effective treatment for involuntary eyelid closure (blepharospasm). A very dilute solution of the toxin can be carefully injected by a neurologist into the eyelid muscles. However, the duration of action is around three months and the injections regularly have to be repeated. Botox can also be used for involuntary turning or bending of the head that occurs in PSP, but injection of botulinum toxin into the neck muscles can sometimes cause slight weakness of the swallowing muscles. In PSP, where swallowing is already impaired, caution should be exercised when considering use of botulinum toxin in neck muscles.
- Experimental Drugs. Physostigmine, idazoxan and efaroxan have all been tried as treatments for PSP and have not been found to be helpful. A trial was carried out in 1996/7 into the possible beneficial effects of nicotine patches in PSP. Unfortunately, there was insufficient evidence to convince neurologists that any symptomatic benefit arises from their use. Aricept, which was found to be helpful in AD, was

trialled in the USA in 1999/2000, but the results did not justify a further - then planned - UK trial. In 2002, funding was made available by the European Union for a trial of Riluzole after it was shown to have mildly beneficial effects in MND. Patients were accepted for this double blind trial up until the end of June 2002, but the results were disappointing and no proven benefit to PSP patients has been established.

- Future Drug Trials. Updates on future studies and drug trials will be published in The PSP Association's Magazine, *PSP Matters*, issued every 4 months. There is recent interest in carrying out trials with lithium and sodium valproate as these drugs have been shown in mice to limit the production of Tau protein. However, although lithium is currently licensed, the required dosages are likely to be significantly higher than currently used and there are concerns about side effects. Interest also remains in the possible value of the complementary substance Co-Enzyme Q10 as a disease modifying treatment.

The PSP or PD Nurse Specialist

The UK PSP Nurse Specialists operate a telephone link to patients and carers who subscribe to The PSP Association and to health professionals who support people with PSP. The PSP Nurses also attend PSP clinics in London (at the National Neurological Hospital), Cambridge, Cardiff, Newcastle and Manchester. PD Nurses throughout the UK often provide an advice service to those with PSP. For PSP Nurse Specialist telephone numbers, see the Contact Details on back cover.

The Physiotherapist

Physiotherapists can provide support and practical advice to patients and their carers in the management of PSP and help the latter in maintaining mobility, as described below:

- Mobility. Features such as falling backwards, hyperextension of the cervical spine, and rigidity can be helped by physiotherapy. Walking aids may be useful for patients with both PSP and CBD, such as wheeled or weighted frames. Individual assessment is always needed as walking aids may not always be appropriate. Gait, balance and falls

education and advice for carers may improve both gait pattern and safety. Physiotherapists also have an important role in footwear advice and identifying, where appropriate, if there is a need for an orthotics review. Referrals to local wheelchair services may be indicated for mobility, when walking becomes unsafe.

- Bed mobility, sit-to-stand transfers and stairs are often difficult due to rigidity, stiffness and balance deficits, therefore re-education of the patient and advice to carers is necessary. Close liaison with occupational therapists to provide advice on the suitability of bed mobility equipment, rails, stair lifts and the home environment may be necessary to ensure safety and promote independence.
- Exercise. Progressive rigidity and slowness of movement causes altered soft tissue length and alignment and joint stiffness, which further contributes to mobility difficulties, alters posture, impairs breathing and coughing and causes pain. Physiotherapy plays an important role in maintaining flexibility through provision of a specific exercise regime based on the individual needs of the patient, including stretches, strengthening, balance and aerobic exercises. This will require ongoing review as the disease progresses. Carers need to be involved if the patient is unable to perform a regime independently or requires support to ensure the regime is completed correctly.

Regular exercise is also often beneficial for an overall sense of psychological well-being, and to guard against the complications of immobility. Certain exercises done in the home on a regular schedule can keep the joints loose. Both physiotherapists and occupational therapists have key roles to play in helping reduce the risk to PSP patients of falls, which can cause fractures. For specific exercises, consult a physiotherapist.

- Walking Assistance. Walking aids are often required. The physiotherapist will perform an individual assessment and advise on the appropriate walking aid. A physiotherapist's advice should be sought before purchasing expensive equipment. Because of the tendency to fall backwards a heavy, sturdy frame may be recommended.
- Chest Care. Physiotherapists, in conjunction with speech and

language therapists, have an important role in the prevention and management of chest infections, including breathing exercises and manual techniques. Postural and positioning advice during feeding may be indicated if the patient's swallowing is impaired.

The Speech and Language Therapist (SLT)

The SLT has a key role to play in the management of PSP, and early referral is important as both speech and swallowing difficulties will occur over time.

