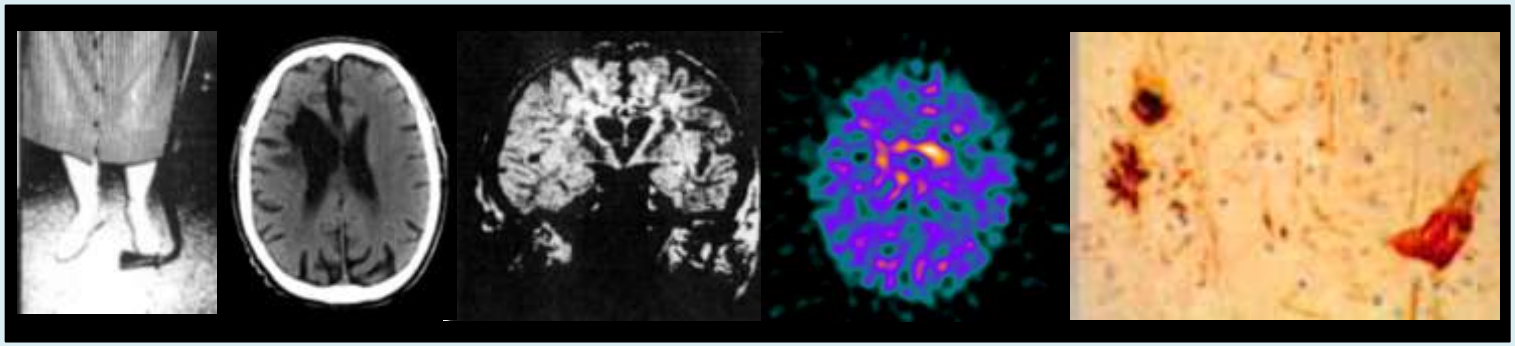
Trastorns de la marxa: no tot és Parkinson…
… o sí?



Dr. Yaroslau Compta Hirnyj (MD; PhD)
Unitat de Parkinson i Trastorns del Moviment
Hospital Clínic / IDIBAPS / Universitat de Barcelona
III Jornada d'atenció a la gent gran en Atenció Primària
Auditori CAMFiC, Barcelona, 30.v.2014

SUMARI

- Breu repàs fisiològic de la marxa
- Classificació dels trastorns de la marxa
- Consideracions en el diagnòstic diferencial
- Paper de les proves terapèutiques (*dx ex-juvantibus*)
- Conclusions

FISIOLOGIA DE LA MARXA

NEUROLOGY 1993

Human walking and higher-level gait disorders, particularly in the elderly

J.G. Nutt, MD; C.D. Marsden, DSc; and P.D. Thompson, MD

Neural control mechanisms	Physiologic mechanisms	Abnormalities
I. Equilibrium		
1. Arising to the erect posture	Righting reactions	Inability to rise
2. Support upright position	Supporting reactions	Inability to stand
3. Correct perturbations and adapt to circumstance	Anticipatory postural reactions Reactive postural responses Rescue reactions Protective reactions	Inability to protect upright stance
II. Locomotion		
1. Initiate steps	Shift center of gravity Start stepping	Gait ignition failure
2. Stepping	Locomotion	Alterations in pattern of stepping
3. Adapting stepping to circumstances	Voluntary	Inability to perform dexterous stepping
III. Non-neurologic factors		
1. The mechanical support system	Bones, joints	Limp
2. General health (cardiorespiratory)	Exercise tolerance	Slowness

CLASSIFICACIÓ DELS TR. DE LA MARXA

I. Lowest-level gait disorders
- A. Peripheral skeletomuscle problems
  - Arthritic gait
  - Myopathic gait
  - Peripheral neuropathic gait
- B. Peripheral sensory problems
  - Sensory ataxic gait
  - Vestibular ataxic gait
  - Visual ataxic gait

II. Middle-level gait disorders
- Hemiplegic gait
- Paraplegic gait
- Cerebellar ataxic gait
- Parkinsonian gait
- Choreic gait
- Dystonic gait

III. Highest-level gait disorders
- Cautious gait
- Subcortical disequilibrium
- Frontal disequilibrium
- Isolated gait ignition failure
- Frontal gait disorder

Nutt et al., Neurology, 1993

Table 3. Final classification of unusual or unexplained gait disorders in 43 elderly patients

Gait disorder	No. cases (men/women)	Age range (yr)	Disequilibrium (impaired balance)	Gait ignition failure (start/turn hesitation) (freezing)	Wide base	Shortened stride
Cautious gait	16 (8/8)	65-98	Mild	Absent	Mild	Mild to moderate
Subcortical disequilibrium	6 (3/3)	70-95	Severe	Variable	Variable	Variable
Frontal disequilibrium	8 (2/6)	69-86	Severe	Variable	Variable	Variable
Isolated gait ignition failure	5 (5/0)	68-75	None	Severe	Absent	Absent*
Frontal gait disorder	8 (8/0)	75-88	Moderate	Moderate to severe	Variable	Mild to moderate

* Steps could be of normal size and rhythm once walking was under way.

