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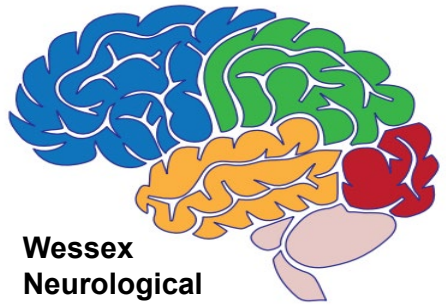

Diagnosing PSP & CBS

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PSPA 20th July 2026

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PSPA

Disclosures

- Lectured for UCB and GSK on atypical Parkinsonism
- Advisory board for NICE, UCB, Ferrer, Immunobrain and Biogen
- Drug trials for Biogen, Ferrer, UCB, Pfizer, TauRx, Novartis, Takeda and Laboratoire Français de Fractionnement et de Biotechnologies
- Trustee and member of research committee for the PSP Association

Aims

- At the end of this session participants will:

1. Know when to consider a diagnosis of PSP or CBS
2. Have an approach to aid diagnosis of PSP and CBS
3. Have some knowledge of management pathways for patients with PSP and CBS (some self study)

Joyce

So how can you diagnose PSP or CBS?

- Rare
- A presentation is not what you would expect
- Look for signs that point you towards PSP or CBS

- Prioritise videos
- Some resources for self study *

Progressive supranuclear palsy

- Tau based disease (cf PD)
- Symmetrical akinetic rigid disease
- Vertical supranuclear gaze palsy
- Bradyphrenia and executive changes
- Bulbar dysfunction

Which saccades are abnormal?

Which saccades were abnormal?

Diagnostic criteria for PSP

- Mandatory inclusion:
  - Age > 40 at first symptom
  - Gradually progressive
  - Sporadic
- Mandatory Exclusion:
  - Signs suggestive of another disease
    - Eg. MND, MSA, AD, DLB, Prion, Encephalitis
  - Imaging suggestive of another disease
    - Eg. NPH
  - Lab findings suggestive of another disease
    - Eg. Neurosyphilis, Wilsons, Niemann Pick

1. Höglinger GU, Respondek G, Stamelou M, et al. Clinical diagnosis of progressive supranuclear palsy: The movement disorder society criteria. *Mov Disord*. 2017;32(6):853-864. doi:10.1002/mds.26987

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	PSP – RS	PSP – PGF	PSP – P	Others
Definite	Defined by pathology			
Probable	Vertical gaze palsy or vertical saccade slowing			
	Unprovoked falls <3 yrs or fall on pull test < 3yrs	Progressive gait freezing in 3 yrs not responsive to dopa	Akinesia and rigidity either axial or limb dopa responsive or not	PSP – F: with frontal cognition (3 of: reduced fluency, apathy, bradyphrenia, dysexecutive, impulsivity)
Possible	Slow vertical saccades			PSP – CBS: Vertical gaze palsy or slow saccades
	> 2 steps back on pull test < 3 yrs	Progressive gait freezing < 3 yrs not dopa responsive		1 Cortical sign (sens loss, apraxia, alien limb) AND 1 MD sign (rigid, akinesia, myoclonus)

1.Höglinger, G. U. *et al.* Clinical diagnosis of progressive supranuclear palsy: The movement disorder society criteria. *Mov. Disord.* **32,** 853–864 (2017).

*

	PSP – RS	PSP – PGF	PSP – P	Others
Suggestive	Macro SWJ or eyelid opening apraxia		Bradykinesia either axial or limb, dopa responsive or not	PSP – PI: Unprovoked falls <3 yrs or fall on pull test < 3yrs
	Fall on pull test or >2 steps back on pull test		SWJ OR EOA OR falls <3 yrs OR fall on pull < 3yrs OR Speech disorder OR frontal cognition OR Dopa resistance OR Hypokinetic dysarthria OR dysphagia OR photophobia	PSP – CBS: 1 Cortical sign (sens loss, apraxia, alien limb) and 1 MD sign (rigid, akinesia, myoclonus)

1.Höglinger, G. U. *et al.* Clinical diagnosis of progressive supranuclear palsy: The movement disorder society criteria. *Mov. Disord.* **32,** 853–864 (2017).