- Speech. The movements of the lips, tongue and throat may all weaken and speech will become quiet and slurred. In some cases, the patient will develop a stammering type of speech. This, coupled with the difficulties of eye contact, forgetfulness and personality change, can make communication difficult. The SLT should recommend appropriate exercises and communication aids, recognising that movement and eye problems may limit the utility of some of the more sophisticated speech aids such as Lightwriters.
- Swallow. Swallowing can be difficult due to weakness of the throat muscles. Slowness in eating may be an early feature and there is a tendency to 'overflow' the mouth, increasing the risk of choking. The cough reflex is often weakened in PSP and therefore, if food or drink is aspirated, it is difficult for the patient to clear the chest. This can result in obstruction of the airway or the slow build up of matter in the lungs leading to chest infections and there is a significant risk of aspiration pneumonia.
- Monitoring. The deterioration in the speech and swallow should be monitored, and advice given on improving communication and the safest consistencies for eating. This may involve recommendations to thicken liquids or to have a soft or pureed diet; advice can be given regarding positioning. Sometimes a videofluoroscopy will be used to assess and monitor the risks. A feeding tube 'PEG' may be recommended, either to supplement oral feeding, or, in the later stages of the disease, as the method of choice for nutrition and hydration.

The Occupational Therapist (OT)

The key priorities for the occupational therapist are to maximise safety of the patient in every day activities, such as self-care, productivity and leisure. An assessment of the patient's functional ability will help the occupational therapist identify the individual's physical and cognitive skills/difficulties and offer help and advice, particularly around the house, including:

- Home safety advice. Since falls are often backwards, serious and without warning, sharp objects or projections could result in serious injury. This is especially likely in areas where a change of direction, step or floor surface is encountered, and will be exacerbated by difficulties in looking down. Walkways in the home should be kept clear of objects such as rugs or low furniture, and good lighting provided to compensate for visual difficulties. A full risk assessment should be carried out.
- Assistance with bathing, transfers or mobilising around the home. Home modifications or equipment will be required as mobility becomes restricted.
- Practical Strategies. Occupational therapists can advise the individual and family members on practical strategies to help compensate for their difficulties, within specific tasks. This may include reviewing the individual's seating and positioning; and raising/adjusting the height of utensils/aids for reading, writing or feeding. Using alternative techniques in meal preparation or car/bed transfers may also be appropriate to accommodate the individual's physical and visual limitation. There are also a number of strategies such as using familiar routine or simple prompts that can be effective in helping carers cope with a decline in the person's cognitive abilities.
- Liaison. Liaison with other therapists such as the speech and language therapist or physiotherapist is important to ensure a thorough assessment and provide a consistent approach across all therapies.

The occupational therapist can assist the family and carers in adjusting to their changing needs and roles over time, initiate referral to other services and provide information on community resources.

The Ophthalmologist and Orthoptist

Eye movement and eyelid problems cannot be corrected by the use of glasses. Problems directing the person's attention to objects located below the eyes present a difficult management problem. Infections of the conjunctiva related to absent blinking will need antibiotic treatment and artificial tears will prevent discomfort. An ophthalmologist and orthoptist can assess vision capability and advise on the ability to drive a vehicle safely and, where appropriate, provide a certificate of blindness (giving access to talking books and other support). Anyone with PSP should also notify the DVLA as soon as a diagnosis is received. (Not to do so is a criminal offence.)

The Social Worker

The Social Worker can assist with a needs assessment for the person living with PSP and carer assessment 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 social worker will also advise on benefits available and accessing local services.

The PSP Association

Formation in the UK

In September 1990, a small number of PSP patients in the USA formed the Society for Progressive Supranuclear Palsy, now known as Cure PSP. Encouraged by their success as a lobby group, a similarly minded PSP family in the UK, with the help of generous friends, set up a sister charity in England in April 1994.

The Association also supports carers of those with PSP and CBD across Europe and encourages other countries to set up their own Associations. France and Germany have their own PSP Associations and there are embryonic PSP groups in Italy and Spain. Elsewhere in Europe, the European Parkinson's Disease Society Newsletter in some countries includes a section on PSP. Patients with PSP have suffered and been isolated, in particular, because of lack of research and effective treatment and lack of knowledge about this disease, even amongst relevant health and welfare professionals.

Objectives

Our mission is the conquest of PSP and CBD.

The objectives of The PSP Association are to:

- **P**romote research worldwide into the cause, effective treatment and eventual cure of PSP and CBD.
- **S**upport patients affected by PSP and CBD, their families and carers.
- **P**rovide information and engender awareness of PSP and CBD, amongst the medical profession and the public at large.

