# Zobrazovací metody v diferenciální diagnostice atypických parkinsonských syndromů

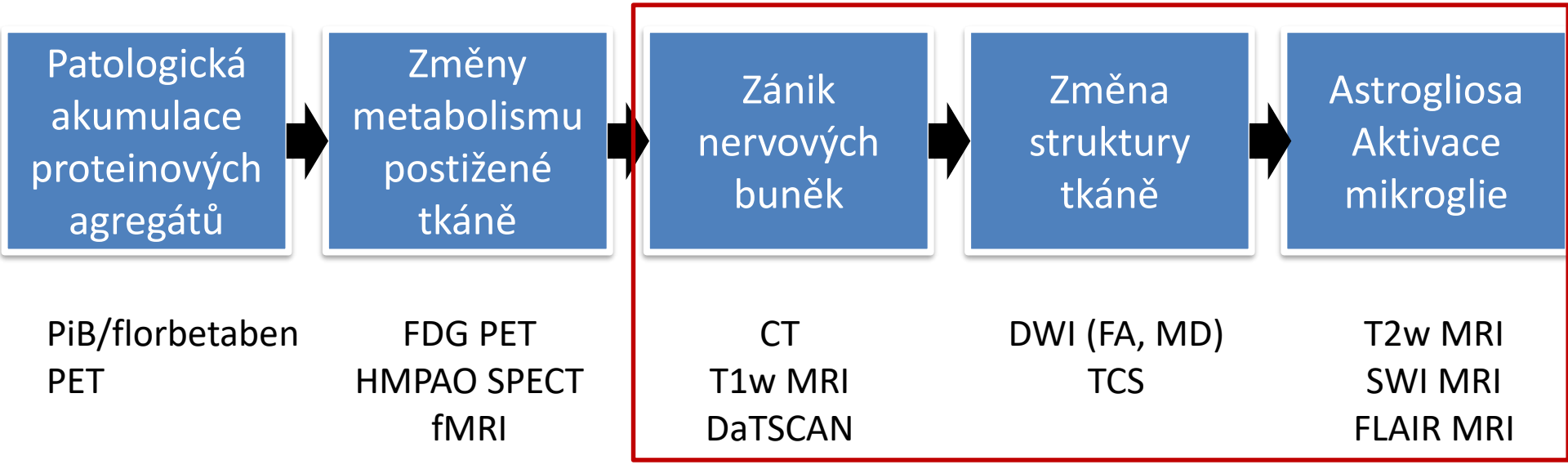
Petr Dušek

Neurologická klinika a Centrum klinických neurověd
Universita Karlova v Praze,
1. lékařská fakulta a Všeobecná fakultní nemocnice v Praze

# Zobrazení u neurodegenerace

Změny nervové tkáně :

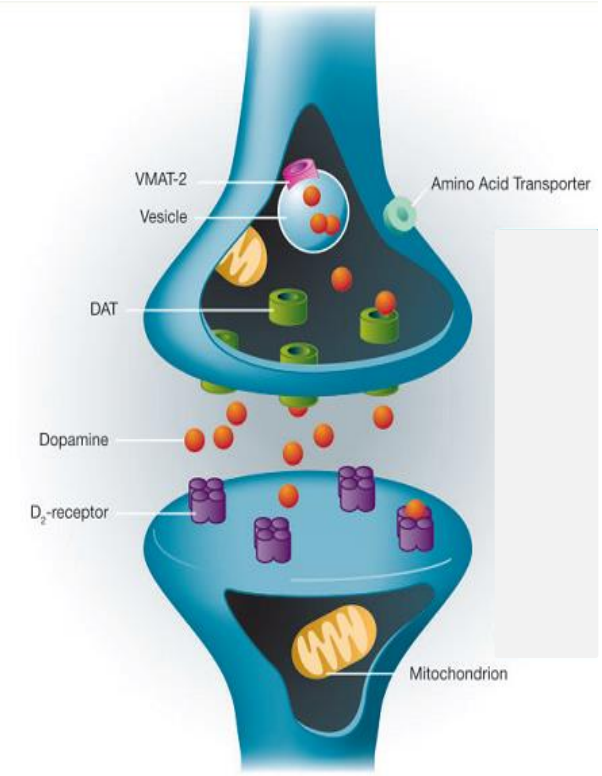
- **makroskopické** (atrofie, gliosa, akumulace ferritinu…)
- **mikroskopické** (Lewyho tělíska, tau protein, amyloidní plaky, dezorganizace axonů…)
- **funkční** (hypometabolismus, hustota receptorů, koncentrace metabolitů…)



# SPECT/PET nigrostriatální dráhy

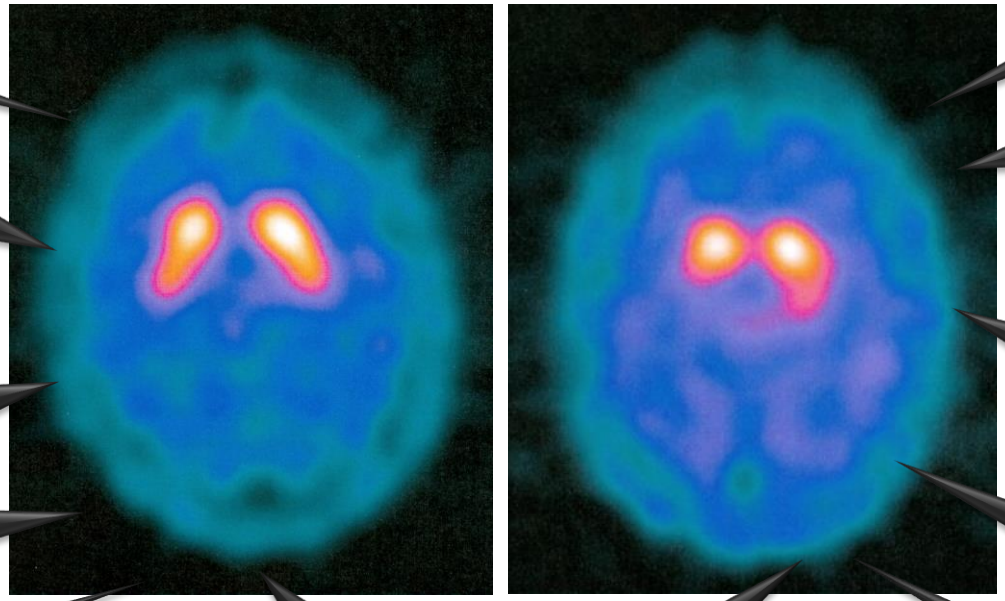
– Presynapticky
  - $^{123}$I-fluopane-CIT SPECT (DAT) = **DaTscan®**
  - $^{18}$F-fluopane-CIT PET (DAT)
  - $^{18}$F-DOPA PET (aktivita AADC)
  - $^{11}$C-dihydrotetrabenazine (VMAT2)

– Postsynapticky
  - [$^{123}$I] – IBMZ SPECT (D2R)
  - $^{11}$C-raclopride PET (D2R)




*Cummings et al., 2011*

# DaTscan®




Esenciální třes

Polékový PS

Funkční PS

DYT-5

Dystonický třes

Vaskulární parkinsonismus

Parkinsonova nemoc

MSA

Lewy-body demence

PSP

strukturální léze BG

# MRI v diferenciální diagnostice parkinsonismu

## Atypické parkinsonské syndromy

- **Multisystémová atrofie**
  - typ P (parkinsonism)
  - typ C (cerebellar)
- **Progresivní supranukleární paréza**
- **Kortikobazální syndrom**

## Sekundární parkinsonismus

- **Normotenzní hydrocefalus**
- **Vaskulární encefalopatie**
- **Léze/ mineralizace bazálních ganglií**
- **Wilsonova nemoc**