Table 4. Associated findings in elderly patients with gait disorders

Type of gait disorder	Dementia	Apraxia*	Frontal release signs†	Parkinsonism‡	Pyramidal signs	Urinary incontinence
Cautious gait	No	No	No	No	No	No
Subcortical disequilibrium	No	No	No	Occasional	Occasional	No
Frontal disequilibrium	Common	Occasional	Common	Occasional	Occasional	Yes
Isolated gait ignition failure	No	No	No	No	No	No
Frontal gait disorder	Common	Occasional	Common	Occasional	Occasional	Yes

* Examined by the ability to perform actions to command or mime gestures with arms or legs.
† Frontal release signs included gegenhalten (a variable increase in muscle tone often accompanied by an inability to relax during passive limb manipulation), grasp reflexes (hand and foot), and rooting responses.
‡ Hypokinesia, difficulty executing rhythmic and repetitive alternating or sequential movements.

Nutt et al., Neurology, 1993

Table 5. Comparison of proposed terms used to describe the clinical patterns of gait in the elderly in this paper with those from previous publications

Proposed terminology	Previous terms	Lesions
Cautious	Elderly gait Senile gait	Musculoskeletal Peripheral nervous lesions Central nervous lesions
Subcortical disequilibrium	Tottering Astasia-abasia Thalamic astasia	Midbrain Basal ganglia Thalamus
Frontal disequilibrium	Gait apraxia Frontal ataxia Astasia-abasia	Frontal lobe and white matter connections
Isolated gait ignition failure	Gait apraxia Magnetic gait Slipping clutch gait Lower half parkinsonism Arteriosclerotic parkinsonism Trepidant abasia (Petren's gait)	Frontal lobe, white matter connections and basal ganglia
Frontal gait disorder	Marché à petits pas Magnetic gait apraxia Arteriosclerotic parkinsonism Parkinsonian ataxia Lower half parkinsonism Lower body parkinsonism	Frontal lobe and white matter lesions

CONSIDERACIONS EN EL DX DIFERENCIAL

Box 1 Most common causes of lower-body parkinsonism.

Normal pressure hydrocephalus
- Idiopathic
- Secondary (meningitis, head trauma)

Vascular parkinsonism
- Binswanger's disease (subcortical ateriosclerotic encephalopathy)
- Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL)
- Dilation of perivascular spaces (also very common in CADASIL patients)
- Multiple lacunar infarcts

Frontal lobe lesions
- Tumors
- Ischemia
- Demyelination

Progressive supranuclear palsy

From Espay et al., Nature Clinical Practice Neurology , 2007

Movement Disorders
Vol. 4, No. 3, 1989, pp. 249–260

Lower Body Parkinsonism: Evidence for Vascular Etiology[1]

Patricia M. FitzGerald and Joseph Jankovic

Department of Neurology, Baylor College of Medicine, Houston, Texas

We studied 10 patients with marked gait difficulty and no or only minimal upper limb involvement, defined here as lower body parkinsonism (LBP). They were compared to a control group of 100 patients with otherwise typical Parkinson's disease (PD)

Group	Symptoms duration	Gait disturbance at onset	Hypertension	L-dopa response
LBP	2.6 ± 1.5 y	90%	70%	22%
IPD	7.5 ± 4.9 y	7%	21%	96%

Movement Disorders
Vol. 4, No. 3, 1989, pp. 249–260

Lower Body Parkinsonism: Evidence for Vascular Etiology[1]

Patricia M. FitzGerald and Joseph Jankovic

Department of Neurology, Baylor College of Medicine, Houston, Texas

(a) Score ≥2 on walking and gait UPDRS-variable;

(b) Score ≤1 in arm tremor, rigidity, and dexterity UPDRS-variables;

(c) No dementia, spasticity, or lateralizing findings;

(d) Other exclusion criteria:
- documented cerebral trauma;
- tumor, stroke or surgery;
- post-enkephalitic parkinsonism;
- drug- or toxin-induced parkinsonism;
- heredodegenerative diseases (parkinsonism plus syndromes);
- age <50 years at time of assessment


J Neurol (2004) 251 : 513–524
DOI 10.1007/s00415-004-0421-4

REVIEW

Vascular parkinsonism

Igor Sibon
Gilles Fenelon
Niall P. Quinn
François Tison

* Incidental vascular lesions occurring in true IPD are up to 10 times more common than VP

* Only 7 VP cases in the UKPDS Brain Bank series of 200 patients clinically diagnosed as IPD

* Vascular“pseudo-parkinsonism” → isolated gait disorders called “LBP”,“frontal-type gait disorders” or “gait ignition failure”

* Usefulness of dopamine transporter SPECT → normal or atypical uptake reduction in VP

	Gait ignition failure	Parkinson's disease	Frontal gait disorders
Standing from seated position	Normal	Impaired	Impaired
Posture during standing/walking	Upright	Flexed	Upright or flexed
Stance and stride base	Narrow	Narrow	Wide
Equilibrium	Normal	Normal (early)	Abnormal
Postural reflexes	Normal	Normal (early)	Abnormal
Protective/rescue reactions	Normal	Normal (early)	Abnormal
Retropulsion	Absent	Present	Present (may fall)
Start/turn hesitation	Marked	Marked	Marked
Stride length	Normal	Short (shuffling)	Short (shuffling)
Freezing	Yes	Yes	Yes
Festination	No	Yes	No
Arm swing	Normal	Reduced	Variable
Leg movement when seated	Normal	Slow	Variable
Apraxia	No	No (bradykinesia)	Variable

Box 1 Most common causes of lower-body parkinsonism.