Research

We cooperate worldwide to act as a catalyst for research, particularly with our sister organisation CurePSP. We:

- Directly fund research into PSP.
- Sponsor and organise an International Medical Workshop every other year. The Workshop attracts researchers and neurologists from across the world with an interest in the condition.
- Founded the Sara Koe PSP Research Centre (SKRC) in January 2002 at the Institute of Neurology. The SKRC, the first such centre in the world, is the focal point for research into PSP in the UK and for coordinating this with research being carried out elsewhere in the world. The SKRC also works closely with the Queen Square Brain Bank for Neurological Disorders. Subscribers (both patients and carers) to The PSP Association are encouraged to make brain donations to the Bank.

Support to those with PSP and their Carers

The PSP Association will respond to enquiries about PSP through its website and discussion forum, through letters and by telephone with basic information on the support it can provide. There is a small subscription charge. We provide for our subscribers:

- a Telephone Link to our PSP Nurse Specialists. Our PSP Nurse Specialists are there to offer advice to subscribers, or anyone directly caring for the person with PSP that they represent.
- a comprehensive Carers' Information Pack, giving practical advice to assist with caring.
- The PSP Association Magazine, *PSP Matters*, issued 3 times a year, that summarises current research into PSP and offers articles on practical care issues.
- access to local Support Groups in more than 20 venues throughout the

UK and Ireland. Some 60 meetings are held each year and they provide an opportunity for The PSP Association to update the Group on current research and support issues, for carers and family to meet others in a similar situation, and for them to have direct discussions with the PSP Nurse Specialist.

- access to local PSP subscriber contact groups.
- an invitation to the annual PSP Symposium that provides a full day of medical briefings.

Awareness

The PSP Association has recruited a number of Development Officers (DOs) based throughout the UK, to raise awareness of PSP across the health and welfare services. The DOs organise regional PSP seminars, provide a PSP presence at other regional and national events and organise teaching sessions by one of our PSP Nurse Specialists.

Alliances and Advocacy

The PSP Association is a member of the:

- Association of Medical Research Charities (AMRC)
- Neurological Alliance (NA)
- Specialist Health Care Alliance (SHCA)
- Primary Care Neurological Society (P-CNS)
- National Council for Palliative Care (NCPC)
- Carers UK
- European Federation of Neurological Alliances (EFNA)
- European Dana Alliance for the Brain (EDAB)

Together with The PSP Association's All Party Parliamentary Group, these groups enable us to influence UK Government and European policy, in particular for increased resources to be devoted to 'fighting' neuro-degenerative diseases. Through these organisations, we made a significant input into the UK Government National Service Framework (NSF) for Long Term Medical Conditions, with a particular focus on neuro-

degenerative diseases. This initiative was announced by the Secretary of State for Health in February 2001 and the policy document was issued in March 2005¹. The NSF provides a Gold Standard for early and accurate diagnosis, and subsequent support for PSP patients and their carers. A major focus in our work is now to monitor and encourage the implementation of the NSF within the Health Service and Local Authorities.

Funding

The PSP Association is almost entirely dependent on voluntary gifts, grants and donations to carry out its work. Some of our funding comes from charitable trusts and a small amount from corporate donors. An important and growing contribution is made by our subscribers through events, committed gifts and in memoriam donations. We also take part in or organise fundraising events such as race days and sponsored sporting activities.

Questions and Answers about PSP

Why have so few people heard of PSP?

This is because it was wrongly considered for many years to be a very rare disease. It was also related inappropriately with Parkinson's Disease and neglected by governmental research bodies. Since Dudley Moore was diagnosed with PSP and research has shown it is far commoner than hitherto believed, public awareness and support for people with PSP has increased.

What causes the brain cells to degenerate in PSP?

Although the cause of nerve cell loss in PSP and CBD is still not known recent research has focused on variations of the tau gene on Chromosome 17, where genetic defects in the tau protein might cause increased susceptibility to neuro-degenerative diseases, such as PSP. One theory suggests that PSP is caused by an as yet unknown virus that causes no symptoms until after it has been slowly incubating in the brain for many years. (There are a few brain diseases proven to be caused by such 'slow

¹ DoH 265109 – available free of charge from dh@prolog.uk.com or 08700 102 870

viruses'.) Another theory is that there is an exposure to an unidentified neuro-toxin present in the environment, which slowly damages certain vulnerable areas of the brain of those genetically susceptible. Patients poisoned with manganese have some symptoms which resemble those found in PSP. On the Pacific island of Guam, there is a neurological disease that has many of the characteristics of PSP and MND. Attention has also been focussed, more recently, on another tropical Island, Guadeloupe, in the Caribbean, where a large cluster of patients, with a disease that presents symptoms remarkably similarly to PSP, have been reported.

