# Síndrome de Rasmussen

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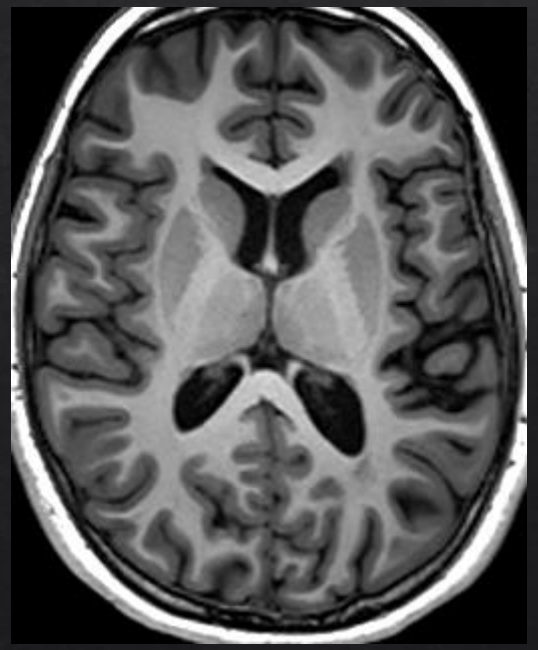
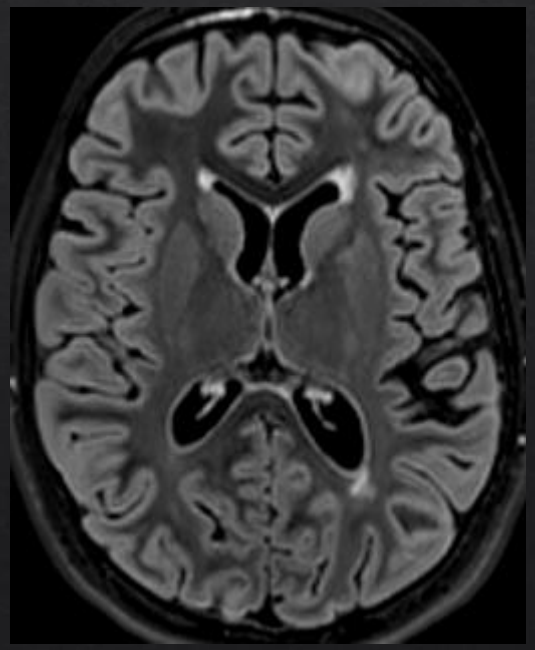
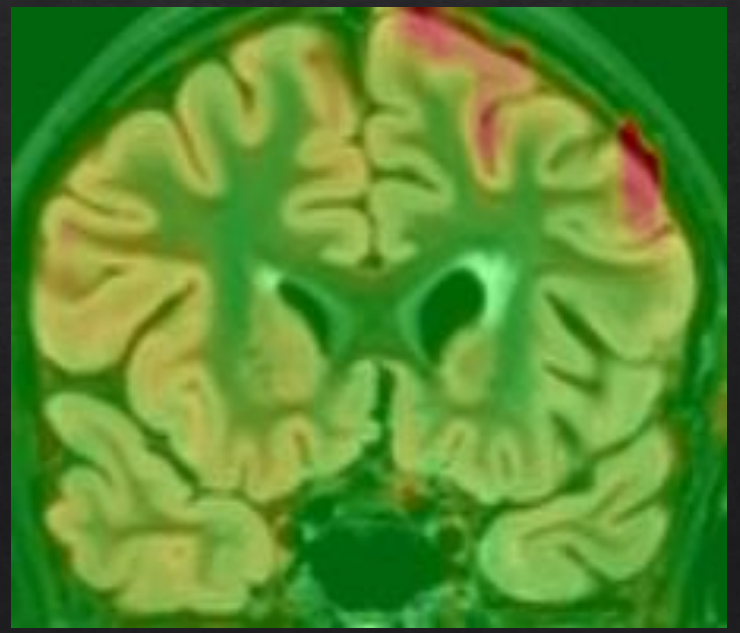




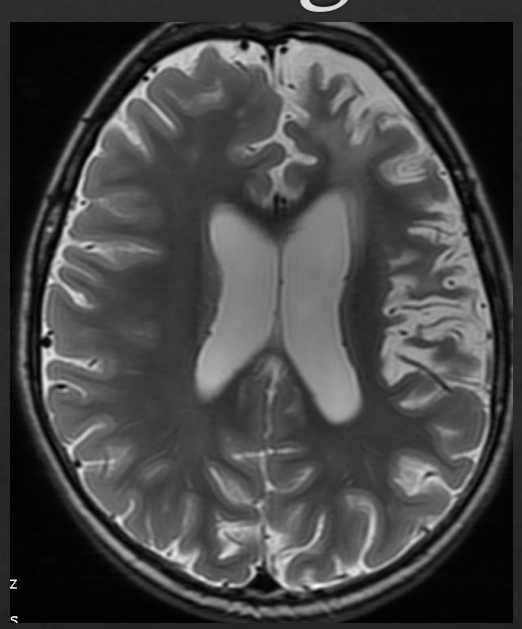


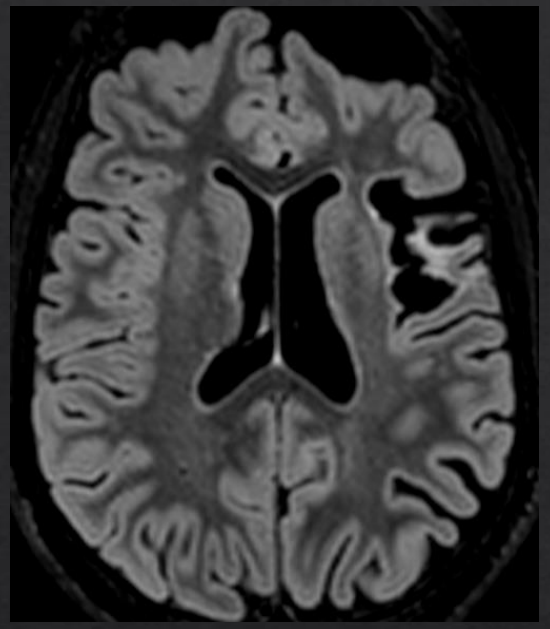
# Resumen

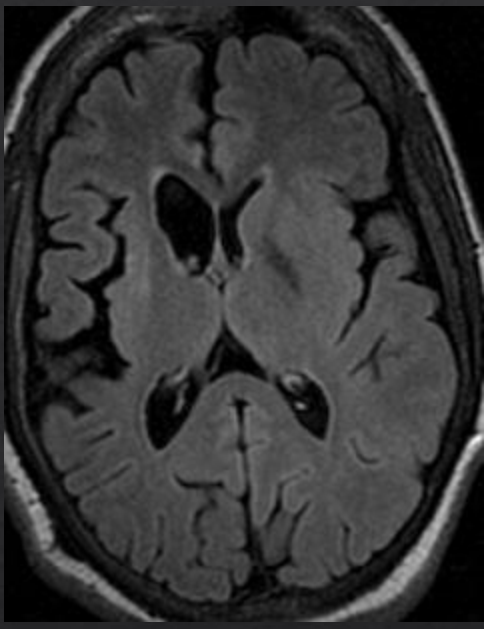
- Síndrome raro (incidencia 2/M), debido a la inflamación unilateral de la corteza cerebral
- Origen autoinmune (Linf T), antecedente de agresión (st viriasis)
- Clínica: Crisis refractarias (CPC frecuentes), déficit (st motor, afasia) y deterioro progresivo
- EEG: ondas lentas con foco(s) ipsilateral
- Imagen: ATROFIA (cortical/estriado) +- Alteración de señal (SG/SB)
- AP: Infilt. linfocitario, nódulos microgliales, astrogliosis. Crónica: datos no concluyentes
- No hay marcadores específicos. Criterios de Bien (2005): Cl+EEG+Imagen +-AP
- Tto curativo: Hemisferectomía. Inmunoterapia (Linf T, Inter-L) en desarrollo, paliativa
- Dx PRECOZ: mejora resultados (Q: plasticidad, IT: mejor en fase aguda), previene deterioro
- Nuevos enfoques diagnósticos:
  - Imagen Funcional (PET, ASL)
  - RM 3T: Atrofia cortical leve/progresiva  y lesiones sutiles (SB periventricular, G Pálido),

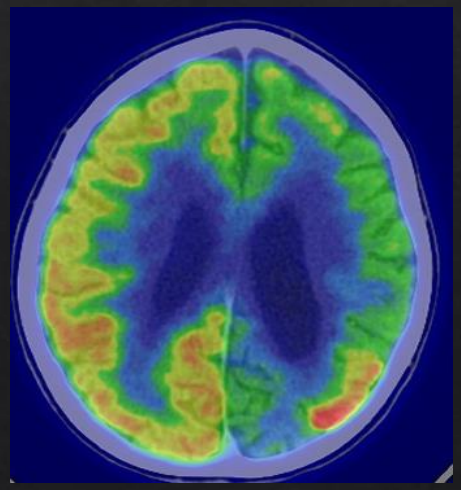
# Criterios Diagnósticos

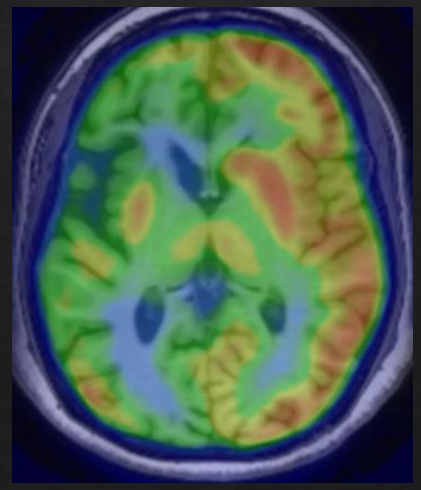
- Parte A (3 de 3)
  - Crisis focales y déficit clínico
  - EEG: Ondas lentas  y foco ipsilateral
  - Atrofia cortical y uno de los siguientes:
    - Alteración de señal (SG o SB)
    - Lesión de N Caudado
- Parte B (2 de 3)
  - EPC o déficit progresivo
  - Atrofia progresiva
  - Biopsia positiva











Pathogenesis, Diagnosis and treatment of Rasmussen encephalitis; a European Consensus Statement
Bien et al **Brain 2005**

No entra en criterios. Complemento?

# Imagen en Rasmussen

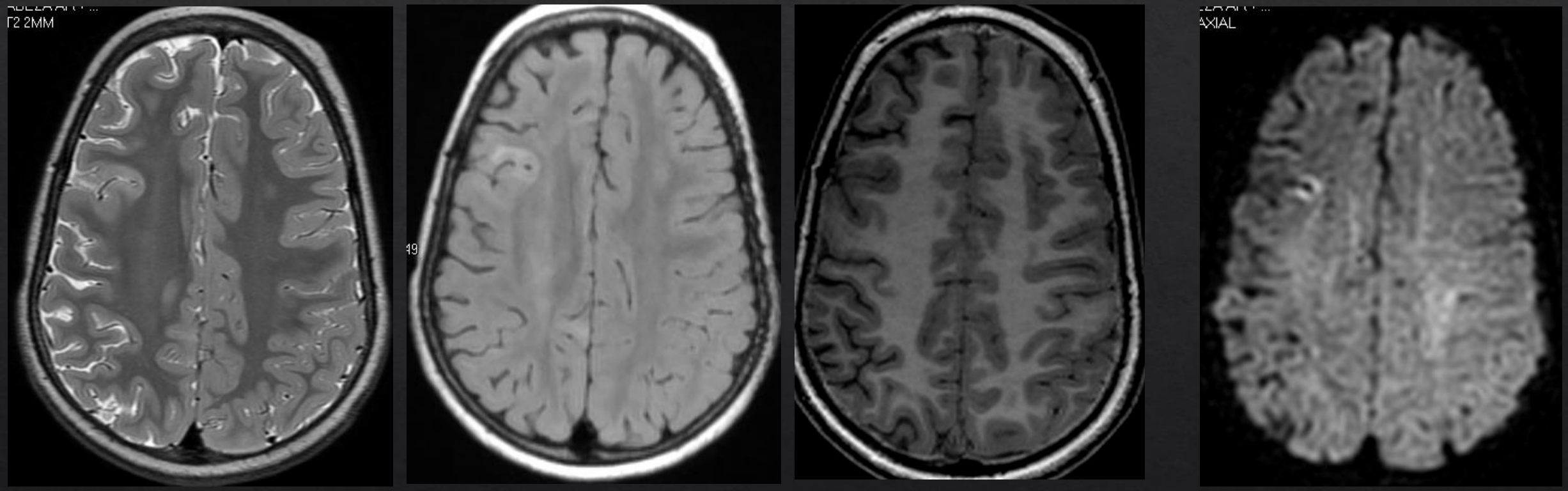
## Formas típicas

- Fase prodrómica (crisis aisladas, poco déficit)
  - Edema cortical (DDx con DCF), +- atrofia leve
- Estadio Agudo (Crisis refractarias/CPC, déficit progresivo)
  - Atrofia (> esp subaracnoideo/ventrículo)
    - Perisilviana
    - Caudado (Putamen)
  - Alteración de señal
    - Sustancia Blanca: YuxtaC, PV, profunda
    - SG: Edema, alt hipocampo (DDx con ETM)
  - Patrones de afectación
    - Giral Difuso: predominio perisilviano/frontal
    - Focal: aspectp geográfico (DDx con infarto)
- Estadio de Secuela (menos crisis, deterioro establecido)
  - Atrofia y menor alteración de señal
  - Atrofia contralateral: marcador de deterioro cognitivo