Normal pressure hydrocephalus

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Frontal lobe lesions

- Tumors
- Ischemia
- Demyelination

Progressive supranuclear palsy

From Espay et al.,
Nature Clinical
Practice Neurology , 2007

Am Fam Physician 2004;70:1071-8,1085-6

Management of Normal Pressure Hydrocephalus

MEG VERREES, M.D., and WARREN R. SELMAN, M.D.
Case Western Reserve University, Cleveland, Ohio

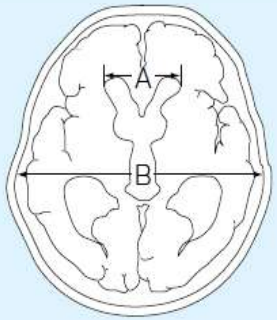
Signs and Symptoms of Normal Pressure Hydrocephalus

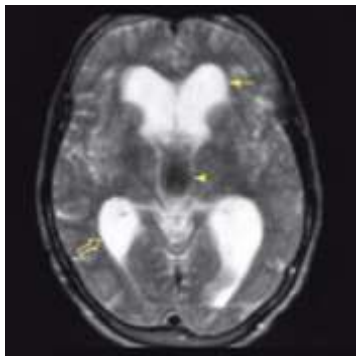
- Ambulation
  - Difficulty or arrest in initiation of ambulation
  - Feet appear "glued to the floor"
  - Gait instability, multiple falls
  - Wide-based stance, shuffling steps
- General
  - Generalized slow movement
- Incontinence
- Mentation
  - Lack of spontaneity in movement, verbal response, and emotion
  - Latency in response to questions or reaction to situations
  - Slowness in processing information

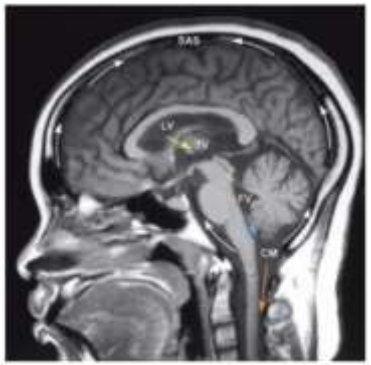
TABLE 2
Differential Diagnosis of Normal Pressure Hydrocephalus

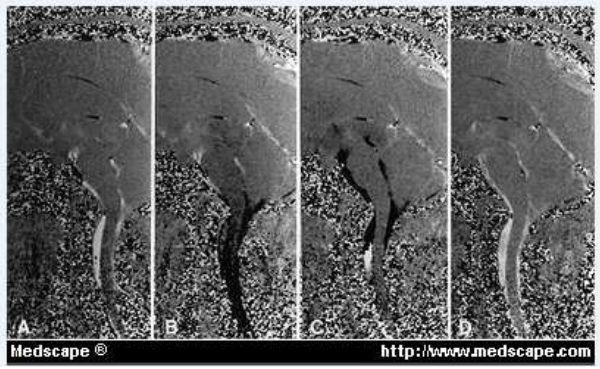
- Alzheimer's disease
- Carcinomatous meningitis
- Chronic alcoholism
- Combinations of conditions affecting gait (e.g., rheumatoid arthritis, cervical stenosis) coupled with conditions that impinge on mentation (e.g., Alzheimer's disease, multi-infarct dementia) and those that have an effect on urination (e.g., prostate disease)
- Intracranial infection (e.g., abscess, subdural empyema, meningitis)
- Multi-infarct dementia
- Parkinson's disease
- Subcortical arteriosclerotic disease
- Subdural hematoma
- Systemic diseases (e.g., hypothyroidism, Addison's disease) or malignancy
- Tumor (benign or malignant)

Evans index : A/B









In NPH, success rates of this procedure vary from 33 percent to more than 90 percent.[4,15,20] This wide range in outcome probably reflects variations in patient selection.[15] The most serious complications of shunt placement include subdural and intracerebral hematomas.

