

Idiopathic Normal Pressure Hydrocephalus (iNPH)

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Pre-survey

- ▶ <https://addenbrookes.onlinesurveys.ac.uk/idiopathic-normal-pressure-hydrocephalus-inph-ahp-survey>



Aims


- ▶ Overview of iNPH covering:
 - Definition
 - Epidemiology
 - Pathophysiology
 - Symptoms
 - Differential diagnosis
 - Treatment
- ▶ The role, challenges, opportunities and reflections



What is iNPH?

- ▶ Adult onset syndrome of uncertain origin that involves the non-obstructive enlargement of the cerebral ventricles, despite cerebrospinal fluid (CSF) pressure being within the normal range
- ▶ As iNPH is potentially reversible by shunt surgery, early and accurate diagnosis is important to optimise treatment outcomes (Gallia et al., 2006)
- ▶ Comprehensive MDT assessment can improve diagnostic accuracy and lead to earlier diagnosis, which is related to better treatment outcomes (Kajimoto et al., 2022). Specialist MDT assessments are particularly important for differential diagnosis, diagnosis of comorbidities and complex presentations



Normal Pressure Hydrocephalus  knowmedge

Mnemonic: "Wacky Wobbly Wet"

NPH Clinical Findings	Wacky (Weird)	Dementia
	Wobbly (Walking)	Wide Gait
	Wet (Water)	Urinary Incontinence

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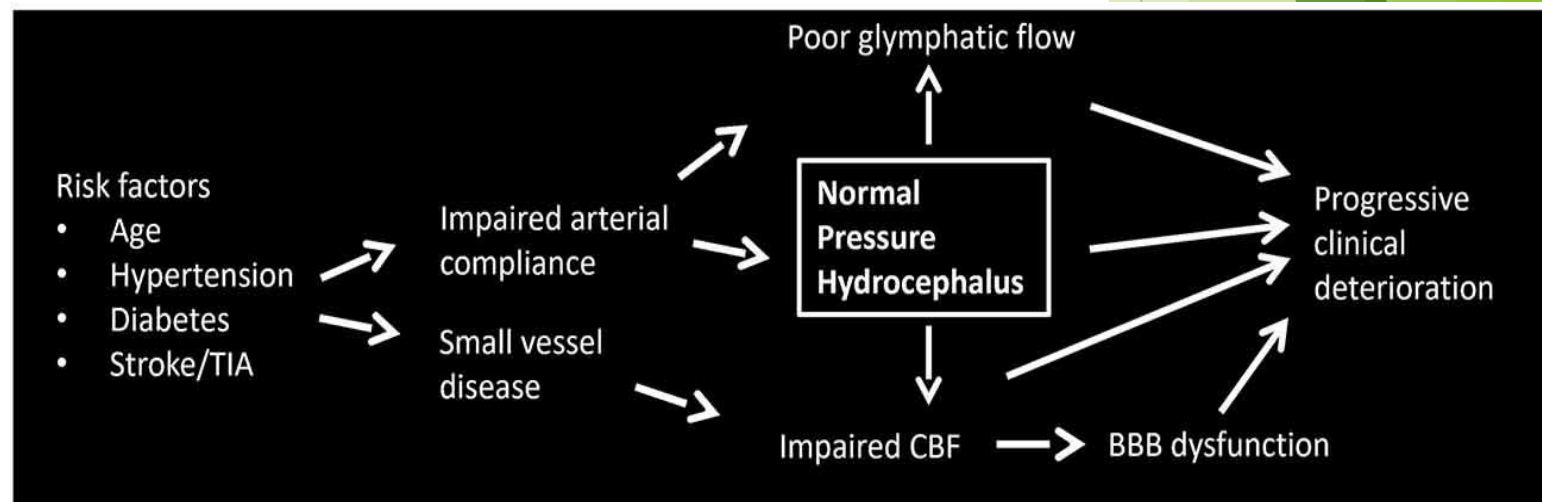
Epidemiology

- ▶ iNPH is estimated to affect up to **3.7% of adults over 65 years** (Anderson et al 2019). The prevalence has been suggested to be as much as 4 times higher in patients 80 years old and over as compared to 65-79. Jaraj et al (2014) suggested prevalence up to 5.9% in people aged 80 years or older.
- ▶ Within the East of England, this would include up to 44,000 people who would potentially benefit from this service. Considering the ageing population, this is likely to increase over time
- ▶ NPH is the most common form of hydrocephalus in adults and a significant cause of reversible dementia amongst older adults (Williams and Malm 2016)
- ▶ Now recognized as the second most common form of dementia and estimated that only 20% of cases are currently diagnosed and treated (Sundstrom et al. 2022)