## Formas atípicas

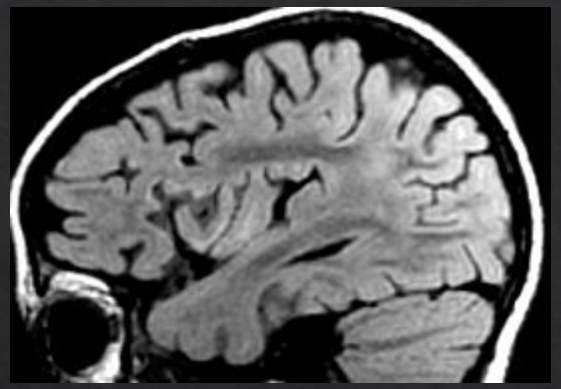
- "Adulto" (10%)
  - Forma menos agresiva (no fase prodrómica)
  - Más lesiones "extrainsulares"
  - Mejor respuesta a IT
- Presentación atípica
  - No déficit inicial
  - Crisis Tardías (hasta 2 años después del inicio)
  - Movimientos anormales (lesión aislada de Estriado)
- Lesión Dual (10%)
  - DCF (tipo IIId ?)
  - Lesiones inespecíficas: Trauma, Vascular
- Atrofia Bilateral
  - La inflamación bilateral es excepcional
  - Secundaria : Excito-toxicidad, Ttos

# Fase aguda

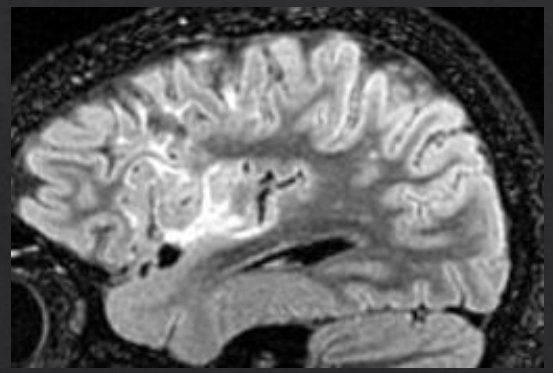



Engrosamiento cortical con alteración de señal, poca atrofia

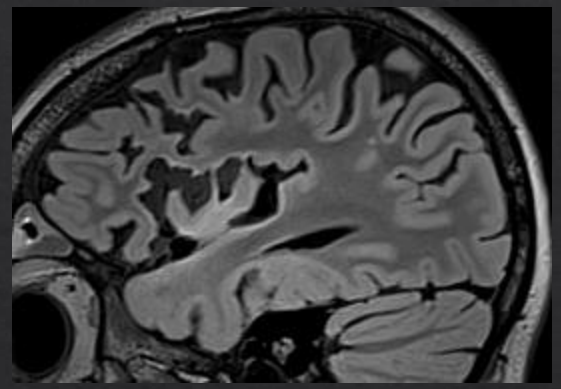
# Evolución en “brotes”



