

# PSP - What is it?



- Comparatively rare neuro-degenerative brain disease
- Cause unknown
- No effective treatment and no cure
- Life expectancy 5-7 Years
- Progressive symptoms of rigidity, bradykinesia, loss of balance with early falls, increasing problem of speech and swallowing, and slowness of eye movement.
- Possible personality change and behavioural problems

# CBD - What is it?



- Cortico Basal Degeneration – 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 frontal lobe deficit

# PSP - Differential diagnosis



- Non levodopa responsive
- No presenting tremor
- Rarely affects people under 40
- Falls are often backwards
- Steady deterioration
- Restricted eye movement
- Neurofibrillary tangles not Lewy Bodies

# PSP – Postural Instability



- Falls often backwards without warning
- Difficulty with down gaze, shuffling, broad based gait
- Stiff arched neck, axial rigidity
- Tiredness, light-headedness
- Sudden change of direction, loss of balance
- O.T. – Grab rails, ramps, stair lift, through floor lift, adaptations to home, equipment
- Physio – Advice, exercise, built up shoes, walking frames, wheelchairs, hoists
- Sitting “en bloc” “rocket sign”
- Limited benefit from PD drugs

# Eye Problems



- Starts with slow saccades
- Later limited voluntary vertical saccades
- Especially down gaze
- Reflex movements remain normal (Doll's eye manoeuvre)
- Indicating supranuclear problem
- Limited up gaze can be normal in elderly but never accompanied by slow saccades

# Sight Difficulties



- Spatial – clipping kerb, door frame
- Non specific – vision blurred
- Double vision
- Vision ‘not right’
- Leading to buying expensive new glasses that do not help

# Downgaze difficulties



- Moving between lines of text
- Using utensils, Ignore food on near half of plate
- Watching TV in bed
- Going downstairs
- Tying shoelaces
- Adjusting to bifocals
- Seeing moving objects

# Management



- Prism glasses
- Ptosis props or tape
- Eye drops
- Artificial tears (clarymist spray)
- Tinted wrap around glasses
- Botox injections
- Educate/Awareness family

# PSP – Dysphagia



- Early involvement of Speech & Language Therapist
- Techniques for safe swallow
- Over filling the mouth, “Dirty Tie Syndrome”
- Weak cough reflex, choking
- Excessive salivation
- Softened diet, thickened liquids
- Dietician, weight loss, supplement drink
- Video-fluoroscopy
- PEG feeding
- Aspiration pneumonia

# PSP – Speech problems



- Early involvement of Speech and Language Therapist
- Exercises for speech
- Delay in response, word search
- Reduced facial expression
- Palalalia, stuttering
- Three types of speech pattern
  - 1) Hypokinetic Dysarthria – like PD, quiet, repetition of sounds
  - 2) Spastic Dysarthria – strained, slow and slurred
  - 3) Ataxic Dysarthria – slurred and imprecise, sounding drunk
- Communication aids: Picture, alphabet boards, amplifiers, electronic aids

# PSP – Cognitive change



- Need for families to understand
- Vague changes in personality
- Intelligence largely intact
- Loss of higher executive function
- Signe d'aplause
- Short term memory problems
- Impaired judgement or reasoning
- Irritability, aggression, apathy
- Emotional lability
- Depression, general lack of interest
- Support for families, recognising emotions
- Antidepressant drugs

# PSP - Pain



- Symptomatic
- Neck (and referred)
- Warmth, Analgesia
- Muscle relaxants
- Massage
- Cramp as stiffness increases
- Possible contractures
- Check with GP for any underlying cause
- Alternative therapies, acupuncture, reflexology, aromatherapy

# PSP – Bladder & bowel problems



- Incontinence
- Continence advice nurse
- Bottle, Commode
- Pads, sheath
- Catheter – either intermittent or permanent
- Nocturnal enuresis
- Difficulty with initiating flow
- U.T.I
- Constipation - diet, roughage, laxatives
- Suppositories and enemas

# PSP – Palliative care



- Improving with awareness of Neurological illness
- Referral to specialist in Palliative care
- Quality of life, patient choice, preferred place of care
- Continuing Care Assessment
- Hospice respite care, outreach team
- Increasing immobility, stiffness, contractures
- Pain control, antibiotics
- U.T.I.
- Respiratory infections – Aspiration Pneumonia
- Carer Support

# PSP - Prevalence



- At least 6 per 100,000 in the UK, as common as Motor Neurone Disease
- Actual figure believed higher:-
  - up to 10,000 living in the UK;
  - up to 15 people per 100,000Affects people of all races  
men and women in roughly equal proportion.
- Evidence PSP existed before 1900 AD

# PSP - Management



- Support from the moment of diagnosis
- Referral to The PSP Association
- MDT approach
- Social Service referral
- Care plan
- Respite
- Palliative care

# PSP - Summary



- Completely immobile
- Restricted vision
- Unable to communicate
- Often fed via PEG
- Incontinent
- Intelligence largely intact

# PSP



**“I have 2 strong legs that I can move  
but when I walk I fall;  
I have a strong voice  
but when I talk no-one understands;  
I have good vision  
but I cannot see – I cannot read;  
My throat is healthy  
but I cannot eat without choking.”**