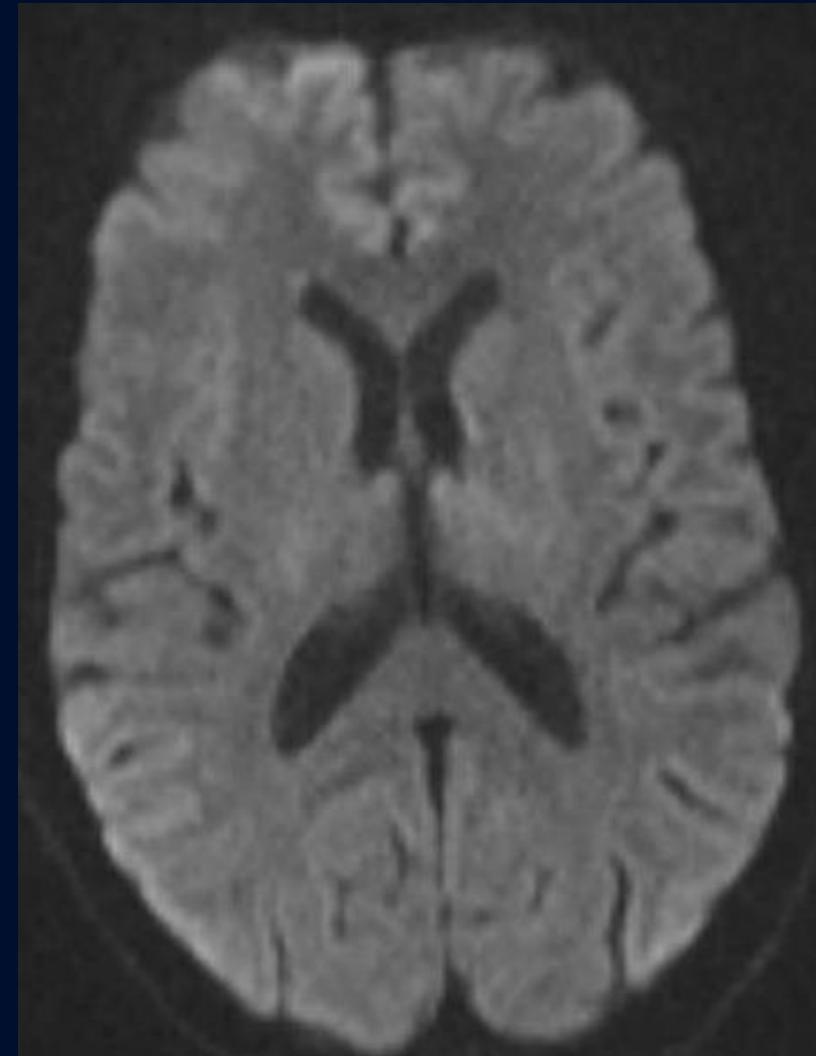
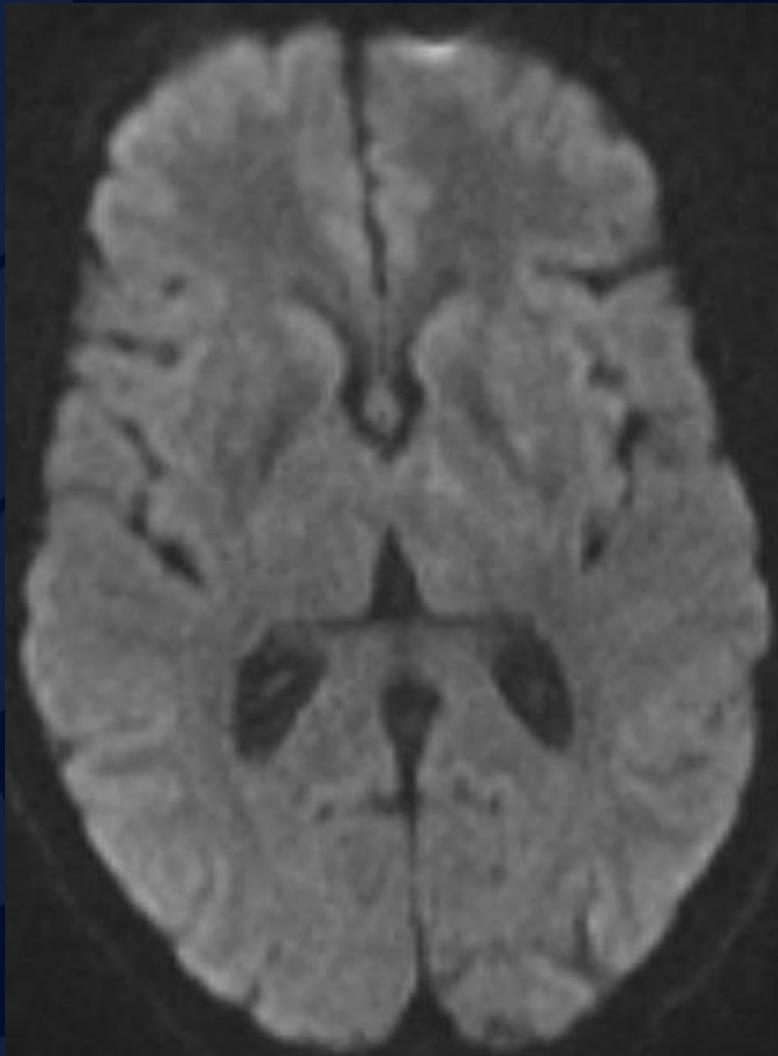




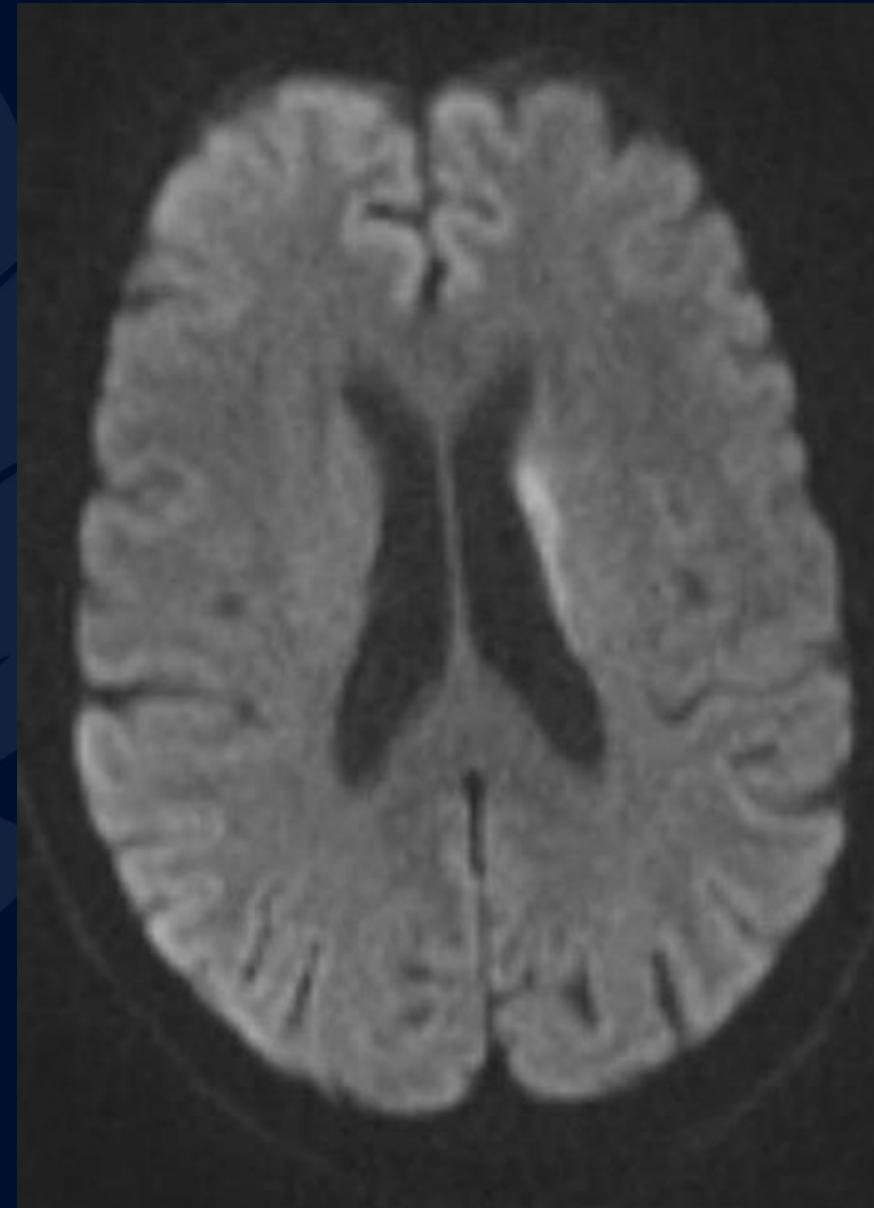
64-year-old female with rapidly
progressing atypical
parkinsonism and cognitive
changes