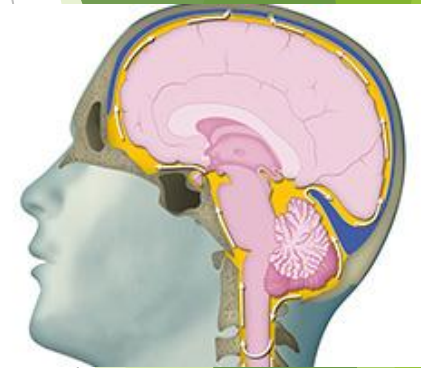
Pathophysiology

- No current firm consensus on the exact undermining pathophysiology causing the symptoms
- Related to an imbalance between CSF formation and removal and thought to predominantly relate to a problem with **CSF return to circulation (reabsorption)**
- Referred to as '**idiopathic**' but the underlying process is thought to relate to cerebrovasculature with impaired compliance triggering a cascade of symptoms that cause deterioration (Bonney et al 2022). It is thought development and further progression of symptoms relate to:
 - ▶ Deficient Cerebral Blood Flow (CBF)
 - ▶ Impaired glymphatic circulation
 - ▶ Loss of blood brain barrier integrity



Neuroanatomic Features

- ▶ 'Possible' role of stretching of corticospinal tracts and the corpus collusum but features of iNPH don't necessarily fit with similar disorders and can be disproportionate to degree of CSF retention (Baker 2018)
- ▶ Dysfunction of cortico-basal ganglia-thalamo-cortical (CBGTC) loop but does not account for pattern or cognitive and urinary deficits (Lenfeldt et al 2008)
- ▶ White matter changes in both motor and sensory pathways at a microvascular level (Tang et al 2021)
- ▶ Poor perfusion of periventricular white matter and prefrontal regions and frontal lobe disturbance
- ▶ Frontoparietal-subcortical-cerebellar circuits may be a vulnerable area for the pathophysiological mechanisms of iNPH (Griffa et al 2020)
- ▶ Thinning of midbrain structures which can respond to shunting (Mocco et al 2006)
- ▶ Stretching of the periventricular sacral fibers of the corticospinal tract causing a loss of voluntary control of bladder contractions (Gleason 1993)
- ▶ Bladder issues though to be the result of a hyperactive detrusor muscle due to reduced or absent central inhibition (Rendtorff et al 2012)
- ▶ **No formal consensus but more likely a combination of the above**

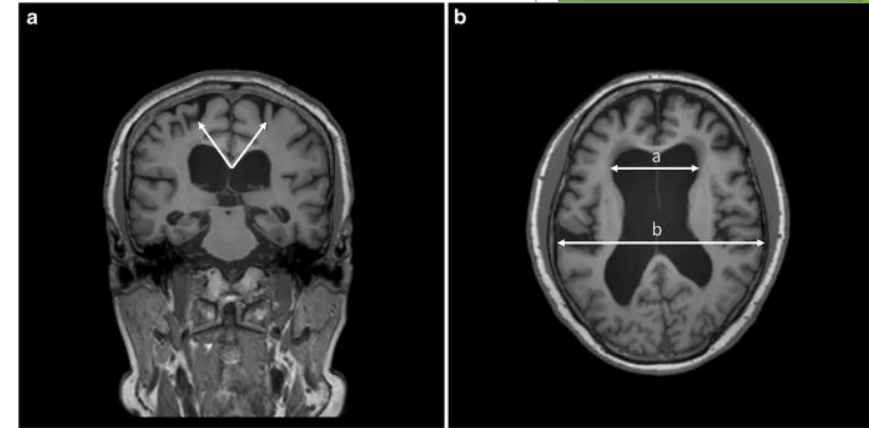


Neuroimaging signs

Callosal angle

Coronal plane. Normal between 100-120 degrees, 50-80 degrees suggestive of NPH.

Appears clinically important cut-off with callosal angles below 63 degrees for shunt response (Virhammar et al 2014).



Evans index

Transverse plane. Normal value between 0.2 and 0.25. Ratio over 0.3 suggestive of ventricular enlargement.

