Mouvements anormaux et pathologie vasculaire cérébrale.

Dr Isabelle BENATRU
CHU de Poitiers

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Introduction

- Spectre clinique vaste de mouvements anormaux, hypo ou hyperkinétiques

- Tous les niveaux et/ou structures des circuits moteurs sous corticaux frontaux peuvent être impliqués :
  - Cortex sensorimoteur
  - Striatum
  - Pallidum
  - Thalamus
  - NST
  - Substance noire
  - Cervelet
  - Voies de connexion

Introduction

- Prévalence
  - Mal connue
  - Dans la littérature : cas cliniques isolés ou petite série de cas
  - 1.1 à 3.9%

- AVC ischémique et hémorragique

- AVC ischémique de toute origine
  - Pathologie des petites artères avec lacunes multiples
  - Athérothrombose
  - Cause cardio-embolique

Ghika-Schmid et al, 1997; Alarcon et al, 2004

Mode de survenue

Transitoire/paroxystique

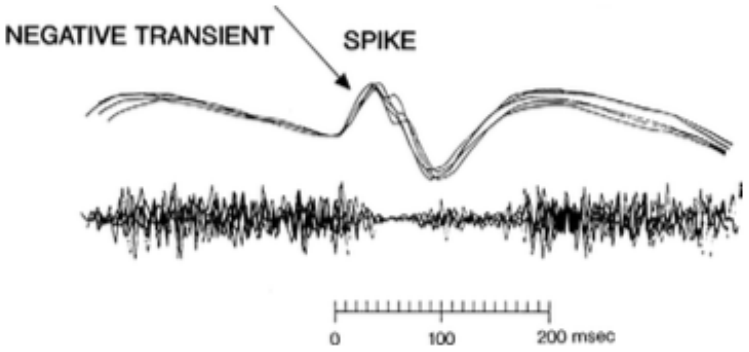
- Astérixis
- « Limbshaking »
- Hémichorée
- Hémiballisme

Permanent

- Dystonie
- Tremblement
- Myoclonus
- Syndrome parkinsonien
- Athétose

Asterixis

- Phase aigu de l'AVC
- Suspensionspontanée et involontaire du tonus musculaire lors du maintien de la posture
- EMG : myoclonies négatives
- Unilatéral, atteinte du membre supérieur

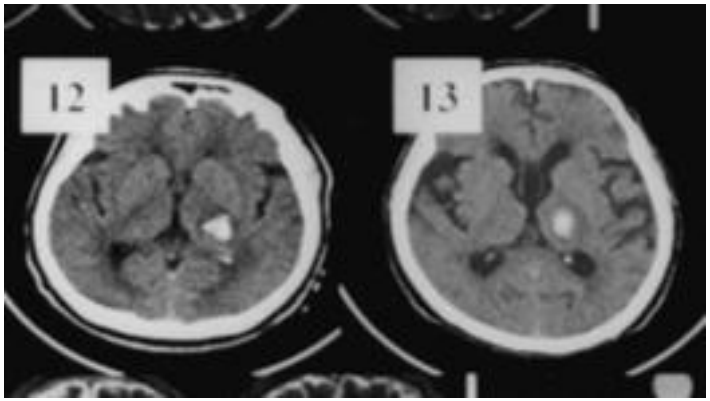


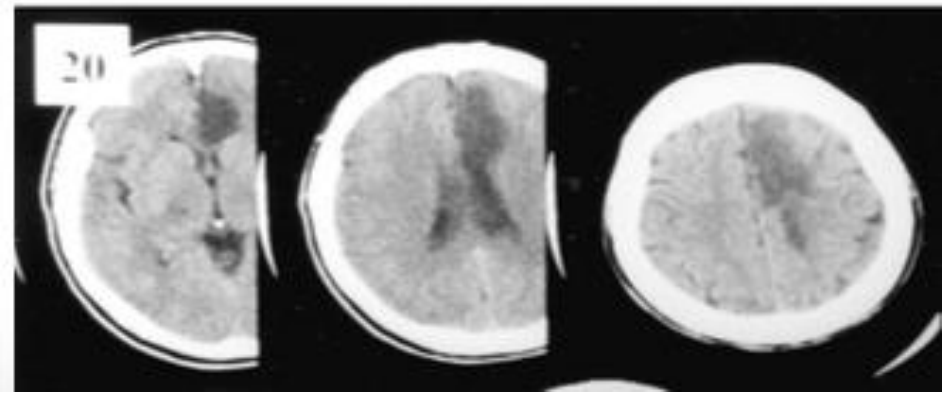
- Traitement :
  - difficile car intermittent et peu invalidant.
  - Traitement anti-épileptique

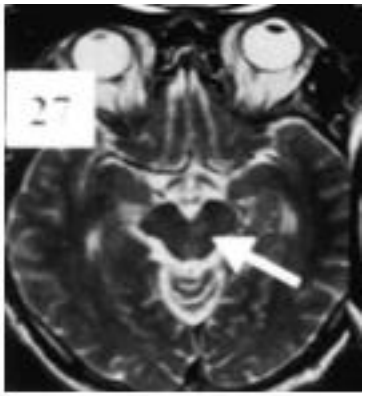
Kim et al, 2001

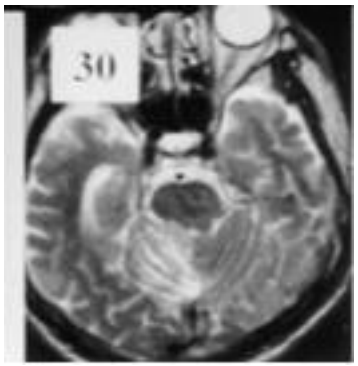
- Lésions du thalamus controlatéral (lobe frontal, noyau lenticulaire, cervelet, tronc cérébral)

- Lésions ipsilatérales de la moelle ou du cervelet









Kim et al, 2001

Chorée, ballisme et athétose

- *Ballisme* : mouvements amples, brusques de grande amplitude, affectant plutôt la partie proximale des membres (membre supérieur)
- *Chorée*: mouvementsinvolontaires plutôt distaux, peu amples, continus, non stéréotypés.
- *Athétose* : mouvements lents, sinueux, reptatoires.

