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

Annie Jin, MS3

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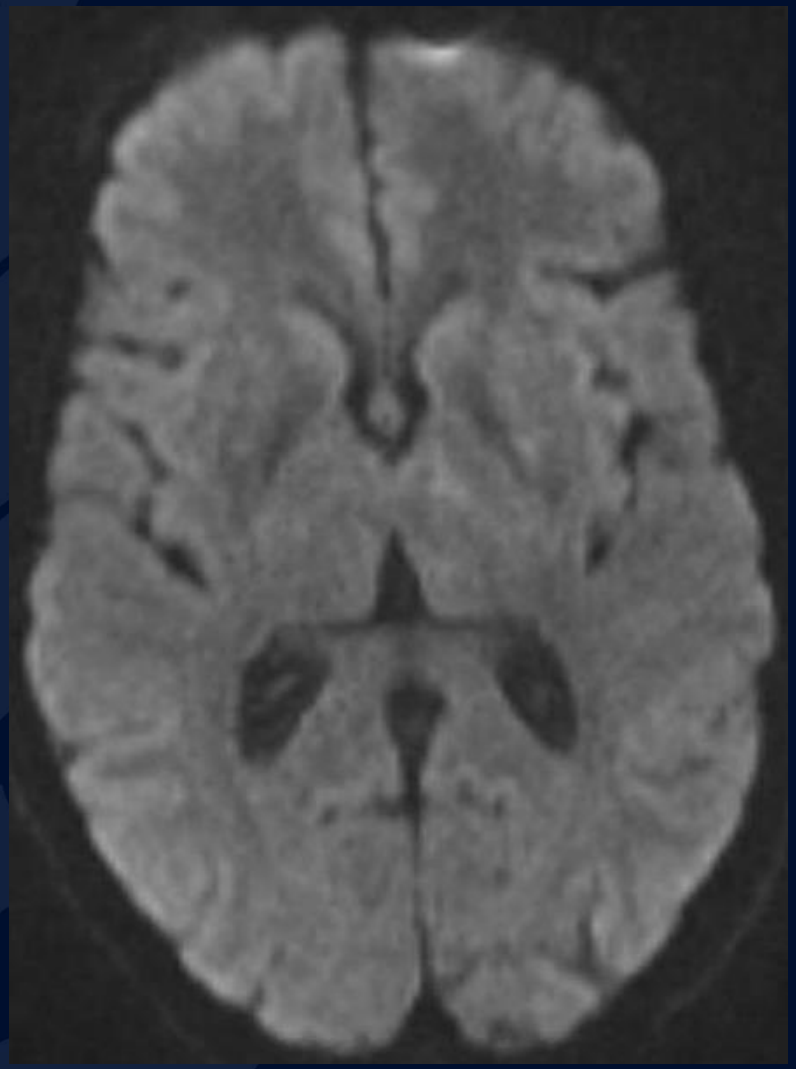
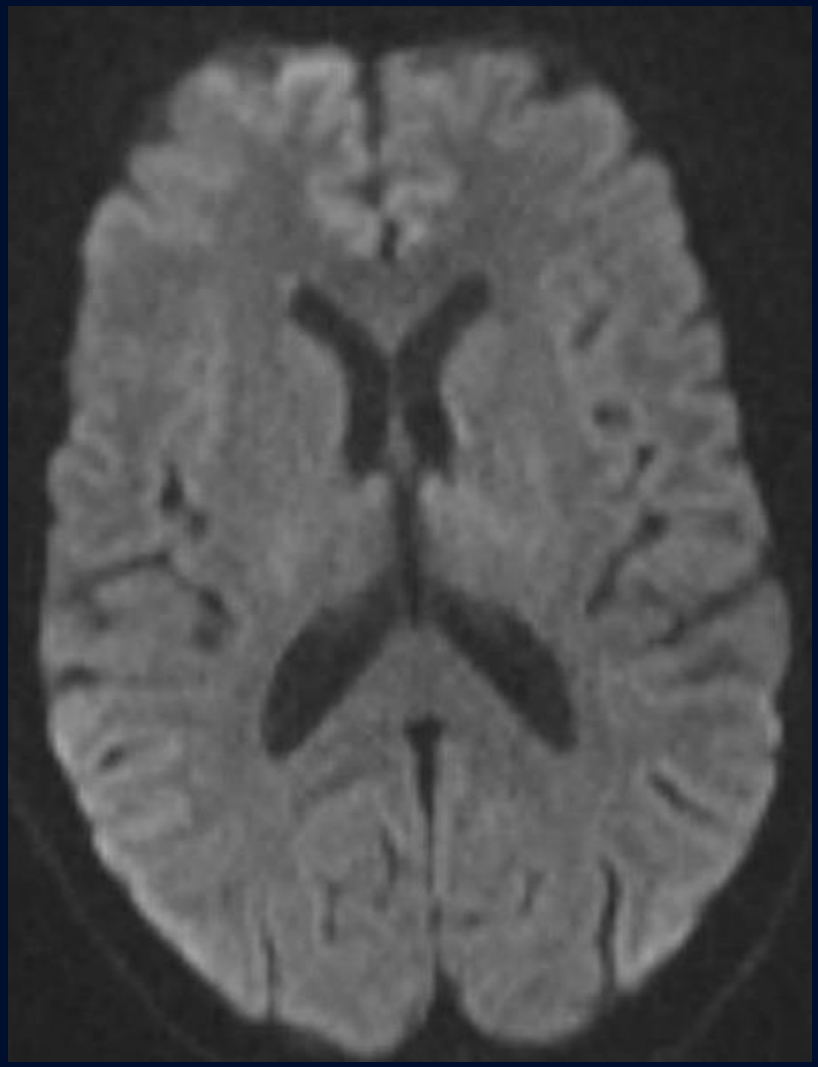
DWI





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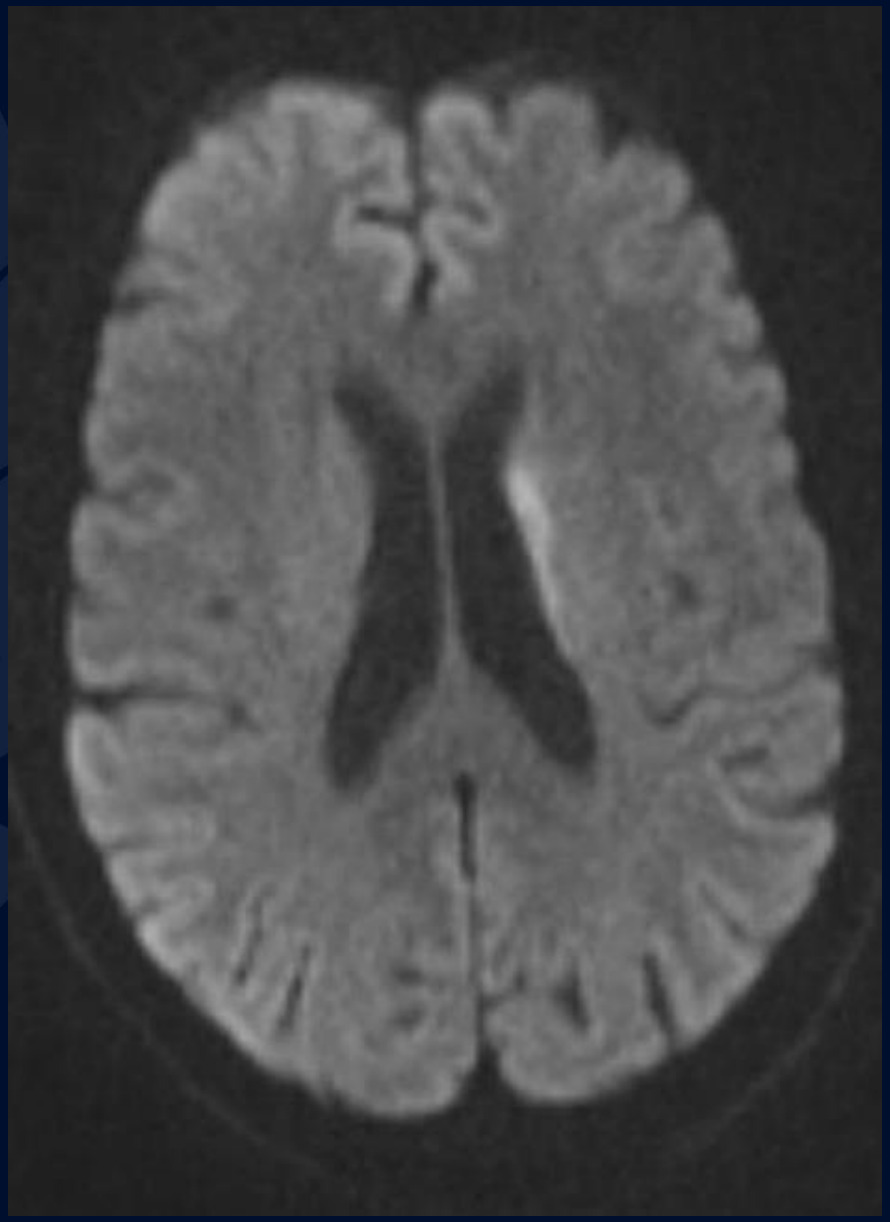