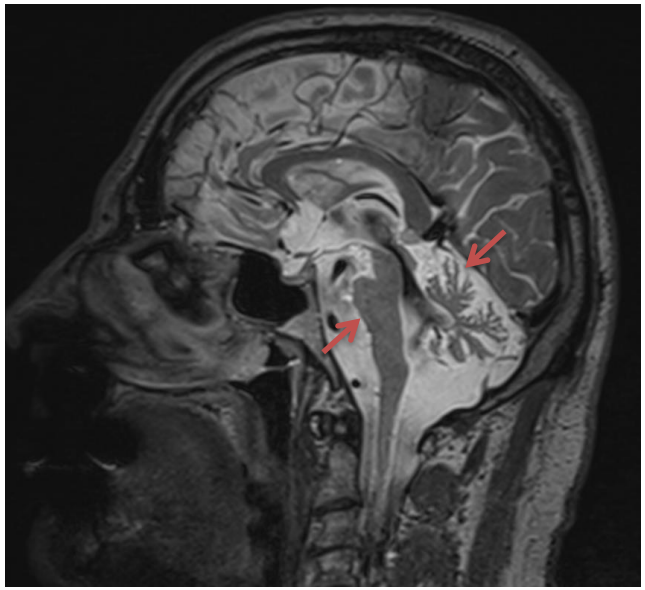
Vzorec atrofie
- Transversální T1
- Sagitální T1

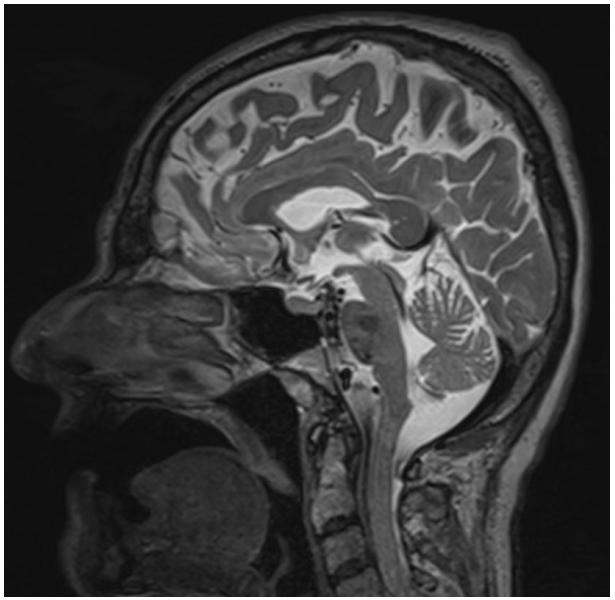
Likvorové prostory
- Komory
- Vertex
- Bazální cisterny

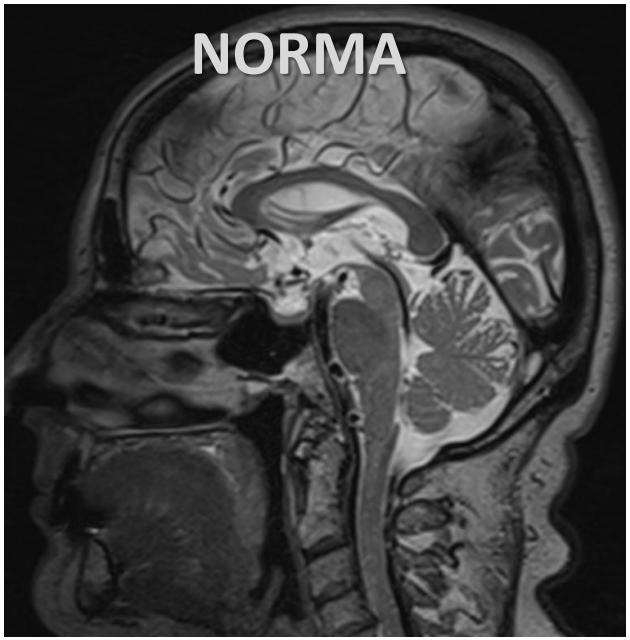
Změny signálu
- Putamen (T2)
- GP (T2)
- Kmen (T2)
- Mozeček (T2)