Annie Jin, MS3

DWI

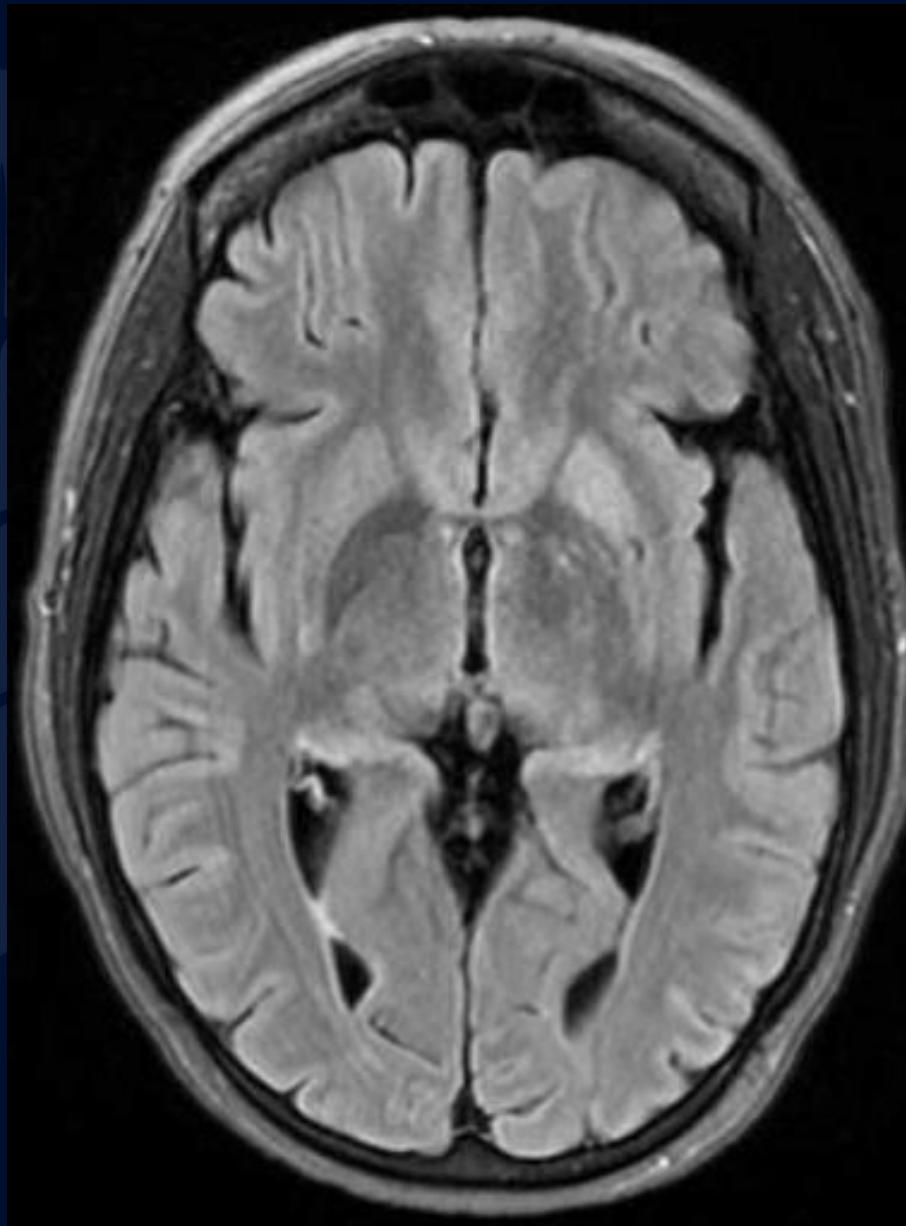


DWI

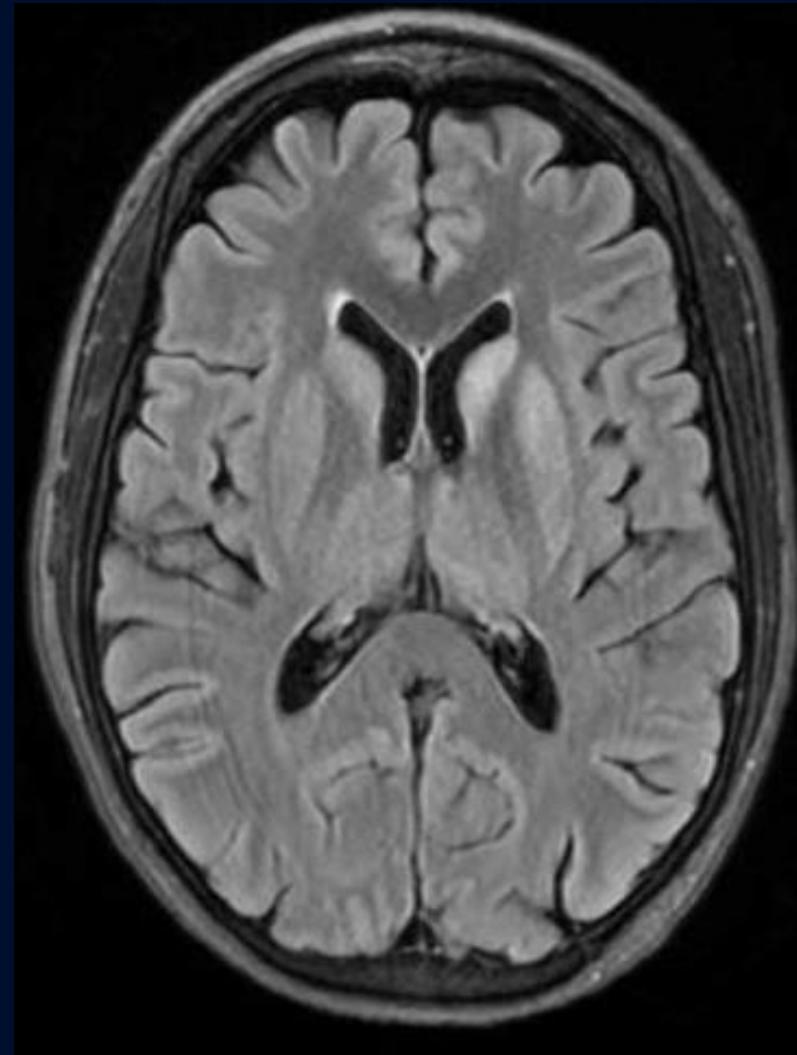
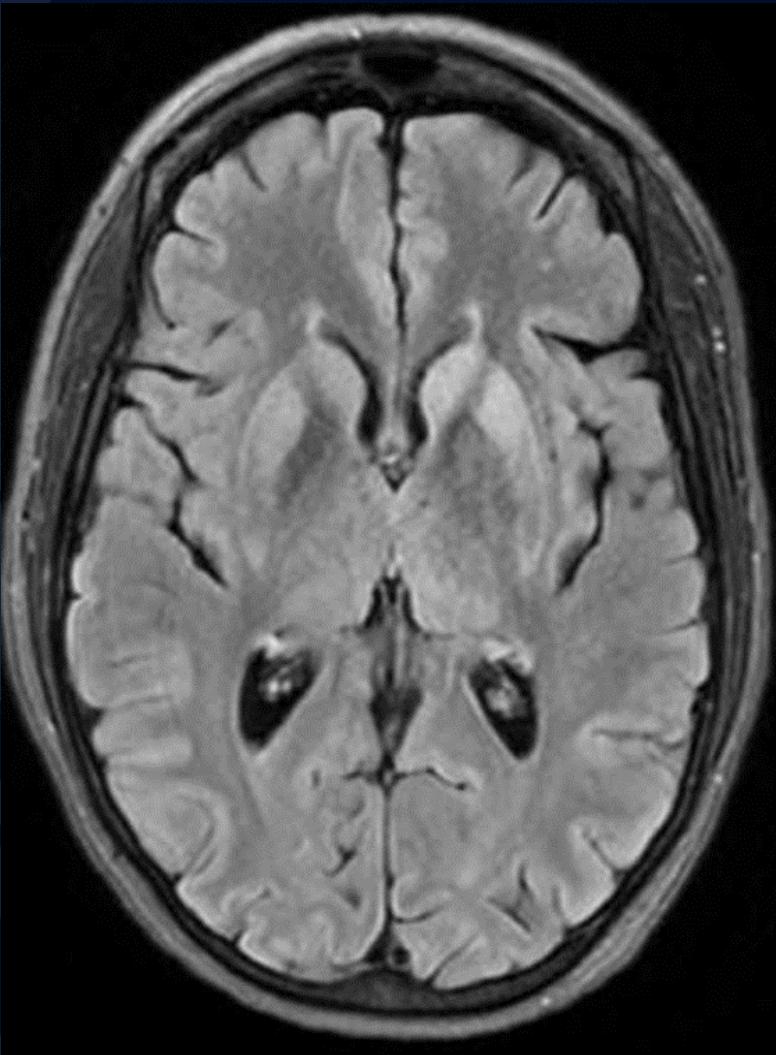