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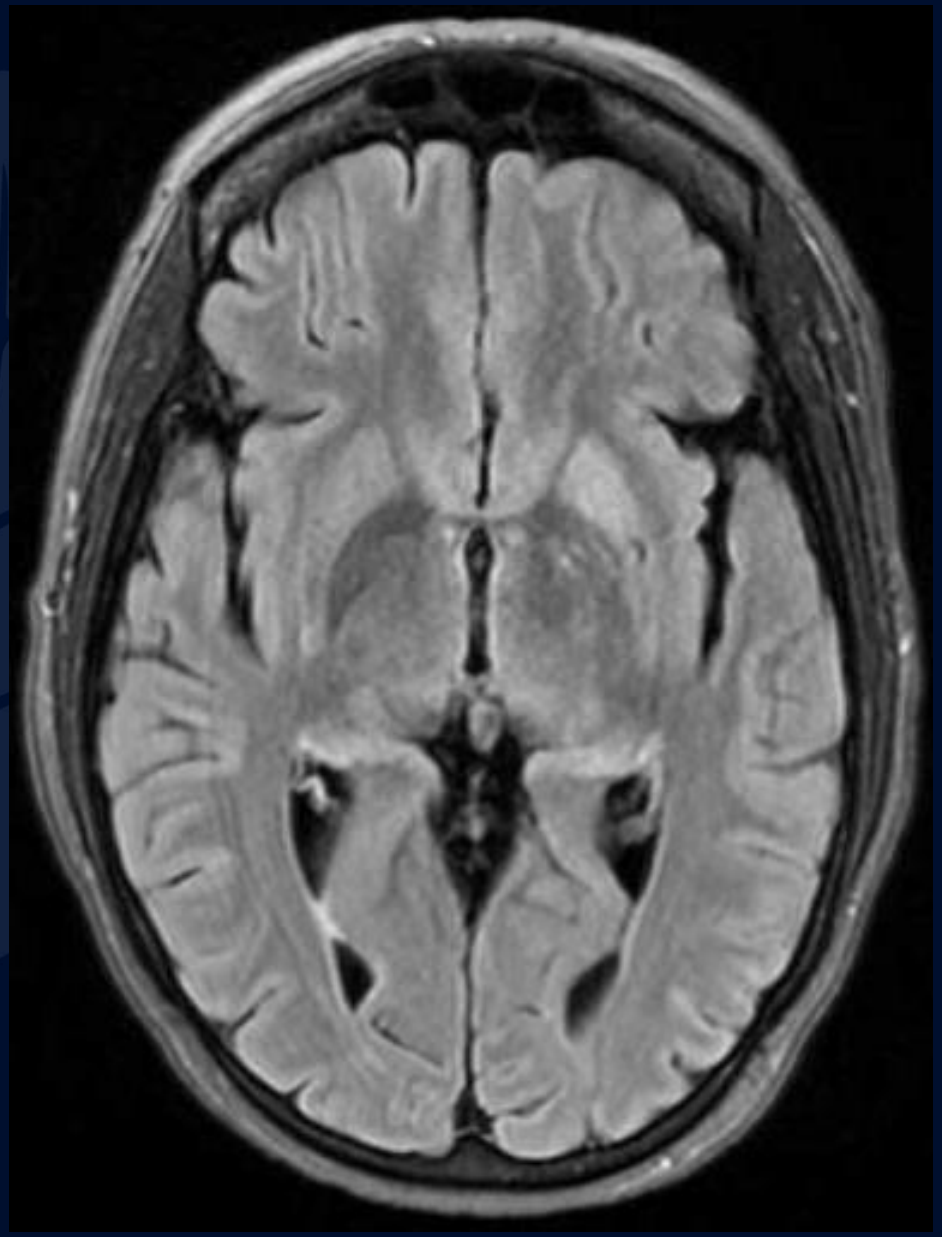
T2 FLAIR