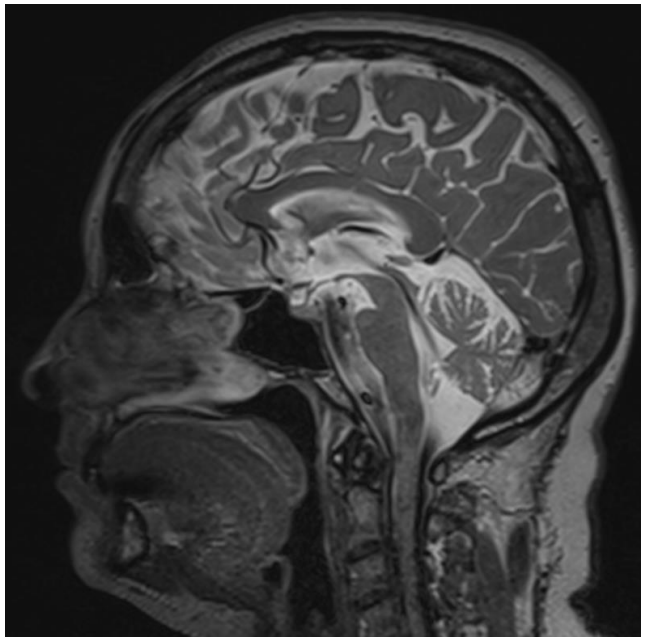
**Mid-sagitální řez**
**u MSA (typ C)**

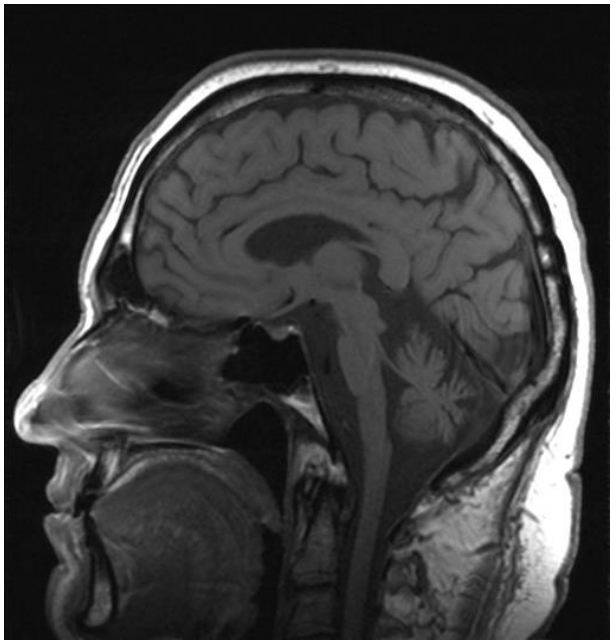
→ atrofie pontu
→ atrofie cerebella





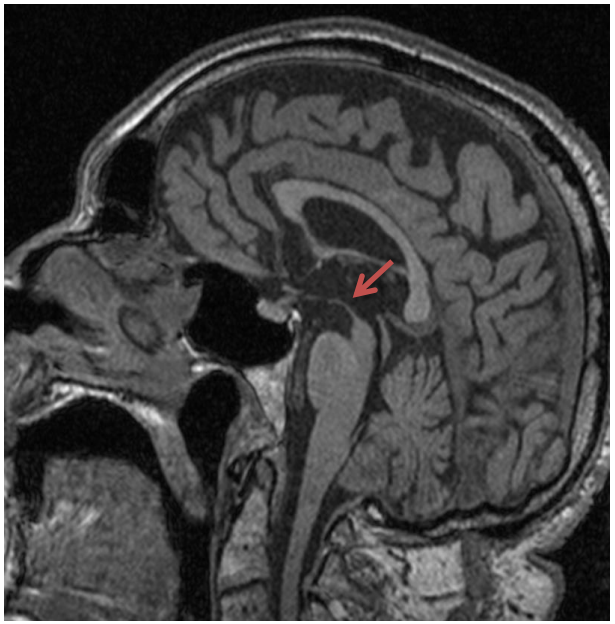


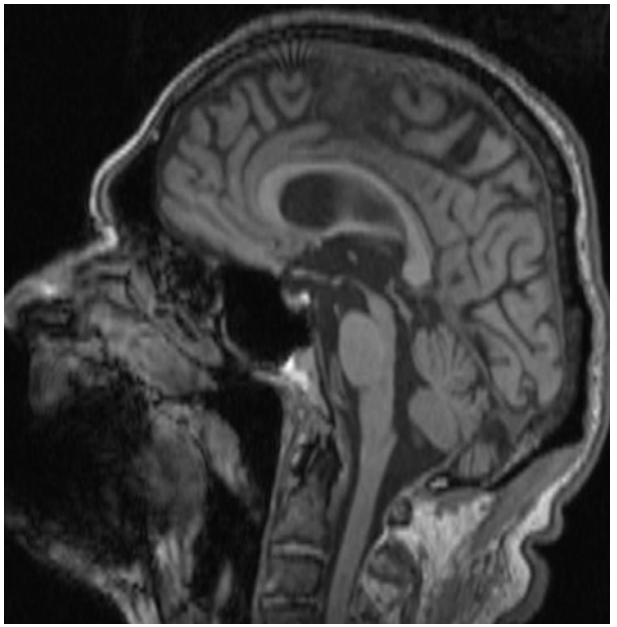



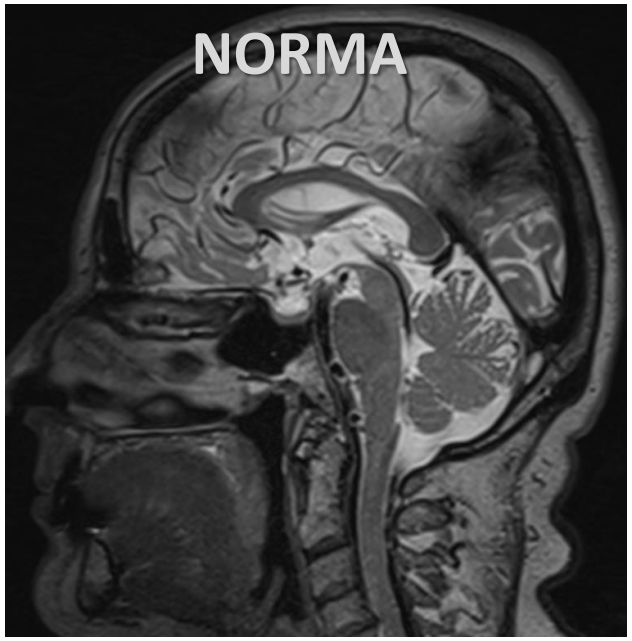


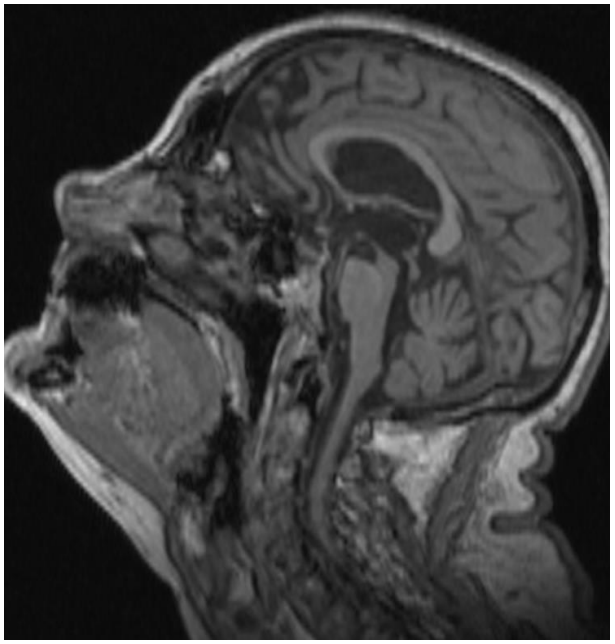
## Mid-sagitální řez u **PSP**

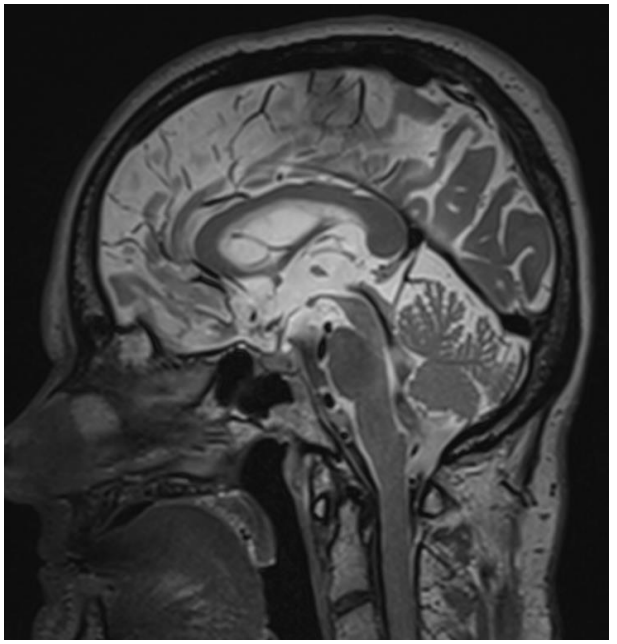
→ atrofie mesencefala
→ extenze šíje





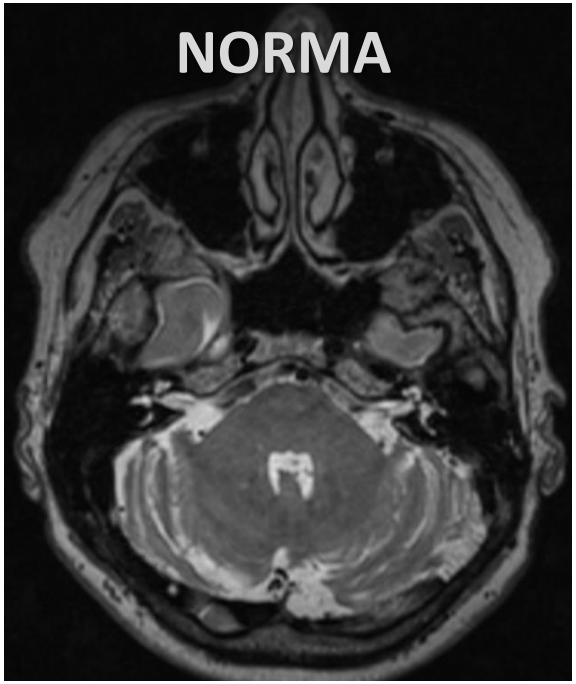


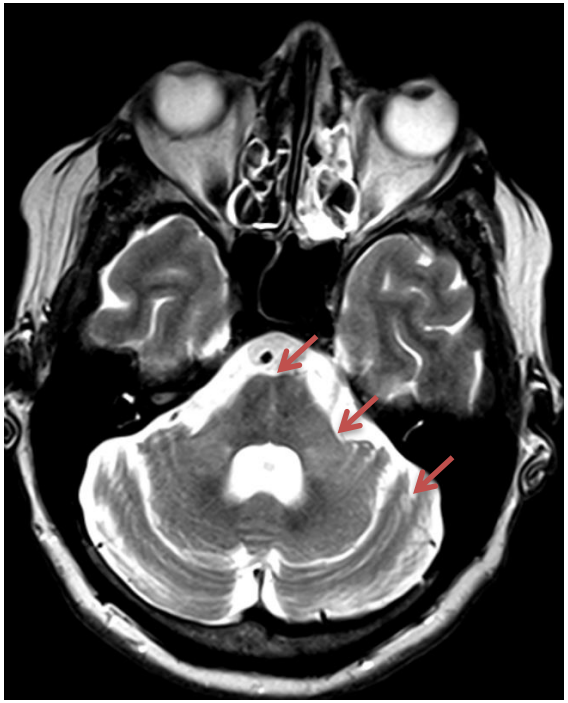