- Présents au repos, majorés par le mouvement, améliorés par la relaxation et disparaissant au sommeil

- Signes associés (variables selon la localisation de l'AVC): dysarthrie, déficit moteur, hypotonie, dystonie

Mitzushima et al, 1997; Bejot et al, 2008; Pantano et al, 1996; Chung et al, 2004; Kenney et al, 2007; Jankovic, 2009

Chorée, ballisme et athétose

- **Lésions du NST** et ses connexions (partie sensorimotrice du putamen)
- Toute interruption des circuits cortex- ganglions de la base
  - Hémiballisme : lésions du territoire de l'ACM frontale, pariétale, corona radiata
  - Hémichorée : striatum controlatéral

- Traitement :
  - Tétrabénazine
  - NL

Mitzushima et al, 1997; Bejot et al, 2008; Pantano et al, 1996; Chung et al, 2004; Kenney et al, 2007; Jankovic, 2009

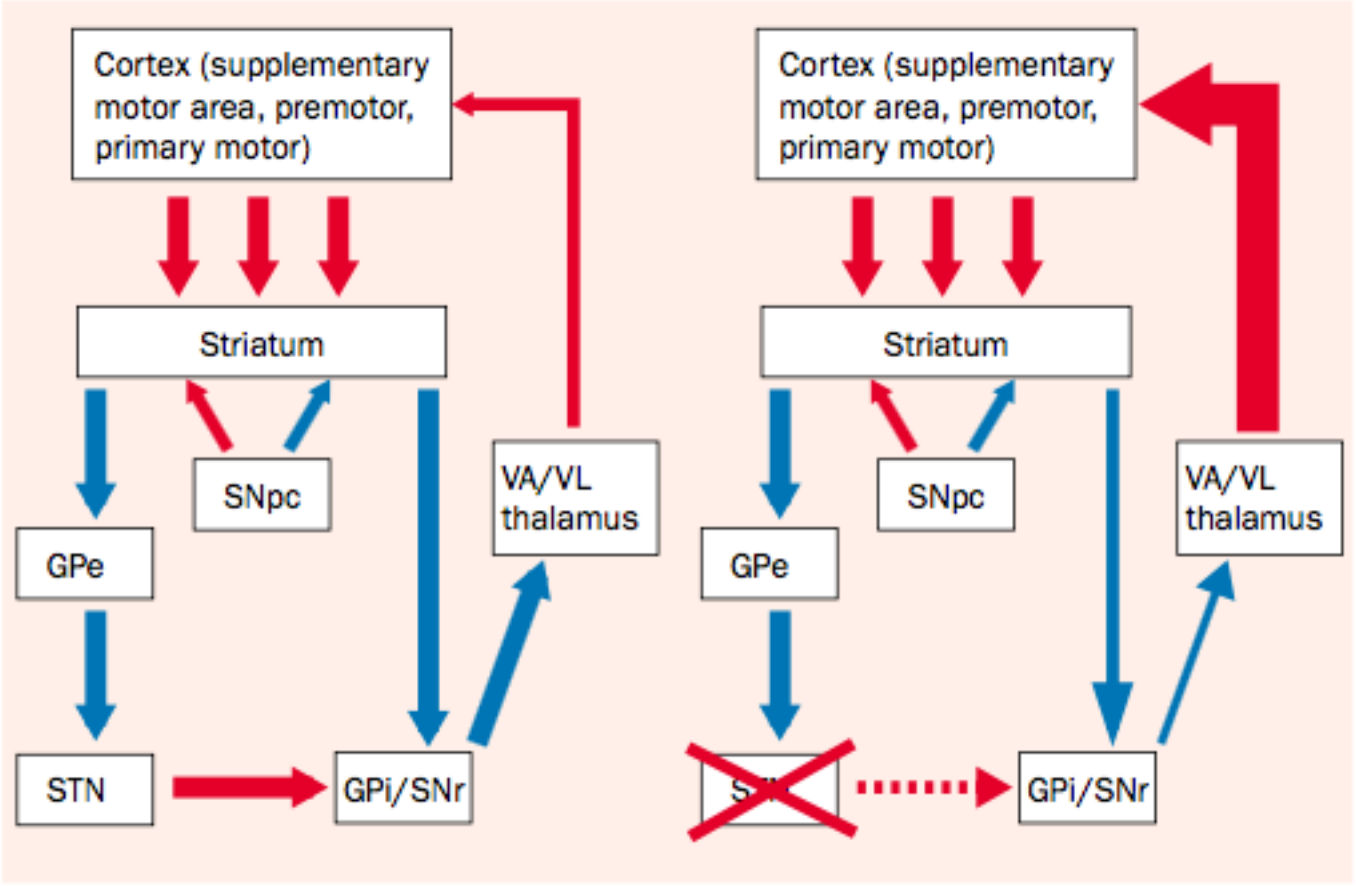


Figure 3. Classic model of hemiballism. A simplified model of basal-ganglia circuitry is shown, with excitatory output indicated by black arrows, and inhibitory output by open arrows. Left: normal basal-ganglia circuitry. Right: basal-ganglia circuitry after a lesion of the STN; the change in arrow width reflects a change in activity in the related pathway. A lesion in the STN causes decreased excitatory innervation of the GPi by the STN, leading to decreased inhibitory output from the GPi to the thalamus, increased excitatory output to the motor cortex from the thalamus, and excessive movement. SNpc=substantia nigra pars compacta; SNr=substantia nigra pars reticulata; VA/VL=ventroanterior/ventrolateral.

Lang et al, 2003

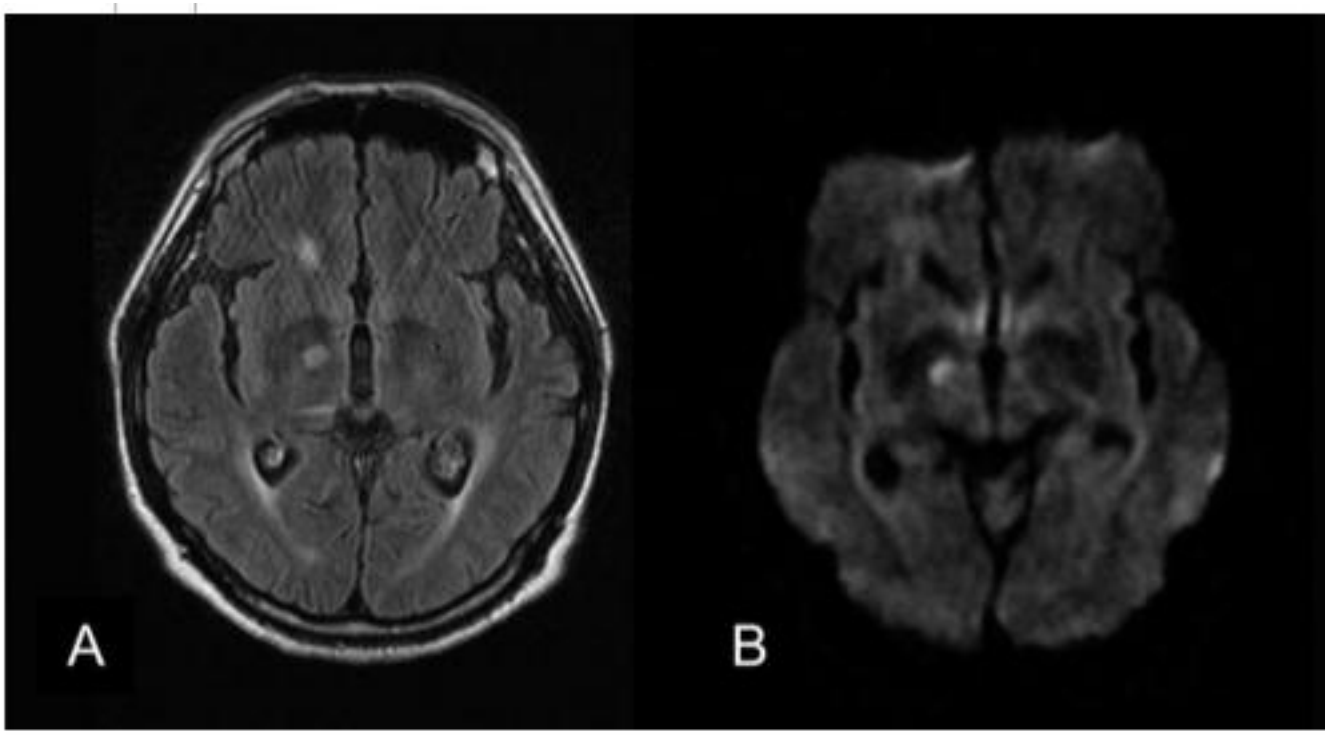


FIG. 1. (**A, B**) Initial fluid attenuated inversion recovery (FLAIR) (A) and diffusion-weighted (B) MR images showed a small acute infarction in the right STN area. (**C**) FLAIR MR image obtained after the second admission showed resolution of the previous STN infarction.