Strength of Recommendations

Key clinical recommendation	Label	References
Examination of the results of multiple studies yields a wide variation in patient response to shunting.	B	2,20
No gold standard test is available to identify patients who will benefit from the shunting procedure.	B	3,7,8

CASE STUDY

www.nature.com/clinicalpractice/neuro

Focus on vascular neurology

Lower-body parkinsonism: reconsidering the threshold for external lumbar drainage

Alberto J Espay*, Raj K Narayan, Andrew P Duker, Edwin T Barrett Jr and Gabrielle de Courten-Myers

80-year-old man → 30 months history of step-wise progressive short-stepped gait and impaired balance with multiple falls along with memory decline

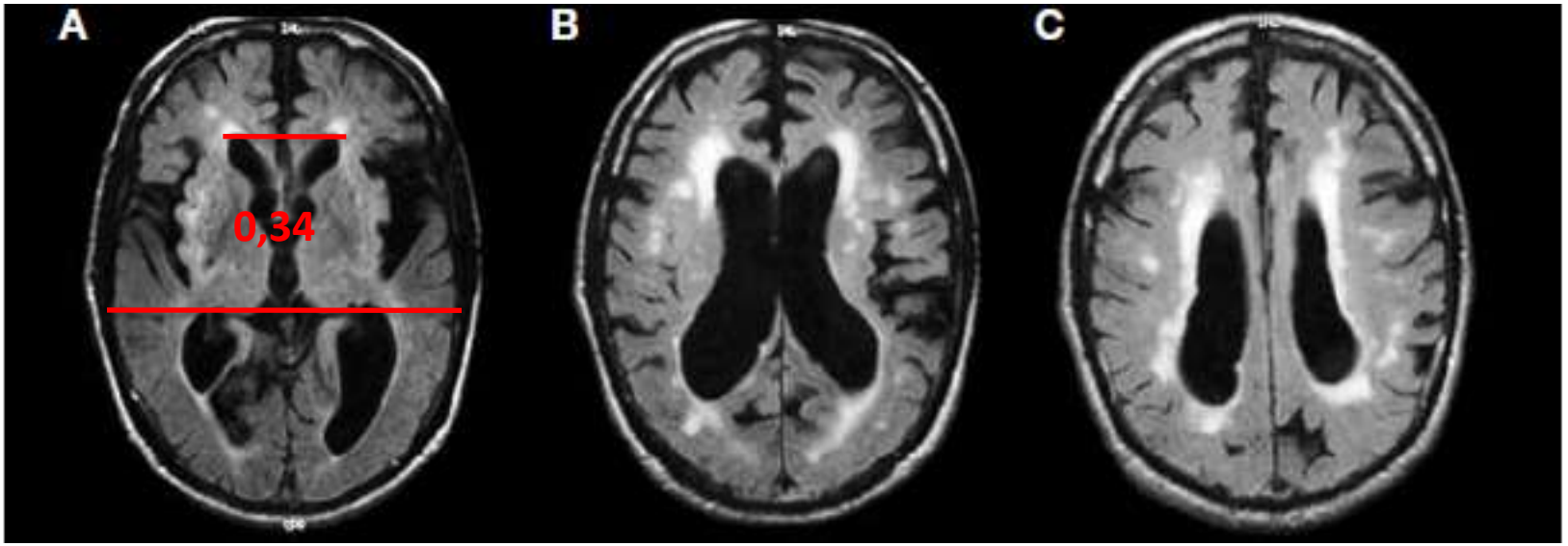
In the last 9 months → urine incontinence

Levodopa up to 600 mg/day failed to alleviate his symptoms.

The patient resided at a rehabilitation facility where he received assistance with gait and transfers, but he was able to bathe, dress, or feed without assistance

Past medical history → + for hypertension, hypercholesterolemia & small vessel disease