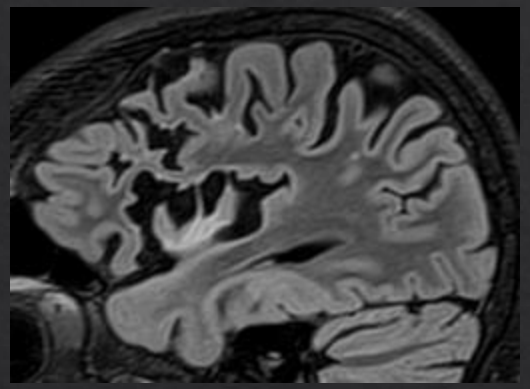
2011



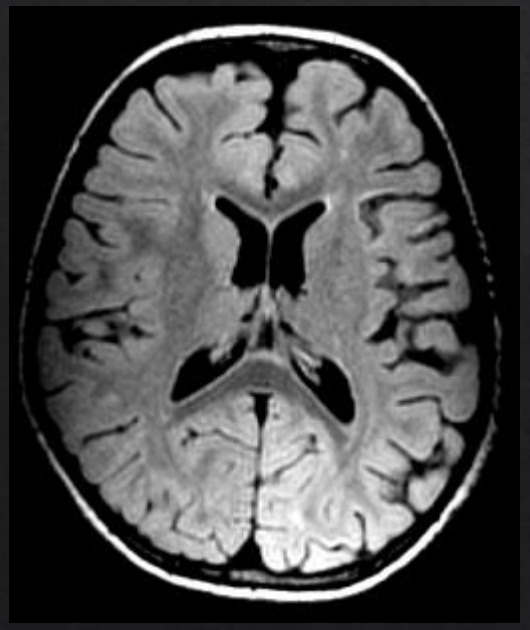
2015

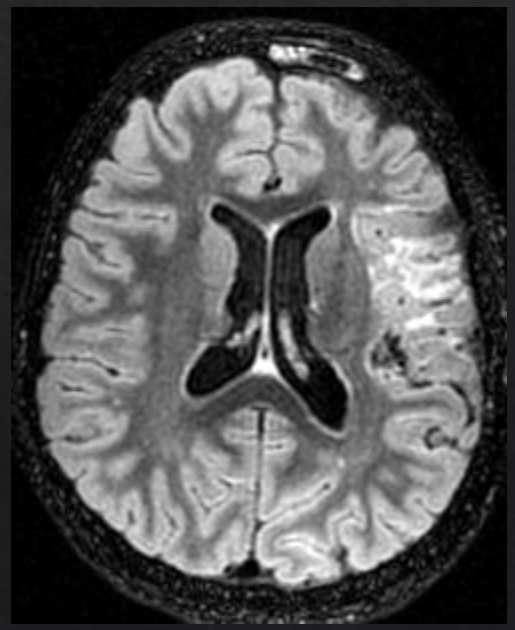


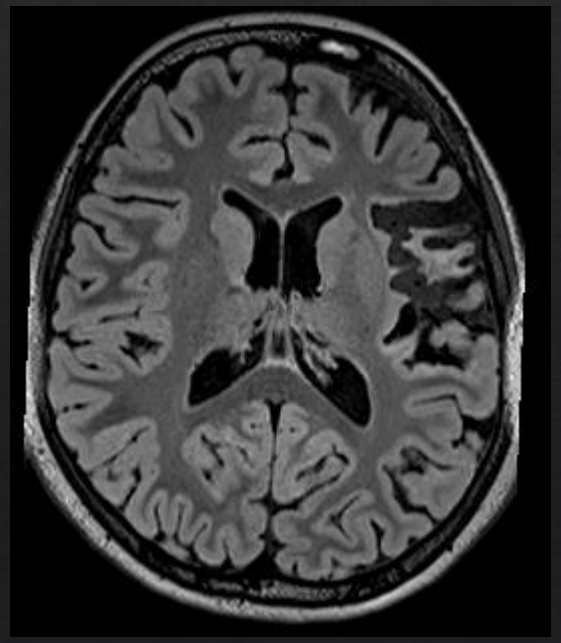
2017

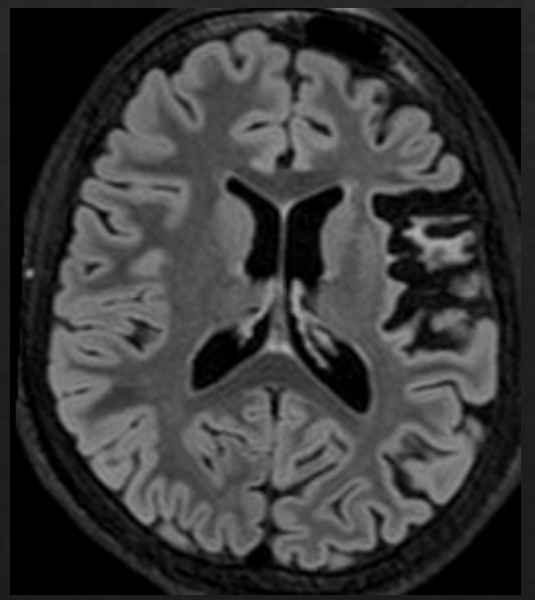


2019

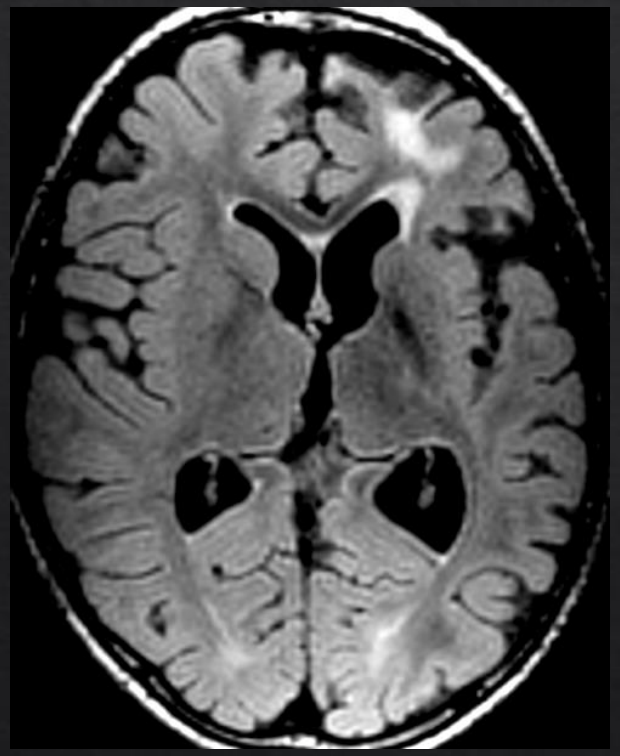


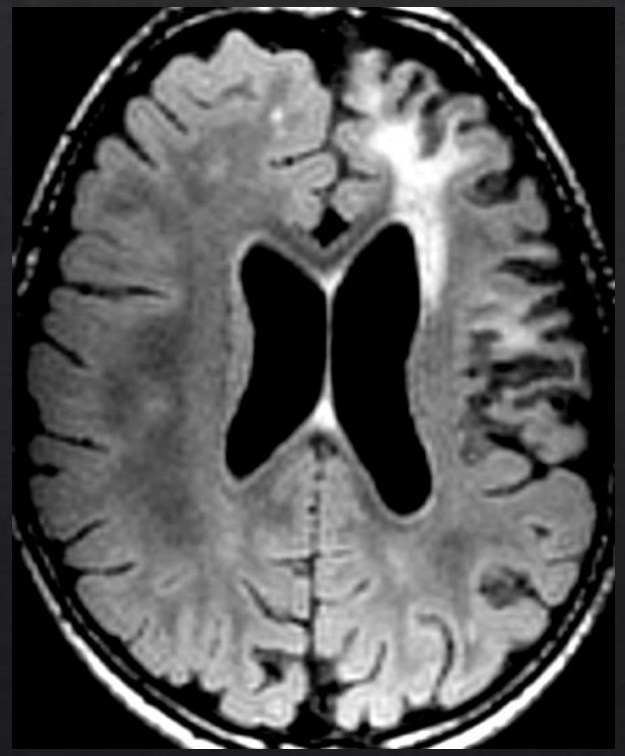


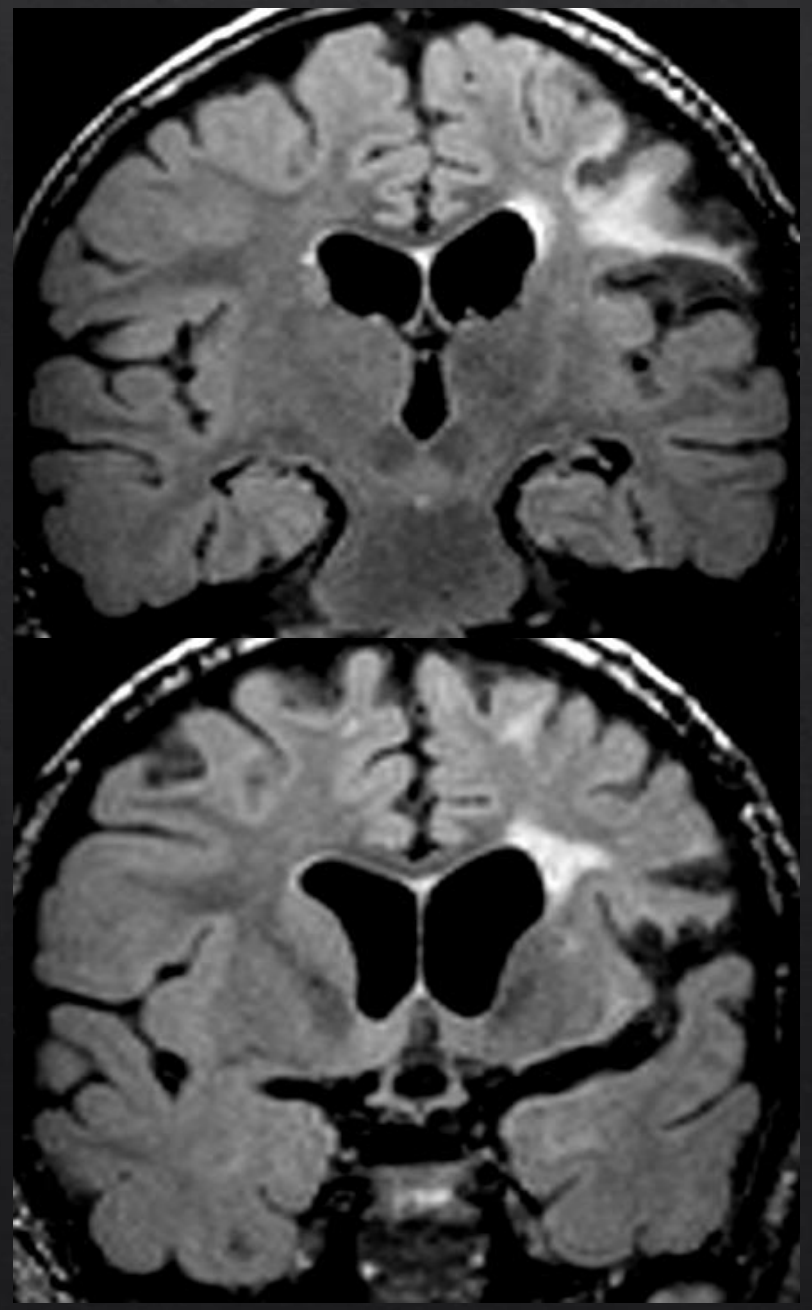




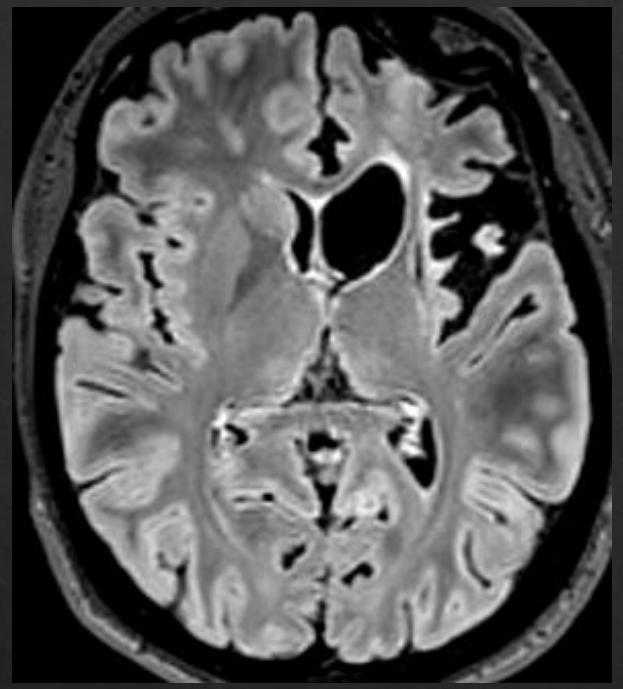
# Hiperseñal SB

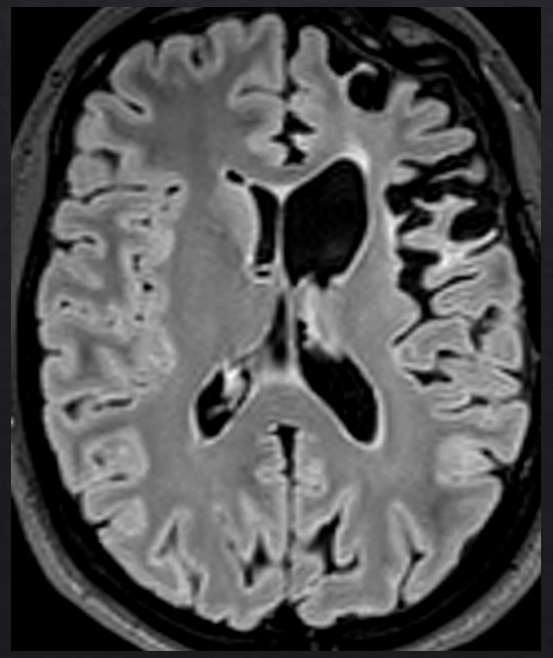


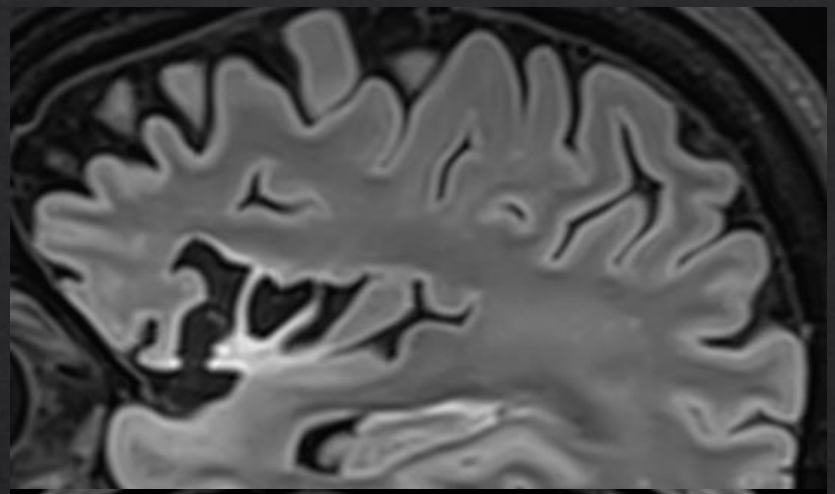


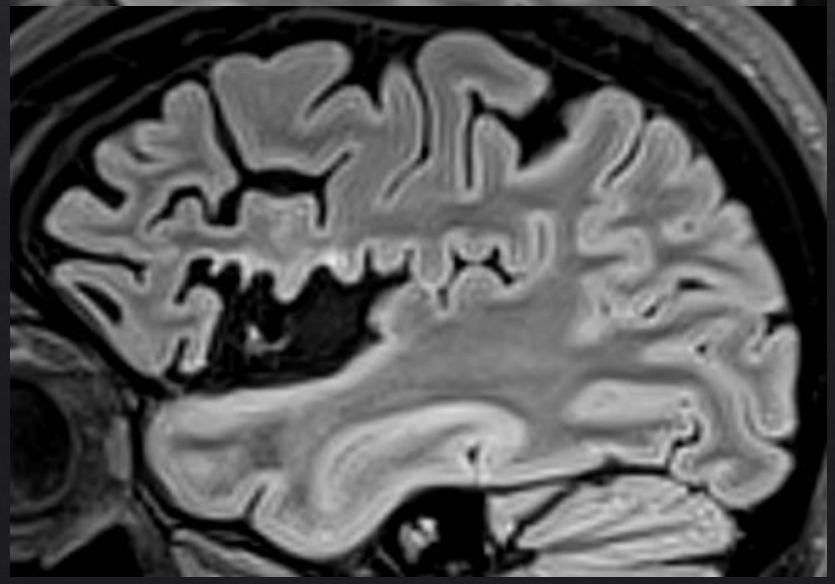


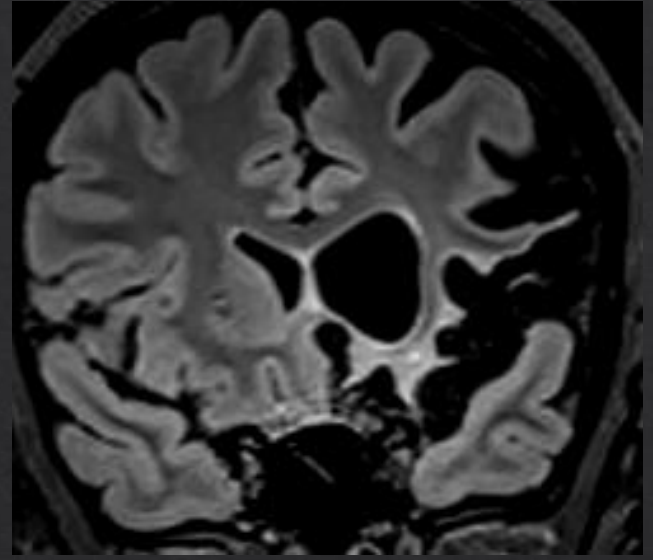
# Hiperseñal SG

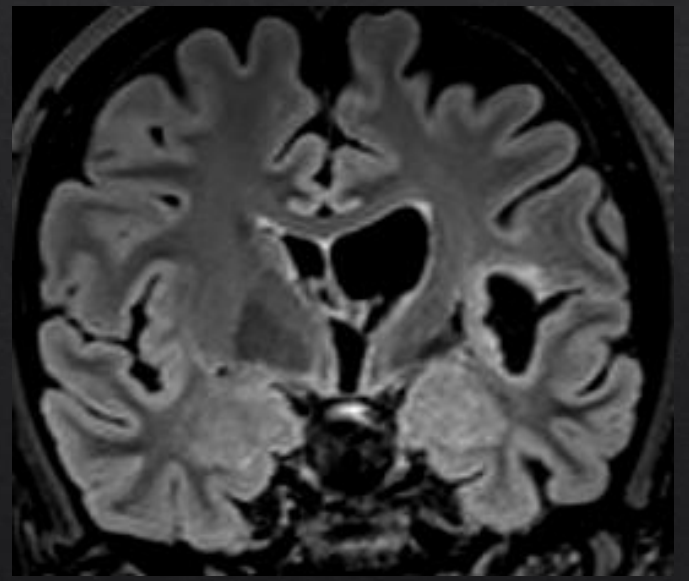




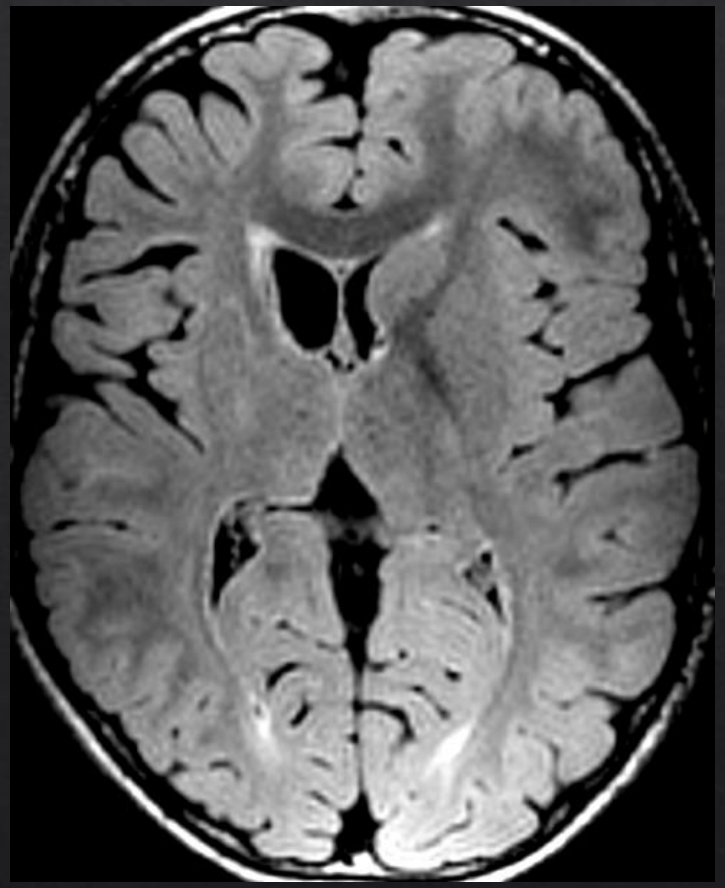