(Damasceno et al 2015)

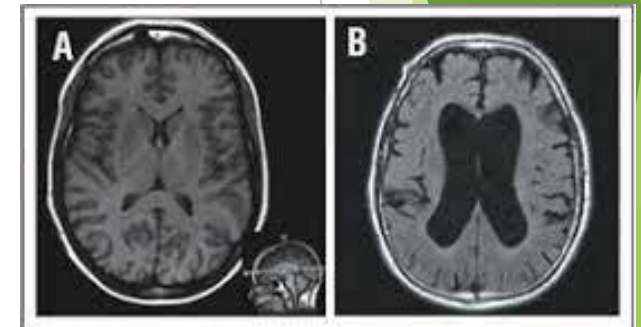
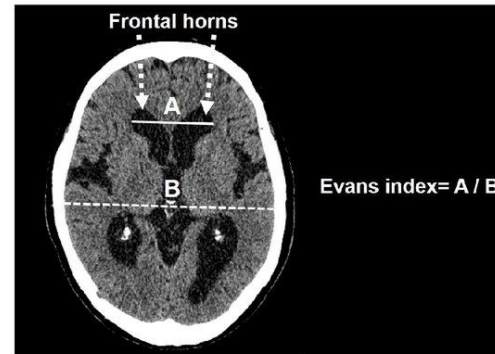
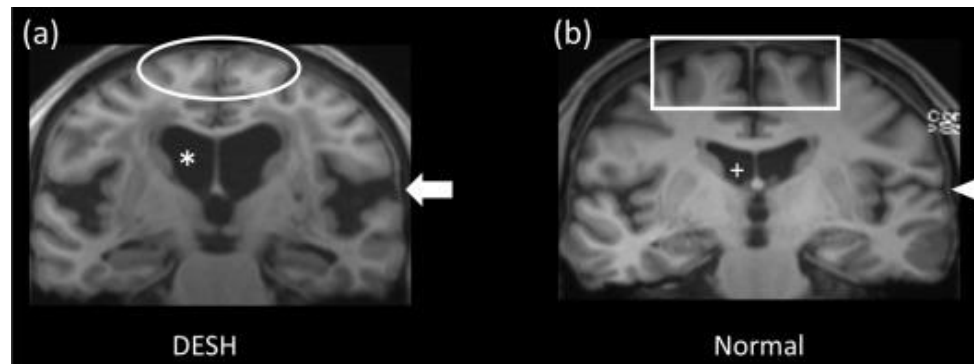


Figure 1. Images of a normal brain (A) and a brain with ventriculomegaly (B).

DESH

Disproportionately enlarged subarachnoid space.

Crowding of the sulci superiorly near the vertex accompanied by enlargement of CSF spaces more inferiorly, particularly in the Sylvian fissures.



Diagnostic tests

Lumbar infusion study

Lumbar puncture with and infusion to assess dynamics. Usually followed with CSF tap.

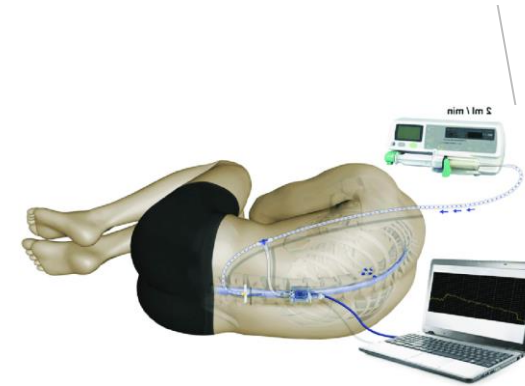
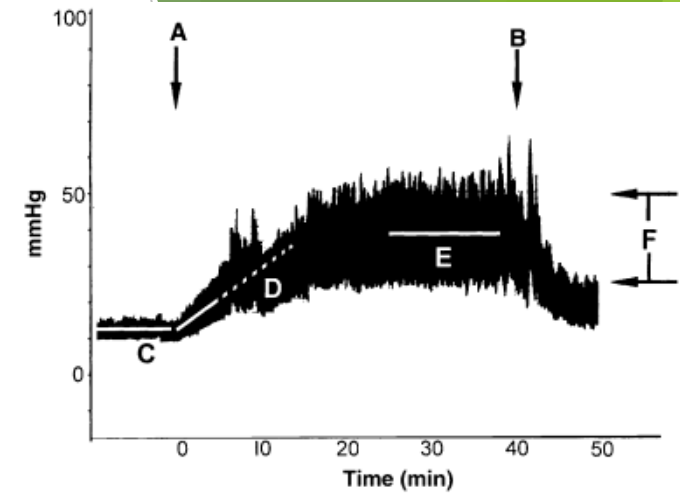
Measure CSF outflow (R-out), above 13+ mmHg/ml/min considered impaired, amongst other metrics.