UCONN
HEALTH
RADIOLOGY

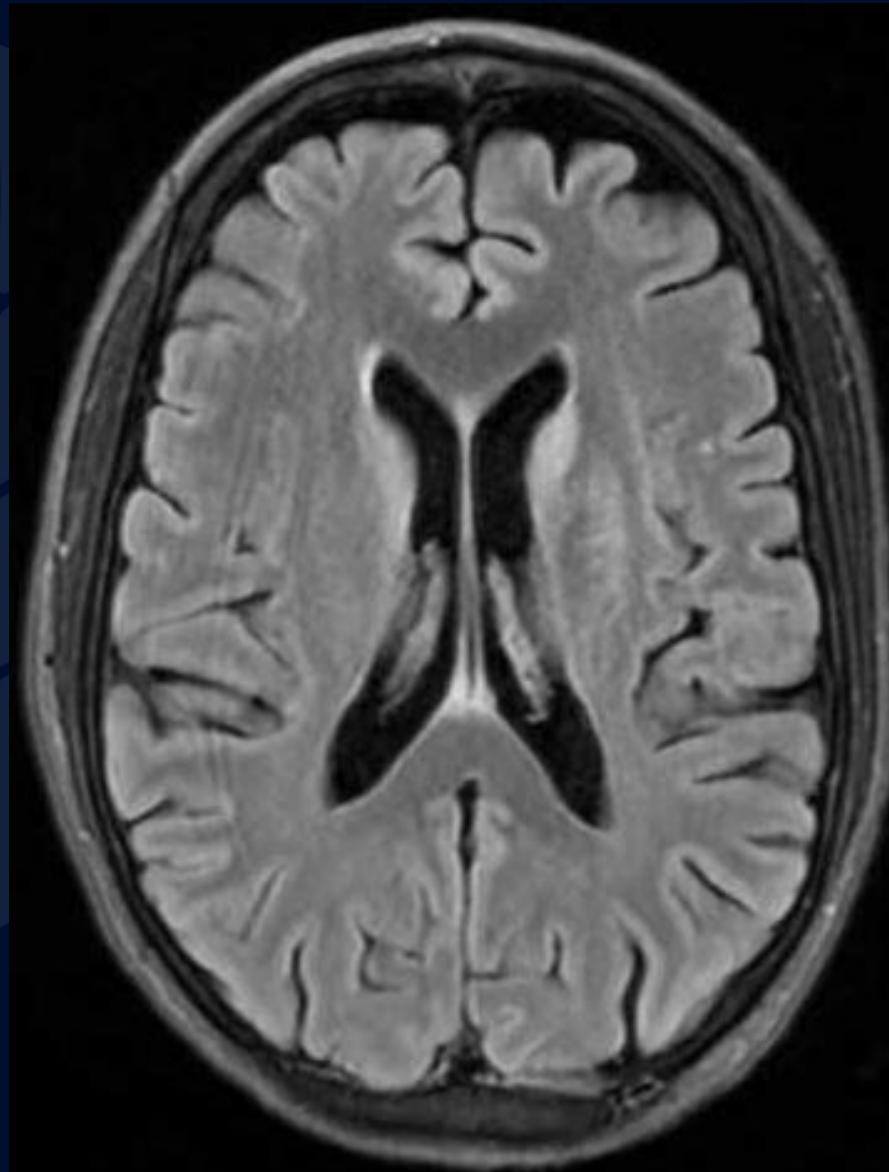
T2 FLAIR



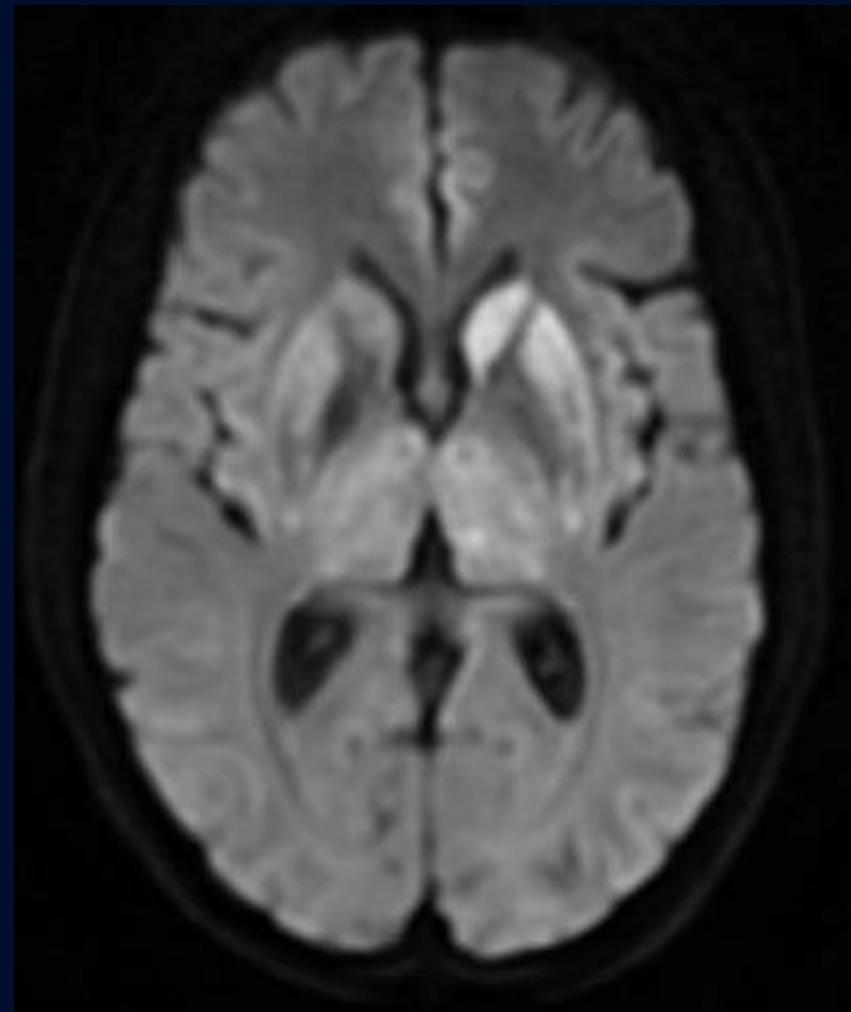