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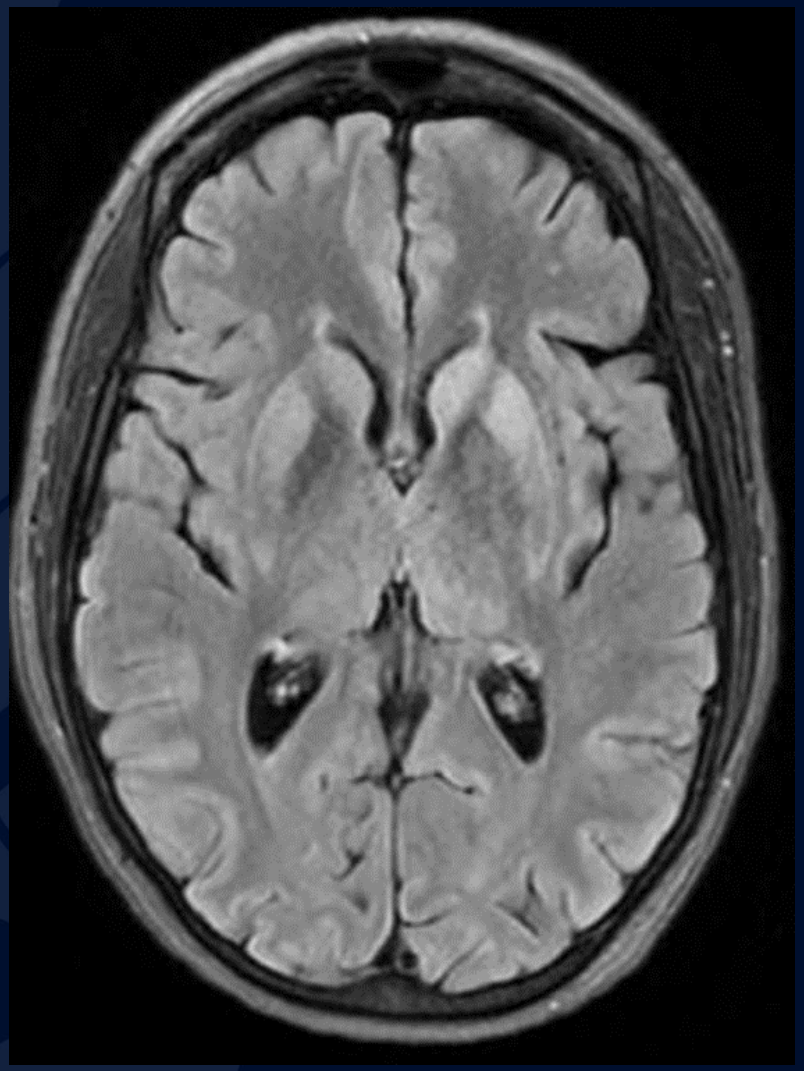
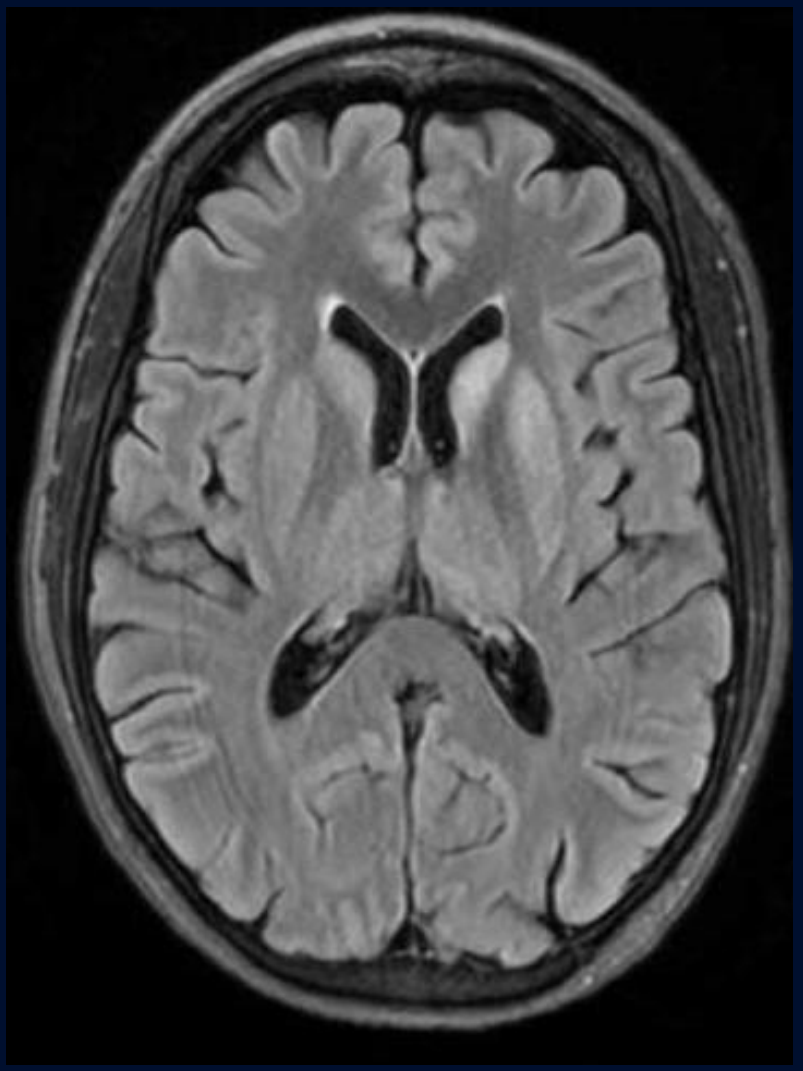
T2 FLAIR





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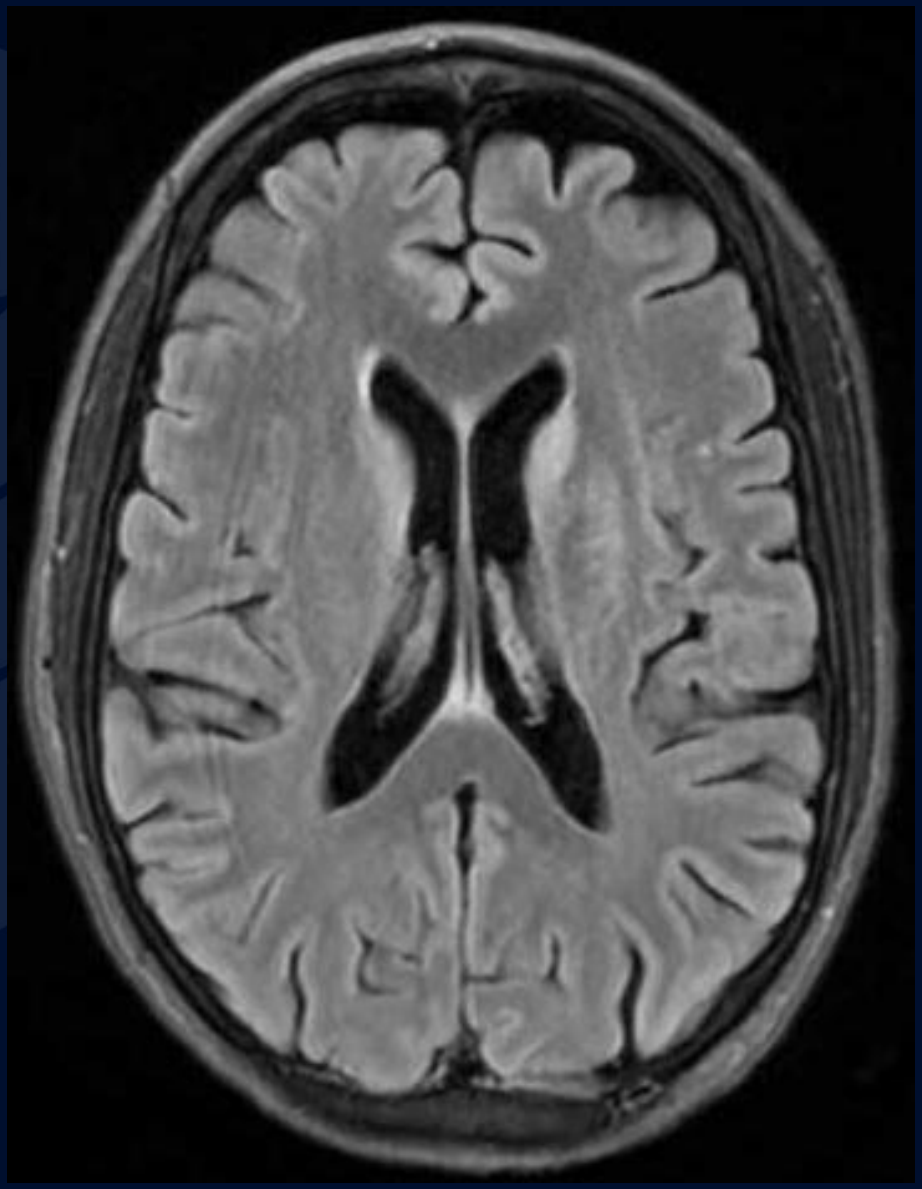
T2 FLAIR