NICE (2008) approved.

PC-MRI

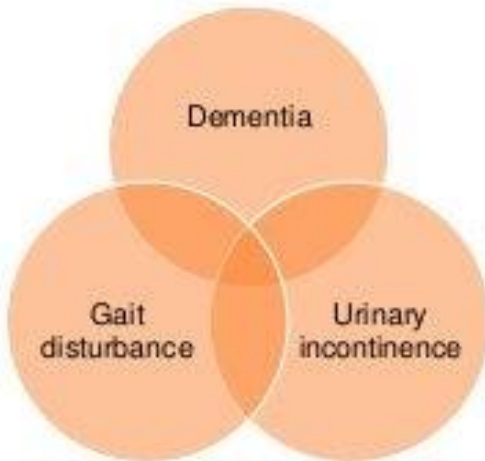
Phase contrast MRI throughout the cardiac cycle.

NPH associated with greater aquaductal outflow of CSF.



Symptoms

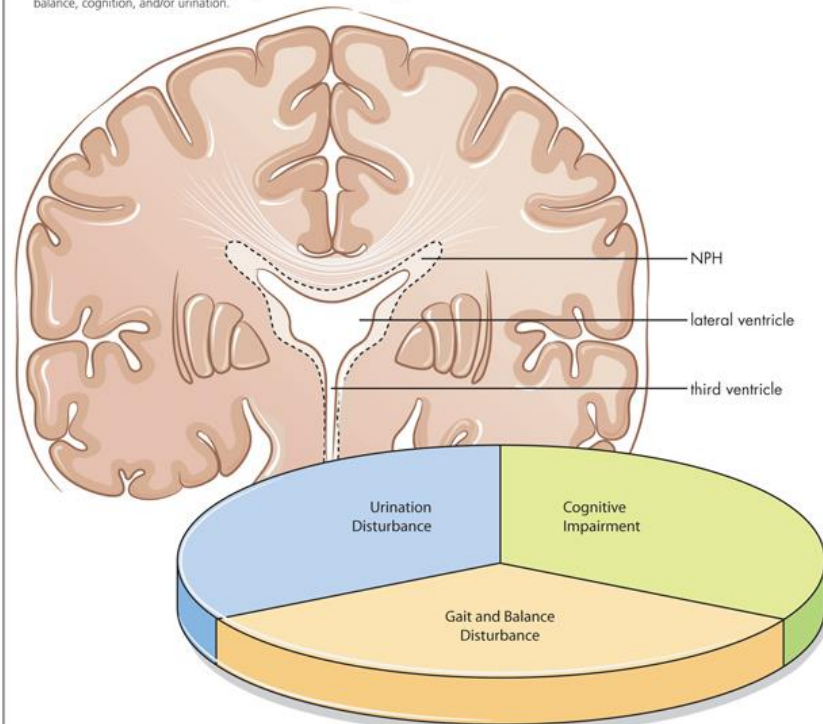
THE CLASSIC CLINICAL TRIAD FIRST DESCRIBED BY HAKIM AND ADAMS IN 1965



Hakim SJ, Adams RD. The so-called clinical triad of symptoms in a patient with normal cerebrospinal fluid pressure in the lateral ventricle. A review. J Neuro (1965) 1:367-371.

Figure 1:
Idiopathic Normal Hydrocephalus Pressure (NPH)

Idiopathic Normal Hydrocephalus Pressure
A condition characterized by chronic nonobstructive enlargement of the cerebral ventricles in association with progressive disturbances of gait, balance, cognition, and/or urination.



"Classic" Clinical Triad of NPH

Gait and balance impairments

- ▶ **Widened base of support** and **outwardly turned feet** (Relkin et al 2005)
- ▶ **Unable to respond to external cues** to correct gait (Morishita et al 2010)
- ▶ **'Magnetic' or shuffled pattern** with low foot clearance
- ▶ Slowness on turning (turning may be 'en bloc) with multiple steps and imbalance
- ▶ May struggle/be unable to respond to external cues to correct gait pattern
- ▶ Difficulties in dual tasking

What do patient's say?