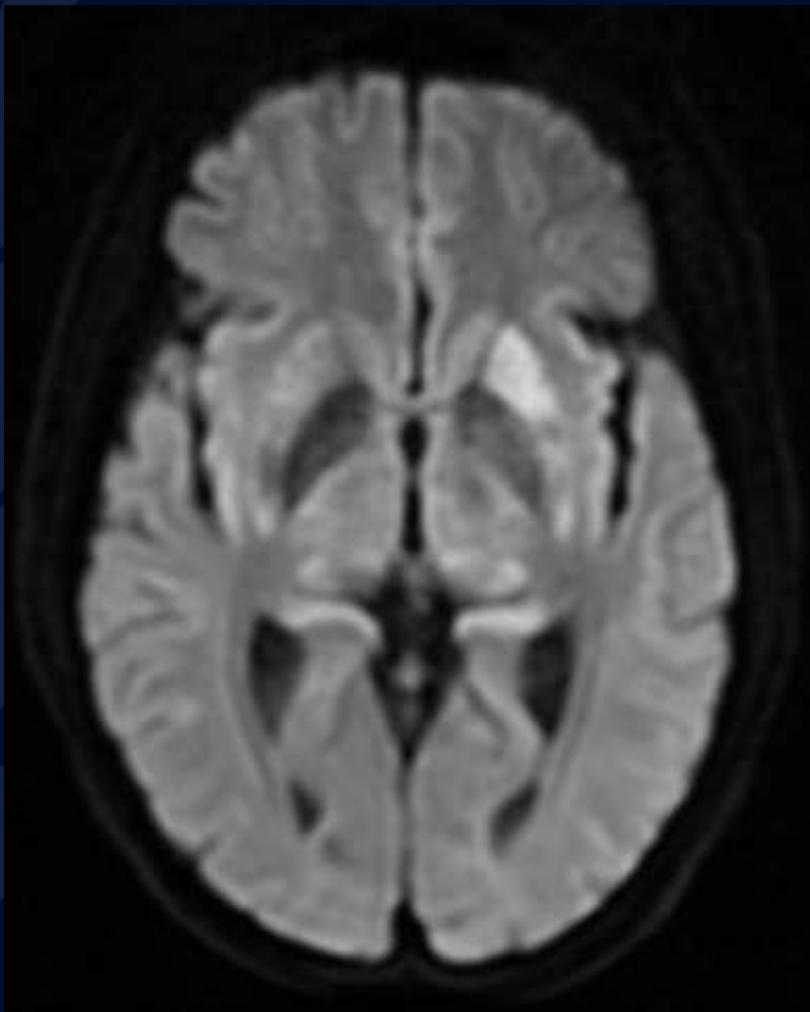
T2 FLAIR



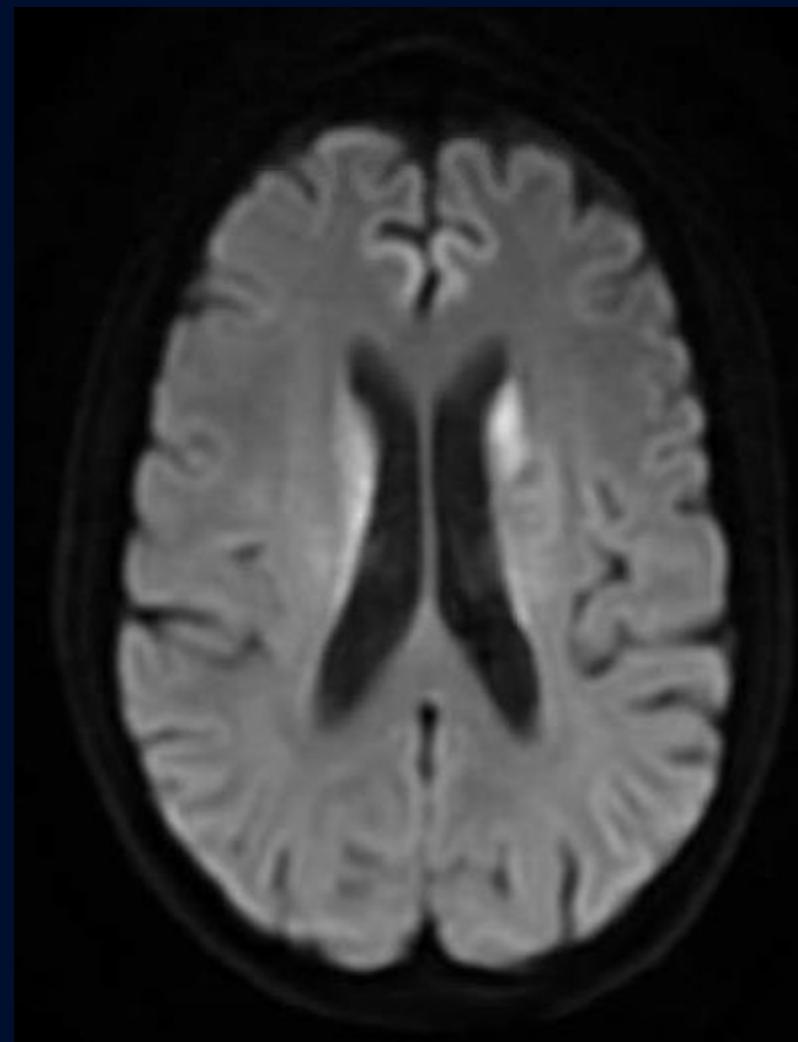
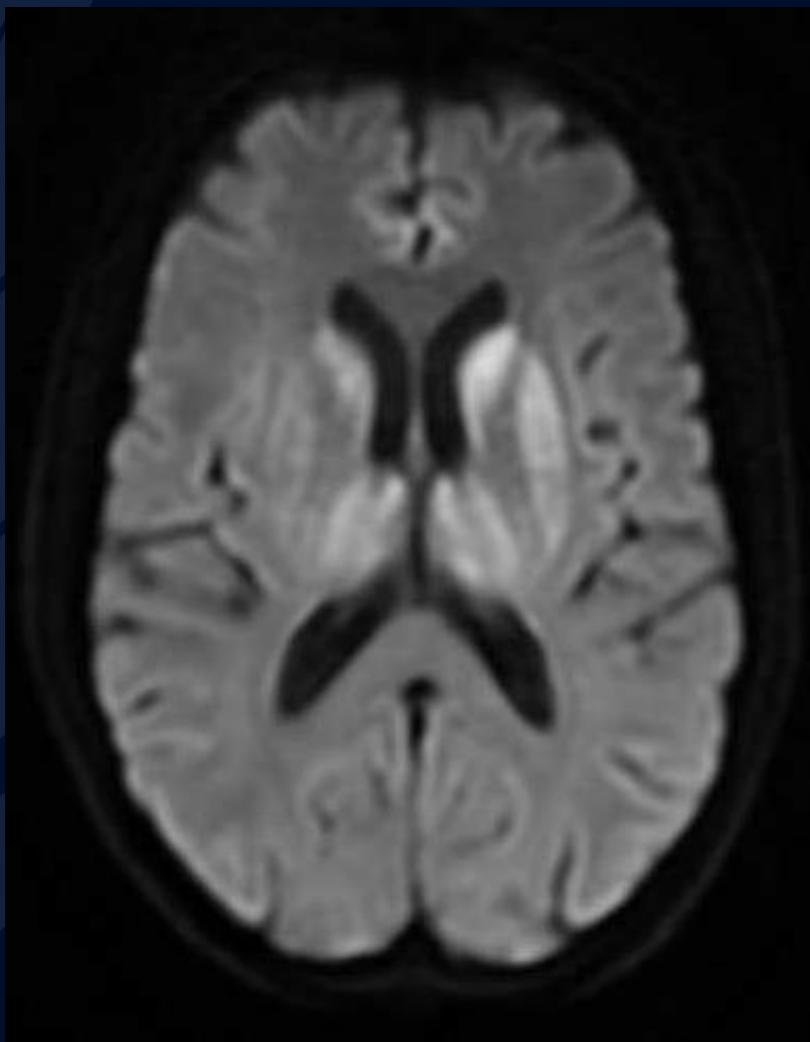
T2 FLAIR