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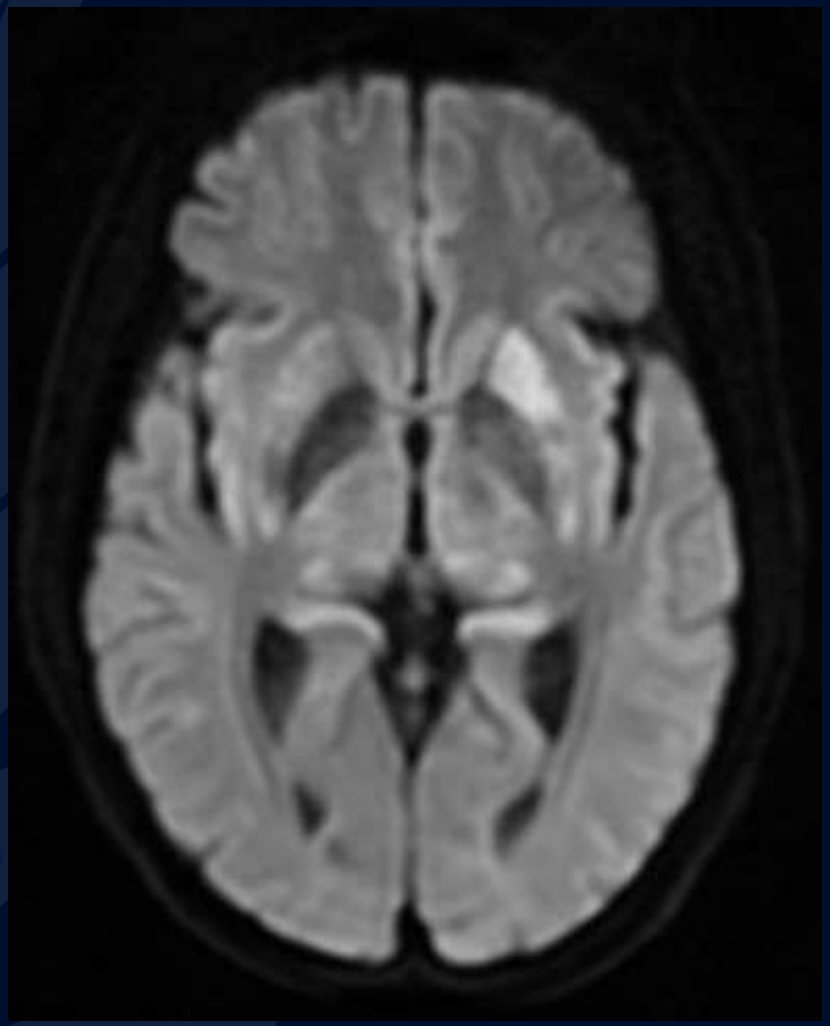

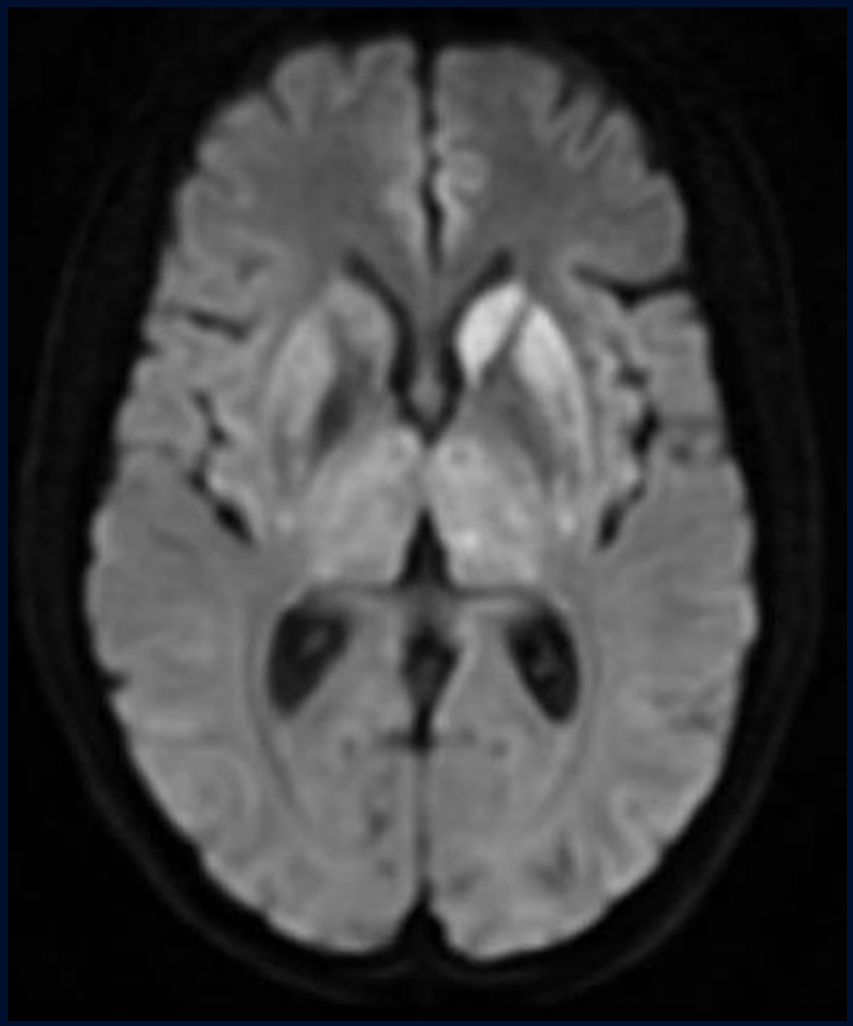

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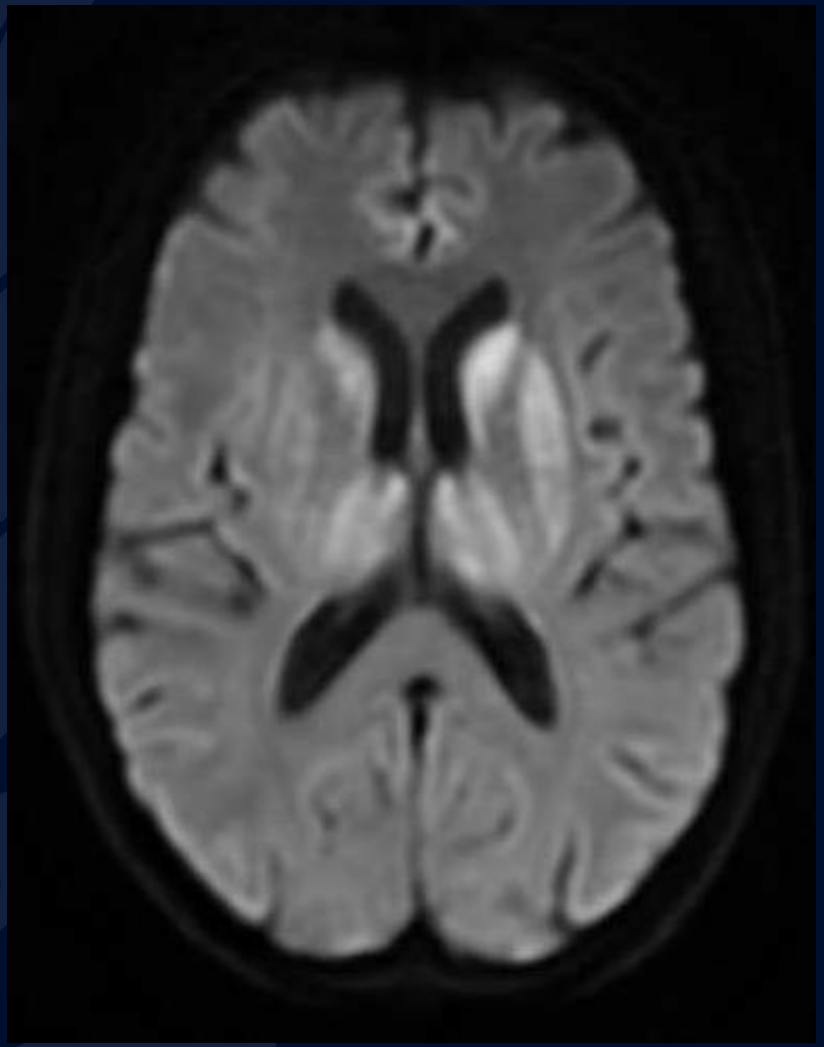