However

- These are research or specialist criteria
- There are also PSP Max criteria which determine what the current diagnosis is
- Not essential for most clinicians……

Multiple types of PSP

	RS	PSP - Park	PAGF	PSP - CBS	PSP -PNFA
Clinical					
Disease duration †	6 years	9 years	-	-	-
Rigidity	Axial more than limb	Axial same or less than limb	Axial	Present	Sometimes
Bradykinesia	Mild	Moderate	Moderate	Present	Mild
Tremor	No	yes/no (rest or jerky postural)	No	No	No
Early falls	Yes	No	No	Sometimes	Sometimes
Early postural instability	Yes	No	Yes	-	-
Early cognitive decline	Often	No	No	No	Yes
Early eye movement abnormalities	Yes	No	No	No	Sometimes
L/dopa response	No	Often	No	No	No
Pathology					
4R:3R ratio*	Higher	Lower	-	-	-
PSP Tau load*	High	Low	Low	-	-

1. Williams, D. R. & Lees, A. J. Progressive supranuclear palsy: clinicopathological concepts and diagnostic challenges. *Lancet Neurology* **8**, 270–279 (2009).
2. Williams, D. R. *et al.* Pathological tau burden and distribution distinguishes progressive supranuclear palsy-parkinsonism from Richardson's syndrome. *Brain* **130**, 1566–76 (2007).
3. Williams, D. R. *et al.* Characteristics of two distinct clinical phenotypes in pathologically proven progressive supranuclear palsy: Richardson's syndrome and PSP-parkinsonism. *Brain* **128**, 1247–58 (2005).

Types of PD

	Young disease onset	Tremor dominant	Non-tremor dominant	Rapid motor progression
Proportion	25%	31%	36%	8%
Age at onset	<55	>55	>55	any
Disease duration	23 yrs	14 yrs	13 yrs	<10 yrs
Tremor	+	+++	+/-	+
Bradykinesia	+	+	+++	+
Falling	14 yrs	9 yrs	8 yrs	6 yrs
Dementia	2%	24%	74%	-
Dementia	16 yrs	10 yrs	9 yrs	-

Selikhova M, Williams DR, Kempster PA, Holton JL, Revesz T, Lees AJ. A clinico-pathological study of subtypes in Parkinson's disease. Brain. 2009 Nov;132(Pt 11):2947–57.

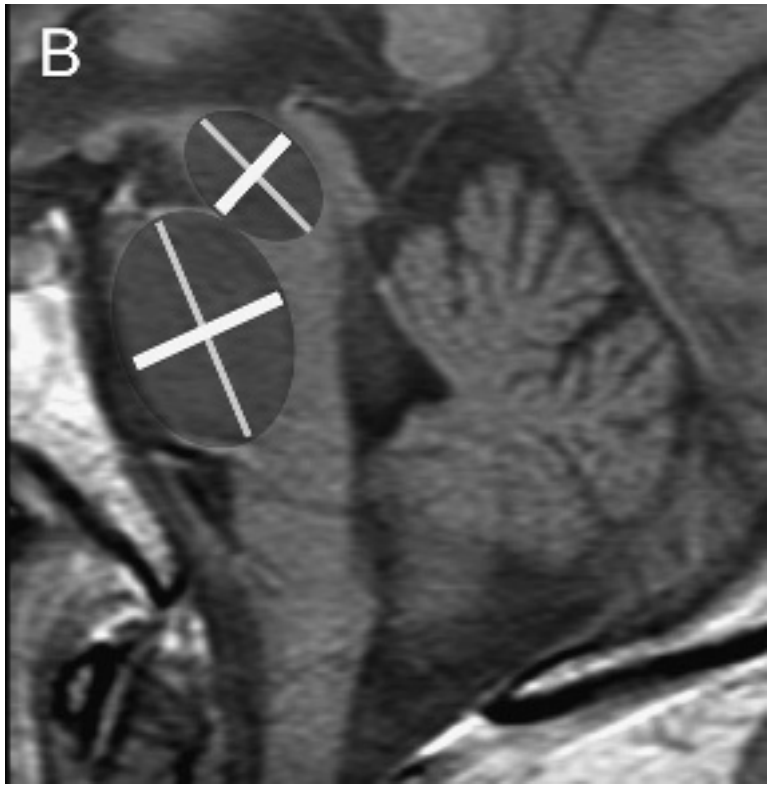
Key features of PSP (RS)