Is PSP Inherited?

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

What are the common early symptoms of PSP?

The most common first symptom is loss of balance while walking. This may take the form of unexplained falls, usually backwards, and/or stiffness, particularly in the neck, which can be arched backwards, with associated awkwardness in walking with a rolling sailor's gait. Sometimes the falls are described by the person experiencing them as attacks of 'dizziness'. This can prompt suspicion of an inner ear problem. Other common early symptoms are difficulties with vision, including blurring and double vision and difficulty looking up or down. There also may be forgetfulness, apathy and changes in personality, with loss of interest in ordinary pleasurable activities or increased irritability and frustration, leading to a lack of co-operation. These mental changes are often misinterpreted as depression or even as dementia. Less common early symptoms are slurring of speech, cramped illegible writing and a drunken lurching gait.

What happens next?

The term 'progressive' was included in the name of the disease because, unfortunately, the early symptoms do get worse and new symptoms develop sooner or later. This is because of the progressive death of nerve cells in an area of the brain called the brainstem.

After a period of perhaps years, imbalance usually worsens to make walking very difficult or impossible. If trouble with eyesight was not present early on, it eventually develops in almost all cases and can sometimes be almost as disabling as the walking difficulty. Speech and swallowing are also likely to be affected as the disease progresses. There can be some incontinence too.

Is the visual problem the most important part of PSP?

In most cases the visual problem is at least as important as the walking difficulty, though visual difficulties do not usually appear until after the walking problem. Because the main difficulty with the eyes is in aiming them properly, reading is often difficult. The patient finds it hard automatically to move the eyes across the line and then shift down to the next line. This is very different from just needing reading glasses.

Another common visual problem is an inability to maintain eye contact during a conversation. This can give the mistaken impression that the patient is confused or uninterested. The same eye movement problem can lead to 'tunnel vision' and can interfere with driving a car. However, the most common eye movement problem in PSP is an impaired ability to move the eyes up or down (upgaze and downgaze). This can interfere with eating or with descending a flight of stairs. This problem is not usually as vexing for the patient and family as the inability to maintain eye contact or to co-ordinate eye movements while reading, but is much easier for the doctor to detect. This reduction in 'willed' vertical eye movement is usually the first clue to the doctor that the diagnosis is PSP. By contrast, the eyes will move up and down if the neck is moved forcibly up and down (doll's eye movement). Other conditions, particularly PD and normal ageing, can sometimes cause difficulty moving the eyes up. Problems in moving the eyes down are, however, very suggestive of PSP.

Yet another eye problem in PSP can be abnormal eyelid movement - either too much or too little. A few patients experience forceful involuntary closing of the eyes for a few seconds or minutes at a time, called 'blepharospasm'. Others have difficulty opening the eyes, even though the lids seem to be relaxed, and will try to use the muscles of the forehead in the effort to open the eyelids ('apraxia of lid opening').

About 20 percent of patients with PSP eventually develop one of these problems. Others, on the contrary, have trouble closing the eyes and blink very little. While about 15 to 25 blinks per minute are normal, people with PSP blink, on average, only about 3 or 4 times per minute. This can cause the eyes to become dry and sore.

What other symptoms occur?

The same general area of the brain that controls eye movement also controls movements of the mouth, tongue and throat, and these movements also weaken in PSP. Speech becomes slurred and growling in most patients after 3 or 4 years, on average, and together with the slight forgetfulness, personality change, and poor eye contact during conversation, can give the erroneous impression of dementia.

Similarly, swallowing food and even drinking can become difficult because of throat muscle weakness. This usually occurs later than the walking, visual, and speech problems, but can become very troublesome if the patient tends to choke on food, which can sometimes pose a danger for the patient of food 'going down the wrong pipe' into the breathing passages. To minimize this risk, food may need to be soft or cut into small pieces and liquids conversely 'thickened'. Inspiratory sighing and involuntary groaning may also occur.

Is gastrostomy feeding advisable for patients with severe swallowing problems?