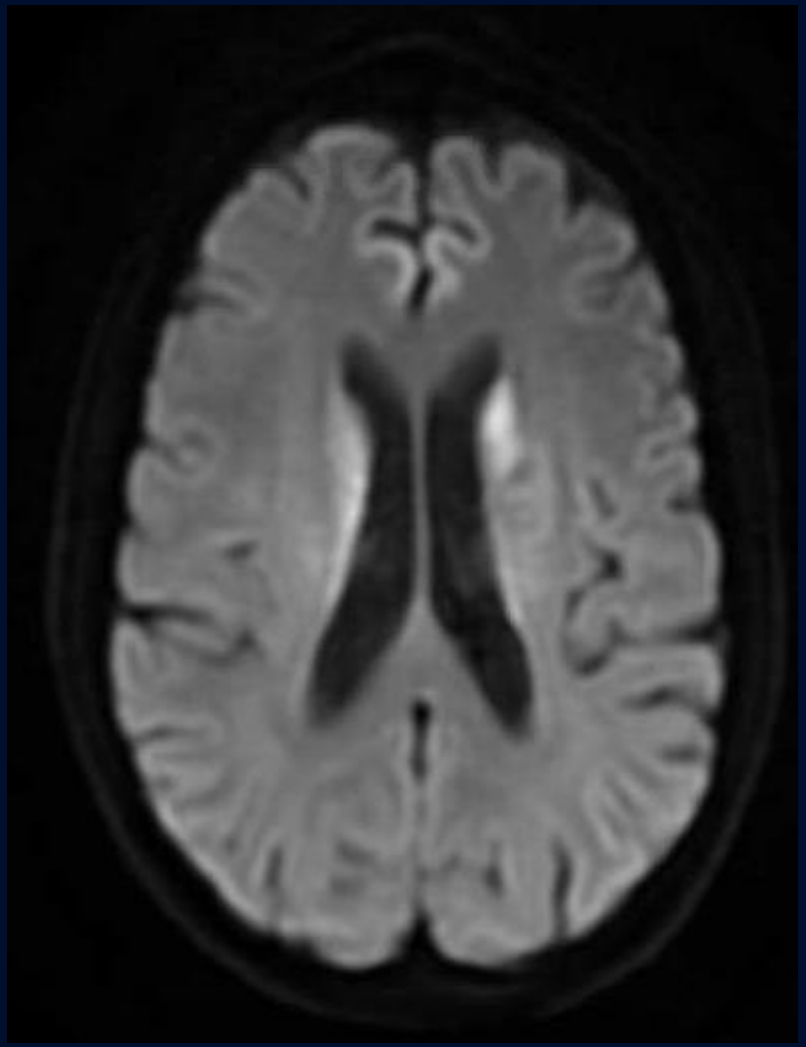

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Cruetzfeldt Jakob Disease (CJD)

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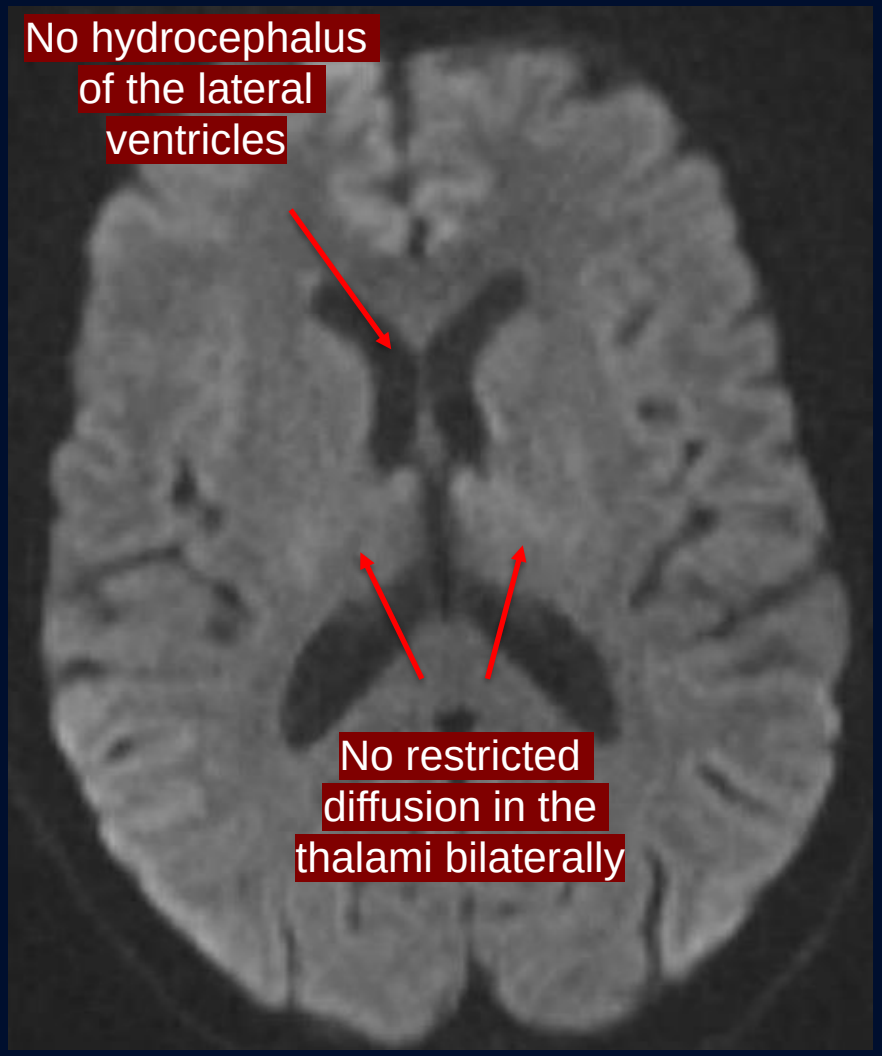