- Early instability
- Eye movement disorder
- Frontal cognitive phenotype
- Altered bulbar function

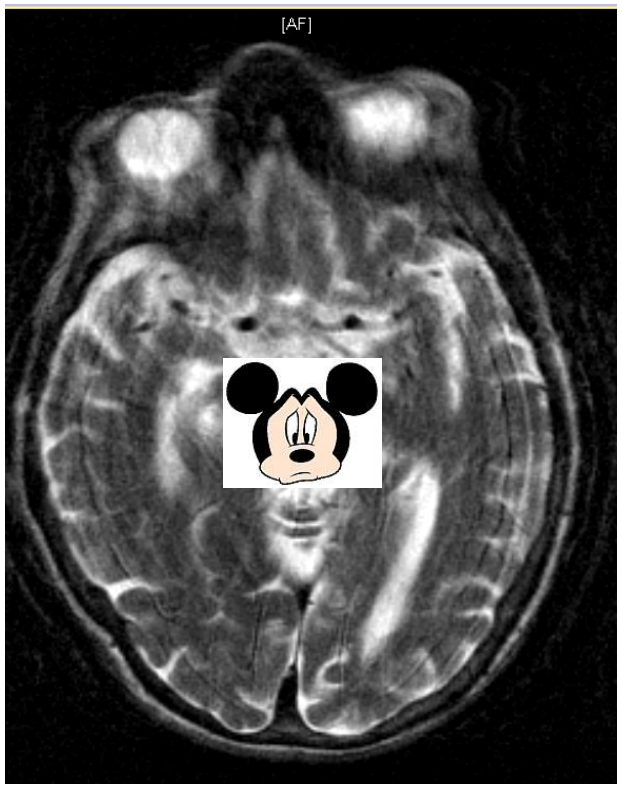
- All types of PSP will tend to converge on one phenotype at the end

Imaging PSP

Humming bird sign



Mickey mouse sign



Diameter of midbrain/pons <0.52 has specificity of 100% and sensitivity of 85.7%

Massey LA, The midbrain to pons ratio: a simple and specific MRI sign of progressive supranuclear palsy. Neurology 2013; 80: 1856–1861.

PSP Problems early on

- Falls
- Gaze problems
- Start of swallowing problems
- Speech
- Impulsivity
- “Bloody mindedness” – executive function
- Social cognition deficits
- Depression and apathy

Falls and mobility

- Get balance on standing and turning
- Levodopa can provide some benefit about 30%
- Amantadine can help – particularly as stimulation
- Watch for hallucinations with amantadine

Gaze problems

- Seen already
- Can’t look down
    - Trips over particularly on stairs - impulsivity
    - Can’t see food on plate – periscope glasses
    - Difficult to read – RNIB aids
- Can’t look up
    - More difficult to participate socially when sitting
- Can’t look sideways
    - Difficult to read
- May contribute to retrocollis

Swallow and speech

- May start “early”
- Meals > 30 mins – cold food
- Coughing and choking – respiratory arrest
- Aspiration
- Think of gastrostomy early
    - Before weight loss
    - Able to open mouth?

- Indistinct speech
- Greater social isolation
- Greater pressure on carer

Impulsivity

- Go up stairs suddenly
- Get up to answer door…..and fall down
- Bend over to pick up fluff ….while being helped to walk
- PD meds exacerbate?

Cognitive function

- Often overlooked
- Can differentiate PSP from other diseases

- Retrieval is a problem – recognition is often better
- Executive dysfunction can frustrate carers – seem bloody minded

Social cognition

- The ability to interpret what others think or feel
- Includes basic emotion recognition eg. sad, angry
- Extends to more complex interpretations eg. bored, disaffected
- Patients with PSP have difficulties….