"My legs won't do what I want them to do"

"I need to hold on to the furniture when I walk"

"Turning is difficult"

"I particularly struggle with slopes"

Cognitive changes

- ▶ Pattern of cognitive deficits most commonly observed in iNPH:
 - ▶ **Slower information processing speed**
 - ▶ Slower psychomotor speed
 - ▶ **Executive functioning** (e.g. flexible thinking)
 - ▶ Attention and working memory
 - ▶ Apathy

Urinary incontinence

- ▶ Initial increased urinary frequency
- ▶ As symptoms progress, urge incontinence usually follows and can progress to an absolute urinary incontinence
- ▶ Faecal incontinence can also occur in the more advanced stages

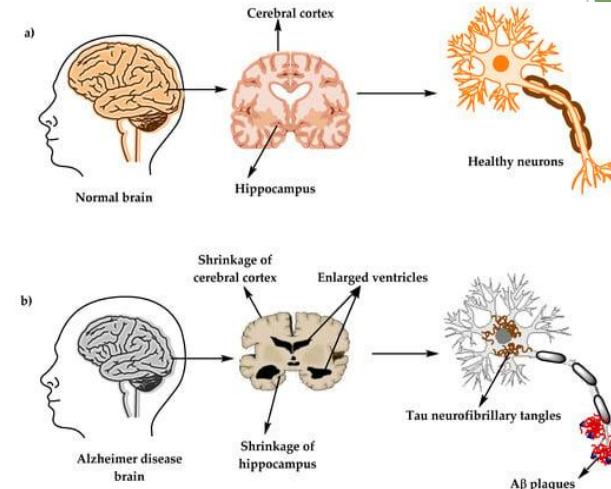
(Rentdorff et al 2012)

Differential diagnosis

Symptoms	PD	iNPH
Gait disturbance	Shuffling gait, retropulsion. External cues might be helpful to reduce freezing. Narrow base. More A/P sway. Kyphotic posture	Shuffling gait, retropulsion. External cues are not usually helpful. Wide based-gait with outward rotation of feet. More R/L sway. More upright posture
Parkinsonism	Bradykinesia, rigidity, resting tremor, freezing, postural instability	Less commonly encountered , rigidity and bradykinesia have been documented
Onset	Asymmetrical	Usually symmetrical
L-dopa Response	Excellent	Poor
Other features	Hyomimia (loss or reduction of facial expression), dysarthria, dysphagia, dystonia	Other PD features rarely encountered
Cognitive symptoms	Cognitive impairment rare in 1-3 years . Processing speed generally well maintained. Not same extent of cognitive slowing	Slowed processing speed and executive functioning difficulties. More rapid deterioration
Autonomic symptoms	Urinary incontinence is rare 1-3 years post diagnosis . Postural hypotension, constipations, urinary urgency/frequency, nocturia, sexual dysfunction, dry eyes, excess sweating	Urinary incontinence/urgency frequently seen in early stages
G-I symptoms	Ageusia (loss of taste), sialorrhea (excess saliva), nausea, reflux, vomiting	Uncommonly reported
Sensory symptoms	Anosmia, pain, paraesthesia (on side of onset)	Uncommonly reported
Sleep disorders	REM sleep disorder, non-REM sleep disorders, vivid dreams, daytime drowsiness, restless legs, insomnia	Daytime sleepiness due to fatigue or sleep disruption more related to urinary urgency
Other	Weight loss, peripheral oedema, diplopia	Uncommonly reported
Family history	Can be present	Not normally present

Alzheimer's/dementia

- Early issues with way-finding
- Episodic memory difficulties (memory for personal events).
- Gait difficulties may be associated with complexity of task and dual-tasking with stride to stride variability
- Language difficulties