Park et al, 2009

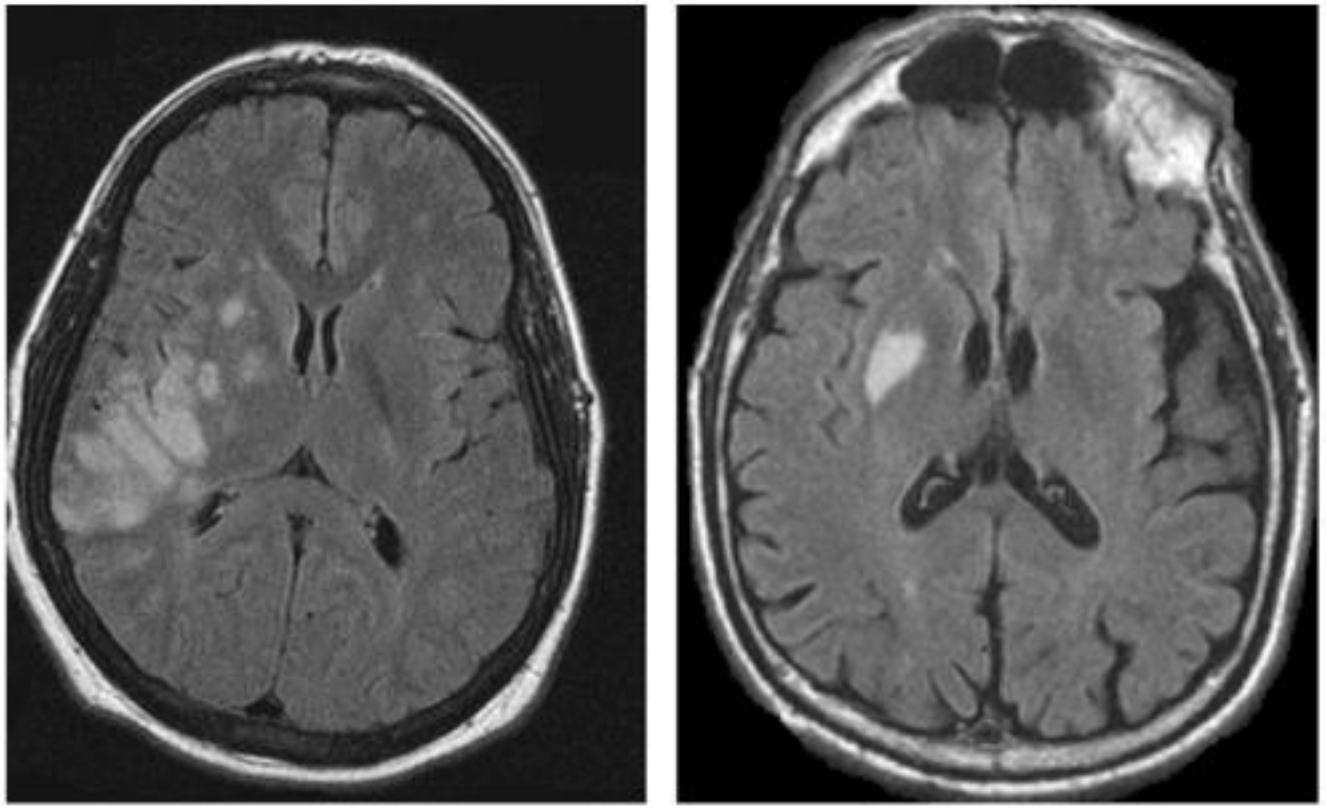


Figure 1. Hemiballism outside the STN. Left: an axial FLAIR T2 image in a 34-year-old man who presented with left-sided hemiballism, showing a large stroke in the right middle-cerebral-artery territory, which was later found to be secondary to endocarditis. There are several ischaemic areas in the basal ganglia and cortex. The region of the STN was spared. Right: an axial FLAIR T2 image in a 69-year-old man with a history of Parkinson's disease who presented with hemiballism of the left limb. An infarct is visible in the right striatum. The region of the STN was spared. This disorder was associated with improvement of the patient's parkinsonism on the left side. Similar improvement of parkinsonian features in association with hemiballism has been reported by many researchers, and these observations were part of the rationale for the development of STN deep brain stimulation in the treatment of Parkinson's disease.[10]

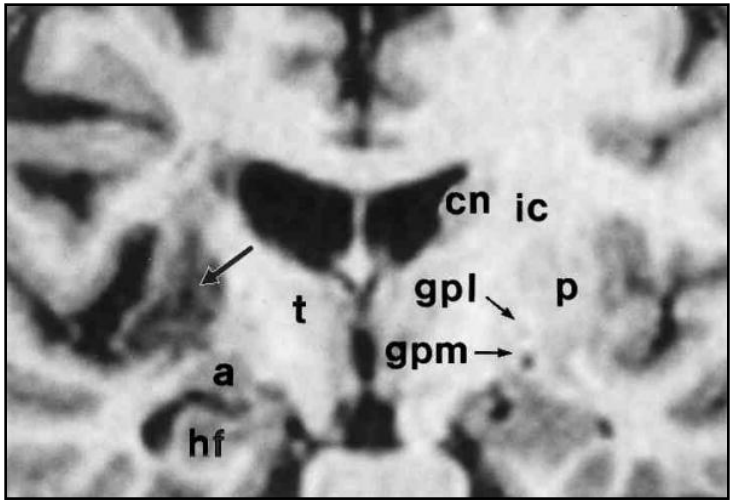
Lang et al, 2003

Dystonie post AVC : localisation des lésions

- **Complexe striato-pallidal** (perforantes profondes lenticulo-striées)
- Thalamus (territoire des perforantes profondes postérieures du thalamus postéro-latéral, artères thalamo-tubérositaires et choroïdiennes postérieures)
- Tronc cérébral (mésencéphale)

Lésions du complexe striato-pallidal

- Dystonie focale, segmentaire ou hémidystonie
- Prédominance distale et au membre supérieur
- Présente au repos, et augmentée par les mouvements
- Apparition différée, après la récupération motrice



- Partie sensorimotrice du complexe striato-pallidal
- Putamen impliqué dans la plupart des cas, +/- pallidum et rarement le caudé
- Atteinte des structures du circuit moteur sous cortico-frontal