Depression

- Patients have apathy
  - Found in 82% [1]
  - Increases with disease progression[2]
- Patients have depression
  - 10-20 % exceeded HADS thresholds for moderate to severe Depression and Anxiety[3]
  - Hamilton Depression rating scale: 100% exceeded threshold 17/54 for moderate depression[4]
  - Our experience – more depressed than previously thought
  - Consider antidepressants

1. Litvan et al. JNNP 1998
2. Bak et al. JNNP 2010
3. Millar et al Movement Disorders 2006
4. Herting et al Movement Disorders 2007

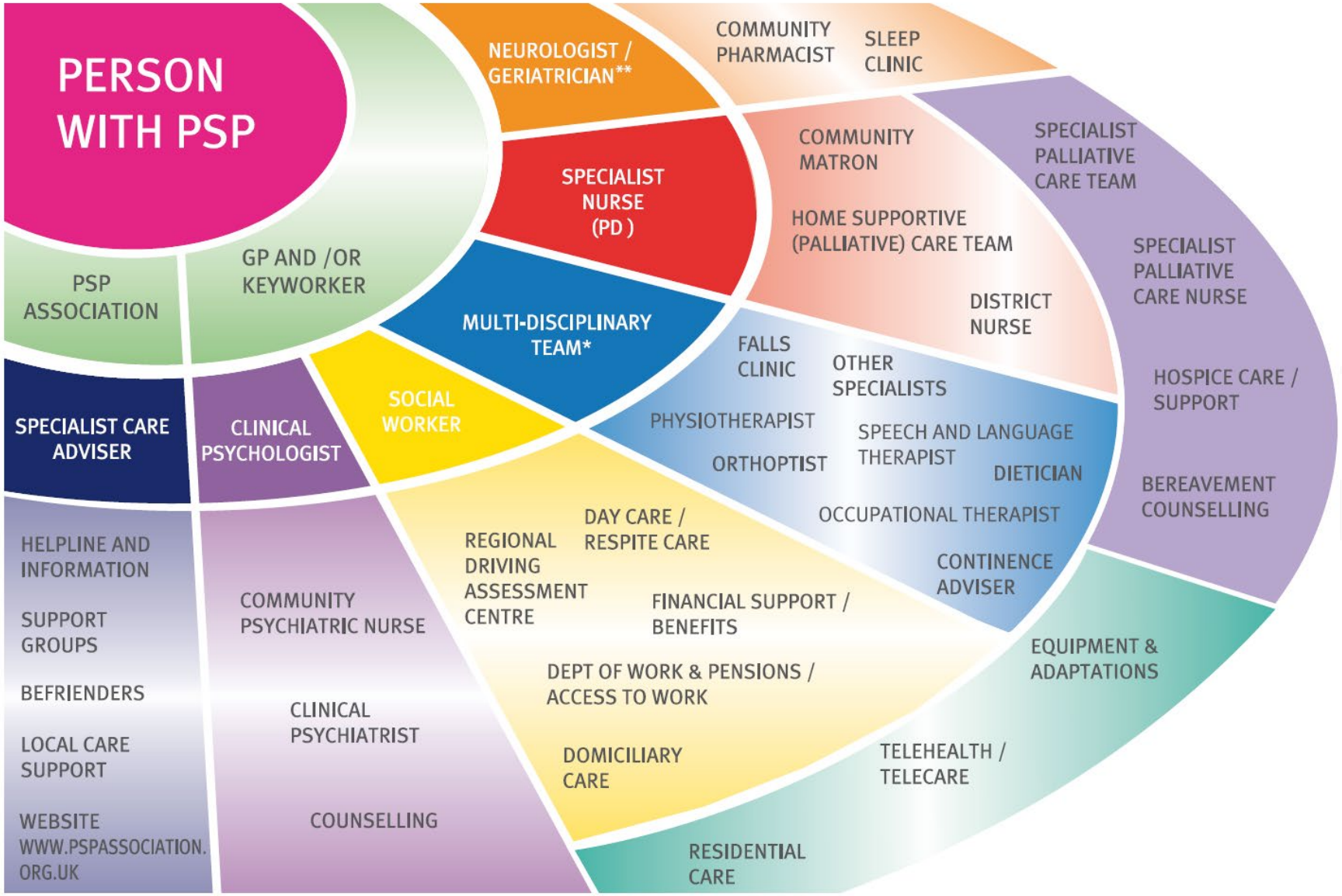
Management of PSP

- Currently no disease modifying treatment
  - Anti Tau treatments in trial

- DIAGNOSIS and Information
- Holistic symptomatic treatment
- Support for patient
- Support for carer