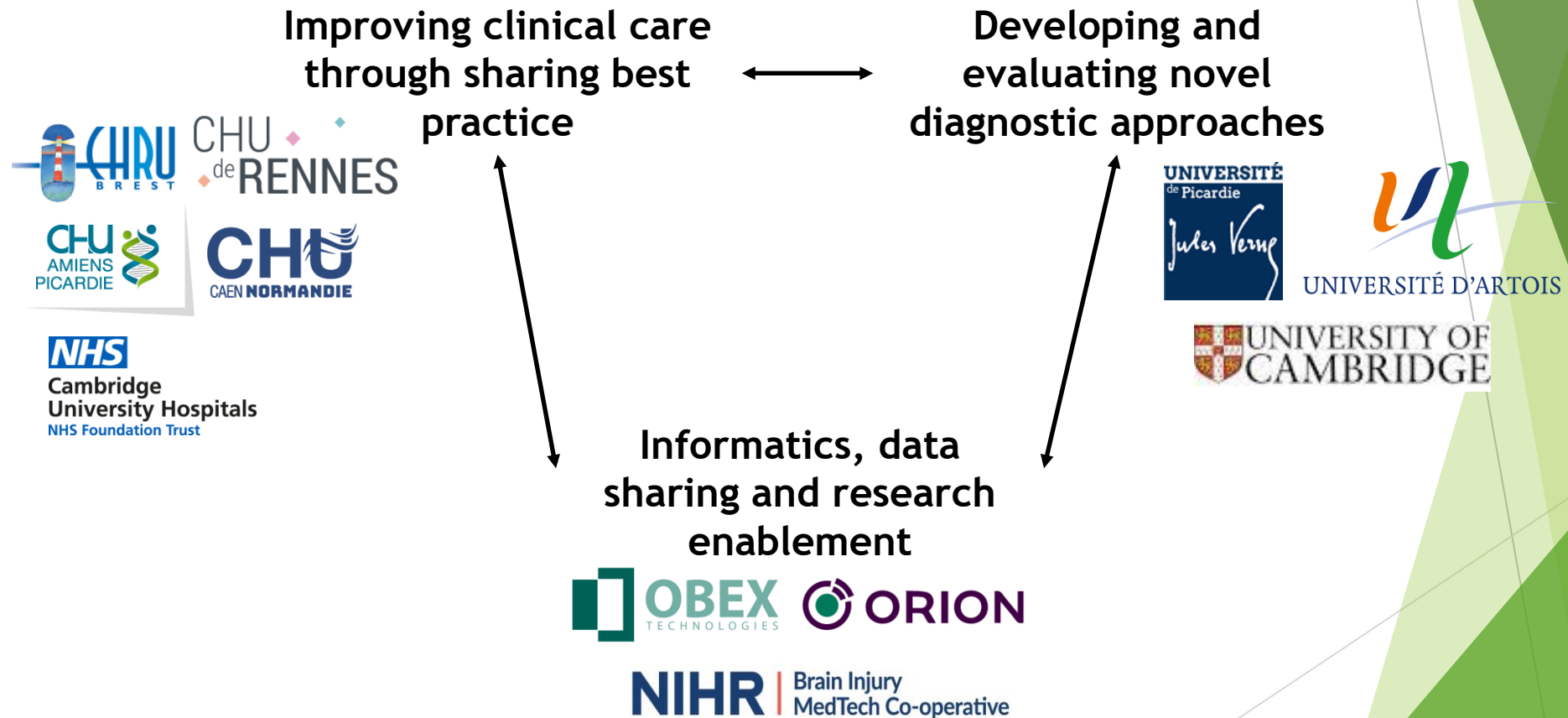
Other PMH

- Considerable PMH
- Cervical myelopathy
- Vestibulopathy
- Previous CNS pathology
- Surgery
- Age-related joint changes (OA, RA, chronic stiffness, posture)
- Peripheral neuropathy

PSP

- Very early on in the condition, the cognitive presentation of PSP can appear similar to iNPH – i.e. Slowed Processing/Psychomotor Speed, Executive Functioning difficulties and Apathy
- Some general slowing of movement, posterior loss of balance, applause sign, axial rigidity, vertical gaze palsy