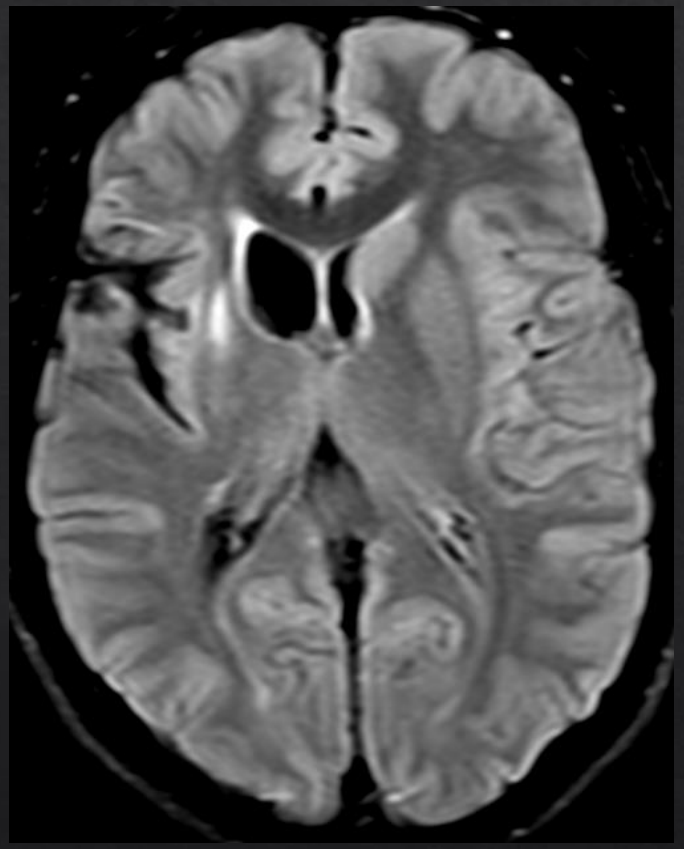




# Lesión del C Estriado

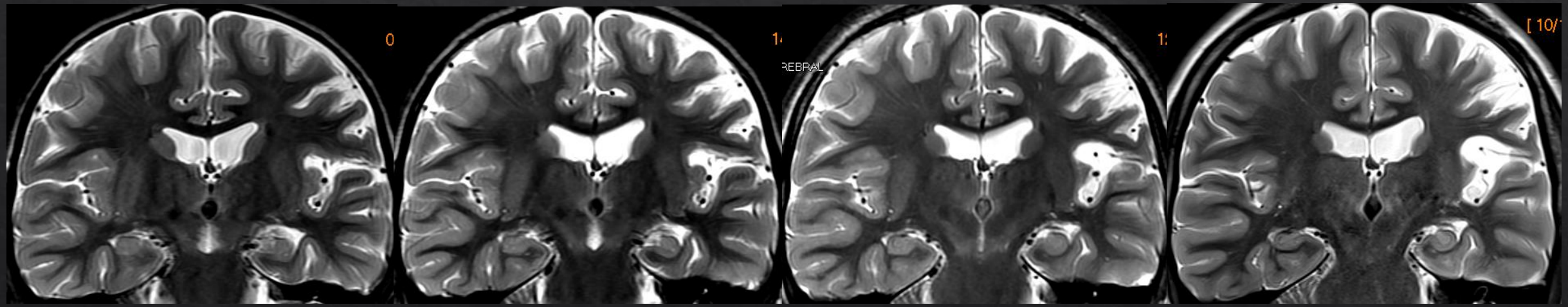


2011

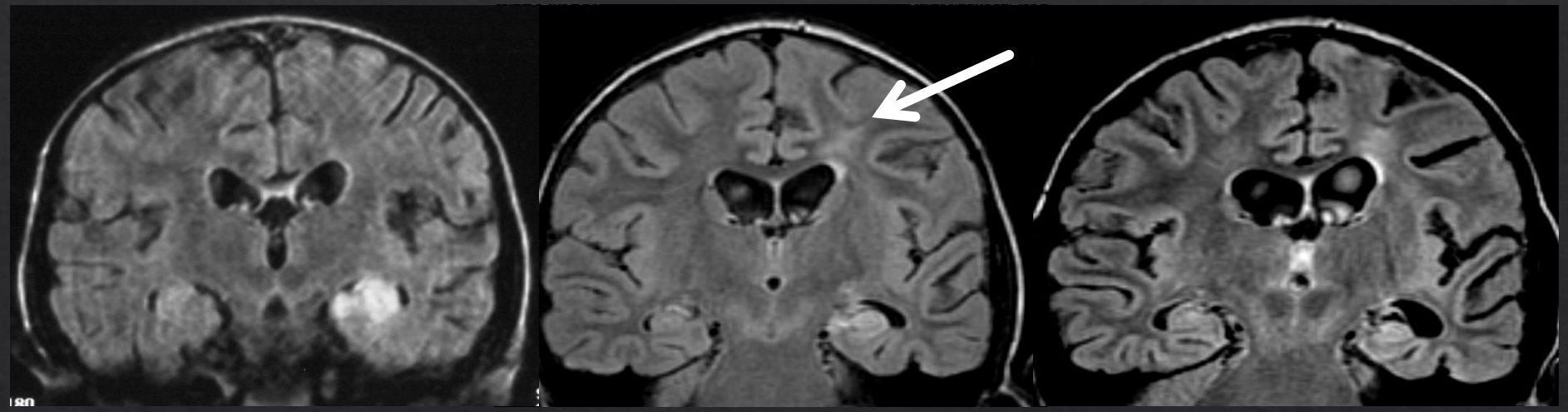


2020

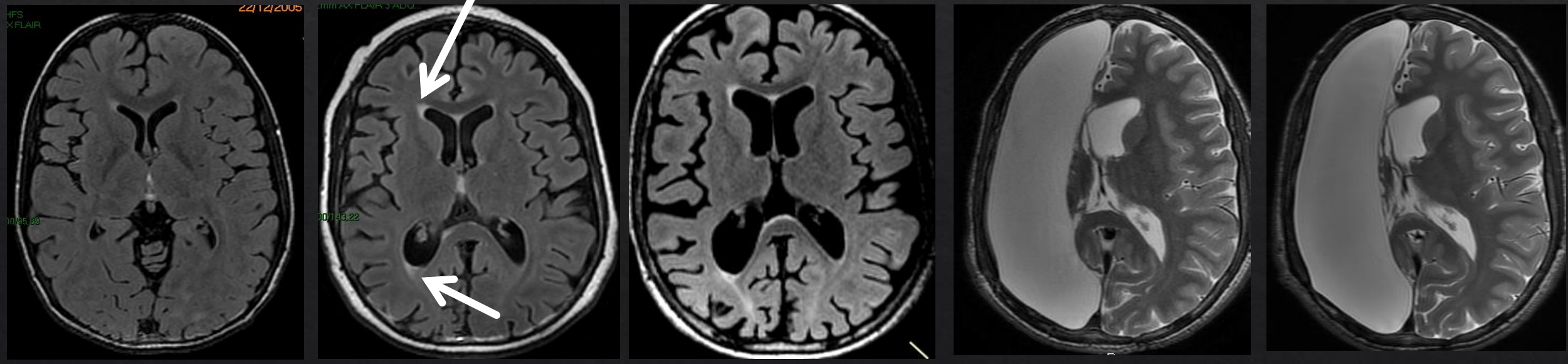
# Atrofia progresiva



# Atrofia Bilateral



11/02/05 3/03/05 11/06/05

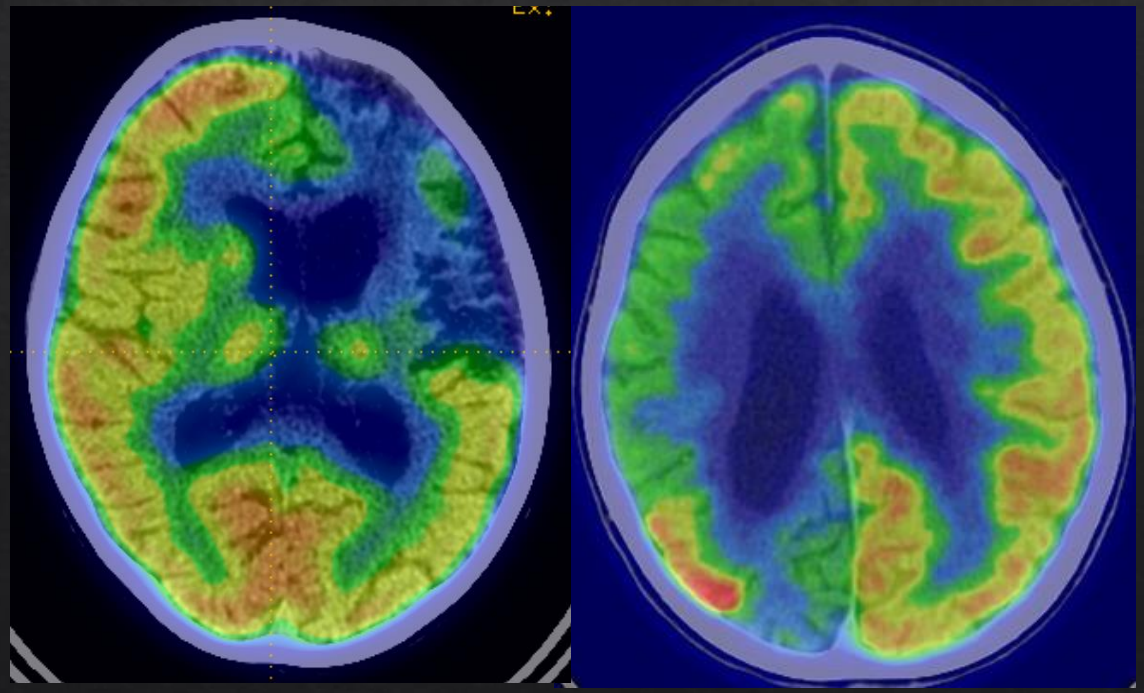


2005 2007 2009 2010 2011

La atrofia contralateral es un marcador de deterioro cognitivo y se detiene tras la desconexión

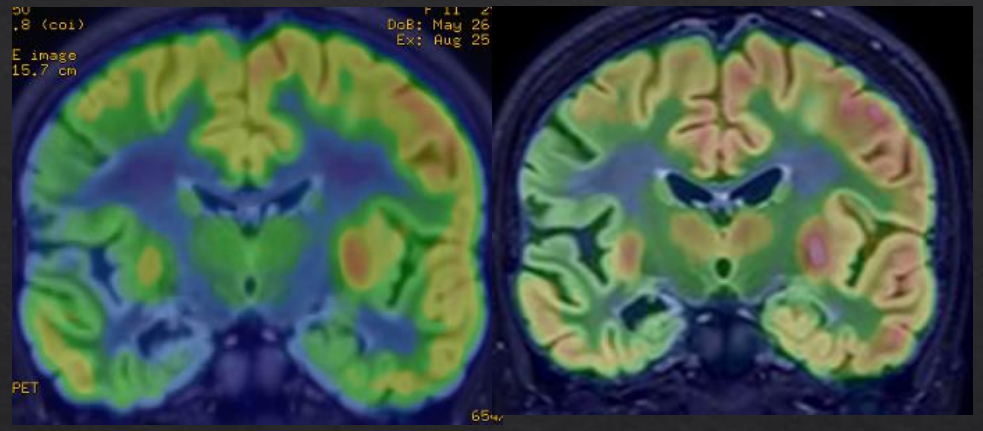
# Imagen Funcional

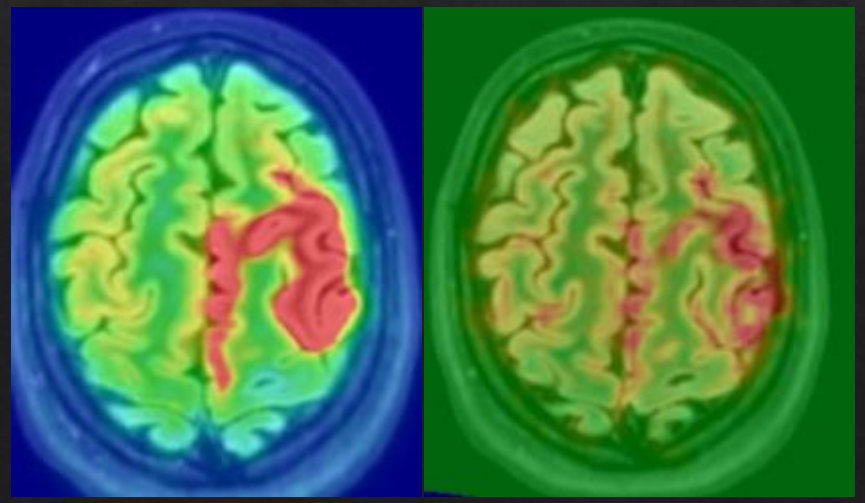
- Complemento de imagen estructural
  - Cuando hay poca atrofia
  - En presentación atípica



Hipometabolismo más extenso que la atrofia
Hipometabolismo con área hipermetabólica (CPC)