Bluett B, Pantelyat AY, Litvan I, et al. Best Practices in the Clinical Management of Progressive Supranuclear Palsy and Corticobasal Syndrome: A Consensus Statement of the CurePSP Centers of Care. *Front Neurol*. 2021;12:694872. doi:10.3389/fneur.2021.694872

Management of PSP



http://www.ncssupport.org.uk/ncs-showcase/psp-care-pathway/

Even in early PSP consider:

- Palliative care register
- Advance directives
- Advanced care planning
- Gastrostomy
- Liaison with Palliative care teams

Cortico-basal Syndrome

- Pathological and clinical diagnosis may be different
- CBD is tau based disease
- CBS may be Alzheimer’s pathology or other
- Predominantly asymmetric

Subtypes of CBD

*

Armstrong MJ, Litvan I, Lang AE, et al. Criteria for the diagnosis of corticobasal degeneration. *Neurology*. 2013;80(5):496-503. doi:10.1212/WNL.0b013e31827f0fd1

Probable CBD	Possible CBD	FBSS	NAV of PPA	PSPS
Asymmetric presentation of 2 of:	Symmetric or asymmetric presentation of 1 of:	2 of:	Effortful agrammatic speech with 1 of:	3 of:
a) Limb rigidity or akinesia		Executive dysfunction	Impaired sentence but preserved word comprehension	Axial or symmetric limb rigidity or akinesia
b) Limb dystonia		Behavioural or personality change	Groping distorted speech production (apraxia of speech)	Postural instability or falls
c) Limb myoclonus		Visuospatial deficits		Urinary incontinence
2 of:	1 of:			Behavioural changes
d) Orobuccal or limb apraxia				Slow or palsy of vertical saccades
e) Cortical sensory deficit				
f) Alien limb phenomena				

Armstrong MJ, Litvan I, Lang AE, Bak TH, Bhatia KP, Borroni B, et al. Criteria for the diagnosis of corticobasal degeneration. Neurology 2013; 80: 496–503.

*

Diagnostic criteria for CBD

	Probable	Possible
Presentation	Insidious	Insidious
Minimum duration (yr)	1	1
Age at onset (yr)	≥ 50	No minimum
Family history (≥2 relatives)	Exclusion	Permitted
Permitted phenotypes	1) Probable CBS 2) FBSS or NAV PLUS one CBS feature	1) Possible CBS 2) FBSS or NAV 3) PSPS PLUS one CBS feature (b-f)
Genetic Tau mutation	Exclusion	Permitted

*

Exclusion criteria

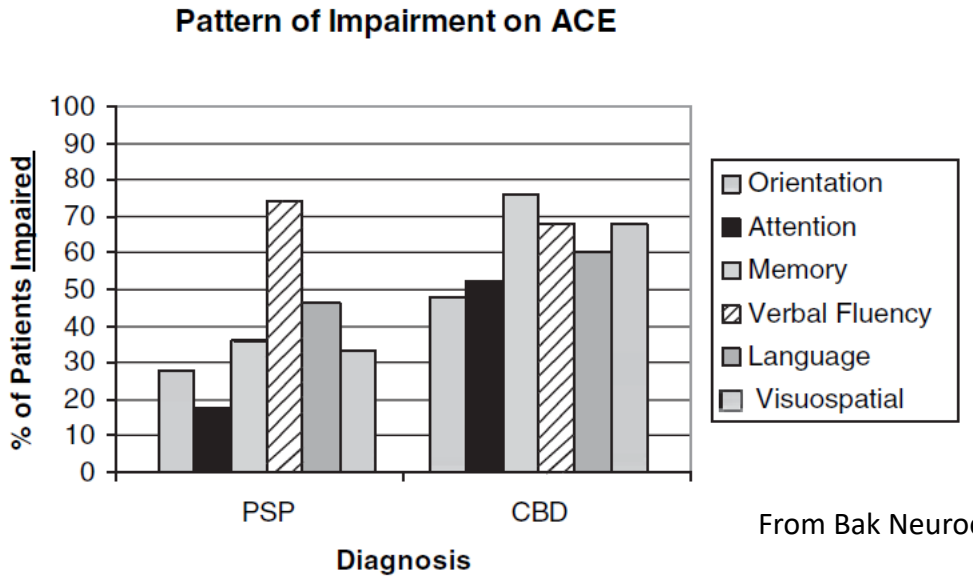
Evidence of Lewy Body disease	Classic PD rest tremor, excellent response to l-Dopa, hallucinations
MSA	Dysautonomia or prominent cerebellar signs
ALS	Both upper and lower motor neuron signs
Alternative PPA	Semantic or logopenic variant PPA
Alternative pathology	Structural lesion suggesting focal cause
Other genetic cause	Granulin, TDP43 or FUS mutations
AD	Low CSF $A^{\beta}42$ to Tau ratio, positive PIB scan, AD genetic mutation