- Traitement : anticholinergiques, toxine botulinique, BZD, baclofene, L-dopa

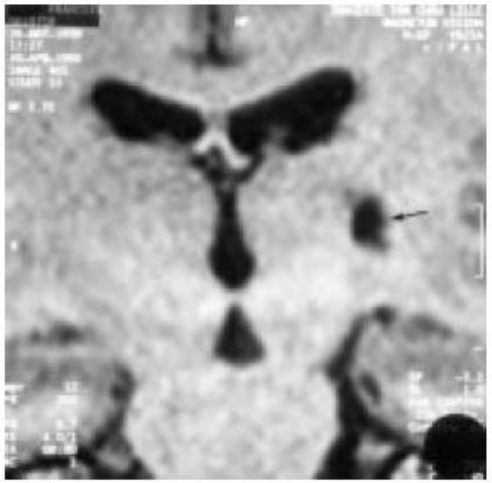


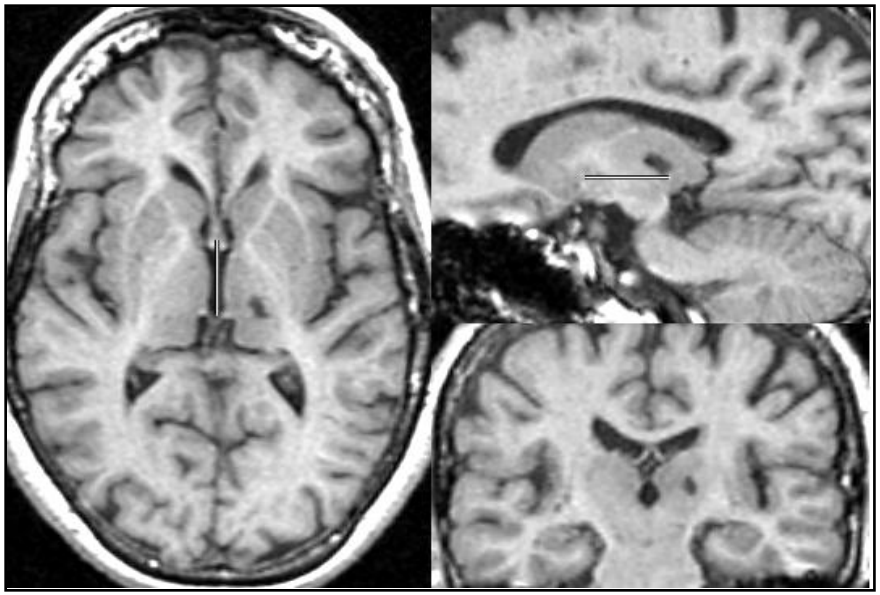
Figure 3 Three dimensional T1 weighted MRI sequence (MP-RAGE): coronal reconstruction according to the bicommissural line, showing the lenticular lesion.

Lehericy et al, 1996; Krystkowiak et al, 1998; Chuang et al, 2002

Lésions thalamiques

- Dystonie +/- **myoclonus**, souvent distal
- Focale ou segmentaire
- Mouvements choréoathétoïdes
- Tremblement lent, postural, note intentionnelle
- Autres signes associés : troubles sensitifs, proprioceptifs, signes cérébelleux

- Traitement : anti-épileptique



Lehericy et al, 1996; Lehericy et al, 2001, Hamasaki T et al, 2008

Lésions du tronc cérébral

- Lésions de plusieurs structures
  - Pédoncule cérébelleux sup
  - Lemniscus médian
  - Lésions dopaminergiques : aire noyau rouge, SN, voies cérébellothalamiques et nigro-striatales
- Dystonie +/- **tremblement**
  - Tremblement postural, augmenté à l'approche de la cible
  - Tremblement de repos, dopasensible
- Syndrome cérébelleux

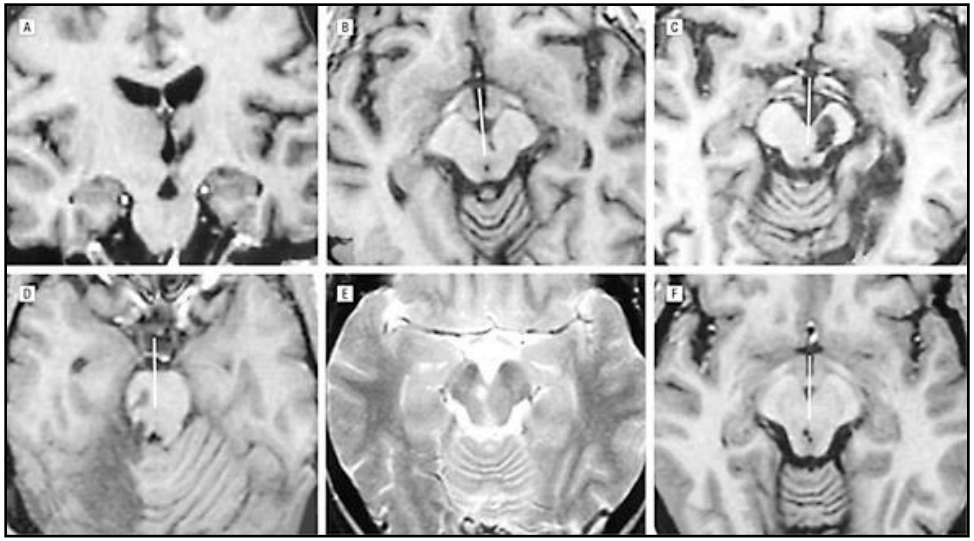
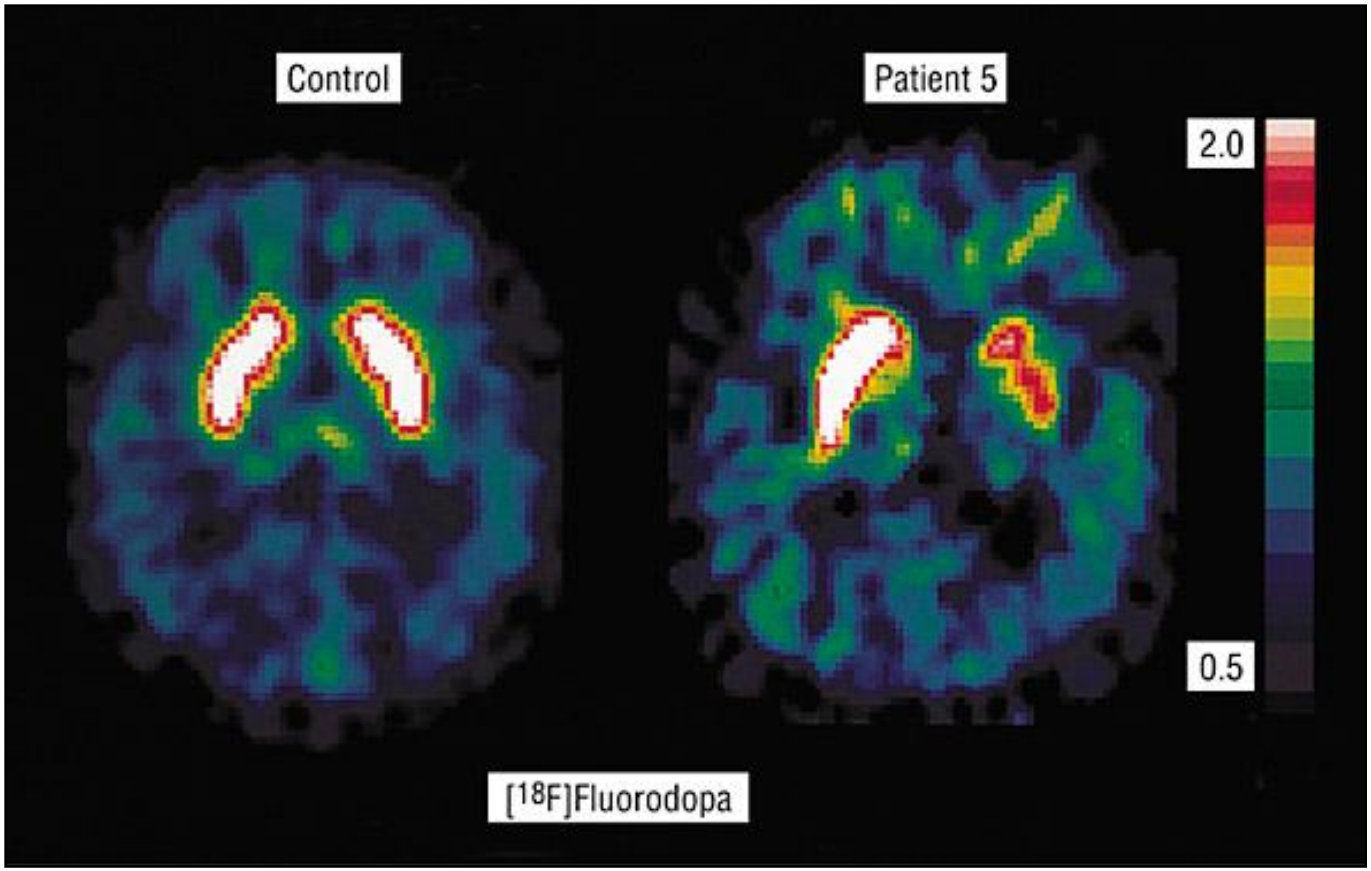