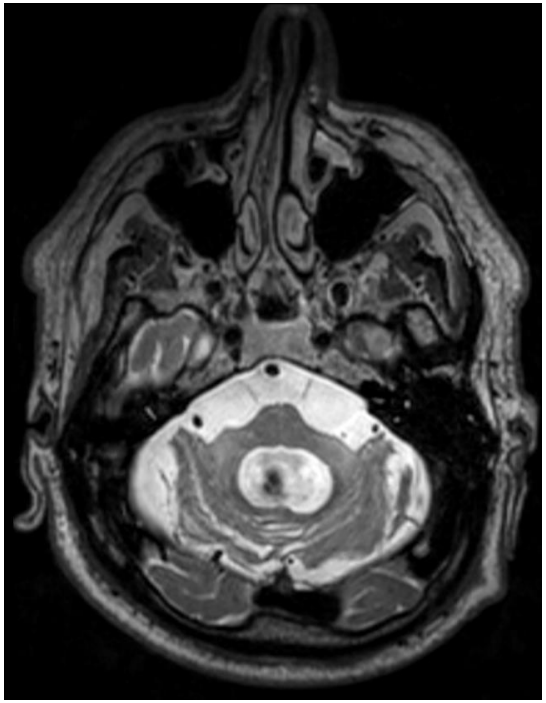


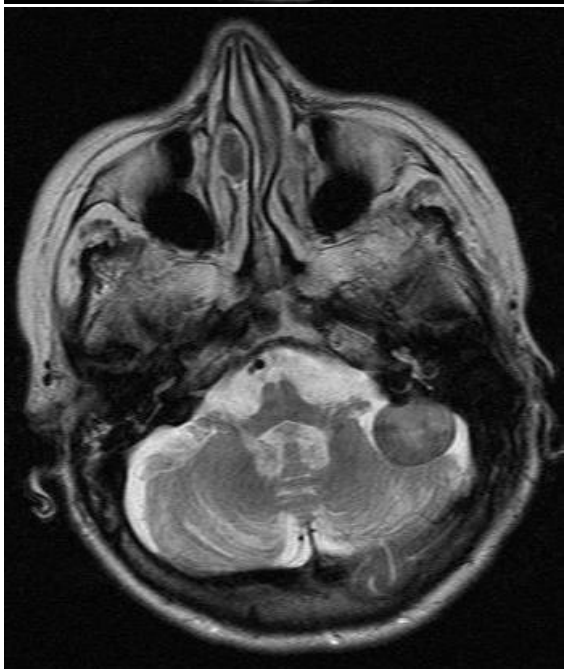
# T2 axiální řez v úrovni pontu u MSA

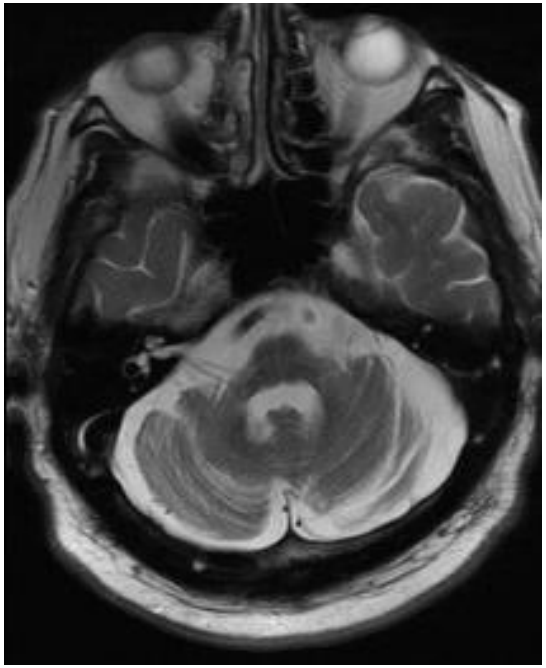
→ atrofie cerebella
→ atrofie (+ ↑T2 signál) středních mozečkových stonků
→ příznak kříže





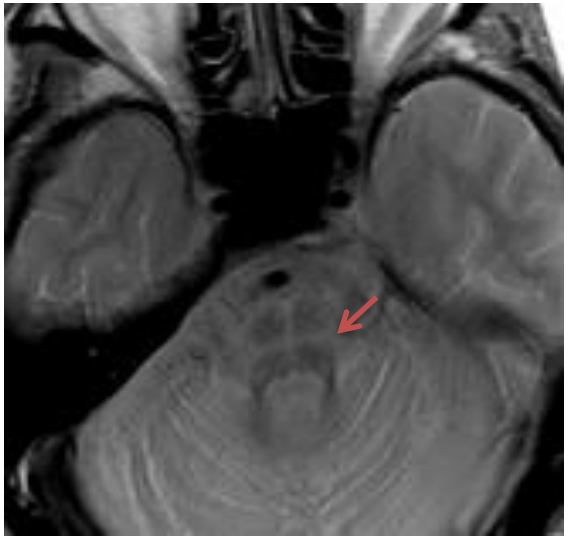


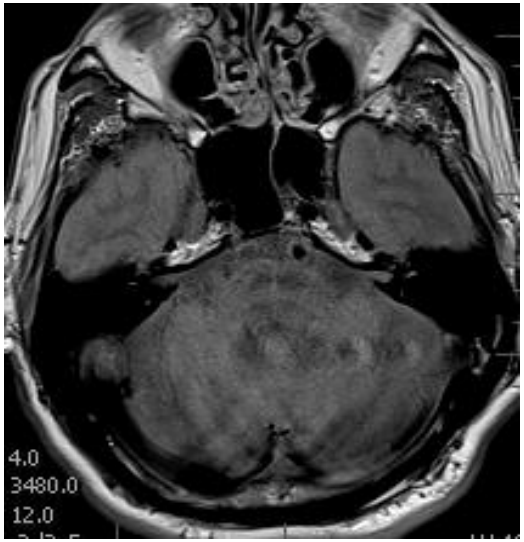


**axiální řez v úrovni pontu u <u>MSA</u>**

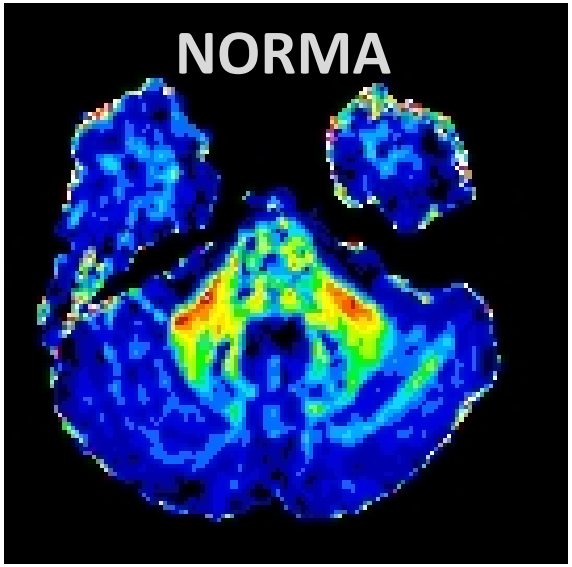
→ atrofie cerebella
→ atrofie (+ ↑T2 signál) středních mozečkových stonků
→ příznak kříže