DWI



DWI





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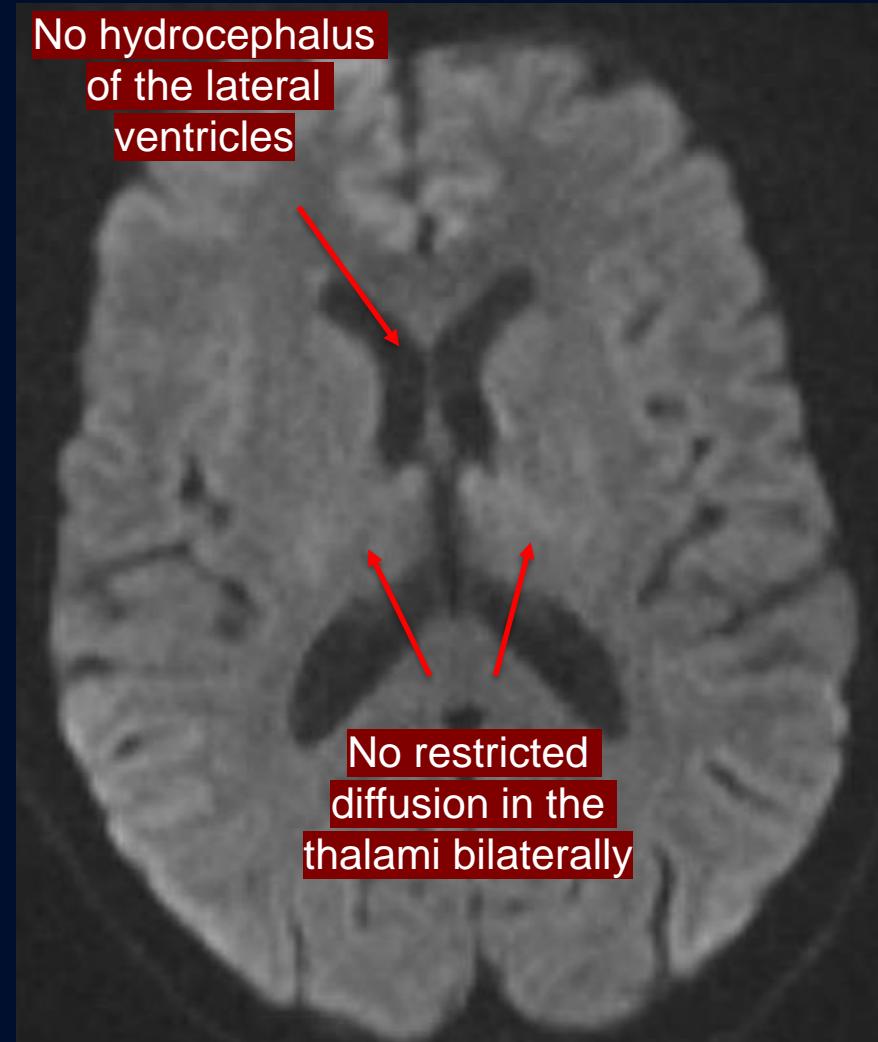


Cruetzfeldt Jakob Disease (CJD)

DWI



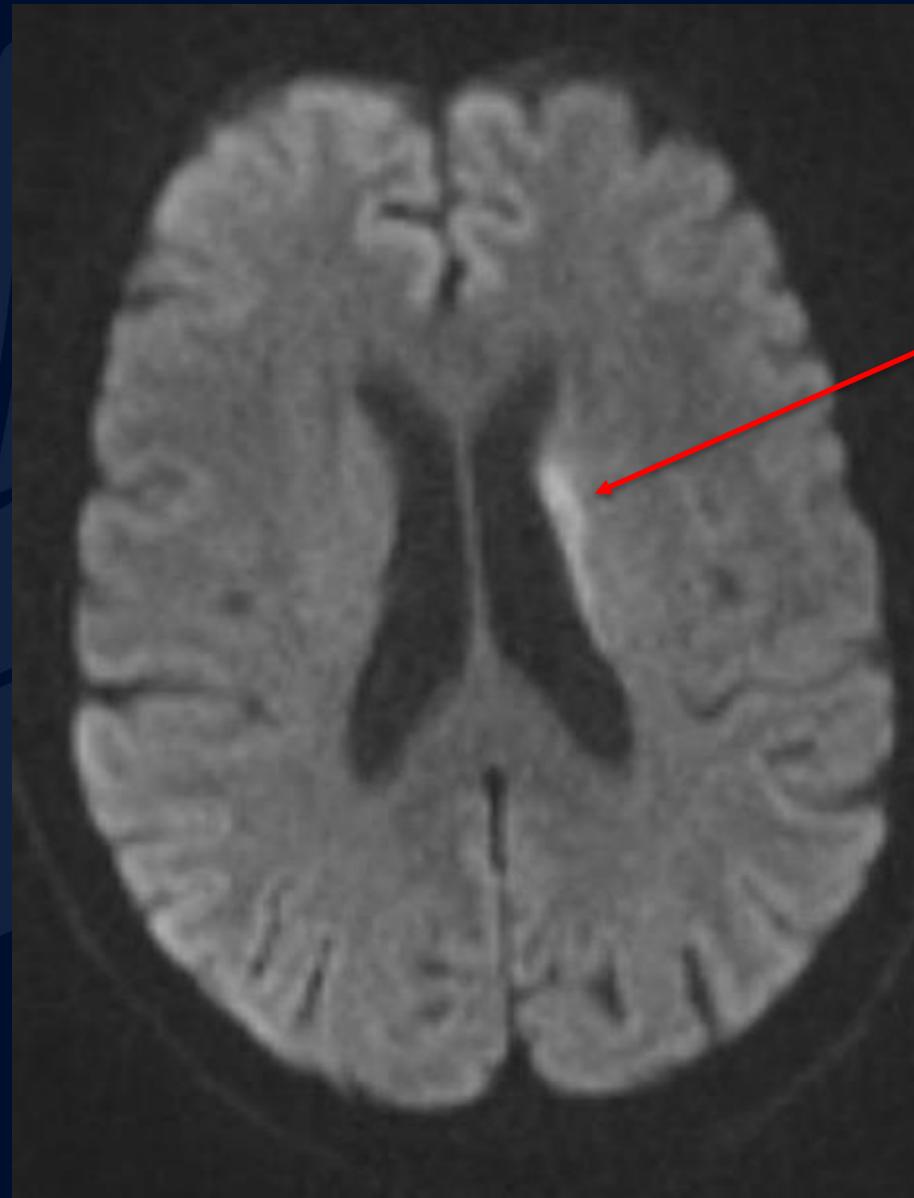
No restricted diffusion in the caudate nucleus bilaterally