Figure 2. Magnetic resonance imaging (MRI) of a coronal T1-weighted scan of patient 1 showing a left paramedianthalamomesencephalic infarct (A); an axial T1-weighted scan of patient 4 showing a left medial mesencephalic infarct (B); an axial T1-weighted scan of patient 5 showing a large left mesencephalic infarct associated with a left temporo-occipital infarct (C); an axial T1-weighted scan of patient 7 showing infarct in the right superior cerebellum, the right superior cerebellar peduncle, and the upper part of the pons (D); and E, an axial T2-weighted scan of patient 3 (4 days after a stroke), showing a right paramedianmesencephalic infarct involving the dopaminergic area. F, A control scan of patient 3, taken 1.5 years after (E), is axial, T1-weighted, and shows a small residual tegmental lesion close to the aqueduct of Sylvius, sparing the nigrostriataldopaminergic system. At the time of the study, dystonia was no longer present. Note a decreased width of the right cerebral peduncle. The white line represents the anterior-posterior commissure.

Vidailhet et al, 1999



TEP scan au 18 F Dopa
Patient avec une lésion mésencéphalique gauche

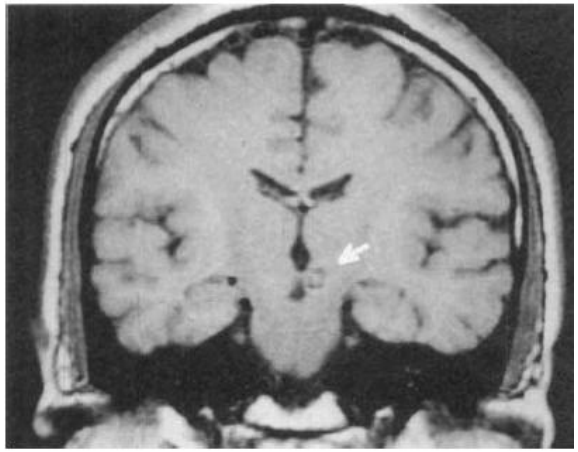
Vidailhet et al, 1999

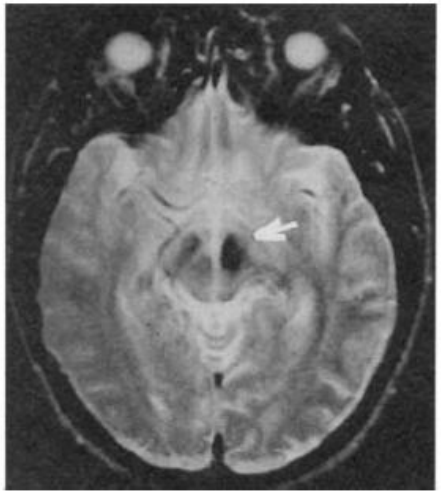
Tremblement de Holmes

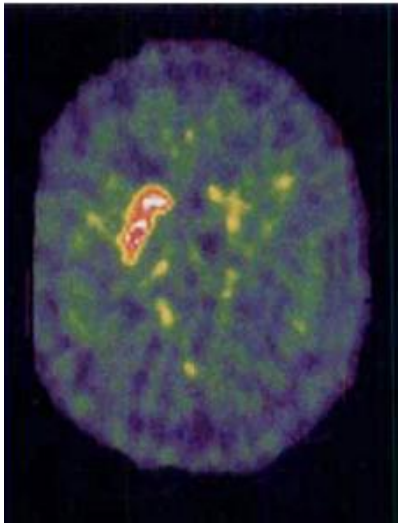
- Tremblement de repos et intentionnel
- Lent (1 à 4 Hz) et de grande amplitude
- Apparition différée de 4 semaines à 2 ans

- Lésions des circuits cérébello-thalamiques et dentato-rubro-thalamiques (noyau dentelé, noyau rouge, pédoncule cérébelleux supérieur, noyau VLp du thalamus)
- Atteinte de la SN et fibres nigrostriatales→ implication du système dopaminergique

- Traitement
  - Propranolol et primidone
  - Clonazepam
  - Sodium valproate
  - Levetiracetam
  - Anticholinergiques
  - levodopa







Remy et al, 1995; Vidailhet et al, 1998; Sung et al, 2009

Tremblement cérébelleux

- Tremblement intentionnel pur ou prédominant +/- postural
- Lent (fréquence < 5Hz)
- Lésions cérébelleuses
- Thalamus postérieur et des voies dentato-rubro-thalamique ou dentato-rubro-olivaire

- Traitement :
  - Propranolol et primidone
  - Clonazepam
  - Sodium valproate
  - Levetiracetam
  - Anticholinergiques
  - levodopa

Vidailhet et al, 1998

Autres tremblements

- Tremblement palatal
  - Lésions impliquant le triangle de Guillain-Mollaret (noyau dentelé, noyau rouge, et olive inférieure), pont, cervelet (PICA)
  - Lent 1 à 3 Hz
  - Peut être associé avec des myoclonies palatines
  - Hypertrophie de l'olive bulbaire inférieure sur l'IRM

- Tremblement à l'écriture (tremblement spécifique d'une tâche)
  - Rare
  - Ressemble au tremblement pur à l'écriture
  - Lésion cortex préfrontal