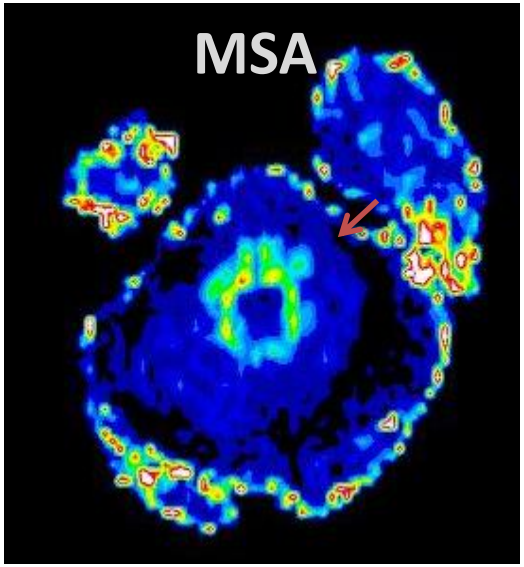
Proton densitně vážený MRI obraz





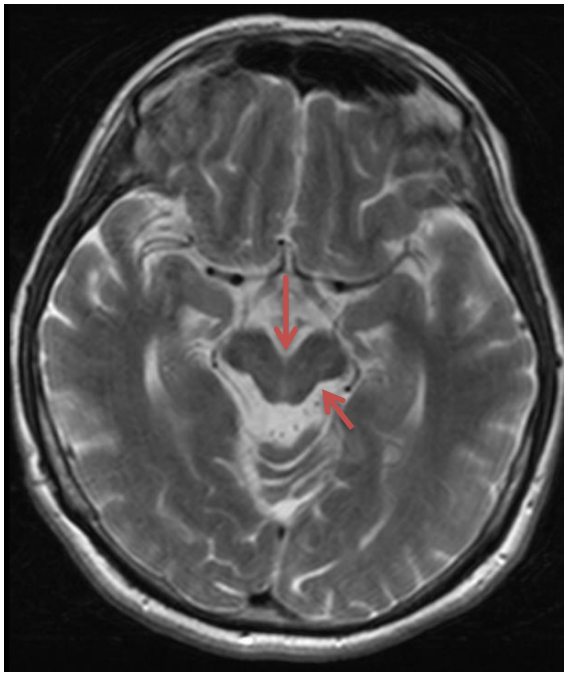

DWI – frakční anisotropie

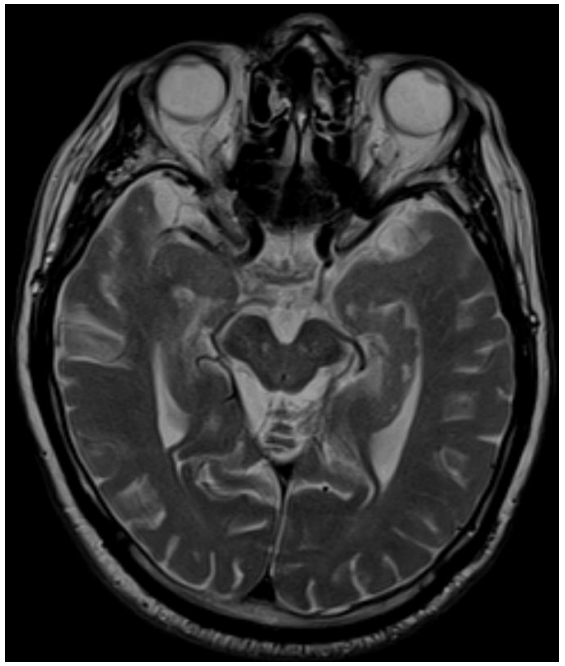


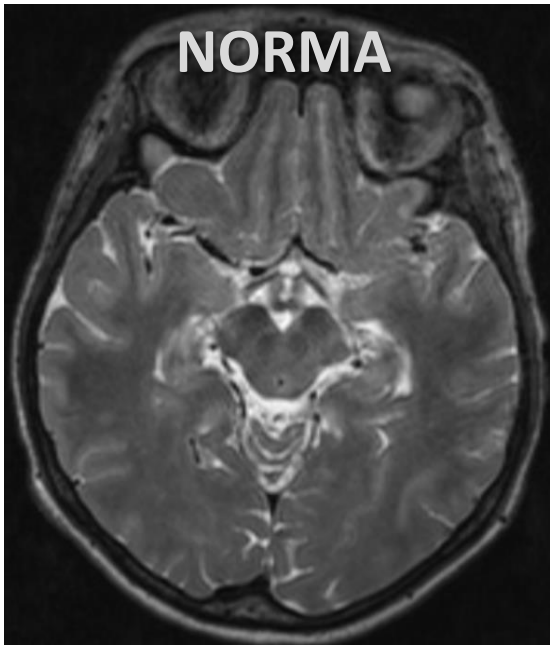



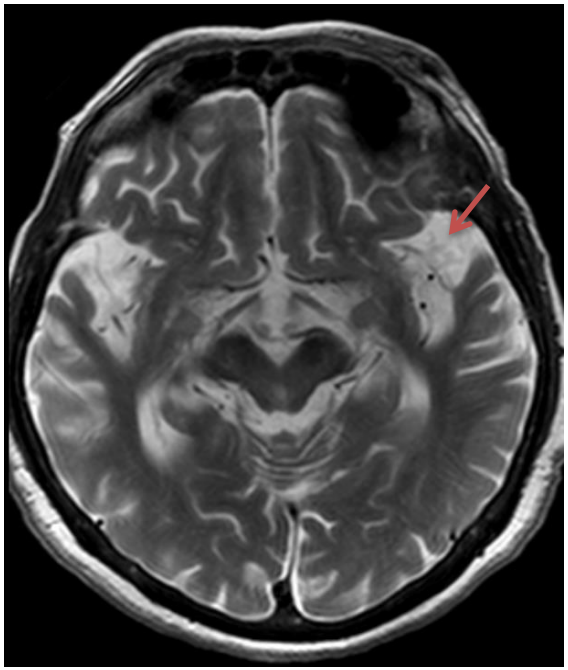
**axiální řez v úrovni mesencefala u PSP**

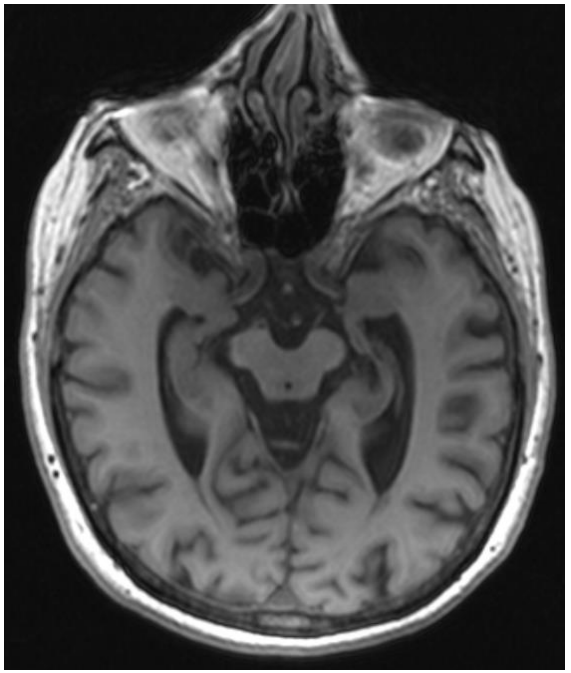
→ atrofie mesencefala
→ atrofie globální