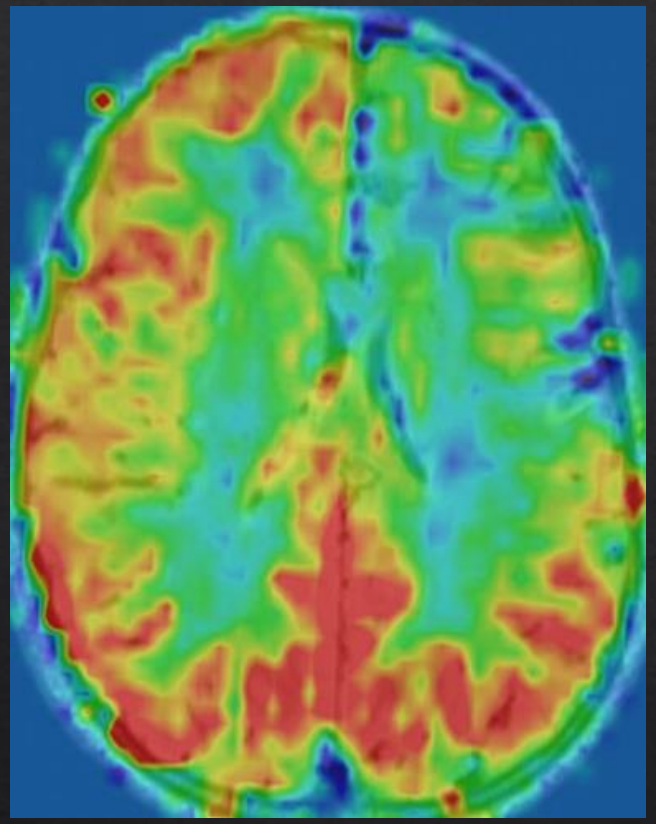
PET vs ASL



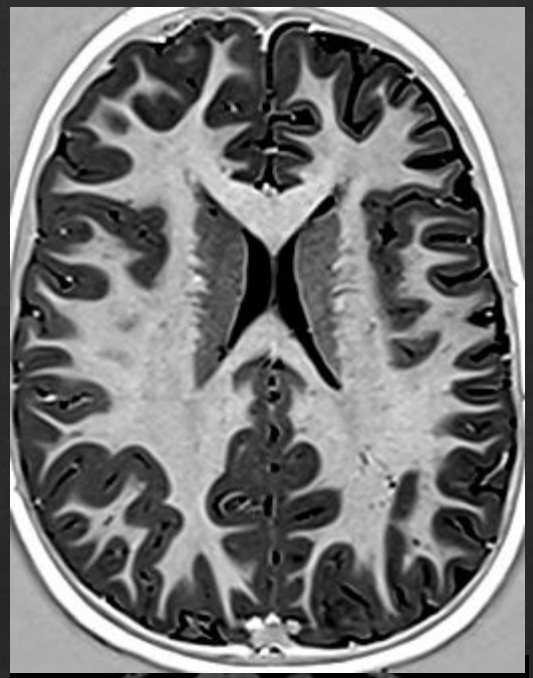



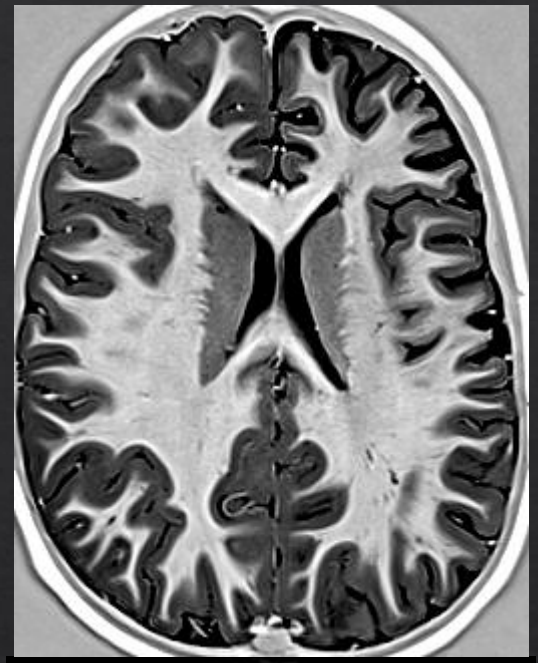
ASL: alternativa st. en seguimiento

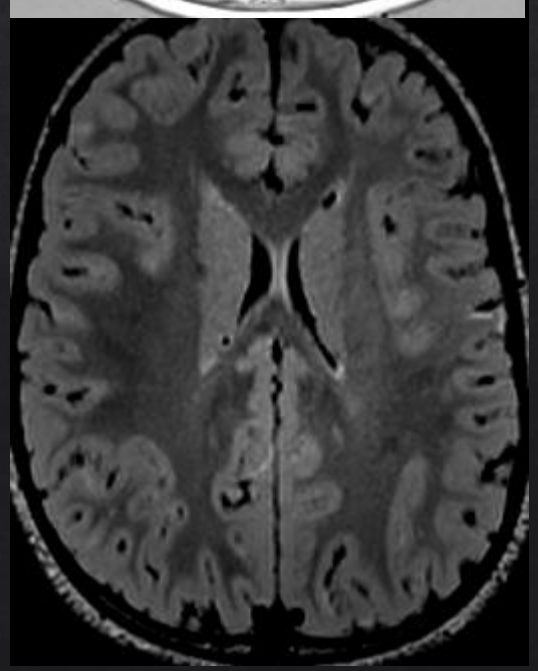
Functional imaging allaws a second look in structural imaging

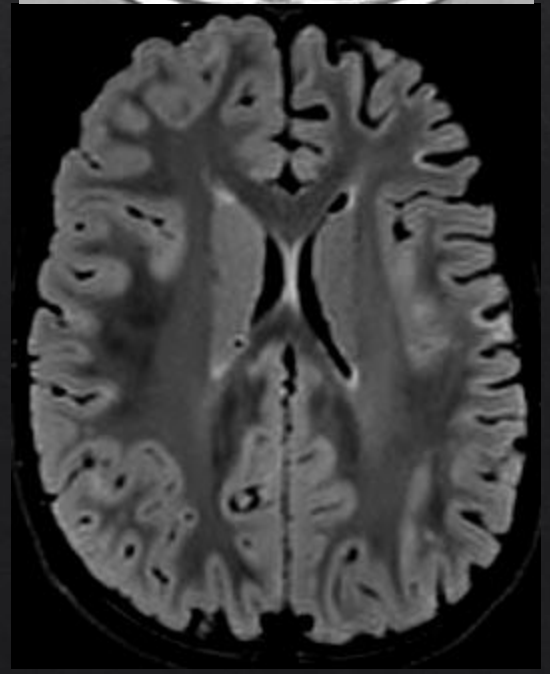


PET/ASL can help to suspect Rasmussen and look for other criteria or recommend  follow up to confirm focal atrophy