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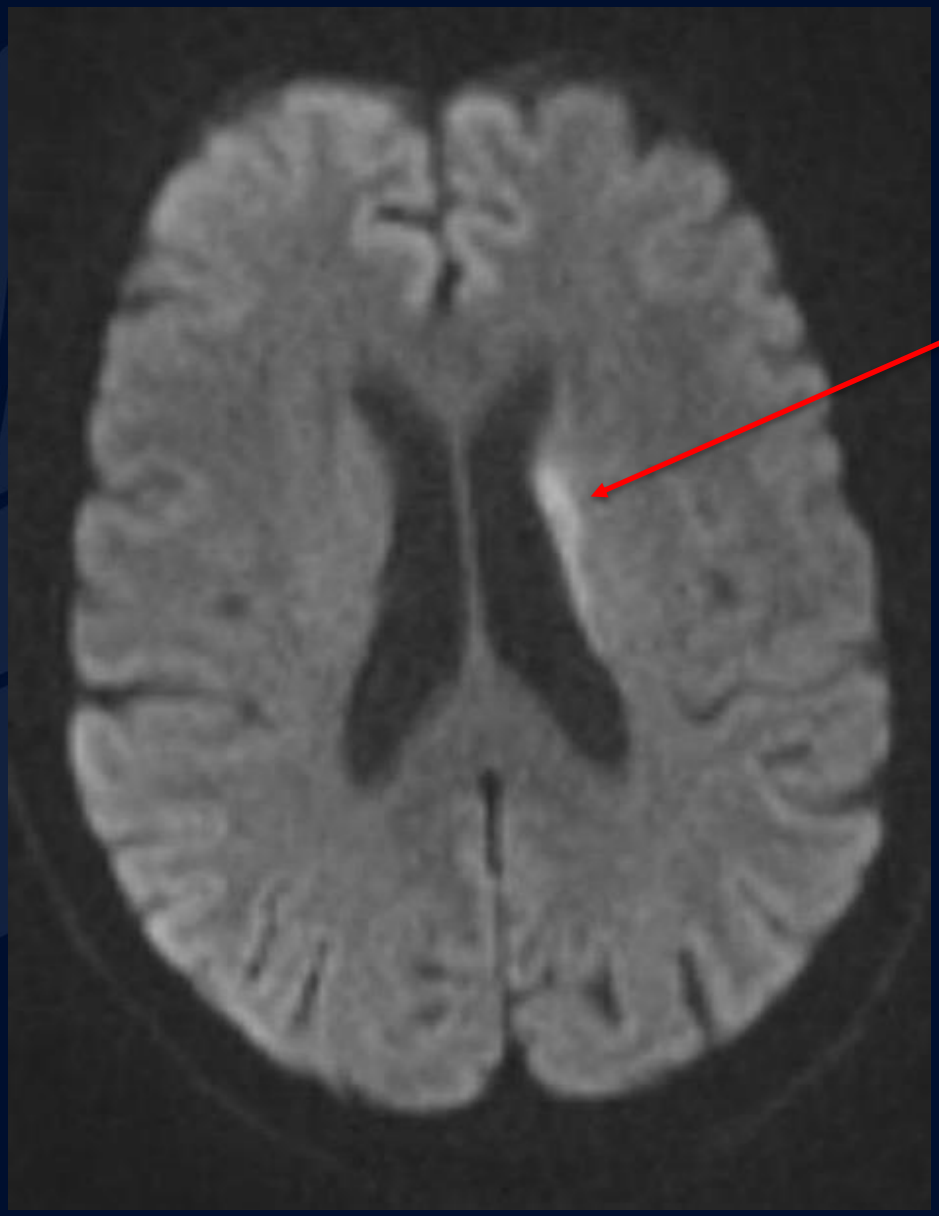

Left centrum semiovale / basal ganglia restricted diffusion

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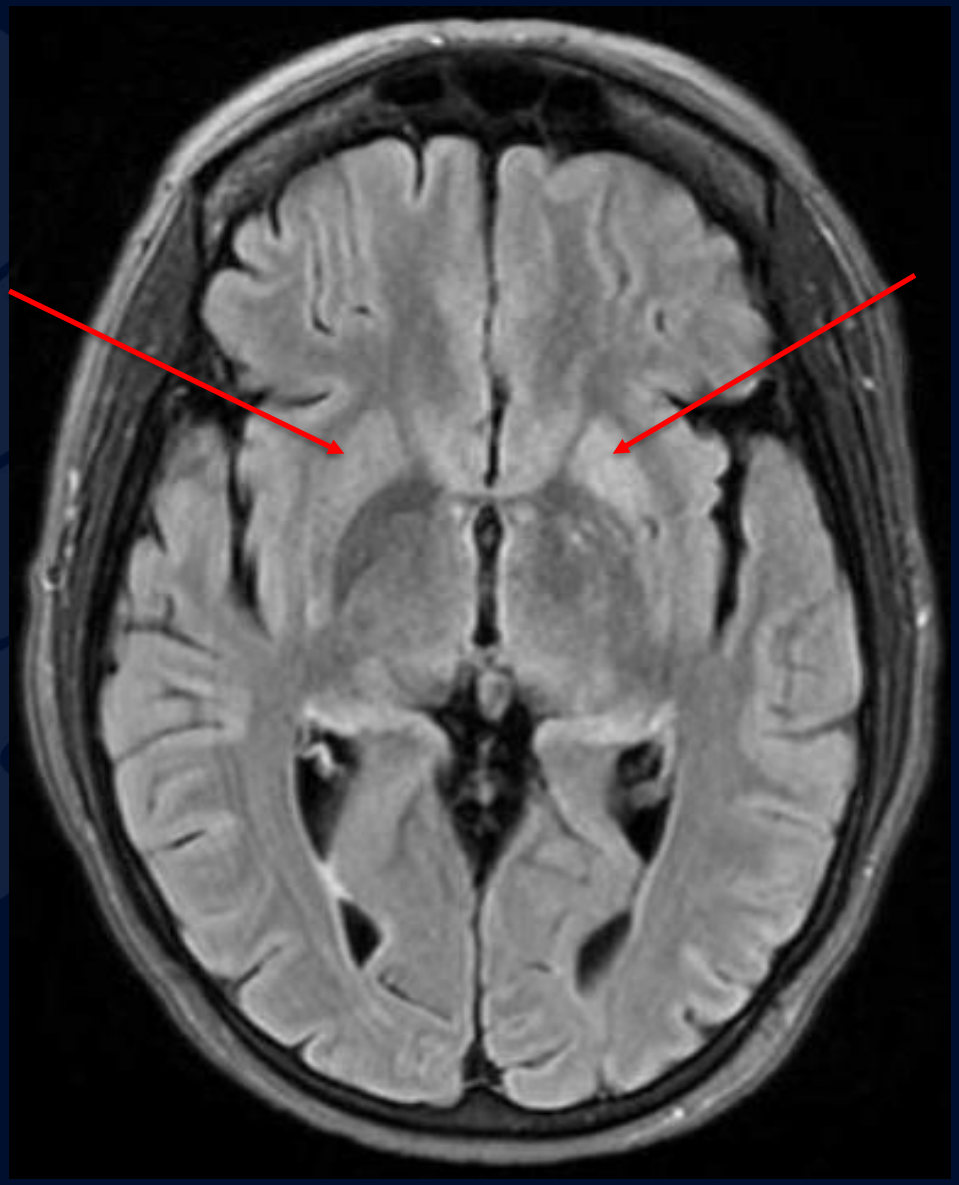
Right putamen less hyperintense than left



Left putamen more hyperintense than right

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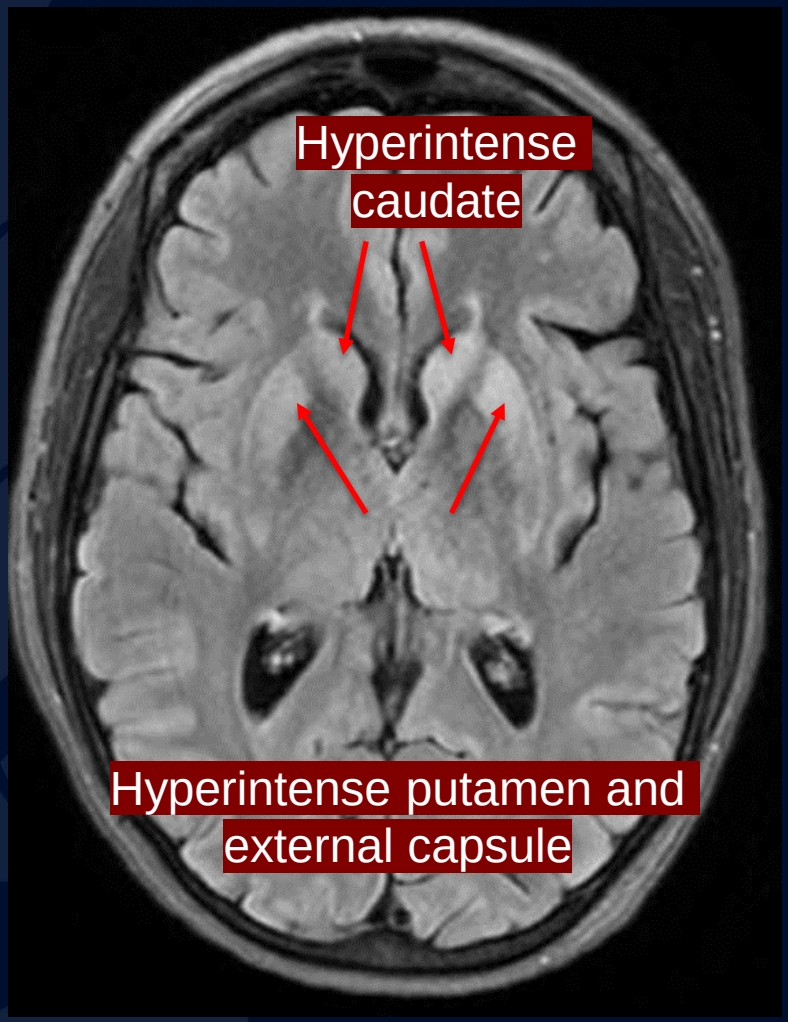