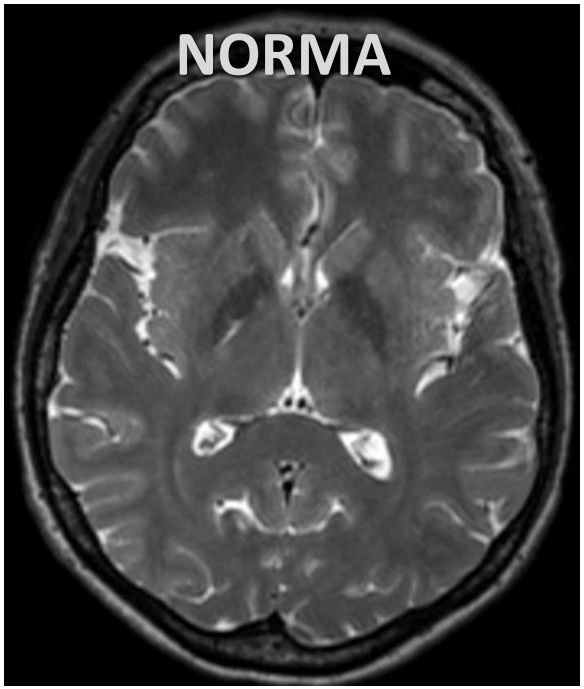


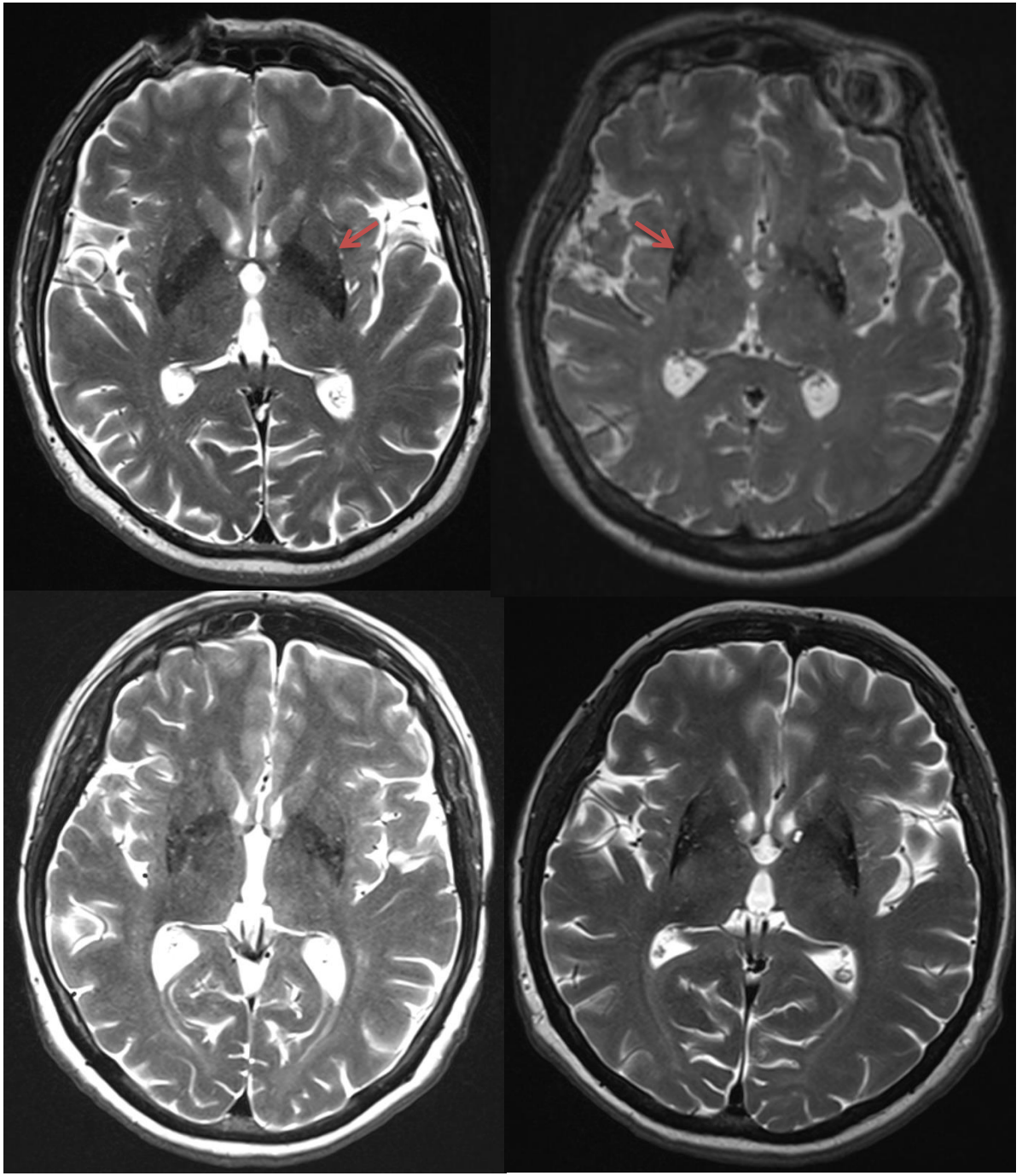





**axiální řez v úrovni BG u MSA (typ P)**

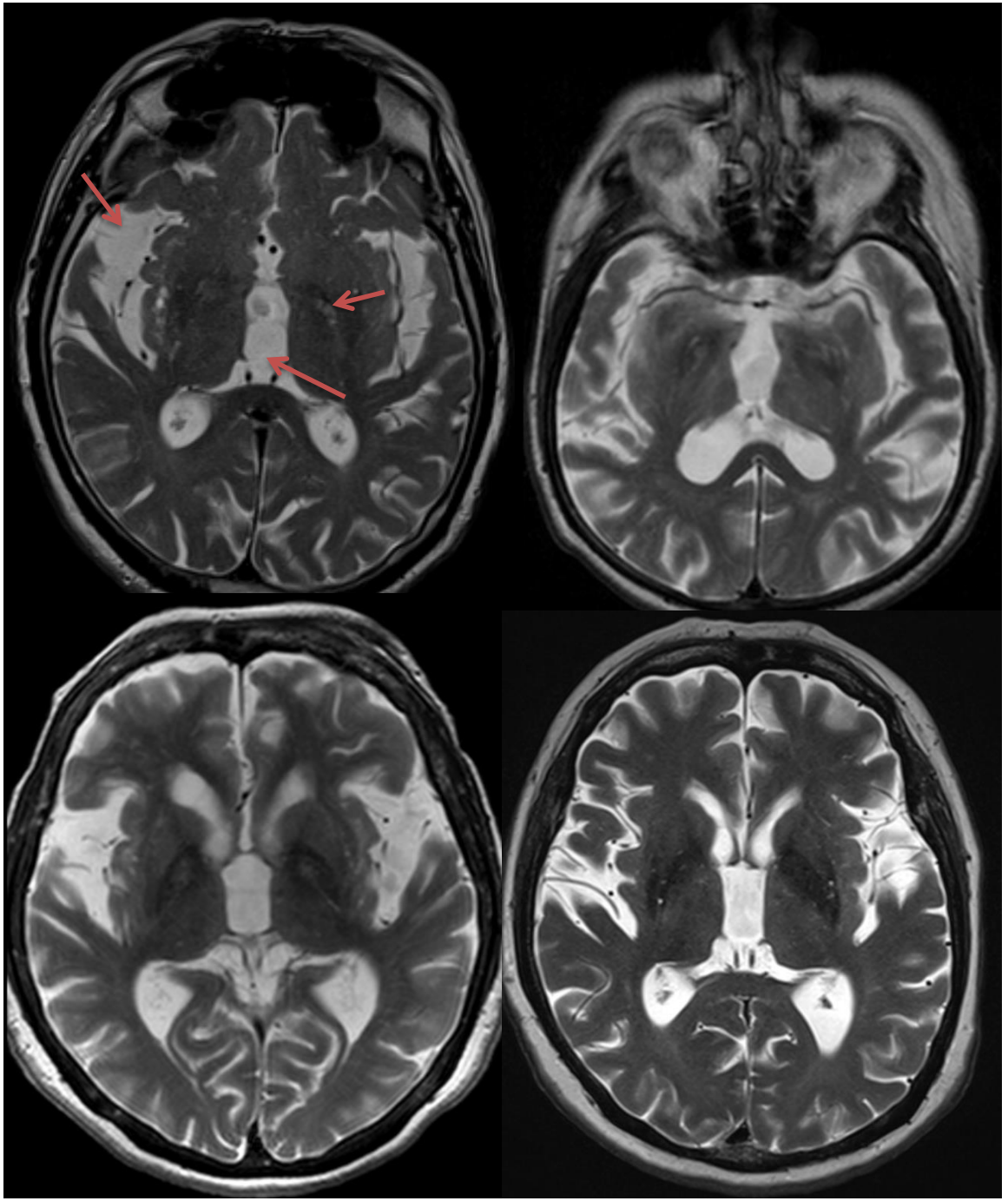
→ atrofie Put (asymetrická, koreluje s klinikou)
→ depozita ferritinu latero-dorsálně v Put
→ T2 hypersignální lem Put

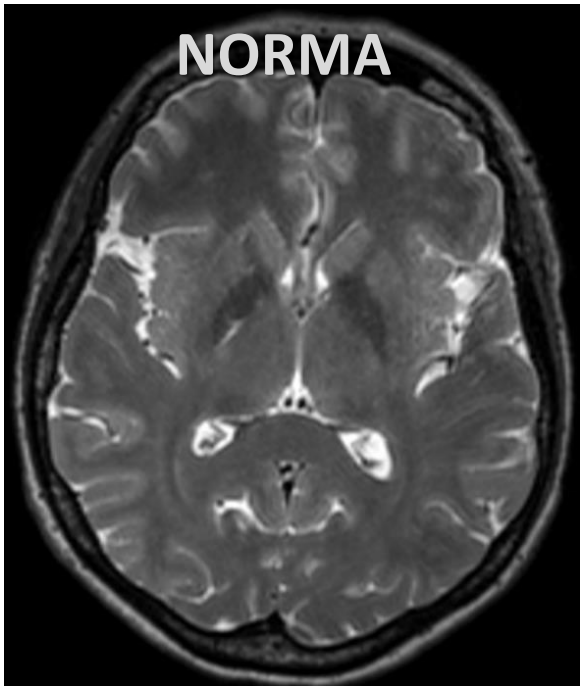


**axiální řez v úrovni BG u PSP**

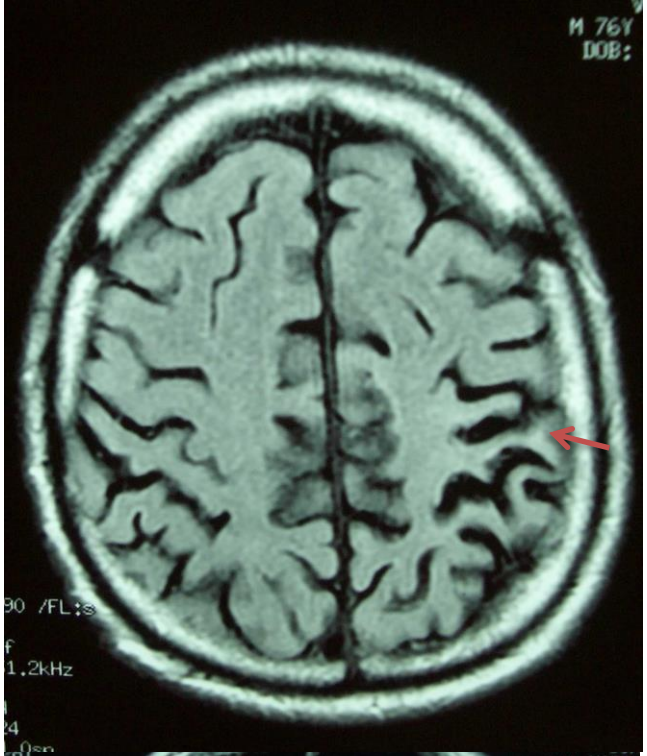
→ atrofie globální (F > O)
→ dilatace III. komory
→ akumulace ferritinu symetricky v GP