Examination → short-stride gait without stooping, shuffling, or festination
→ MMSE 25/30
→ frontal release signs
→ neuropsychological testing → multiple domain involvement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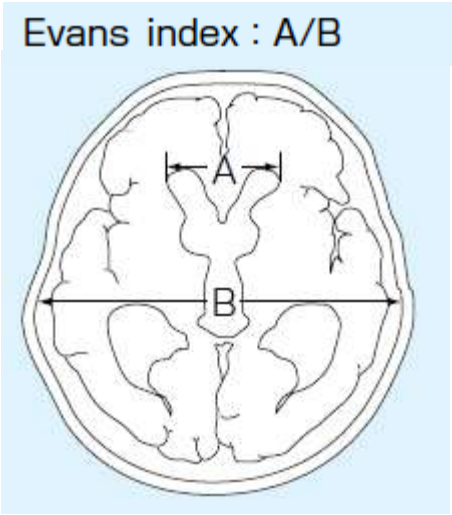
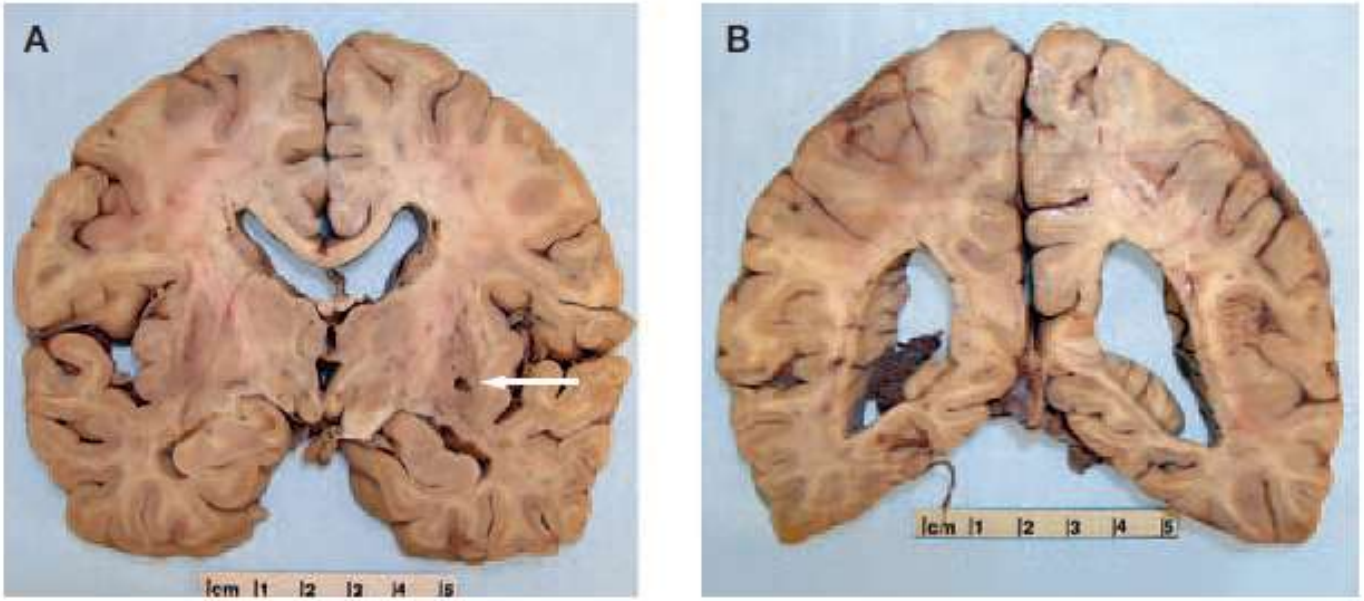

Evans index : A/B
A
B

Table 2 Gait assessments before and after ELD.

Parameter	Straight path (2.6 m)			Turning path (0.7 m)		
	Pre-ELD	Post-ELD	Percentage change	Pre-ELD	Post-ELD	Percentage change
Timed gait (s)	22	13	40.9	15	6	60
Cadence (steps/min)	76	92	17.4	68	100	47.1
Gait velocity (m/min)	7.2	12	40	2.8	7.0	250
Step length[a] (cm)	9.2	12.9	28.7	4.1	7.0	70.7



Box 2 Clinical features of gait in VaP and NPH.

Common to NPH and VaP

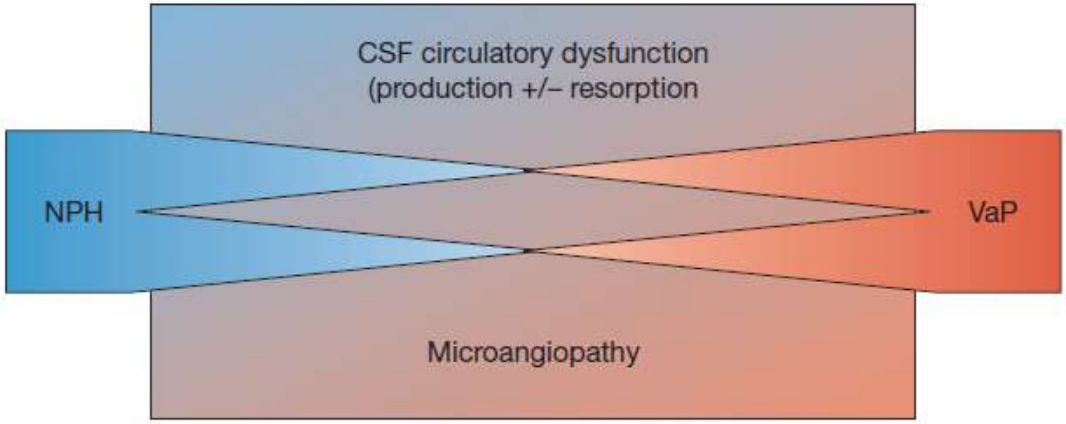
- Reduced gait velocity
- Reduced stride length
- Diminished step height
- Increased step width
- Freezing of gait
- Gait apraxia ("frontal ataxia" or "magnetic foot")
- Preserved arm swing
- Poor response to external cues[a]
- Poor response to levodopa

Presumed unique to NPH

- Broad-based gait with outwardly rotated feet[17]

Presumed unique to VaP

- Upright posture with "wooden appearance,"[6] reduced hip extension, reduced knee flexion[18]
- Lack of festination (i.e. hastening steps with progressively shortened stride)



Cohort of 347 patients with LBP

Freezing was seen at comparable rates in VaP & NPH

VaP & NPH could be similar 'frontal gait disorders'

[Giladi N *et al.* (1997)]