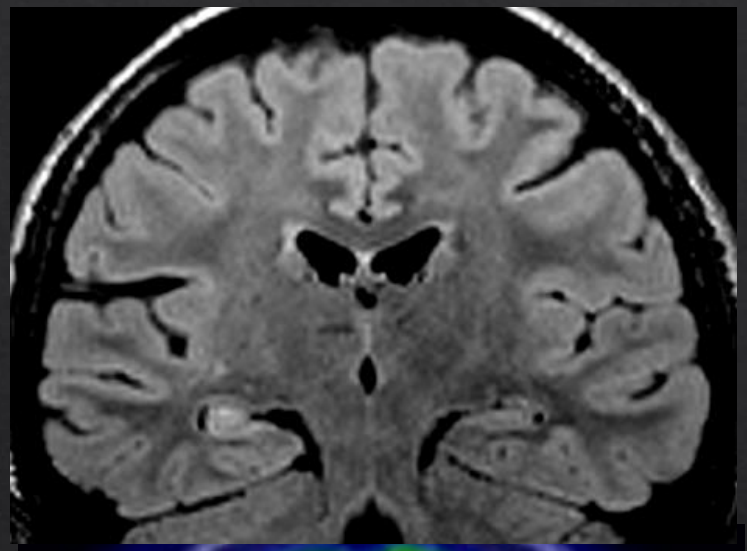


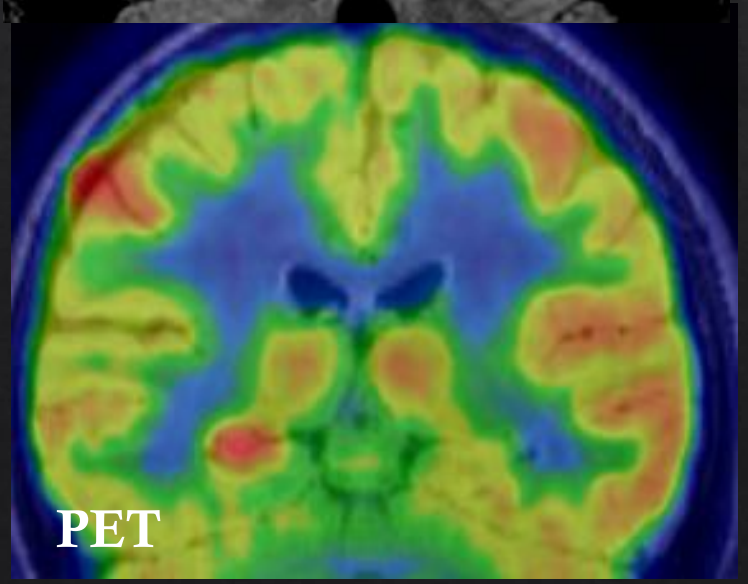




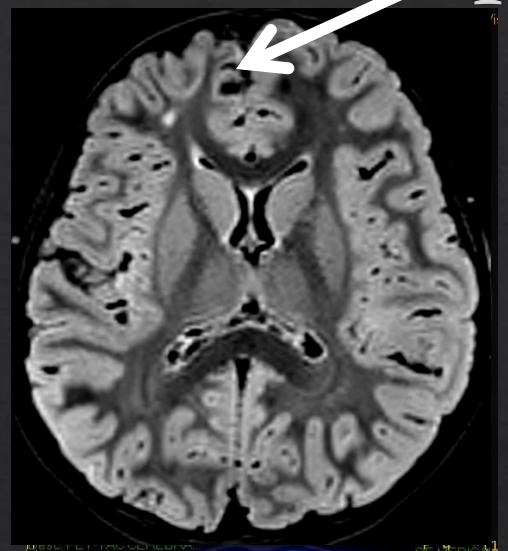


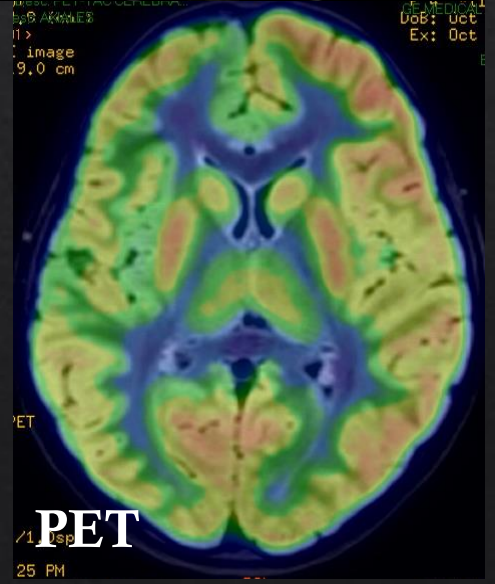
# Localización atípica



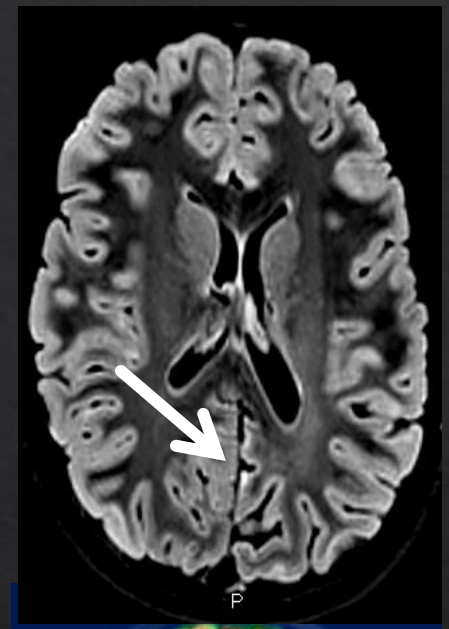


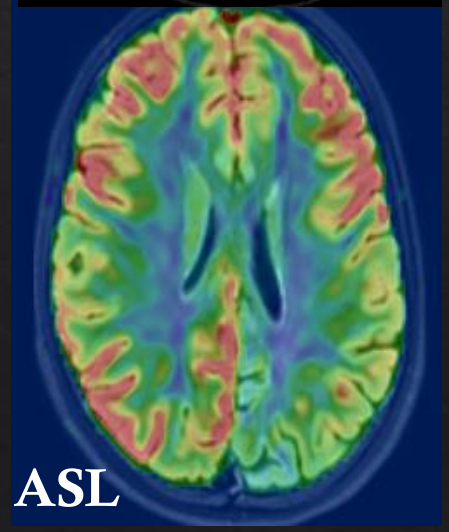

ETM



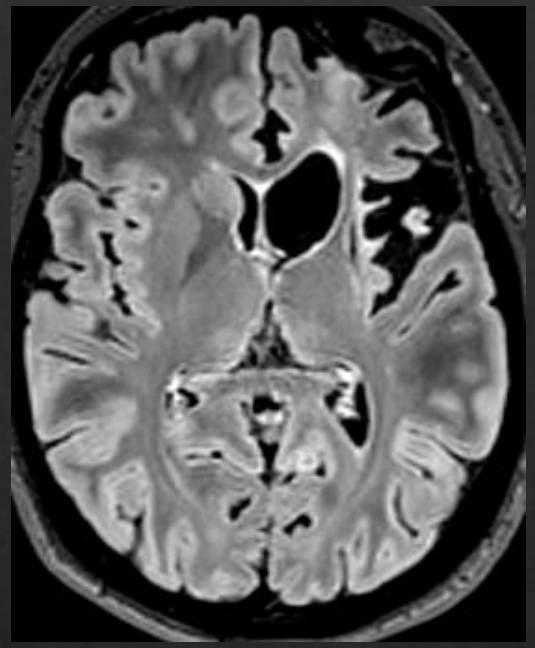


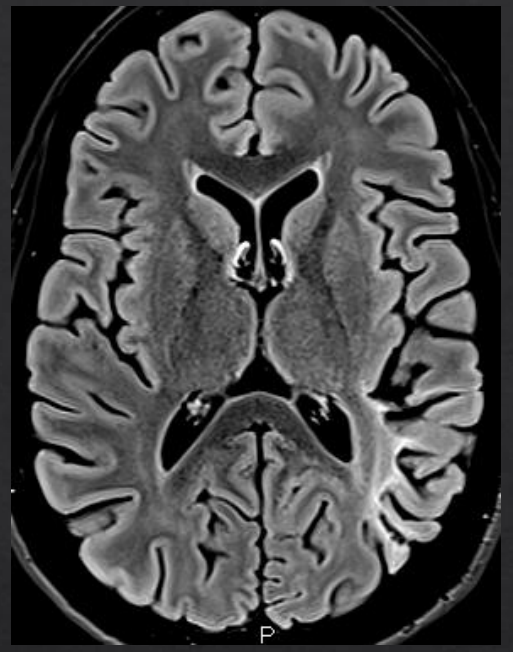

Frontal

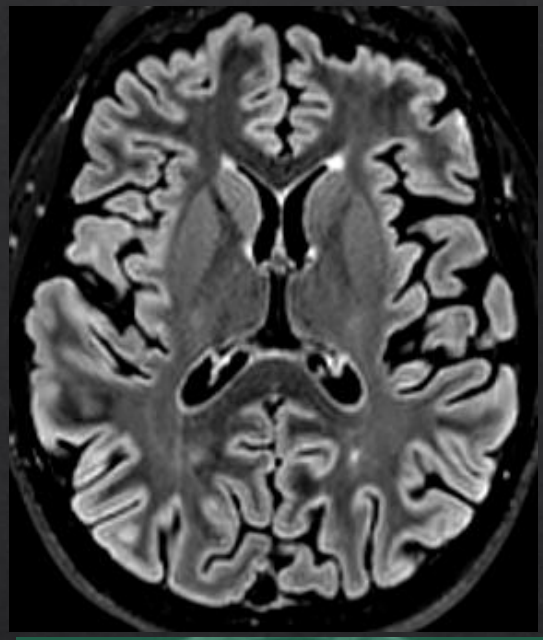


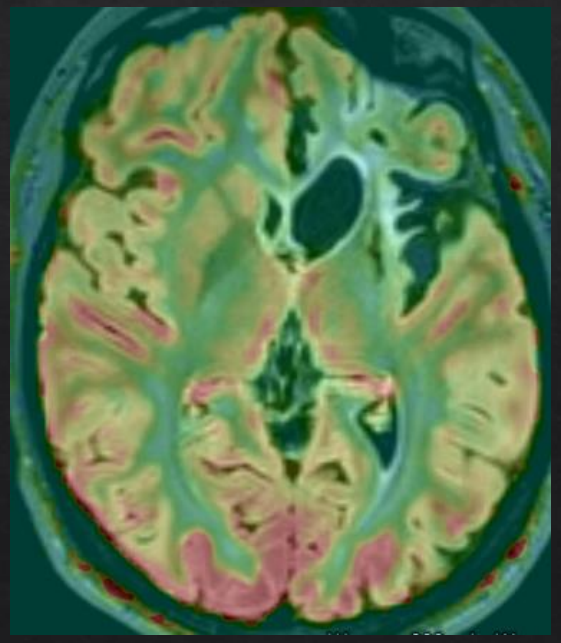



Occipital

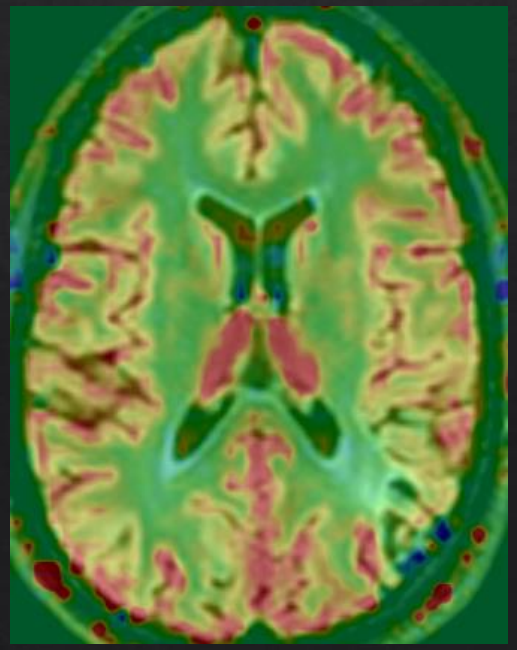




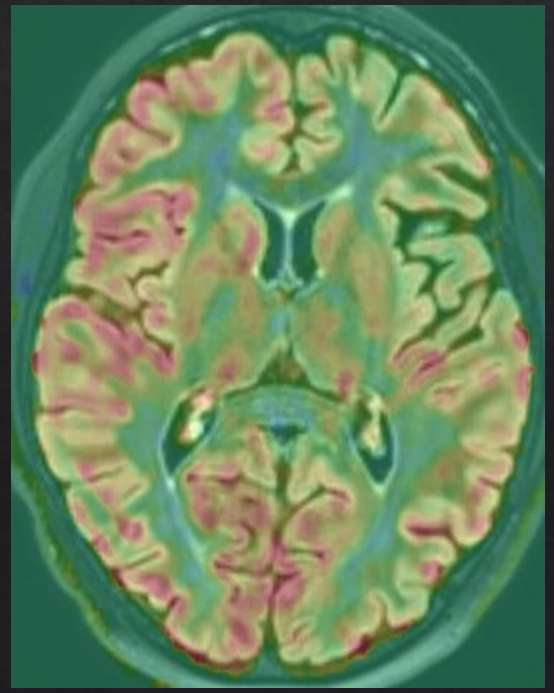




Rasmussen



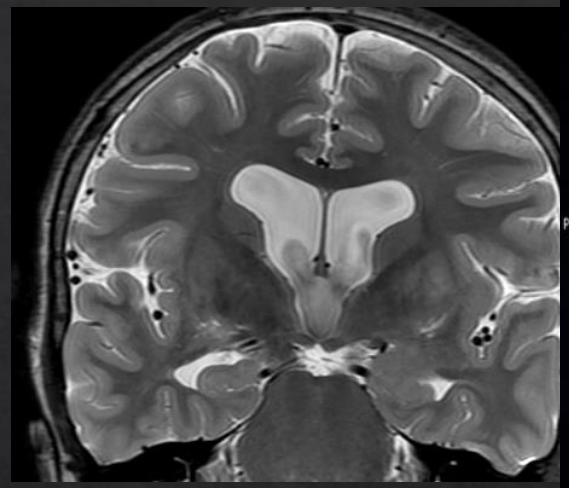
Ulegiria

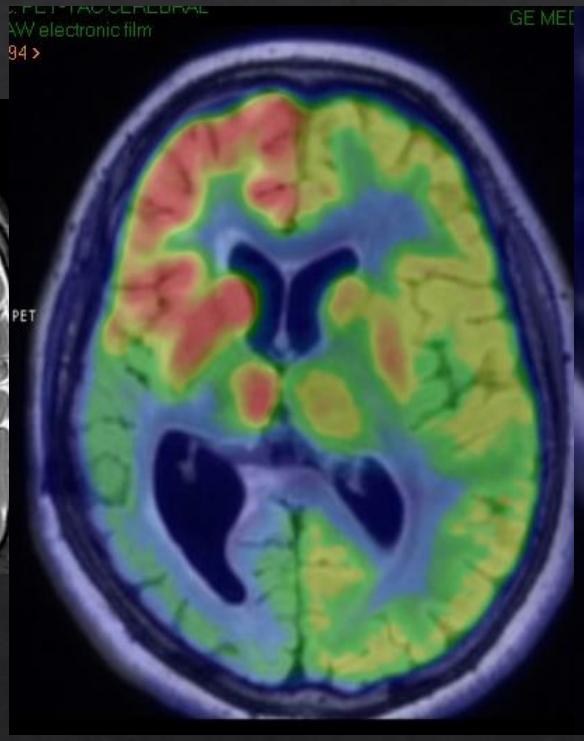


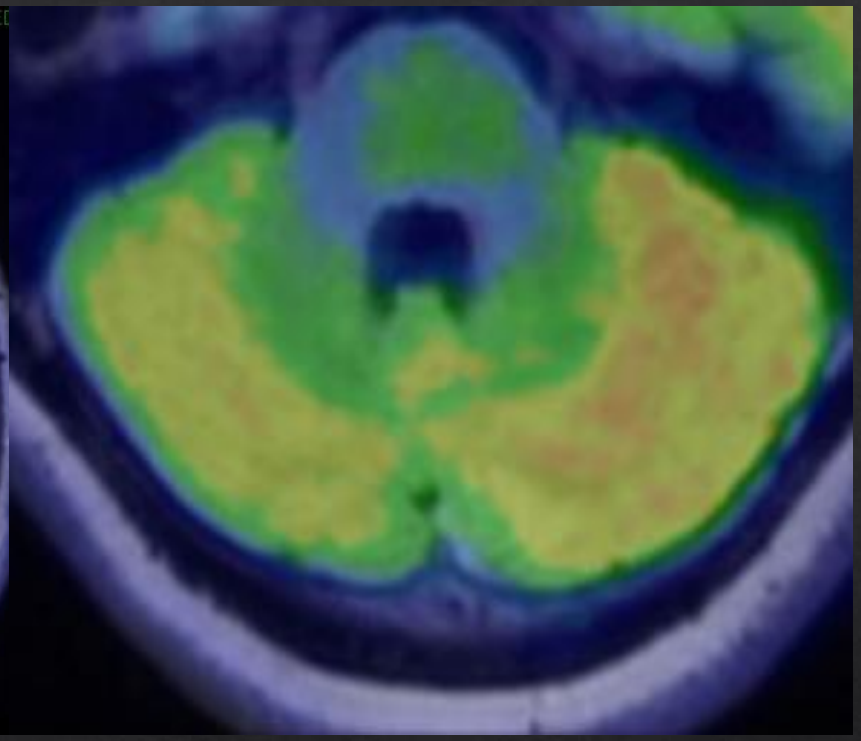
Rasmussen

# Evolución: cambios funcionales

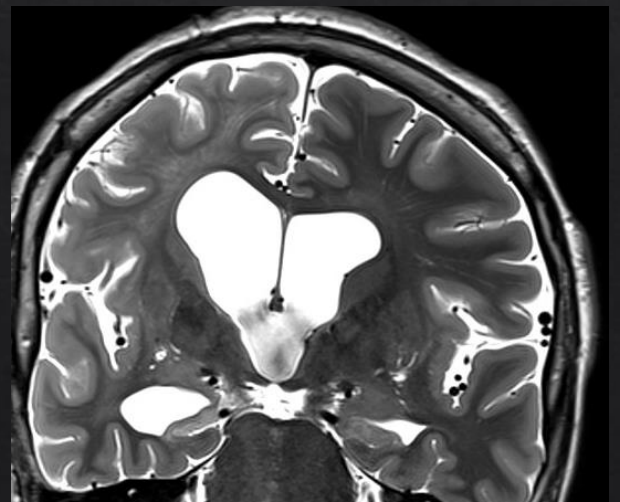