Yanagisawa et al, 1999; duffey et al, 2007; Nishie et al, 2002

Limb shaking

- Décrit en 1962 par Miller et Fisher
- Mouvements involontaires répétitifs, stéréotypés, brusques, durant moins de 5 minutes (« episodic paroxysmal dyskinesia »)
- Touchent la main et le bras +/- jambe
- Associé au déficit moteur
- Aggravé avec les facteurs d'hypoperfusion cérébrale

- Sténose serrée ou occlusion de la carotide interne (10% des patients)
- Hypoperfusion cérébrale focale transitoire (zone frontière entre ACA et ACM)

- Existe aussi sur le système vertébro-basilaire, ou dans le MoyaMoya et dans les TVC

Gálvez-Jiménez et al, 2002; Persoon et al, 2010;SalahUddin et al, 2004; Ali et al, 2006

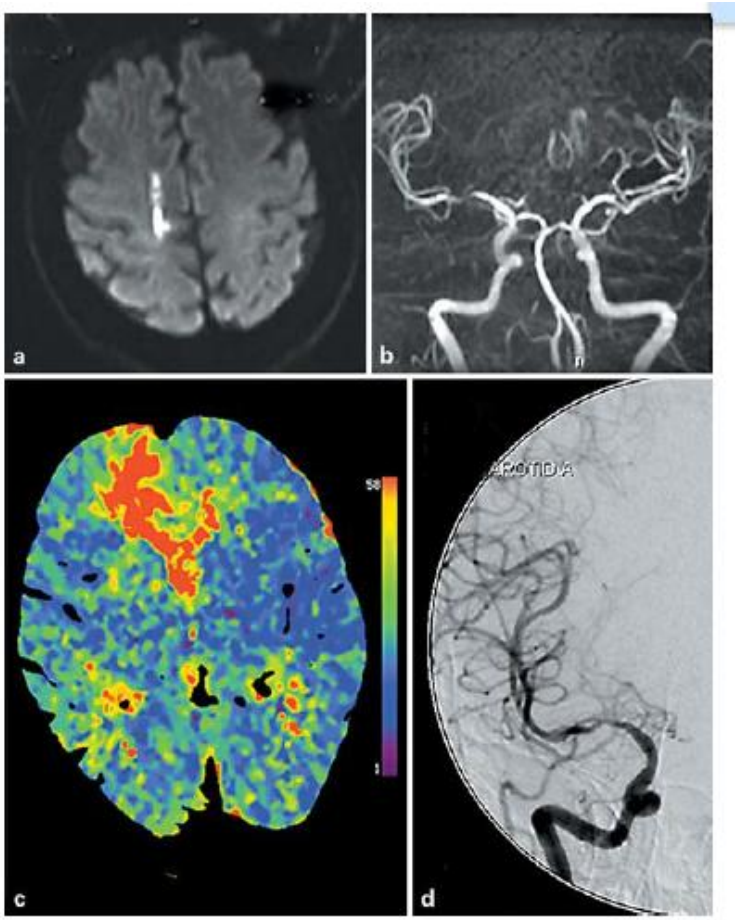


Fig. 1. a Diffusion-weighted MRI revealed high signal intensity lesions in the right watershed territory between the ACA and MCA. **b** Magnetic resonance angiogram showed severe focal stenosis of the right MCA and absence of bilateral ACA. **c** Perfusion CT showed delayed mean transit time in the right anterior frontal lobe, the right anterior and posterior border zone, and the right basal ganglia. **d** Digital substraction angiography showed severe focal stenosis of the M1 segment of the right MCA and absence of the ACA. The right border zone (between the ACA and MCA) shifted internally, suggestive of the right ACA territory being compensated partially by the leptomeningeal collateral vessels from the right MCA.

Jiang et al, 2006

Syndrome parkinsonien vasculaire

- Clinique
  - Défaut d'initiation de la marche, instabilité posturale précoce
  - Syndrome parkinsonien prédominant aux membres inférieurs
  - Absence de tremblement de repos (rare)
  - Faible réponse au traitement dopaminergique
  - Signes associés : signes pyramidaux, incontinence urinaire, syndrome dysexécutif, démence vasculaire

- Hémisyndrome parkinsonien (lésion vasculaire des GB controlatérale)

- 2.5 à 8.8% des syndromes parkinsoniens
- Age moyen de début : 70 ans

Critères diagnostiques

Possible Criteria for the Clinical Diagnosis of Vascular Parkinsonism

a. *Parkinsonism: bradykinesia* (slowness of initiation with progressive reduction in speed and amplitude of repetitive actions in either upper limb or lower limb, including the presence of reduced step length) and at least one of the following: rest tremor, muscular rigidity, or postural instability not caused by primary visual, vestibular, cerebellar or proprioceptive dysfunction

b. *Cerebrovascular disease,* defined by evidence of relevant cerebrovascular disease by brain imaging (CT or MRI) *or* the presence of focal signs or symptoms that are consistent with stroke

c. *A relationship between the above two disorders. In practice* this is met with infarcts in or near areas that can increase the basal ganglia motor output (GPe or substantia nigra pars compacta) or decrease the thalamocortical drive directly (VL of the thalamus, large frontal lobe infarct). The parkinsonism at onset consists of a contralateral bradykinetic rigid syndrome or shuffling gait, within 1 year after a stroke (VPa). (2) An insidious onset of parkinsonism with extensive subcortical white matter lesions, bilateral symptoms at onset, and the presence of early shuffling gait or early cognitive dysfunction (VPi).

Exclusion criteria for VP: History of repeated head injury, definite encephalitis, neuroleptic treatment at onset of symptoms, presence of cerebral tumor or communicating hydrocephalus on CT or MRI scan, or other alternative explanation for parkinsonism.

Zijlmans et al, 2004

- Imagerie
  - IRM encéphalique
    - Lésions diffuses de la SB
    - Lacunes multiples des NGC
    - Lésion vasculaire unique (hémisyndrome parkinsonien)
  - DAT-scan : réduction de la captation striatale

- Physiopathologie
  - Réduction des voies thalamo-corticales et/ou des voies nigro-striées
  - Sous activation des aires de planification et d'éxécution motrice
  - Lésions focales des NGC causent un PV dans 8% des cas

- Attention à la co-existence MPI et lésions vasculaires…

- Traitement :
  - Lévodopa,
  - Kiné, ergo, orthophonie..
  - Prise en charge des FRCV