• Extensive basal ganglia MRI abnormalities → present in controls

• By contrast, and counter-intuitively, striatal infarcts are rarely followed by clinical parkinsonism

• Post-mortem MRI–pathologic correlations of asymptomatic subjects with periventricular hyperintensities:

* myelin pallor
* dilatation of perivascular spaces
* increase in extracellular spaces
* discontinuity of ependymal lining
* subependymal gliosis

Periventricular WMHs on MRI might not always indicate microangiopathy

→ In some instances might correspond to NPH

Movement Disorders
Vol. 17, No. 1, 2002, pp. 91–97

Published by Wiley-Liss, Inc.
DOI 10.1002/mds.1270

Vascular Parkinsonism: Clinical Correlates Predicting Motor Improvement After Lumbar Puncture

William G. Ondo, MD,[1]* Ling Ling Chan, MD,[2] and Joel K. Levy, PhD[1]

* Any positive response to levodopa ($P < 0.001$)
* Lack of vertical gaze palsies ($P < 0.05$)
* Lack of a pure freezing gait ($P < 0.05$)
* Lack of hypotensive episodes ($P < 0.05$)

Predictors of good response to removal of 35-40 CSF mL via lumbar pucture in 40 PD patients

Blinded MRI interpretation did not find any clear-cut response-predictors

VP patients clinically resembling IPD improve after LP

Non-responders more closely resemble PSP

¿¿¿Possibility of testing CSF drainage in IPD patients with subcortical WMHs???

Box 1 Most common causes of lower-body parkinsonism.

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- Idiopathic
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Frontal lobe lesions