Differentiating features

- Alien limb syndrome
- Cortical sensory loss
- Apraxia
- Cognition

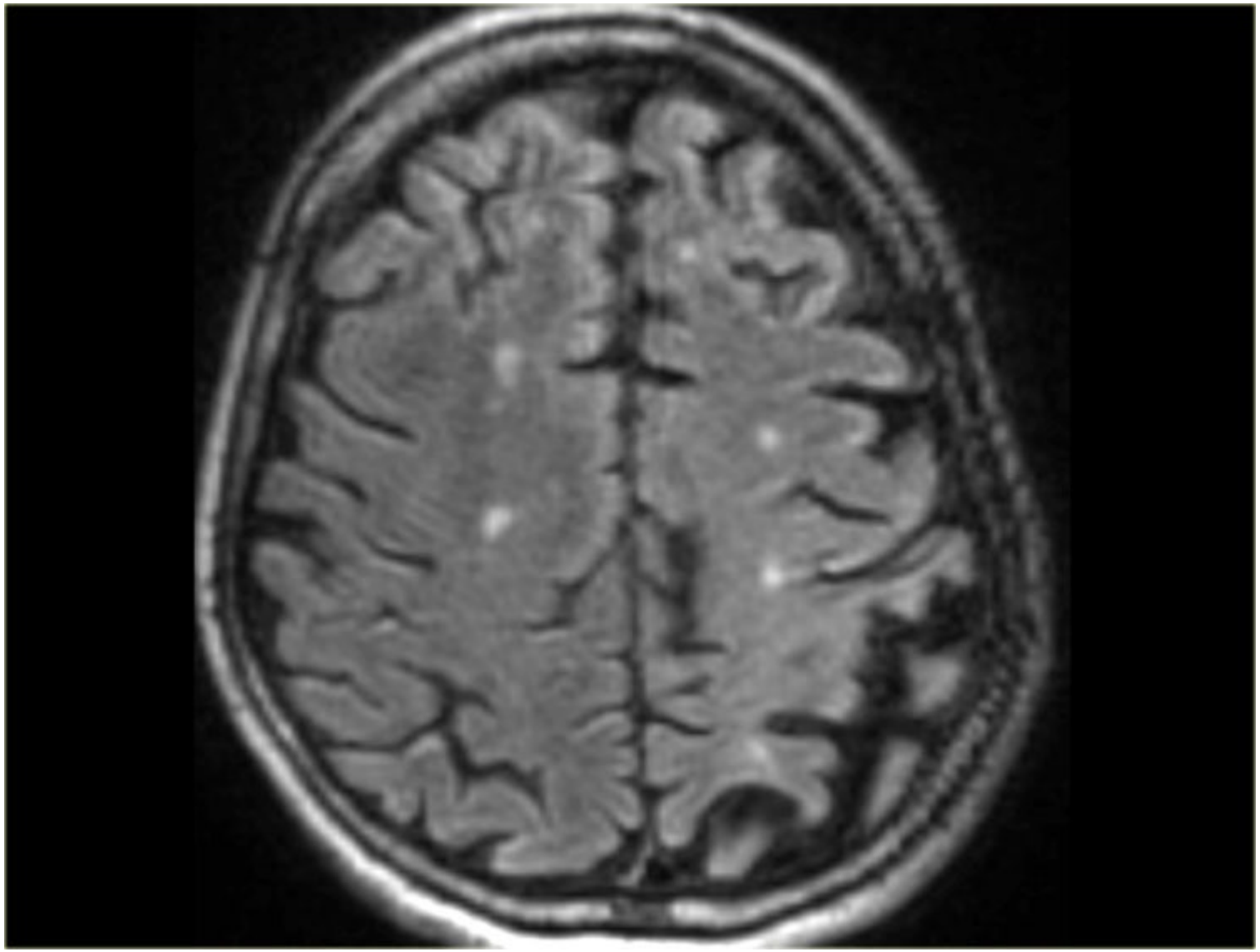
CBD v PSP on the ACE-R

Diagnosis	CBD	PSP	Control
MMSE	23/30	25/30	>28/30
ACE-R total	62/100	71/100	>88/100
Memory	10/26	18/26	26/26
Attention/orientation	15/18	15/18	18/18
P-words	12	2	>17
Animals	9	6	>21



From Bak Neurocase 2005

Imaging



1. Whitwell, J. L. *et al.* Imaging correlates of pathology in corticobasal syndrome. *Neurology* **75,** 1879–1887 (2010).

Management

- As for PSP
- Botox for dystonia
- Cholinesterase inhibitors for cognition?

Improve APS

Be Part of Our Research!

University of Hertfordshire UH

NHS University Hospital Southampton NHS Foundation Trust

Recruiting Health and Social Care Professionals now!

Share your view!

Reimbursing time with shopping voucher.

Aims

- What matters to people affected by APS?
- How people experience diagnosis and ongoing care?
- How services could work better for patients and carers?

Phase 1

- Review of published and lay evidence of how services work
- 24 Interviews with people affected by CBD, MSA, PSP
- 3 focus groups with Health and Social care Professionals

We want to hear from you about your experience of health and social care services!

Phase 2

1st Workshops

- Create a map of networks and services
- Find together possible solutions, based on the best examples

Phase 3

2nd Workshops

- Create a questionnaire to ask people about their preference among the potential solutions
- Planning the next phase of putting recommendations in practice

Contacts

IF YOU ARE A PATIENT, A CARER, A PROFESSIONAL, PLEASE GET IN TOUCH

- ImproveAPS@herts.ac.uk
- Call - 0794 6077 082

APS: Atypycal Parkinsonian Syndromes

Community of Practice: A forum to share research findings to improve care for people living with Atypical Parkinsonian Syndromes

Why are we creating this forum?

- Part of the Improve APS study knowledge mobilisation plan.
- Knowledge mobilisation: how to get research evidence used in the real world!
- Support professionals to use research findings
- Foster collaborations for future projects

Funded by the University of Hertfordshire Impact Acceleration Account awarded by The Economic and Social Research Council.