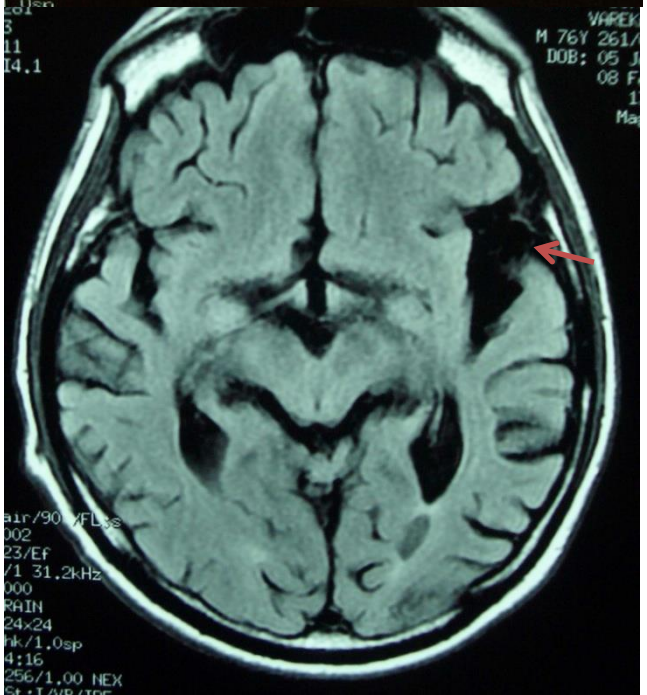




**axiální řezy v úrovni kortexu a mesencefala u <u>CBS</u>**

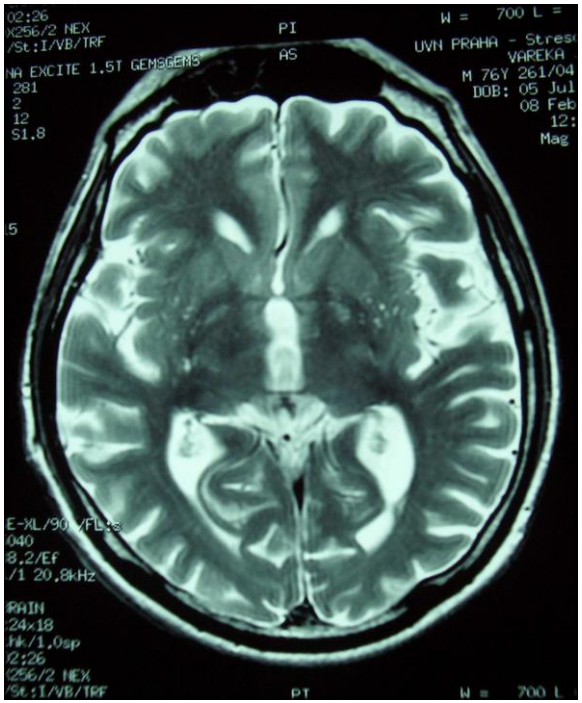
→ atrofie globální (asymetrická, koreluje s klinikou)
→ kortikální hypometabolismus (asymetrický)
→ nález podobný PSP

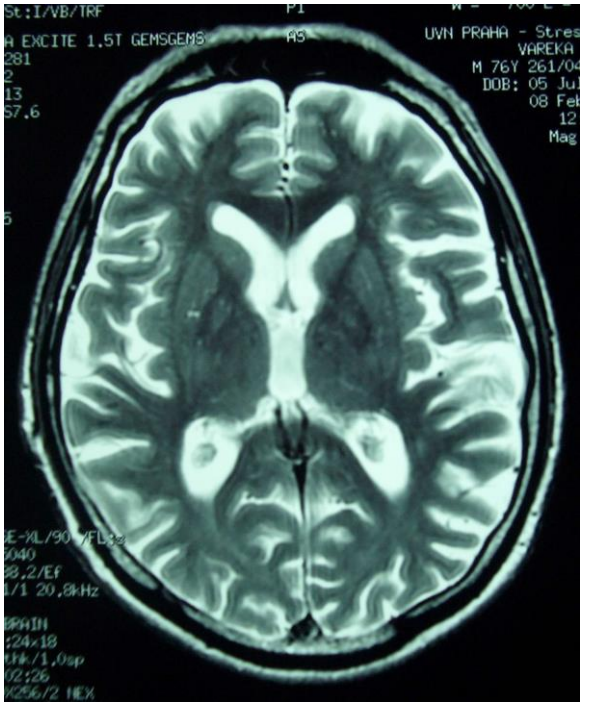



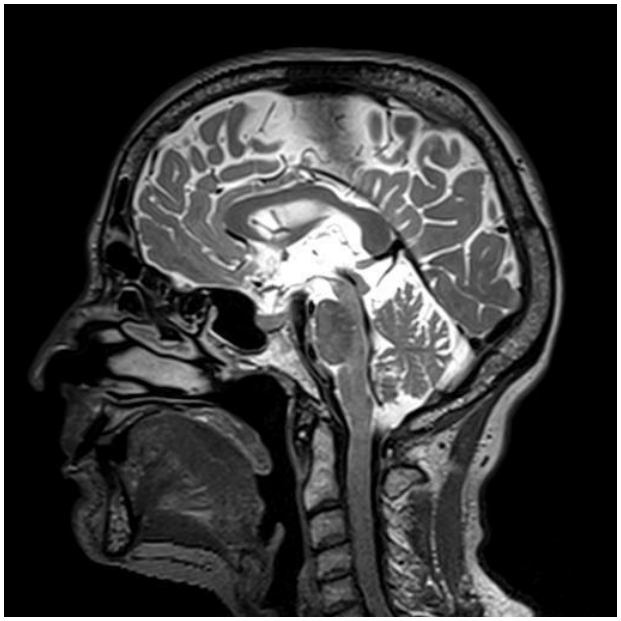



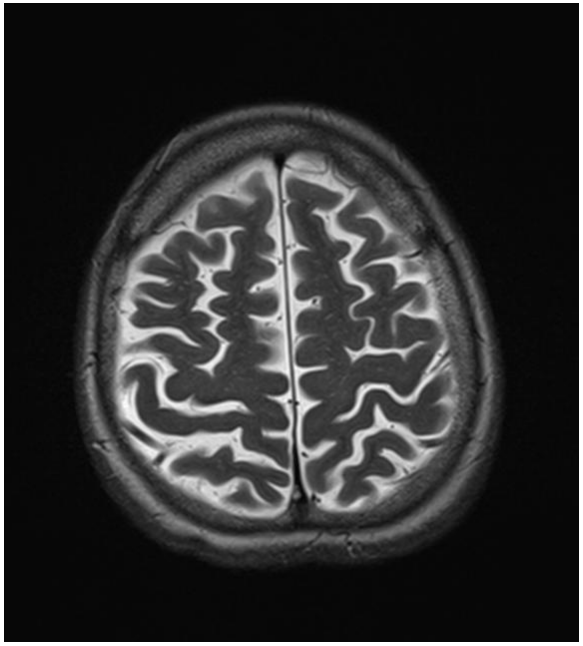
# axiální řezy v úrovni kortexu a mesencefala u CBS