- Tumors
- Ischemia
- Demyelination

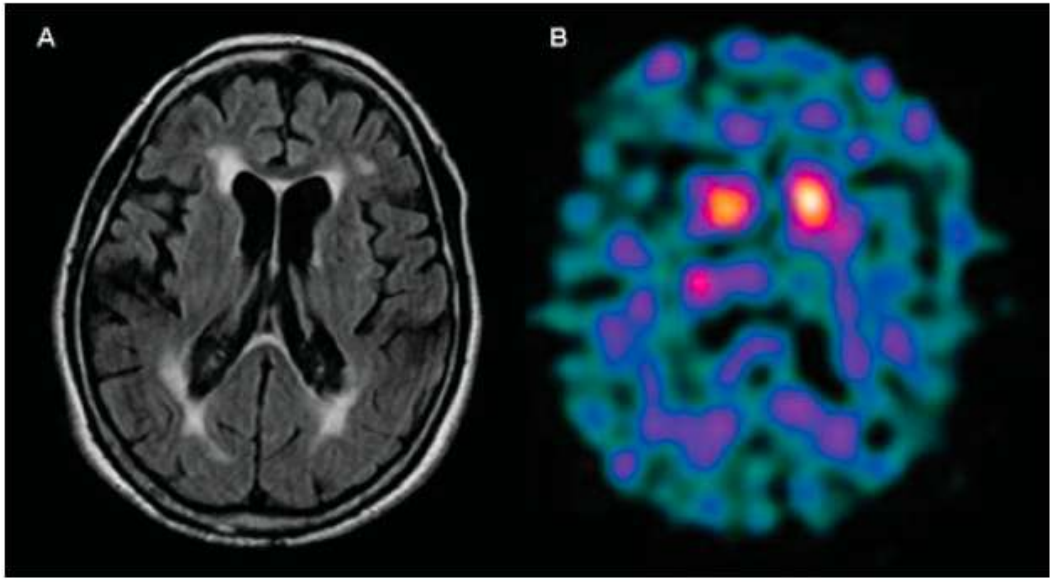
Progressive supranuclear palsy

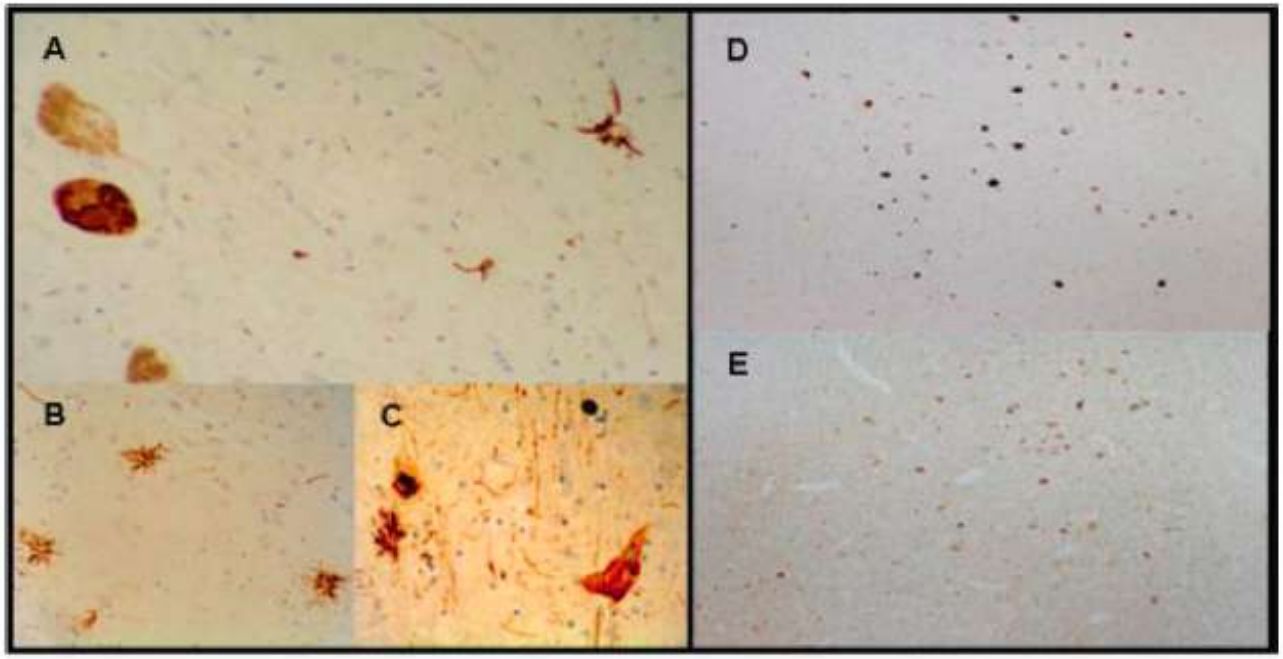
From Espay et al.,
Nature Clinical
Practice Neurology , 2007

Movement Disorders, Vol. 22, No. 13, 2007

Long Lasting Pure Freezing of Gait Preceding Progressive Supranuclear Palsy: A Clinicopathological Study

Yaroslau Compta, MD,[1]
Francesc Valldeoriola, MD, PhD,[1]*
Eduardo Tolosa, MD, PhD, FRCP,[1]
María Jesús Rey, PhD,[2] María José Martí, MD, PhD,[1]
and Josep Valls-Solé, MD, PhD[1]





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Progressive supranuclear palsy

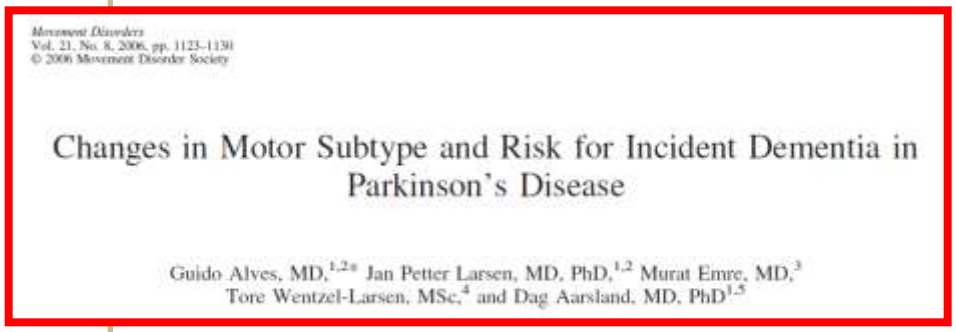
From Espay et al., Nature Clinical Practice Neurology , 2007

Parkinson's disease ???