No hydrocephalus
of the lateral
ventricles

No restricted
diffusion in the
thalami bilaterally

DWI

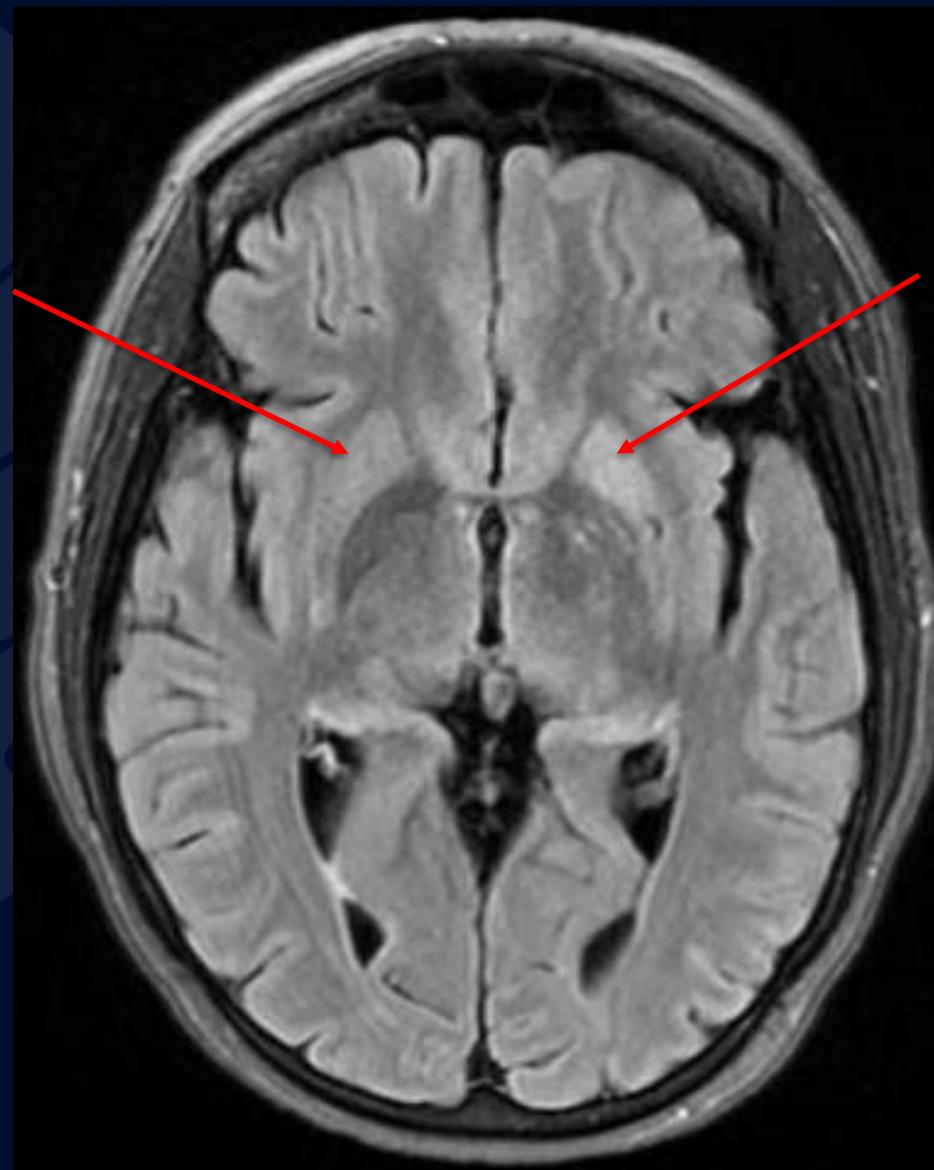


Left centrum
semiovale / basal
ganglia restricted
diffusion

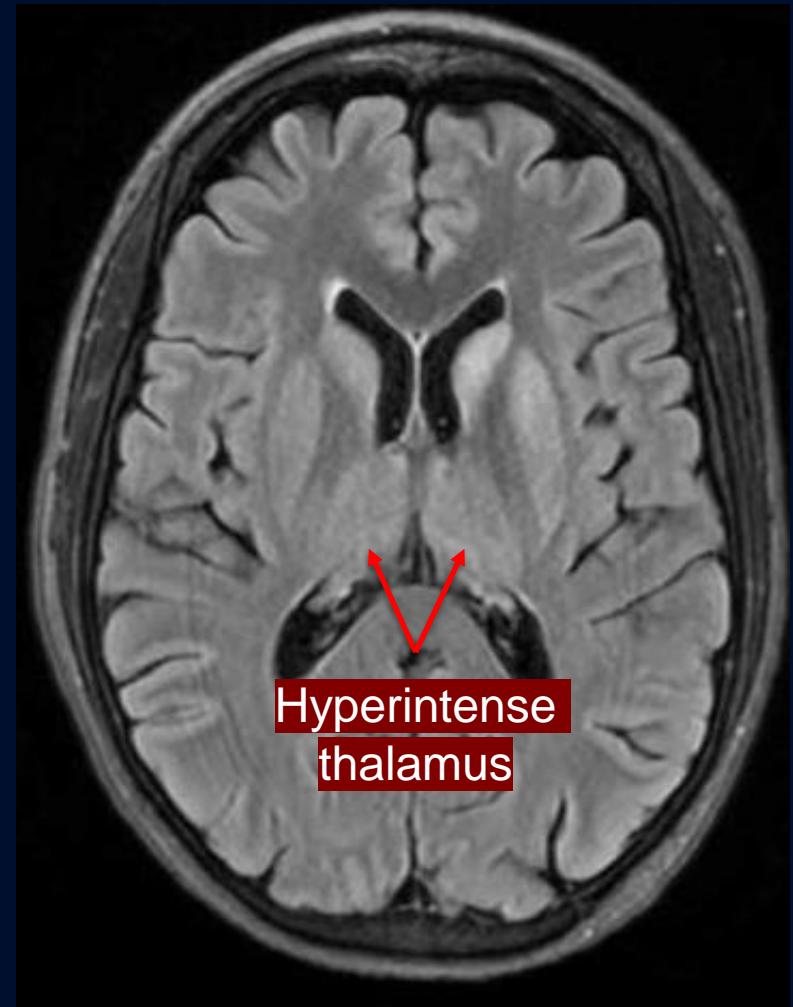
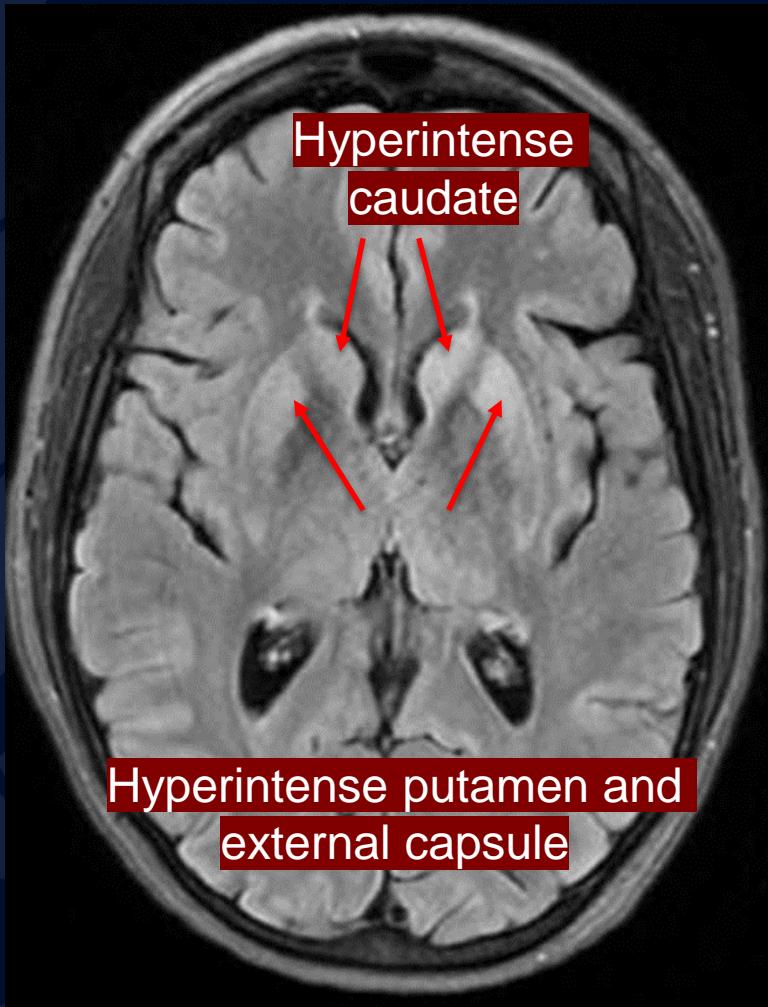
T2 FLAIR