Fase aguda

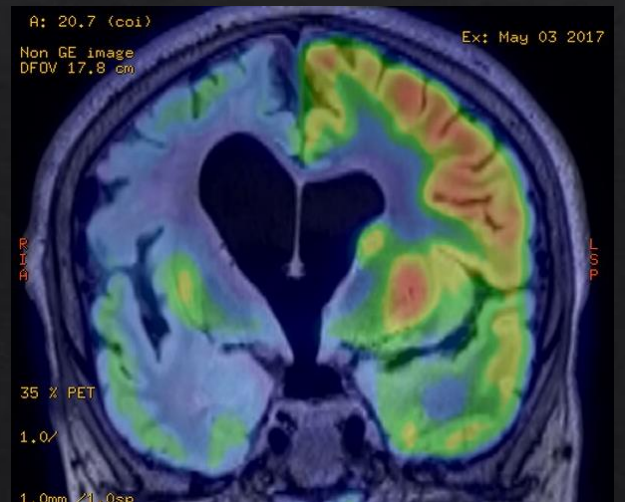


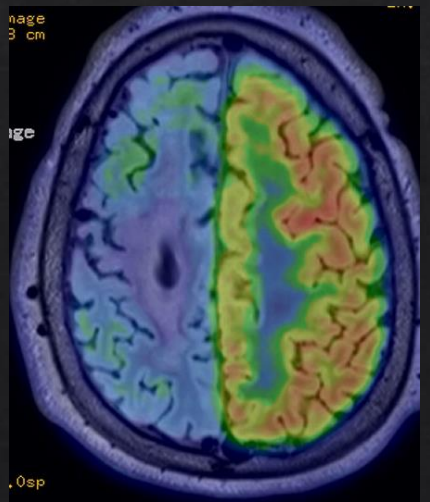


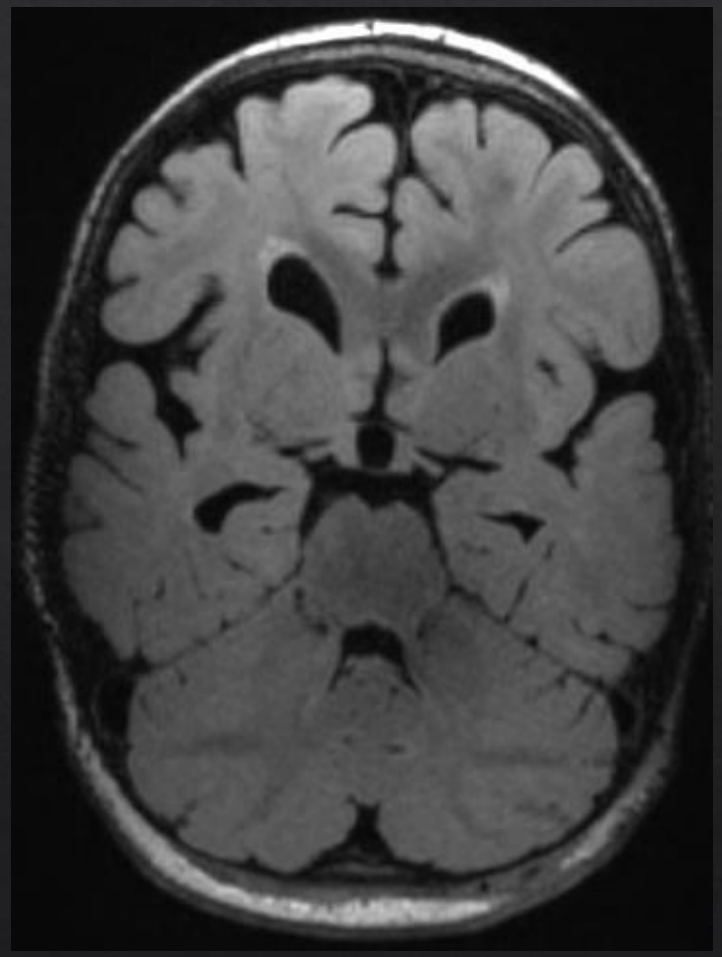
Fase de secuela

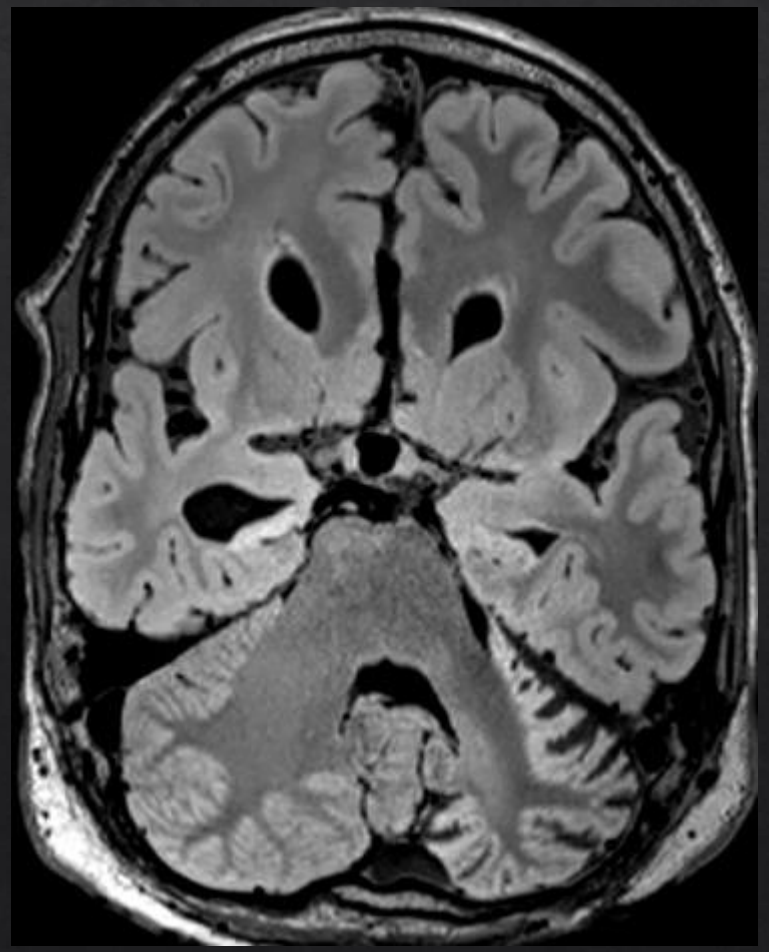




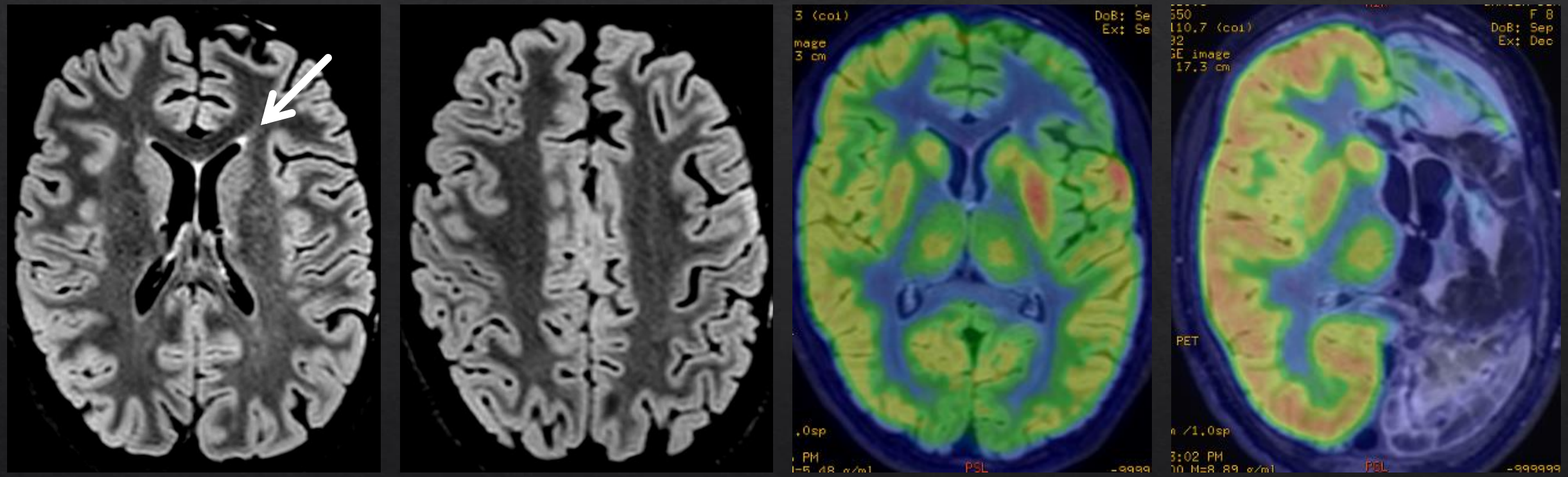



# Evolución: atrofia secundaria





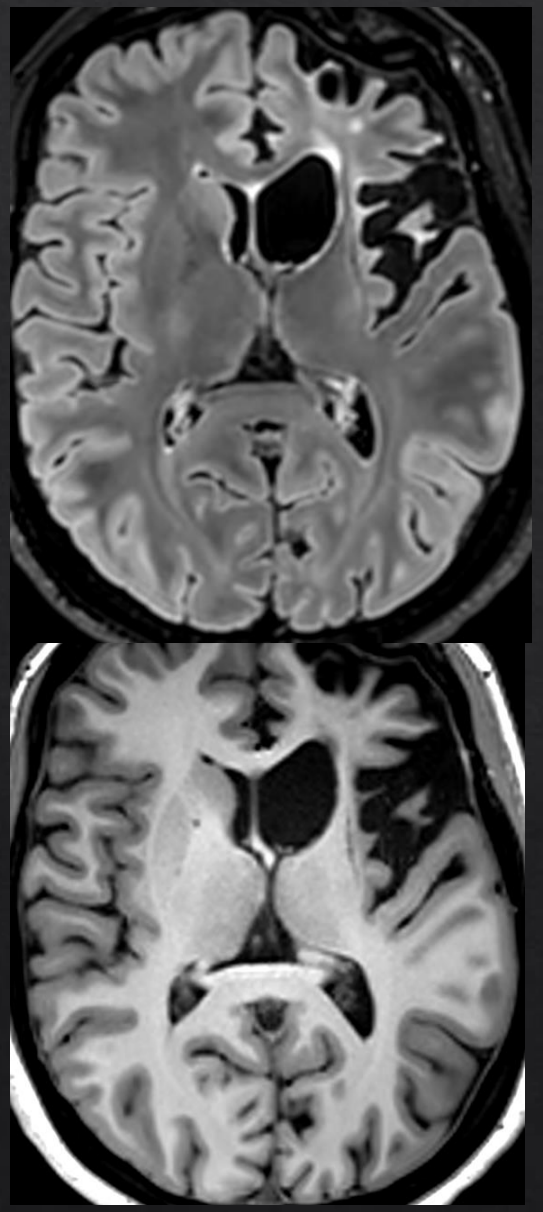
# Control postQ: recuperación funcional

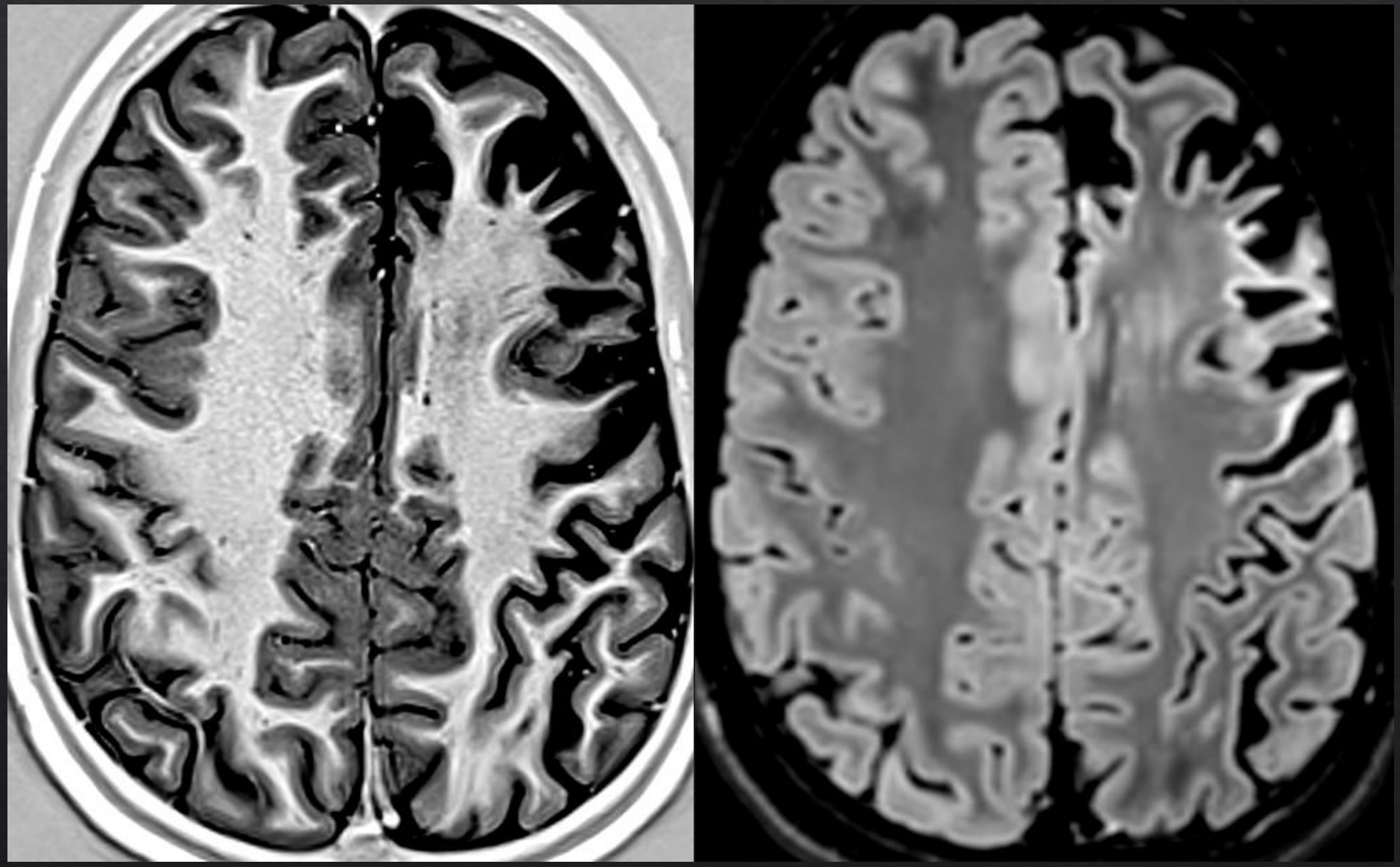


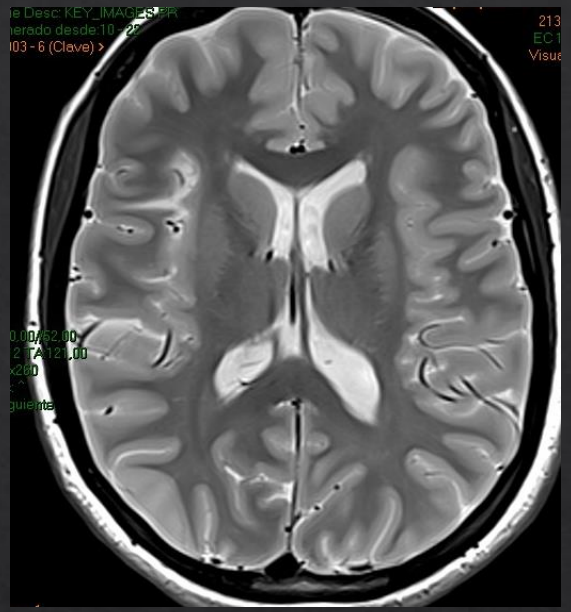

Afectación estructural discreta con gran afectación funcional

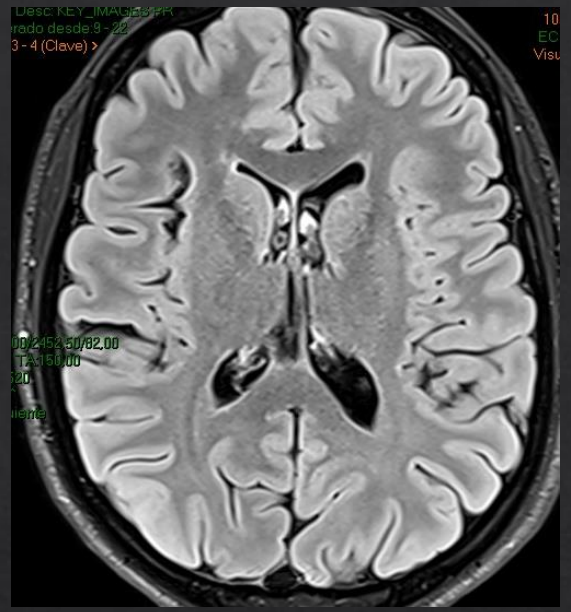
# Signos en RM de alta resolución

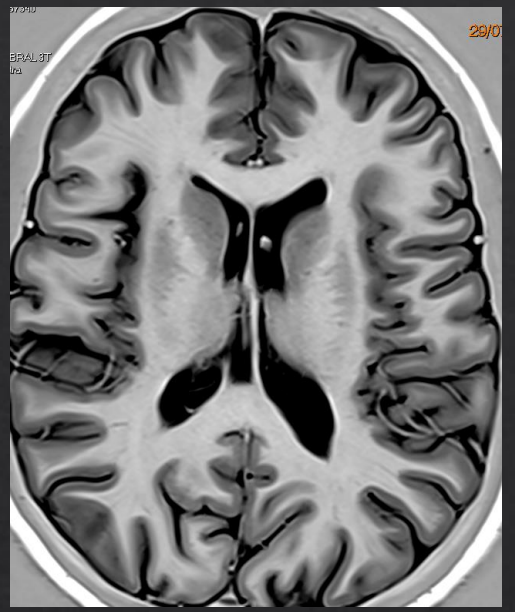
- Atrofia (hallazgo precoz, inicialmente sutil, frecuentemente progresiva,distingue de DCF)
  - Prominencia de espacio subaracnoideo