The Revert Project



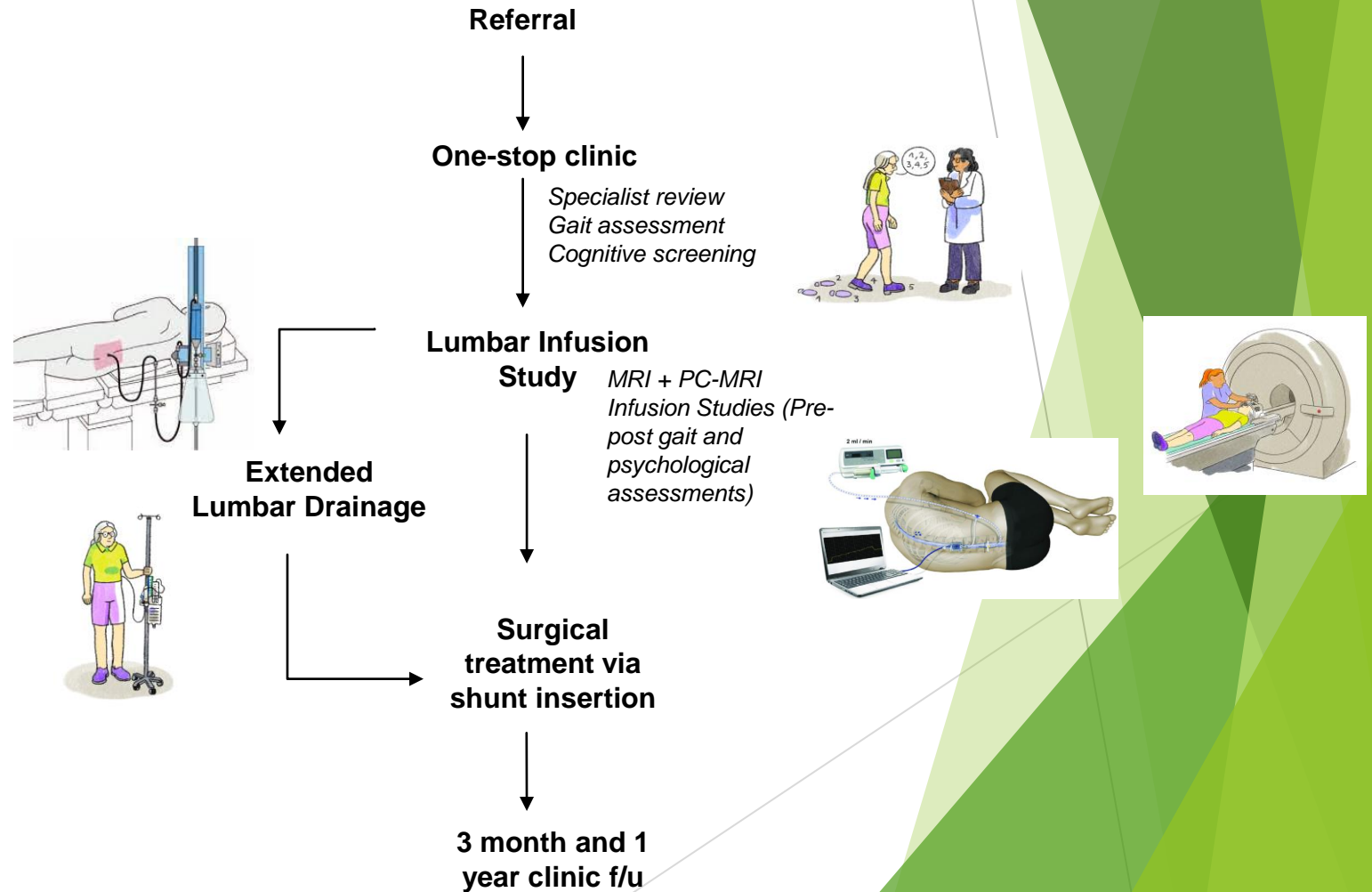
<https://revertproject.org/>

The CUH NPH Service

- ▶ MDT (Neurosurgeon, Specialist Nurse, Infusion Technician, Physiotherapist, Neuropsychologist)
- ▶ Jointly working to support patients across whole clinical pathway from referral to follow-up post surgery
- ▶ Provide in-reach support within teams at CUH
- ▶ Research focus to ensure treatment approach is evidence based and to support future developments in the treatment of iNPH
- ▶ Exploring role for a formal rehab service

Treatment Pathway

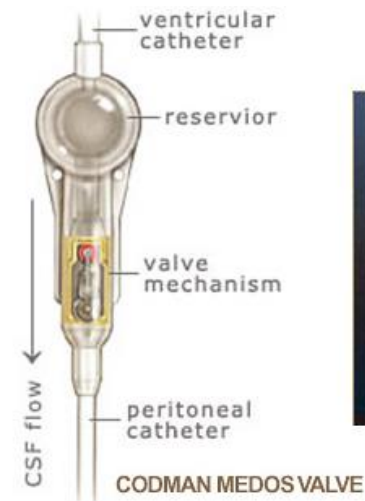
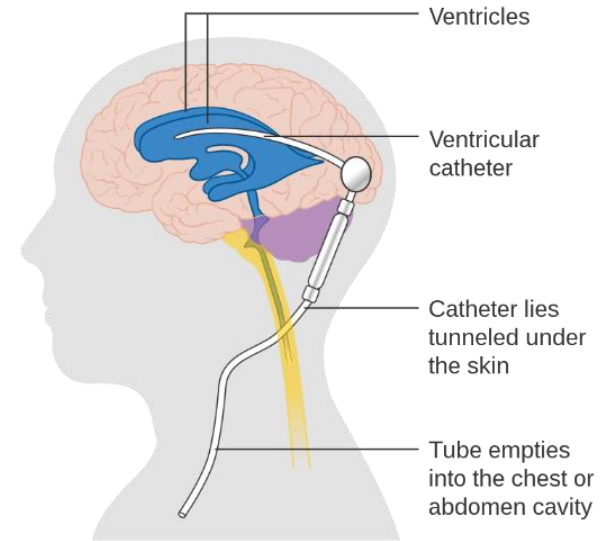
Patient's assessed at each stage of clinical pathway under MDT