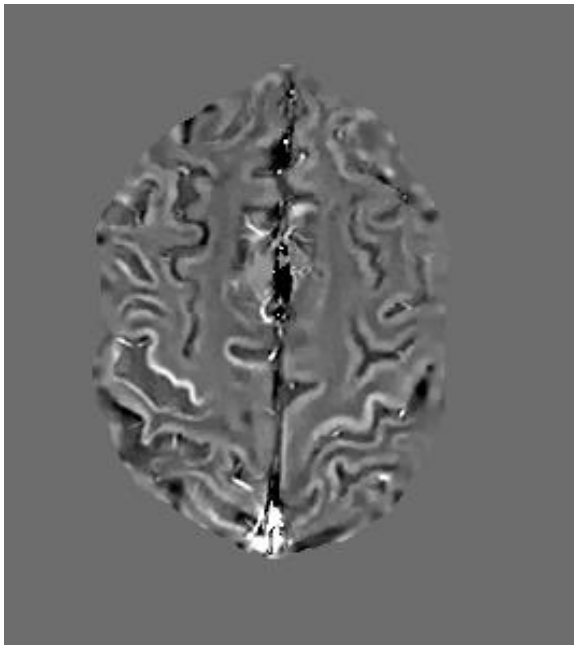
→ atrofie globální (asymetrická)
→ kortikální hypometabolismus (asymetrický)
→ nález podobný PSP



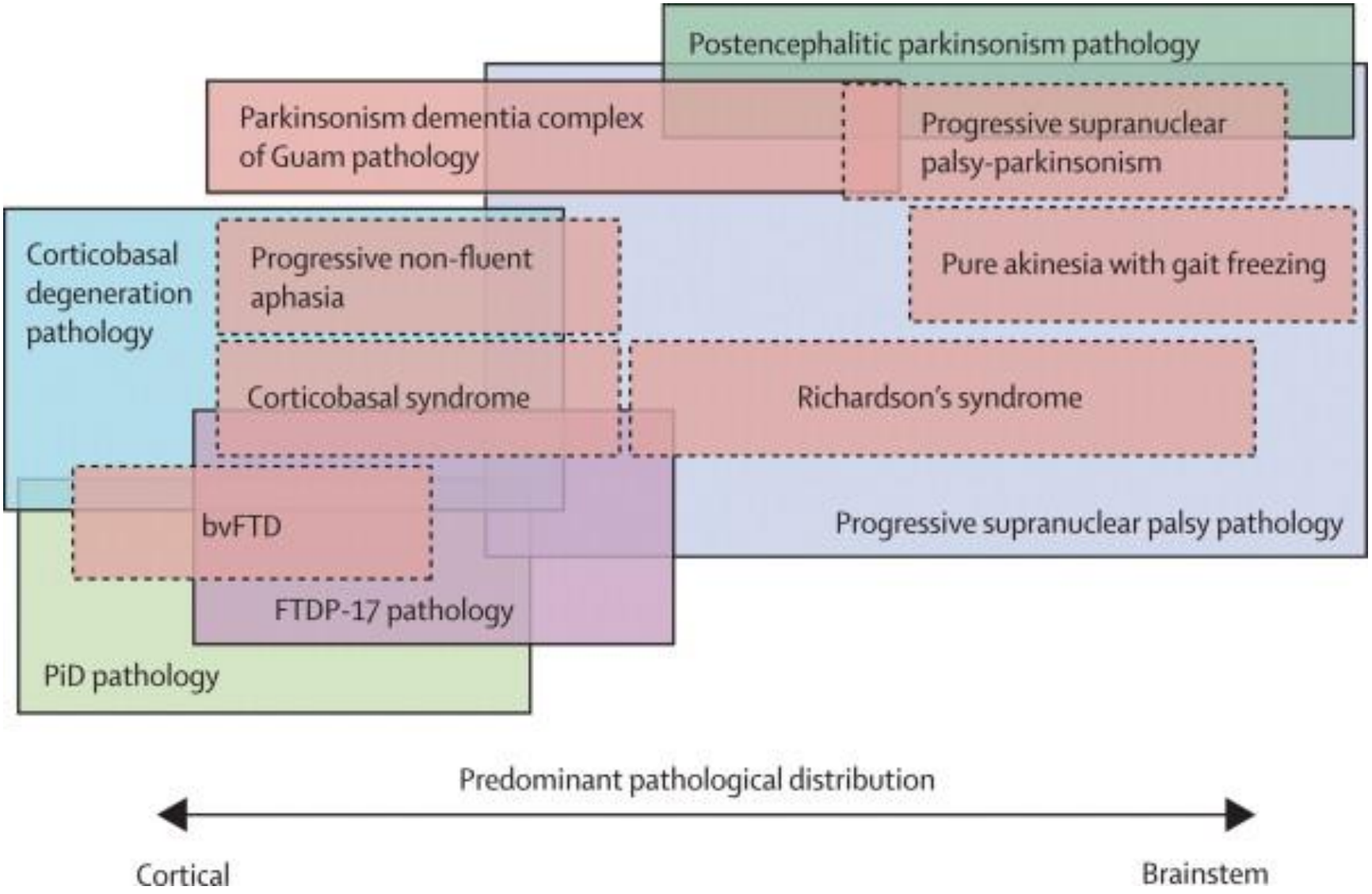







# PSP a CBS




*Williams and Lees, Lancet, 2009*

RESEARCH ARTICLE

# Conventional Magnetic Resonance Imaging in Confirmed Progressive Supranuclear Palsy and Multiple System Atrophy

Luke A. Massey, MRCP,[1–3]* Caroline Micallef, MD, FRCR,[4] Dominic C. Paviour, PhD,[1] Sean S. O'Sullivan, PhD, MRCPI,[2,3] Helen Ling, BScMed, BMBS, MSc,[1–3] David R. Williams, PhD,[5] Constantinos Kallis, PhD,[6] Janice L. Holton, PhD, FRCPath,[1,2] Tamas Revesz, MD, FRCPath,[1,2] David J. Burn, MD, FRCP,[7] Tarek Yousry, Dr med Habil, FRCR,[4] Andrew J. Lees, MD, FRCP,[1–3] Nick C. Fox, PhD, FRCP,[8] and Hans R. Jäger, MD, FRCR[4]

**ABSTRACT:** Conventional magnetic resonance imaging (cMRI) is often used to aid the diagnosis of progressive supranuclear palsy (PSP) and multiple system atrophy (MSA), but its ability to predict the histopathological diagnosis has not been systematically studied. cMRI from 48 neuropathologically confirmed cases, including PSP (n = 22), MSA (n = 13), Parkinson's disease (PD) (n = 7), and corticobasal degeneration (n = 6), and controls (n = 9) were assessed blinded to clinical details and systematically rated for reported abnormalities. Clinical diagnosis and macroscopic postmortem findings were retrospectively assessed. Radiological assessment of MRI was correct in 16 of 22 (72.7%) PSP cases and 10 of 13 (76.9%) MSA cases with substantial interrater agreement (Cohen's kappa 0.708; $P < .001$); no PSP case was misclassified as MSA or vice versa. MRI was less sensitive but more specific than clinical diagnosis in PSP and both more sensitive and specific than clinical diagnosis in MSA. The "hummingbird" and "morning glory" signs were highly specific for PSP, and "the middle cerebellar peduncle sign" and "hot cross bun" for MSA, but sensitivity was lower (up to 68.4%) and characteristic findings may not be present even at autopsy. cMRI, clinical diagnosis, and macroscopic examination at postmortem have similar sensitivity and specificity in predicting a neuropathological diagnosis. We have validated specific radiological signs in pathologically confirmed PSP and MSA. However, the low sensitivity of these and macroscopic findings at autopsy suggest a need for imaging techniques sensitive to microstructural abnormalities without regional atrophy. © 2012 *Movement* Disorder Society

**Key Words:** pathological confirmation; conventional MRI; progressive supranuclear palsy; multiple system atrophy

# Shrnutí

- Neuroimaging je pomocnou metodou v diferenciální diagnostice neurodegenerací
- MR známky APS mají vysokou specificitu, ale omezenou sensitivitu
- **MSA**: současné **postižení BG** (asymetrická atrofie putamen s depozity ferritinu) **a ponto-cerebellární oblasti** (atrofie a T2 hypersignál středního mozečkového stonku)
- **PSP: globální atrofie** s maximem v oblasti **mesencefala** a symetrická akumulace Fe v GP
- **CBS**: **asymetrická atrofie kortikálně** a nález podobný PSP