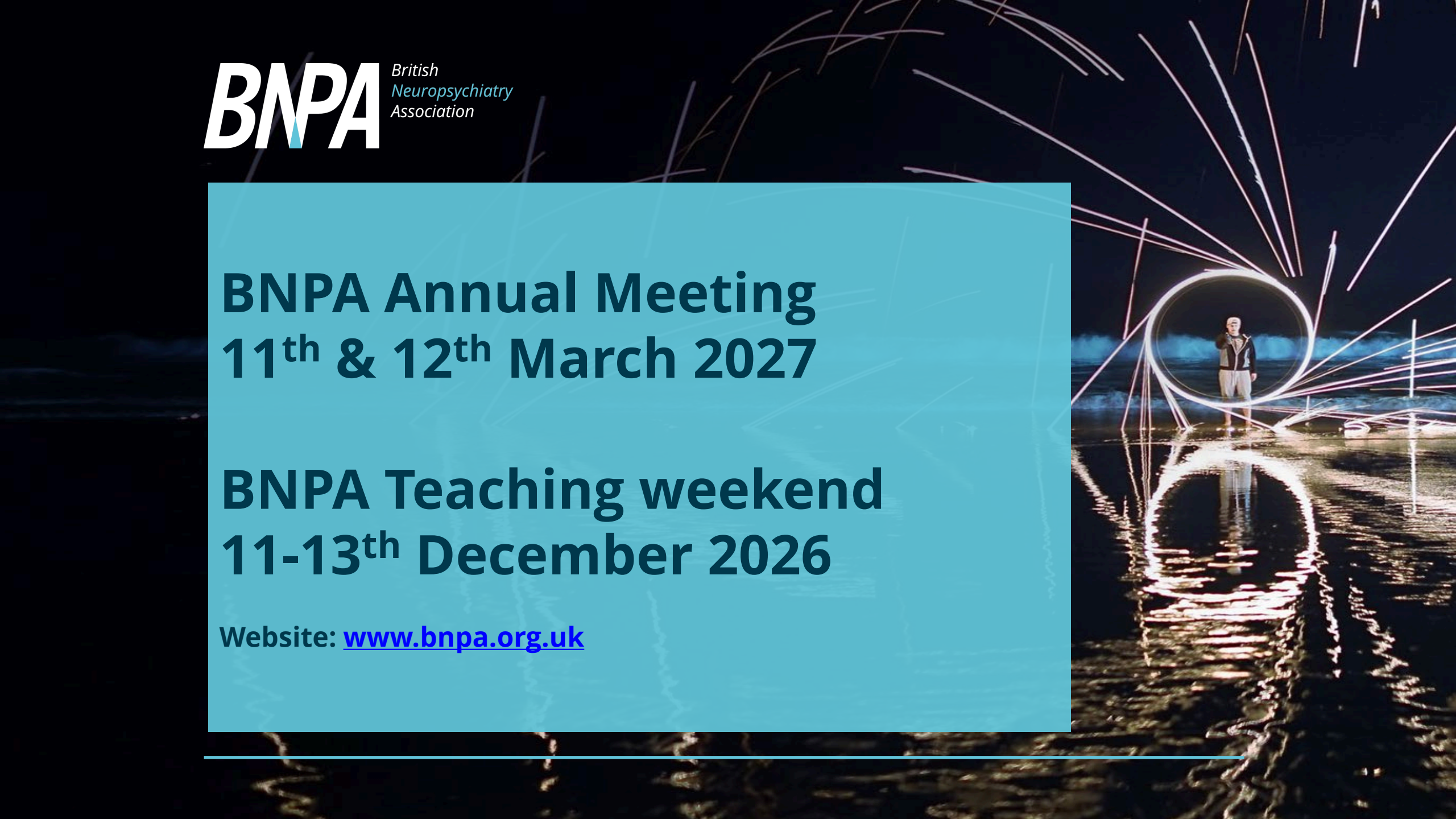
Join us if you can on the Mon 27th of July at 12pm
for details email APSresearchCoP@herts.ac.uk

BNPA British Neuropsychiatry Association

BNPA Annual Meeting
11th & 12th March 2027

BNPA Teaching weekend
11-13th December 2026

Website: www.bnpa.org.uk



* Resources to look at

- Höglinger GU, Respondek G, Stamelou M, et al. Clinical diagnosis of progressive supranuclear palsy: The movement disorder society criteria. *Mov Disord*. 2017;32(6):853-864. doi:10.1002/mds.26987
- Bluett B, Pantelyat AY, Litvan I, et al. Best Practices in the Clinical Management of Progressive Supranuclear Palsy and Corticobasal Syndrome: A Consensus Statement of the CurePSP Centers of Care. *Front Neurol*. 2021;12:694872. doi:10.3389/fneur.2021.694872
- Armstrong MJ, Litvan I, Lang AE, et al. Criteria for the diagnosis of corticobasal degeneration. *Neurology*. 2013;80(5):496-503. doi:10.1212/WNL.0b013e31827f0fd1
- Wenning GK, Stankovic I, Vignatelli L, et al. The Movement Disorder Society Criteria for the Diagnosis of Multiple System Atrophy. *Mov Disord*. 2022;37(6):1131-1148. doi:10.1002/mds.29005
- Goh YY, Saunders E, Pavey S, et al. Multiple system atrophy. *Practical Neurology*. 2023;23(3):208-221. doi:10.1136/pn-2020-002797

Any questions?

- Thank you for your attention

Aims

- At the end of this session participants will:

1. Know when to consider a diagnosis of PSP or CBS
2. Have an approach to aid diagnosis of PSP and CBS
3. Have some knowledge of management pathways for patients with PSP and CBS (some self study)