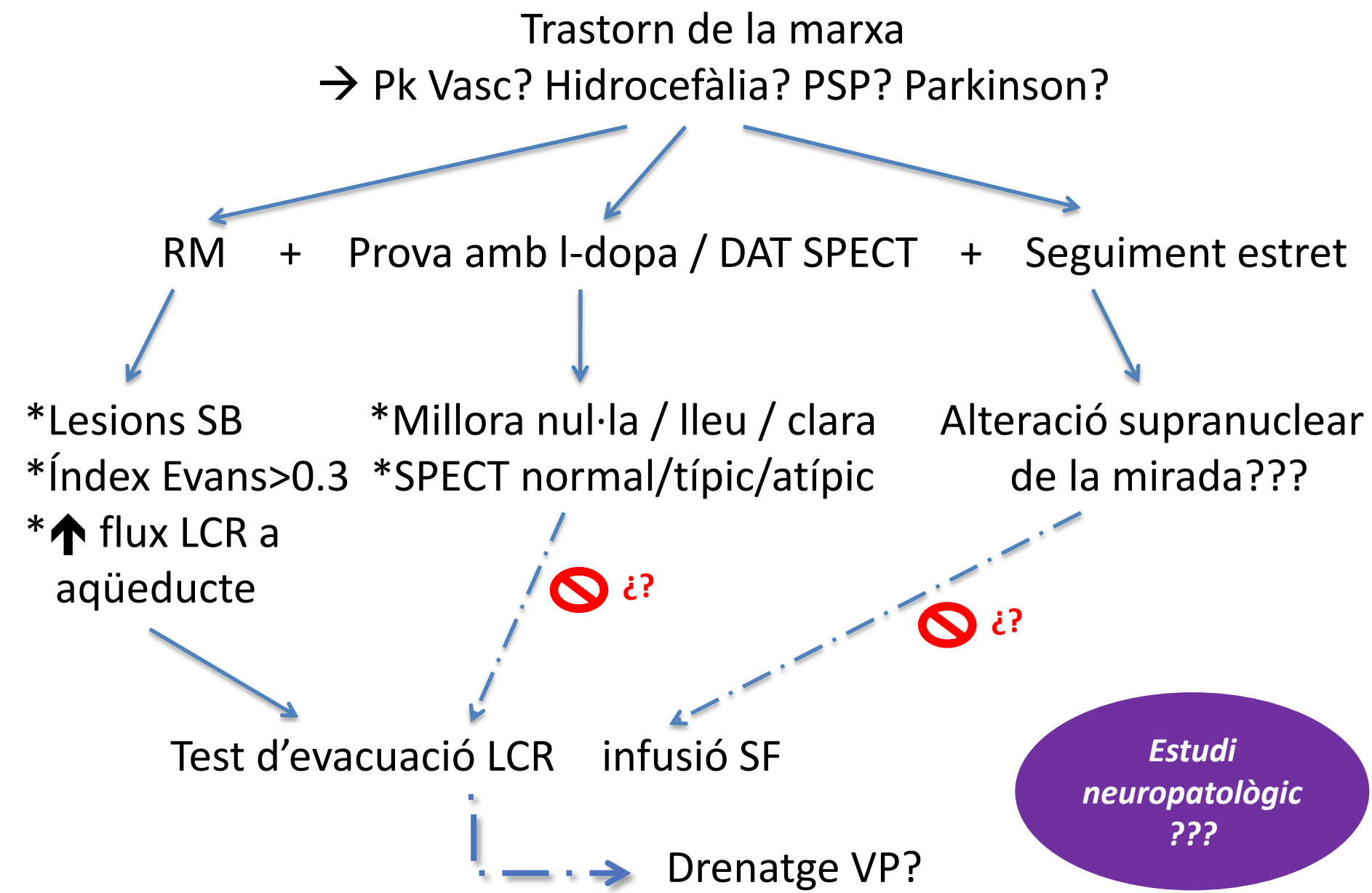
Postural instability & gait disturbance (PIGD)

*A PD phenotype proposed by Jankovic and further defined in 1990 (DATATOP study)

*Related since then to poorer prognosis including higher risk of dementia as opposed to tremor-dominant PD



PAPER DE LES PROVES TERAPÈUTIQUES EN EL DX



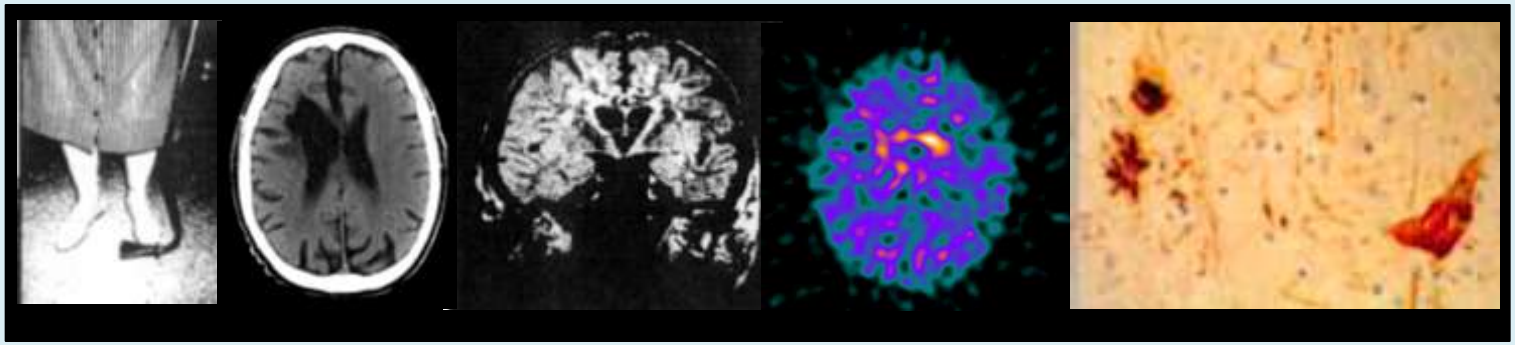
CONCLUSIONS

- Els trastorns de la marxa poden ser de nivell:
  - ***inferior*** → causes mecàniques / osteoarticulars / SNP / sensorials;
  - ***intermedi*** → tronc / GGBB / cerebel → atàxia, parkinsonisme, corea;
  - ***superior*** → alteracions de connexions córtico[fronto]-subcorticals
    - → marxa cautelosa
    - → desequilibri subcortical i frontal
    - → fracàs de l'inici de la marxa

- El dx dcial del nivell superior és complex:
  - causa vascular,
  - causa hidrocefàlica,
  - causa degenerativa,
  - causa autoimmunitària?

- Considerar sempre causes tractables:
  - ***hidrocefàlia***...
  - ... però també el ***Parkinson*** !!!

Paper de
dx ex-juvantibus i
mesures "agressives"



Moltes gràcies