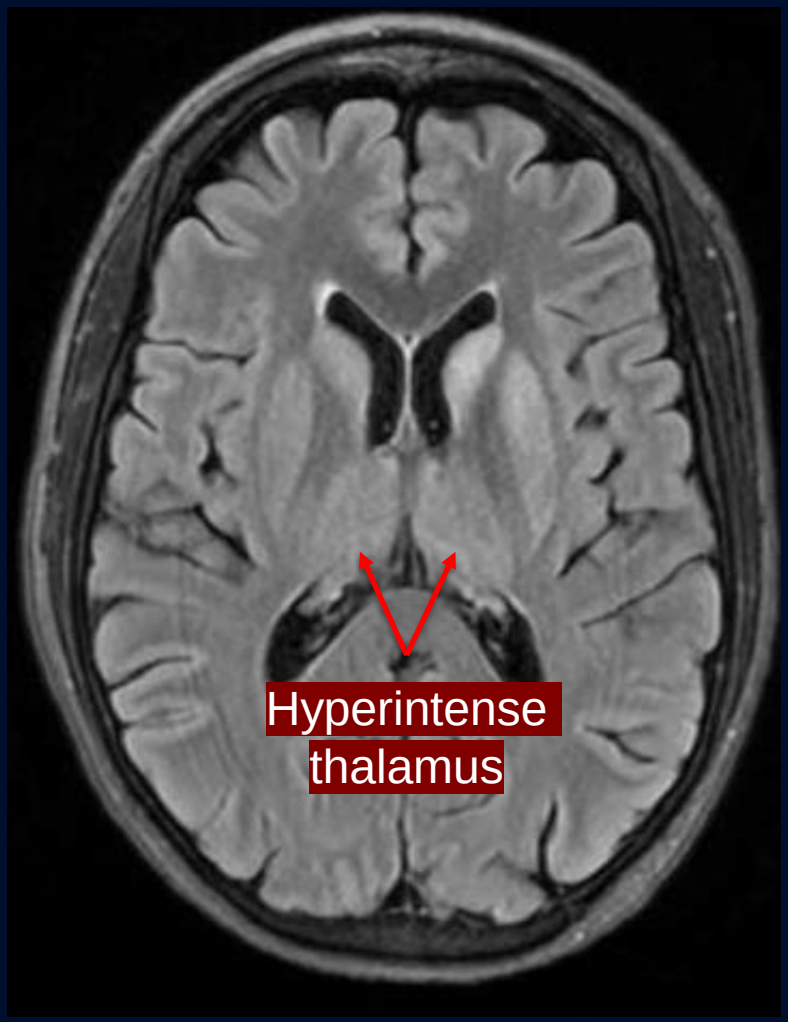

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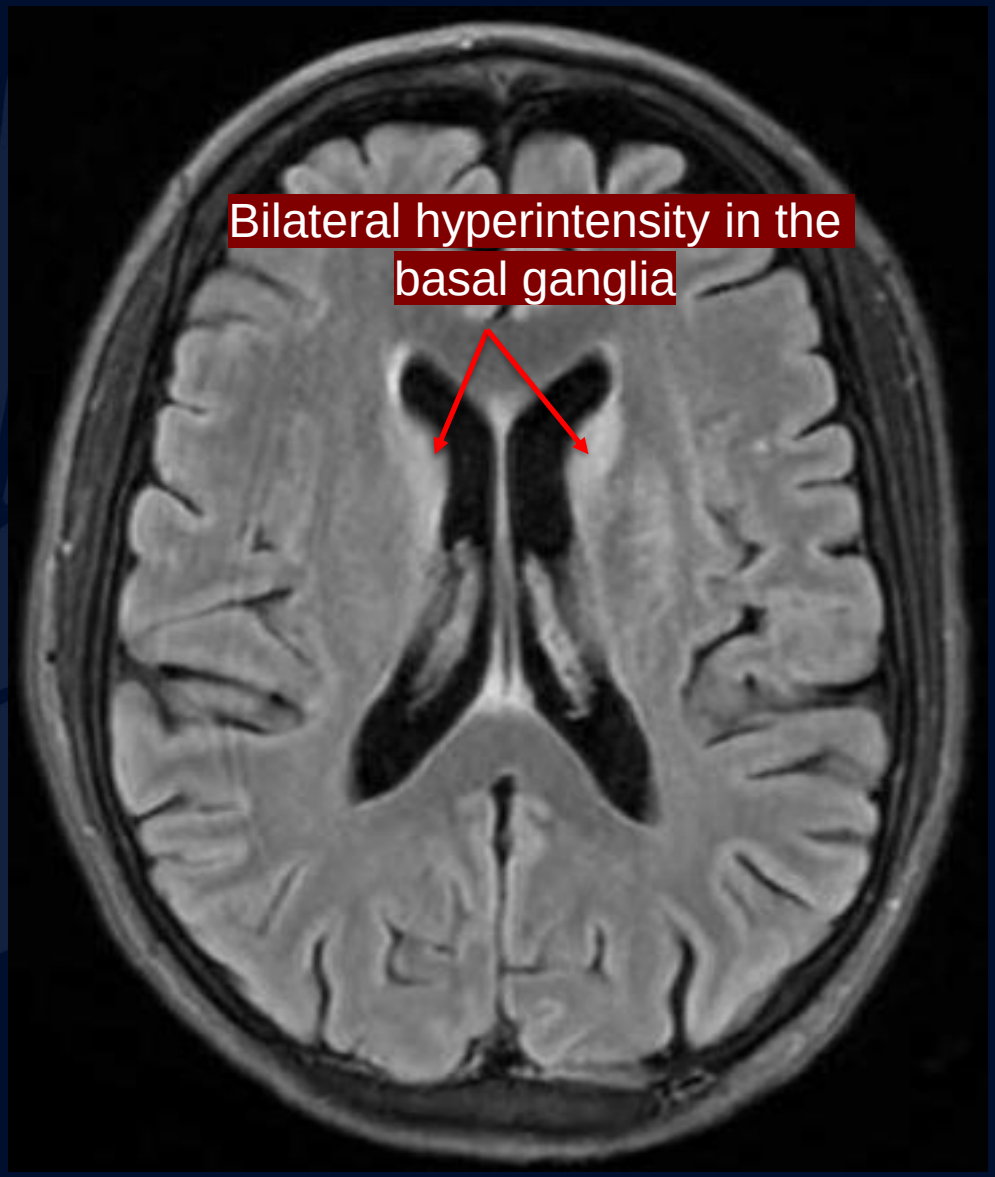