Treatment

What is a shunt?

- ▶ An implanted shunt diverts CSF from the ventricles within the brain to another body region where it will be absorbed
- ▶ Creates an alternative route for removal of CSF
- ▶ Usually restores the physiological balance between CSF production, flow, and absorption



MEDTRONIC STRATA VALVE

Prognosis and follow-up

- ▶ Better prognosis associated with earlier diagnosis and treatment (Kajimoto et al 2022, Andren et al 2021)
- ▶ 30 studies published since 2006 included in the systematic review by Toma et al (2013) demonstrated an overall improvement rate of **82% at 1 year** and **73% at 3 years+**
- ▶ Gait improvement seen in 75% of patients, cognitive function improvement in more than 60%, and improvement of incontinence in 55% (Giordan et al 2018)
- ▶ Overall improvement after **12 months** was present in **91.2%** of patients. An overall complication rate of 8.8% and a reoperation rate of 9.4% were recorded (Trungu et al 2022)
- ▶ Aiming to further develop and evidence post surgery rehab for this patient group but growing supportive evidence (Sun et al 2022, Nikaido et al 2022)




How should patients be referred?

- ▶ GP or Consultant based referrals but have had several referrals triggered from letters from Allied Health Professionals.
- ▶ At least **1 of the clinical triad** of symptoms that is **not explainable** by other pre-existing medical diagnoses.

1. In the absence of imaging, GP to refer for possible imaging investigations (CT/MRI) either directly or via neurology

2. With imaging, GP to refer directly to Neurosurgery at CUH via Mr Joannides or Mr Garnett (can accept out of area referrals)

Clinical background

- ▶ Broad band 5 and 6 rotations
- ▶ Specialism within neurology and neuro-rehab
- ▶ Develop experience in leadership, clinical and service development via static and team lead roles
- ▶ Previous experience in research projects through contacts at work (Cochrane review)
- ▶ Research support at CUH via clinical research physiotherapy colleagues and departmental EBP
- ▶ Post initially funded via REVERT, application by CUH  data to support business case and long-term funding
- ▶ External training: masters module, wider research skills courses (NIHR, MOOC, University Library, CAHPR East Anglia), peer support
- ▶ Transferable skills via previous roles to utilise into wider job role



Developing my role

- ▶ Reviewed evidence based to better understand the condition and how to assess and diagnose it
- ▶ Contacted other services and discussed current provision and PT input
- ▶ Liaised locally within our MDT and more widely across the hospital to identify gaps and service need
- ▶ Worked closely within team to collaborate and develop research ideas
- ▶ Ongoing adaptation and evolution of role as have started

What does my role entail

- ▶ Day to day assessment and management of patients through service and trouble-shooting
- ▶ Attend weekly MDT clinic for follow-ups and new patients
- ▶ Clinically oversee in-patients both with confirmed and possible diagnoses
- ▶ Teaching/networking to increase awareness of iNPH
- ▶ Work with wider partners to build resources and links
- ▶ Develop clinical research to help build the evidence base for treatment of iNPH
- ▶ Collect and utilise service level data to help the iNPH service grow and build evidence to support funding
- ▶ Develop wider skills of research, leadership and service development

Challenges and opportunities

- ▶ Working on edge of clinical scope
- ▶ Previous lack of research knowledge and experience
- ▶ Understanding more broadly how services operate
- ▶ Clinical leadership – moving away from just people management to leading and promoting services
- ▶ Building wider skills
- ▶ Increased degree of self-management and self-control
- ▶ Managing broader projects over longer periods of time
- ▶ Working in a narrower clinical specialism – less direct departmental support
- ▶ Uncertainty with secondment
- ▶ Transferable skills



Post-survey

- ▶ <https://addenbrookes.onlinesurveys.ac.uk/inph-postteachingsurvey>



Any questions?



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