Right putamen
less hyperintense
than left

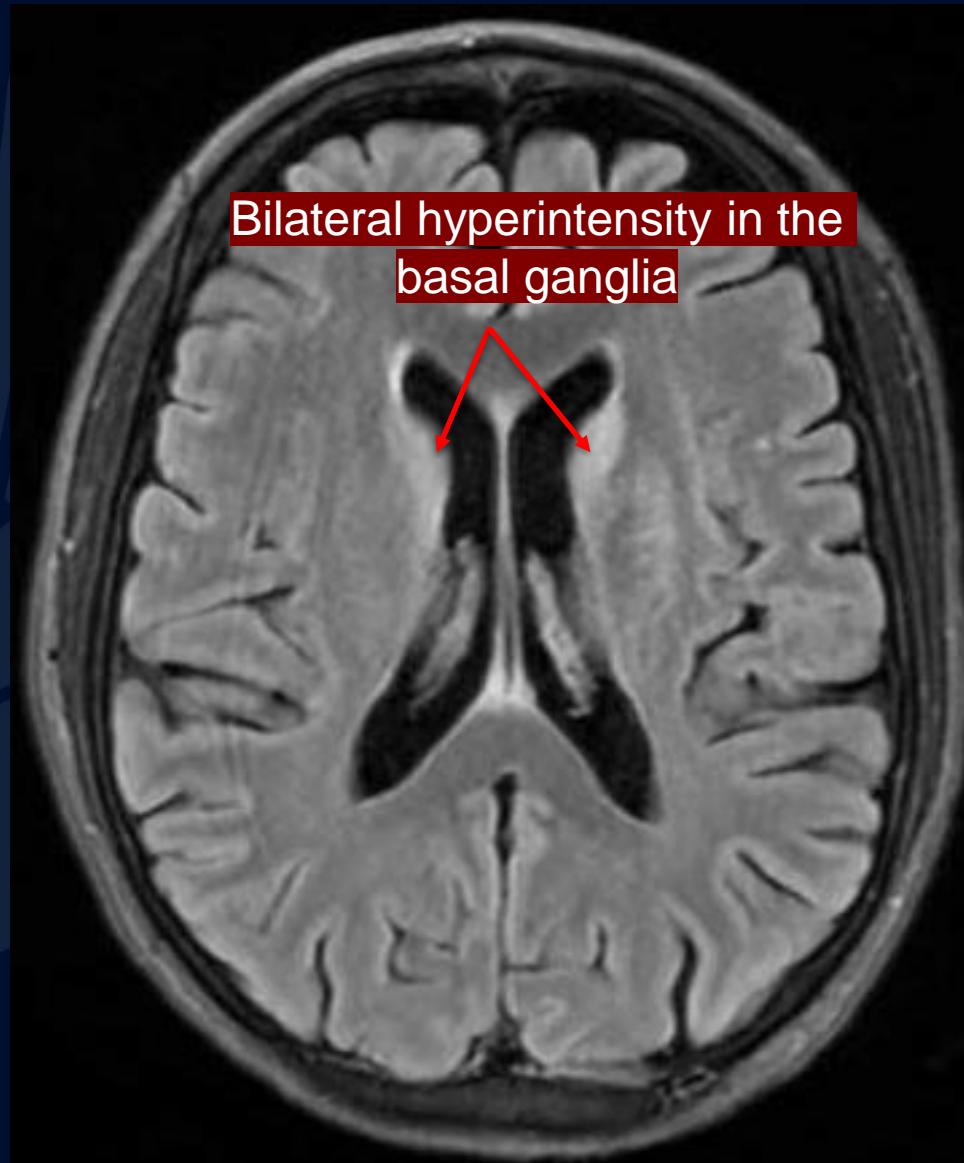
Left putamen
more hyperintense
than right