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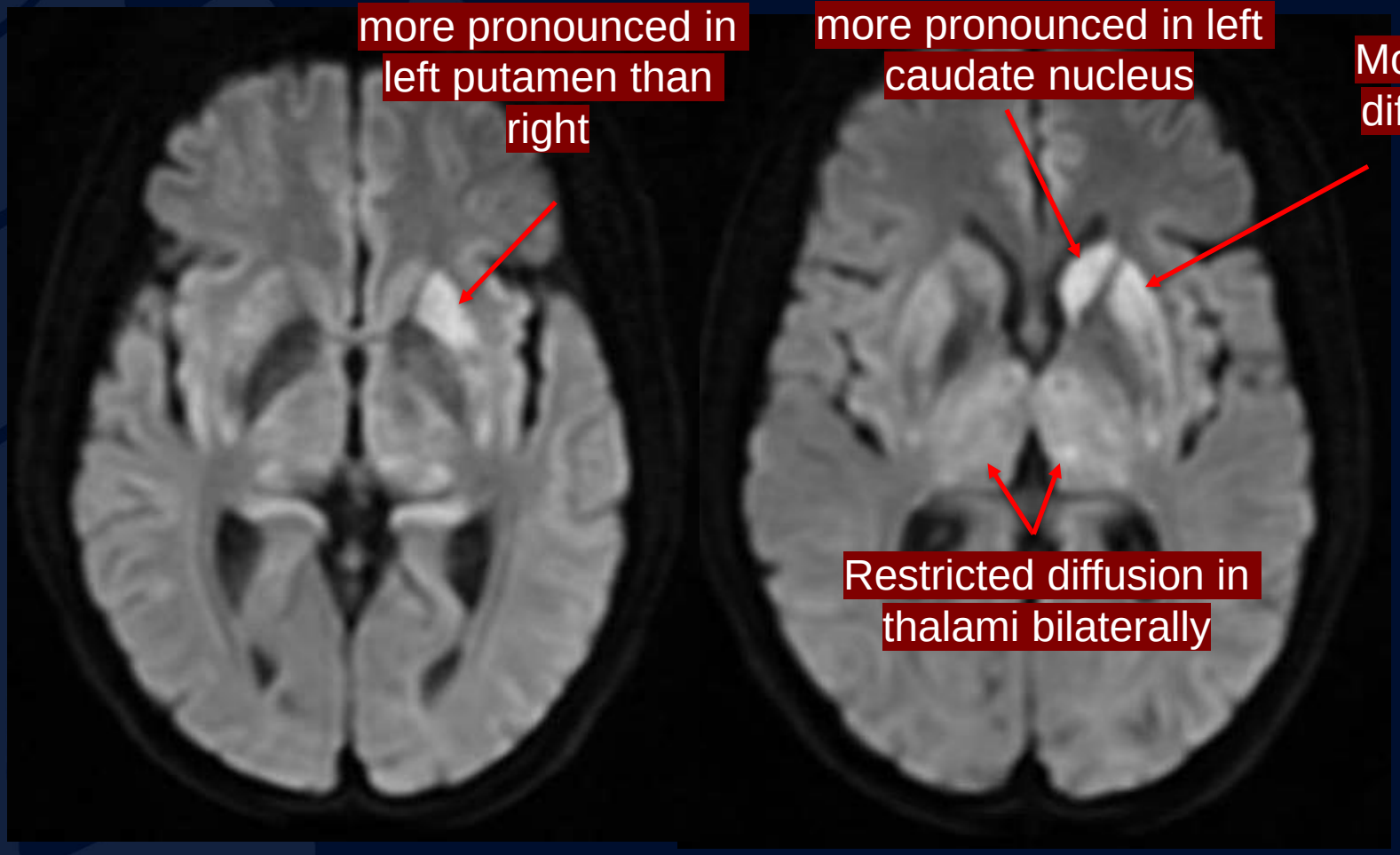

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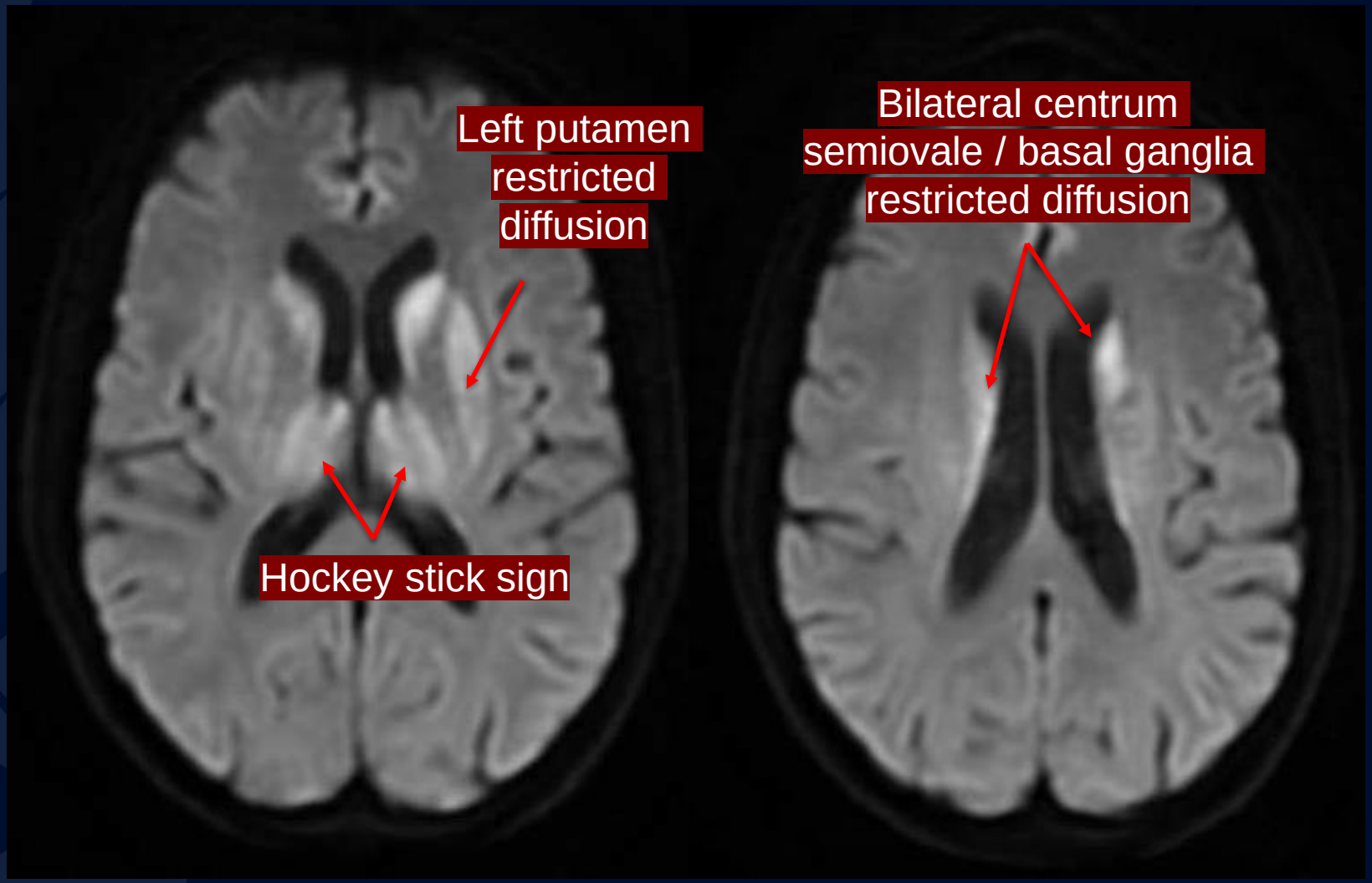

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Cruetzfeldt Jakob Disease (CJD)

- 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 thalami
  - 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

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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 thalami
  - Seen on DWI, FLAIR and T2 weighted MRI

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