When swallowing becomes troublesome, the amount of food and drink that can be taken often decreases; this may result in weight loss, dehydration and lack of energy. Gastrostomy, or PEG feeding, is a successful way of providing good quality nutrition via a feeding tube especially in those who have severe swallowing problems. It has been designed for people who require tube feeding over a longer period of time and provides the extra calories and fluid to help build up energy levels and strength. A PEG is a small feeding tube, which is inserted directly into the stomach and is hidden discreetly under the clothes. A speech and language therapist can discuss the suitability of having a PEG fitted with the patient and carers, as can The PSP Association Nurse Specialists.

The Percutaneous Endoscopic Gastrostomy (PEG) insertion procedure requires a minor operation and is usually carried out under local anaesthetic or a light general anaesthetic. This is done in the endoscopy suite (x-ray department) in the local hospital. The whole process takes about 30 minutes to complete and the patient will feel drowsy or be asleep whilst this is happening. As this is a minor surgical procedure, the patient may have some pain for a few days afterwards. This can be controlled with regular pain killers. Most patients will need a short stay in hospital after the procedure; this could vary from two to five days.

Following PEG insertion, many people report an improved sense of well being with increased energy levels. They also report that the good quality nutrition provided reduces fatigue and exhaustion. Difficulty with swallowing can be distressing for the patient, family and carers. The PEG can help remove the anxiety and pressure, which often accompanies meals by providing all the nutrition you need from a specially prepared liquid feed. PEG feeding can also have additional benefits of helping manage symptoms such as constipation, urine infections and low blood pressure. There are two main ways that the patient can be fed, via:

- a pump – this is the standard way of feeding in hospital and at home. The bag or bottle of liquid feed is hung on a drip stand and pumped through the PEG at a set rate.

- bolus (syringe) feeding – liquid feed is drawn up in to a 50ml syringe and then slowly syringed into the PEG tube, at various intervals during the day. However, to obtain the full amount of feed per day, this may require up to 8 bolus feeds per day. This can be time consuming and therefore not suitable for everyone.

A registered dietician will help guide the patient, family and/or carer's decision on which is most suitable for feeding the patient.

If the PEG tube falls out or is pulled out, it is strongly advised that the patient is taken to their nearest Accident & Emergency department as soon as possible to have another tube put in. If you leave it too long the incision could start to close up, making it more difficult to insert another tube. Do not try to re-insert the peg tube yourself.

How is PSP different from Parkinson's Disease?

Early on, PSP may be difficult to distinguish from PD. PSP is still sometimes referred to as an unusual form of Parkinsonism though it is recognised as clinically, biologically and pathologically different. Both PSP and PD can cause stiffness, slowness, and clumsiness. However, shaking at rest ('rest tremor'), while prominent in most people with PD, is rare in PSP. When it does occur in PSP, it is usually mild and present only when the hand is in use ('action tremor').

Patients with PSP often stand with their head arched or tilted backward and tend to fall backwards unexpectedly, while those with PD may have a stooped posture and falls can occur in any direction, later in the disease. The problems with vision, speech and swallowing are much more common and severe in PSP than in PD. Importantly, the main treatment for PD, L-dopa, may benefit only a proportion of people with PSP, and then, for a shorter time period. Pathological examination of brain tissue reveals neuro fibrillary tangles of tau in PSP and characteristic Lewy bodies in PD.

Is there a brain operation for PSP as in Parkinson's Disease?

A brain 'transplant' operation for PD (actually an implantation into the brain of a piece of the patient's own adrenal gland) was tried, in a few patients ten years ago, without much long-term success. This procedure has now been abandoned. Although the newer foetal brain cell implantation has shown some promise in dopamine replacement for PD, it has not been considered as likely to help in PSP. Thalamotomy and Pallidotomy, two operations that freeze small parts of the brain that are overactive in PD, have been tried in PSP without success and deep cerebral stimulation with pacemakers (DBS), although of benefit in severe cases of PD, has also been found, to date, to be ineffective in PSP.

Why is Palliative Care important?

Community nurses have a very important role to play in supporting the patient living at home and they can often signpost to other professionals for review or advice when symptoms change.

It is never too soon to involve palliative care, as people living with PSP will deteriorate at different rates. Sometimes this can occur fairly rapidly, over a few months.

Caring for someone with PSP does become a 24 hour job and carers can easily become exhausted. Regular periods of respite are therefore important to enable them to continue in their role. This respite can be offered within a nursing home environment or a hospice setting. The PSP Nurse Specialists are available to give teaching sessions to local nursing homes or hospices if required.

Ideally a referral to the palliative care team will allow access to the local hospice for respite periods, day care where appropriate, outreach services and support for the family or carers. It will also mean that when terminally ill, the patient can decide where to receive care to ensure a peaceful and comfortable death.

Notes

Notes

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