T2 FLAIR



T2 FLAIR



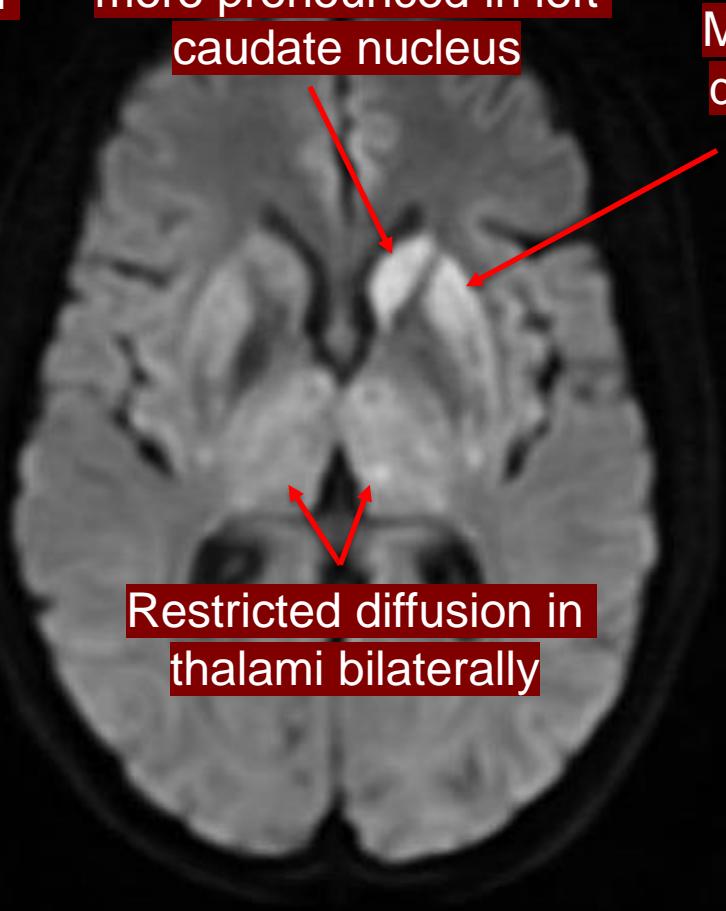
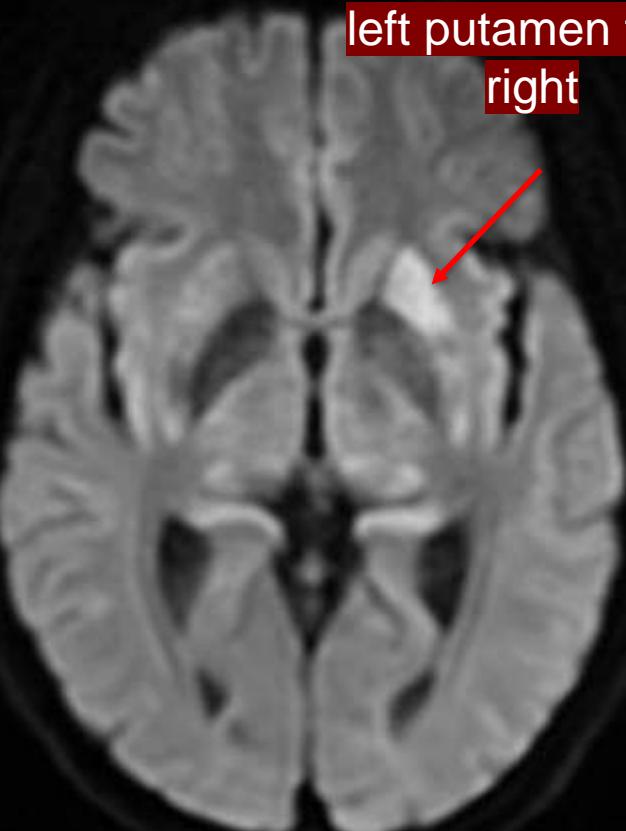
Bilateral hyperintensity in the
basal ganglia

DWI

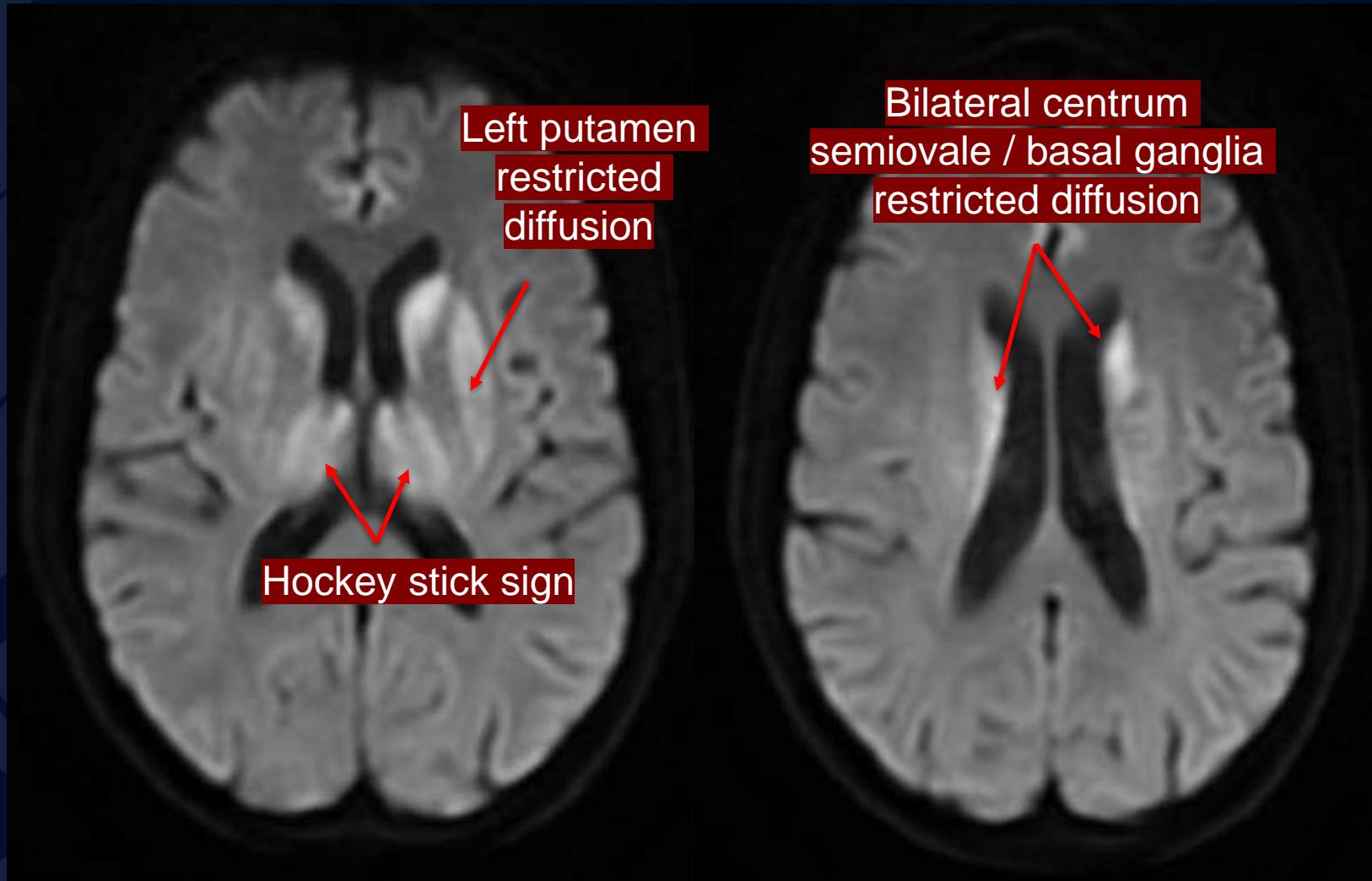
Restricted diffusion
more pronounced in
left putamen than
right

Restricted diffusion
more pronounced in left
caudate nucleus

More restricted
diffusion in left
putamen



DWI



Cruetzfeldt Jakob Disease (CJD)

- CJD is a rare disease, with an occurrence rate of 1 per 1 million population per year
 - Sporadic CJD thought to be the most common 85-95%
 - 5-15% are due to genetic CJD, iatrogenic CJD
 - 1% variant CJD
- Prions: infectious protein containing particle PrP^{SC} that replicate, replace normal prion proteins and cause neurotoxicity
- **MRI:** abnormal hyperintensities are seen in the head of the caudate, putamen, sometimes the thalamus
 - Variant CJD: hockey stick sign, involving the pulvinar and dorsomedial thalamic nuclei bilaterally
- EEG findings: periodic sharp wave complexes
- CSF samples:
 - Real time quaking-induced conversion RT-QuIC: most sensitive
 - 14-3-3 protein test: nonspecific test
- Prognosis is poor, there is no effective treatment and death occurs within one year of symptom onset

MRI Imaging Findings

Hyperintense signals can be seen on DWI, FLAIR and T2 images

- DWI: **most sensitive** MRI sequence to detect CJD related lesions
 - Best to observe cortical and striatal changes
 - Correlates with spongiform change and vacuolization of neuropil
 - Rare cases signal in cerebellum can occur
- T2:
 - Spontaneous CJD: increase T2 signal in basal ganglia involve caudate and putamen
 - Thalamic hyperintensities can occur
 - Late atrophy or white matter change
- FLAIR:
 - Rare cases cerebellum involvement can be seen
- Variant CJD:
 - Double hockey stick or pulvinar sign: confluent hyperintense signal in the mesial and dorsal thalamus
 - Seen on DWI, FLAIR and T2 weighted MRI

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