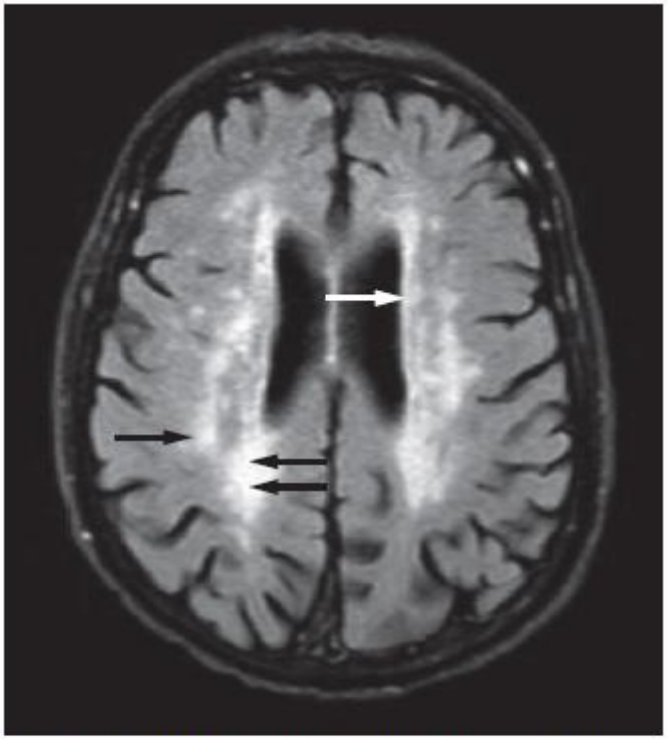


Figure 1 | Brain MRI in a patient with parkinsonism. The T2-weighted image shows a section of the brain of a 63-year-old male with lower body parkinsonism. Disruption of the periventricular and deep white matter (white and black arrows, respectively) is seen. Permission obtained from Y. Balash, Tel Aviv Sourasky Medical Centre, Tel Aviv, Israel.

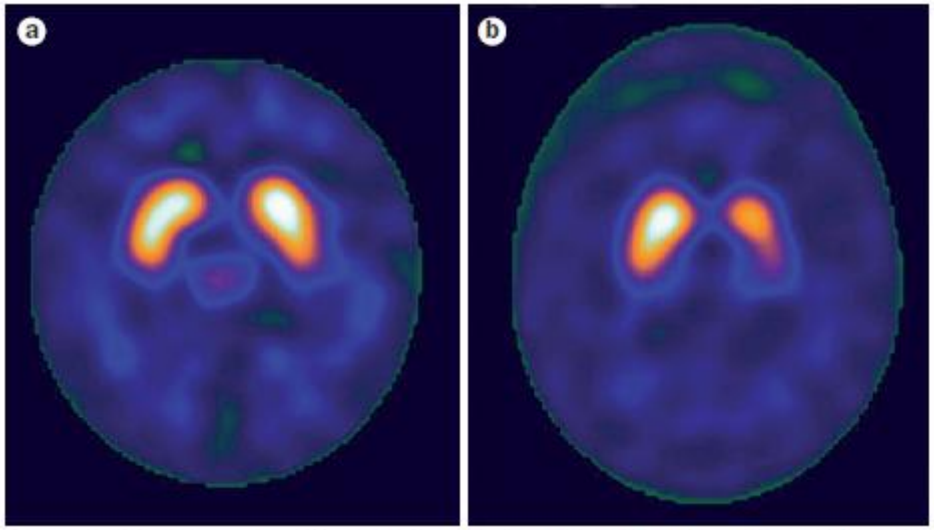


Figure 2 | SPECT imaging of the brain in patients with parkinsonism. The radioligand ^{123}I-FP-CIT was used to quantify dopamine transporter activity (yellow). **a** | Brain SPECT scan from an 83-year-old woman with lower body parkinsonism, who had good uptake of ^{123}I-FP-CIT in the striatum and a poor response to levodopa therapy. **b** | Brain SPECT scan from a 79-year-old woman with right hemiparkinsonism, who demonstrated markedly diminished uptake of ^{123}I-FP-CIT in the left striatum and a good response to levodopa therapy. Abbreviation: SPECT, single-photon emission CT. Permission obtained from M. Lorberboym, Department of Nuclear Medicine, Edith Wolfson Medical Centre, Sackler Faculty of Medicine, Tel Aviv University, Israel.

Korczyn 2015

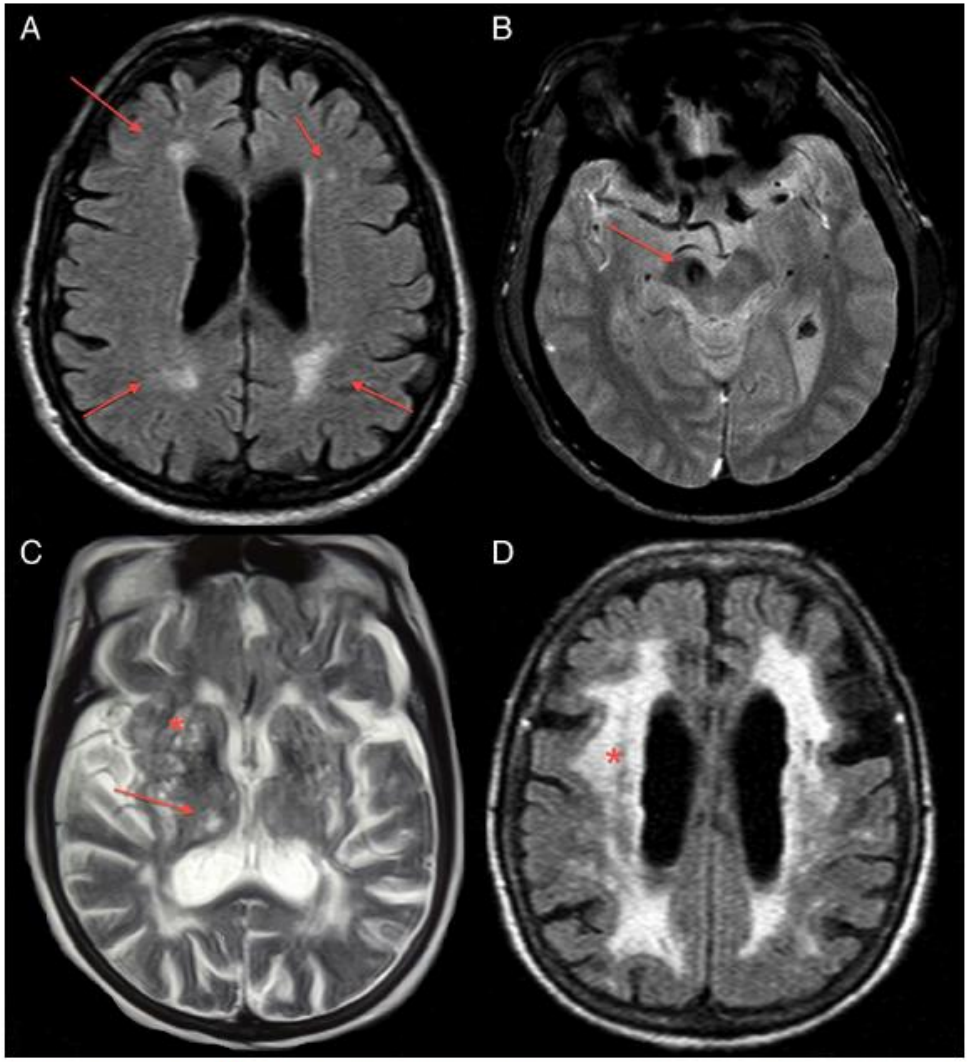


Figure 1 (A) T1-weighted-sequenced brain MRI showing mild cortical atrophy and non-specific periventricular white matter changes (red arrows) in a 65-year-old patient with hypertension and Parkinson's disease; (B) Spin-echo-sequenced brain MRI showing a single right midbrain lacunar infarct (red arrow) in a 62-year-old patient with vascular parkinsonism; (C) A T2-weighted-sequenced brain MRI showing multilacunar infarcts in the basal ganglia (red asterix), thalami (red arrow) and internal capsules in a 76-year-old patient with vascular parkinsonism; (D) fluid-attenuated inversion recovery-sequenced brain MRI showing extensive white matter lesion (red asterix) in a 72-year-old patient with vascular parkinsonism.

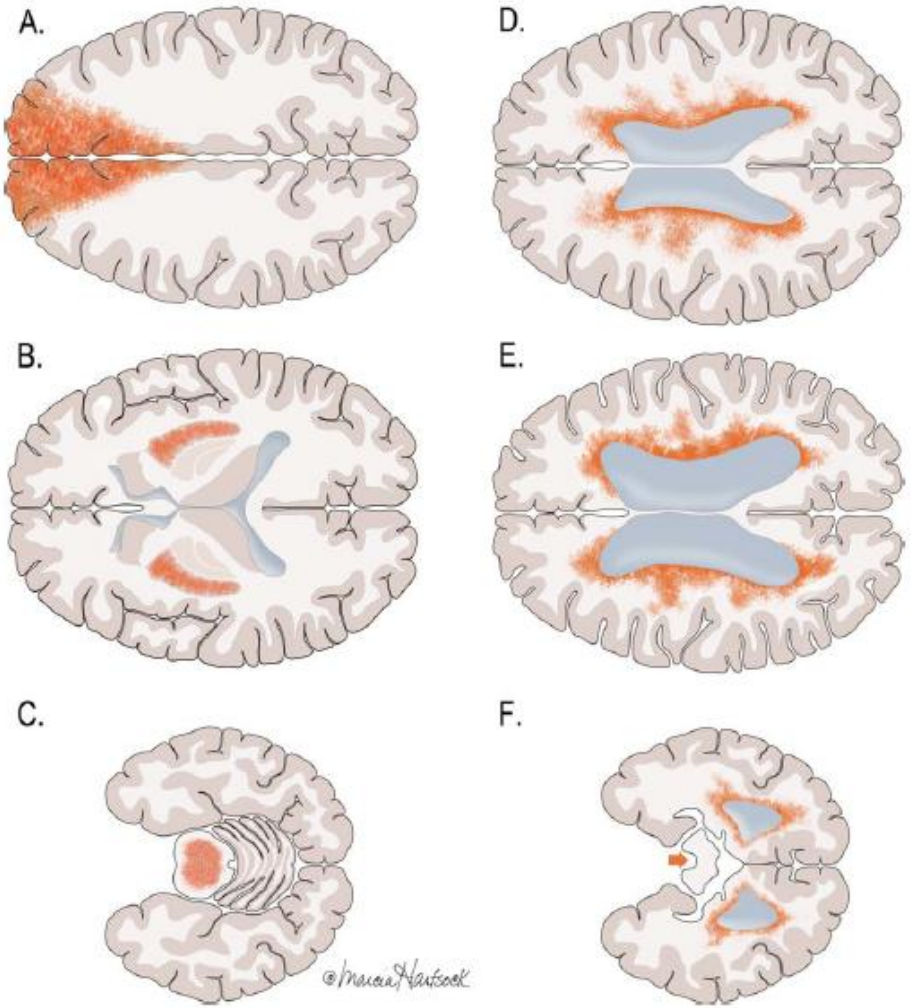


FIG. 3. Anatomical patterns in VaP. Vascular pseudoparkinsonism arises in the context of bilateral mesial frontal strokes resulting from anterior cerebral artery territory infarcts (A, akinetic mutism); bilateral striatal lacunar infarctions (B, apathetic depression); or small-vessel ischemic disease affecting the pons (C, pyramidal weakness and slowness). Pseudovascular pseudoparkinsonism occurs in the context of periventricular and deep WM signal abnormalities in isolation (D, higher-level gait disorder) or in association with ventriculomegaly (E, higher-level gait disorder in NPH). Pseudovascular parkinsonism can be documented in patients with PD (with a pattern similar to D) or PSP (F, arrow point to atrophic midbrain). This diagrammatic representation does not include mixed-pathology parkinsonism (e.g., PD with true microangiopathic brain disease), presumably accounting for a minority of patients with VaP. [Color figure can be viewed in the online issue, which is available at wileyonlinelibrary.com.]

Vizcarra 2015

Autres mouvements anormaux

- Tics
  - Tics vocaux simple après AVC du noyau caudé (Gomis et al, 2008)
  - Tics moteurs et vocaux complexes après hémorragie sur MAV du lobe frontal gauche (Yochelson et David, 2000)

- SJSR (pont, striatum) :15% des AVC/AIT vs 3% contrôles
  (Sechi et al, 2008; Schlesinger et al, 2015; Ruppert et al, 2014 et 2015)

- Hyperekplexie(pont) (Kimber et al, 1997)

- Akathisie(pont, thalamus postérieur controlatéral)

(Ghika-Schmid et al, 1997; Han et al, 2014)

- Stéréotypies(pariétal, lenticulostrié, thalamus, tronc cérébral, territoire de l'ACM) (Ghika-Schmid et al, 1997; Maraganore et al, 1991)

Conclusion

- Les mouvements anormaux sont rares en phase aigue d'AVC sauf hémiballisme et « limb shaking »
- La plupart du temps, apparition retardée par rapport à l'AVC
- Tous les mouvements anormaux peuvent être observés
- Corrélations clinico-radiologiques
- Physiopathologie : dysfonctionnement des voies reliant le cortex aux GB

Movement Disorder	Acute or Delayed Onset	Localization	Therapy
Ballism/chorea	Acute	STn, caudate, putamen, thalamus	Dopamine receptor antagonists, tetrabenazine, clonazepam, diazepam, topiramate, botulinum toxin, functional neurosurgery
Chorea	Acute	Putamen, STn, caudate, cortex	Dopamine receptor antagonists, clonazepam, diazepam, botulinum toxin, functional neurosurgery
Dystonia	Delayed	Putamen, caudate, pallidum, thalamus, midbrain	Anticholinergics, benzodiazepines, baclofen, tetrabenazine, botulinum toxin, functional neurosurgery
Myoclonus	Acute	Hemispheres, basal ganglia, midbrain, pons, spinal cord, post-anoxic	Clonazepam, valproate, piracetam, levitiracetam
Asterixis	Acute	Thalamus, frontal lobes, basal ganglia, midbrain, cerebellum	Clonazepam, valproate, piracetam, levitiracetam
Holmes' tremor	Acute	Brainstem, cerebellum, thalamus	Levodopa, clonazepam, propranolol, valproate, levitiracetam, functional neurosurgery
Palatal tremor	Delayed	Triangle of Guillain–Mollaret	Botulinum toxin, sumatriptan, oxitriptan, carbamazepine, clonazepam
Tics	Acute	Basal ganglia, frontal lobe	Alpha-agonists, dopamine receptor antagonists
Vascular Parkinsonism	Delayed	subcortex	Levodopa, dopamine agonists

Bansil et al, 2012

Merci de votre attention