  - Adelgazamiento cortical marcado
  - Patrón espiculado si es extremo
  - Estriado
    - Disminución de volumen/Alteración de señal
  - Pálido
    - Alteración de señal (T1, T2/FLAIR)
    - Depósito de hierro ocasionalmemte sin atrofia (T2*, SWI)
- Hiperseñal T2/FLAIR
    - Subcortical/cortical
      - Lineal: característica
      - Si es transitoria: muy específico
    - Periventricular: "cup" unilateral o asimétrico

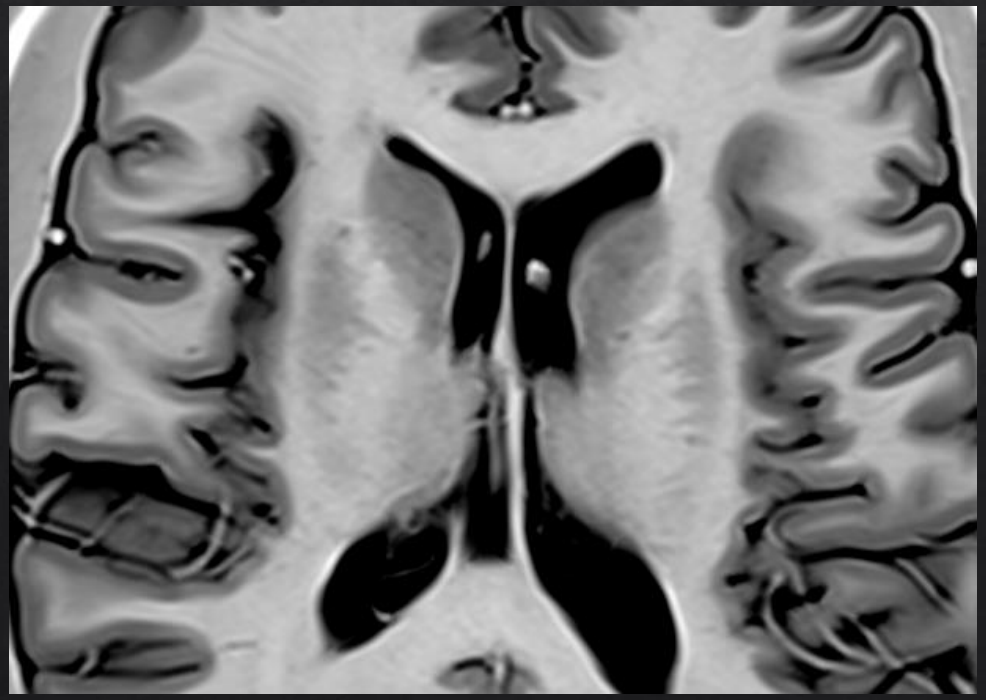


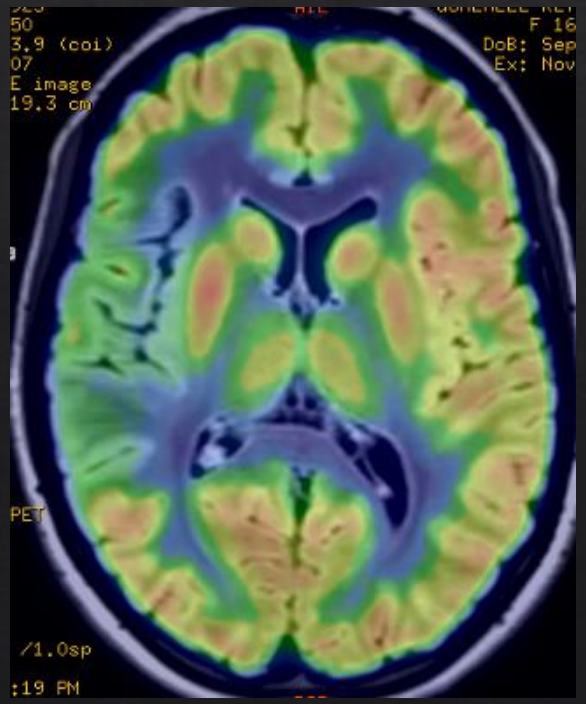








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Ex: Nov
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PET
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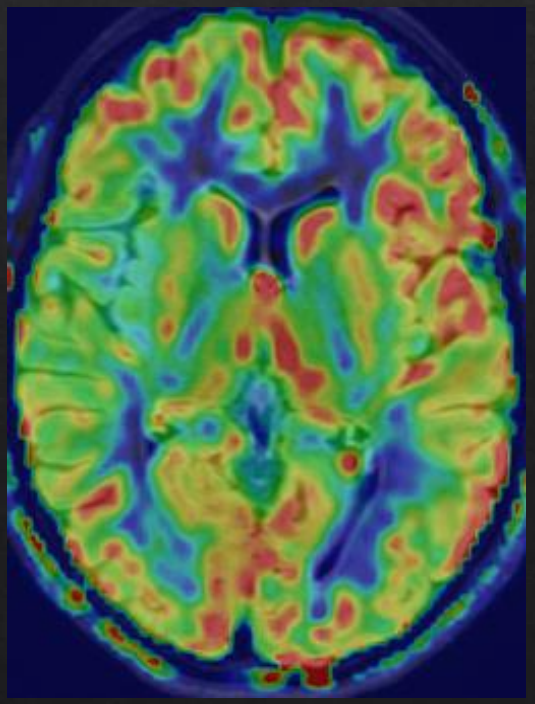
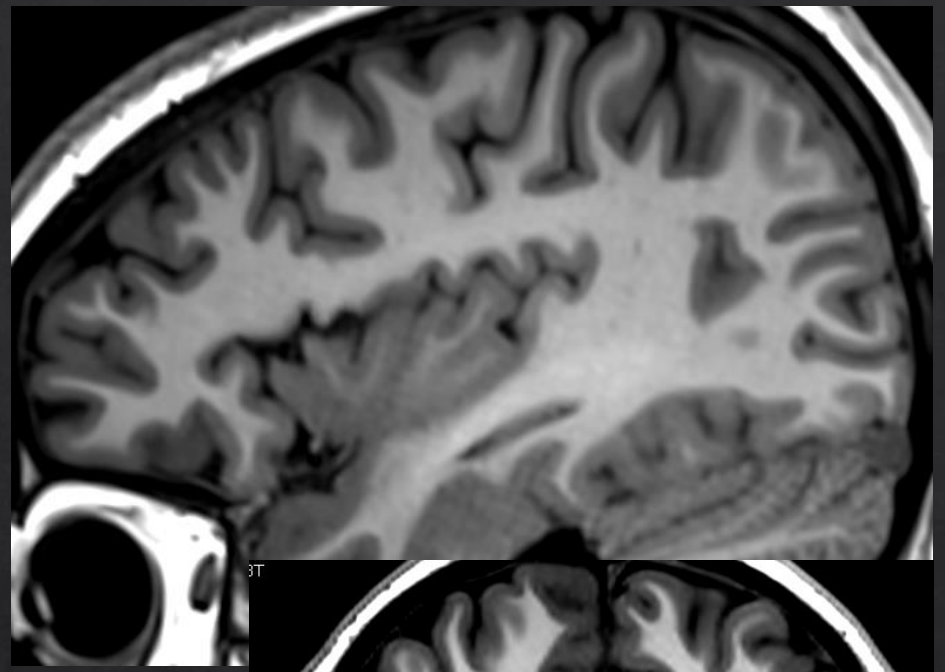
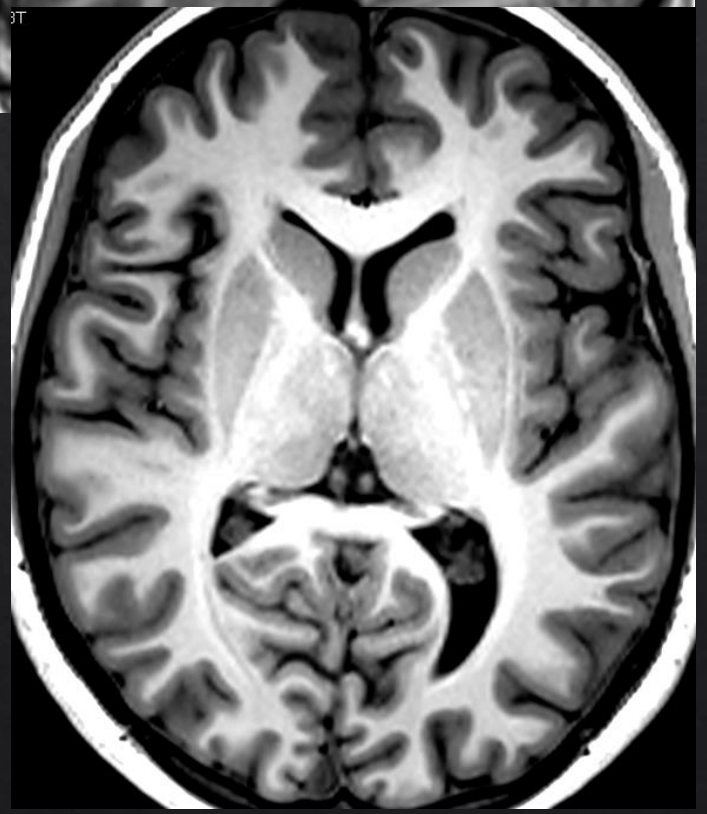
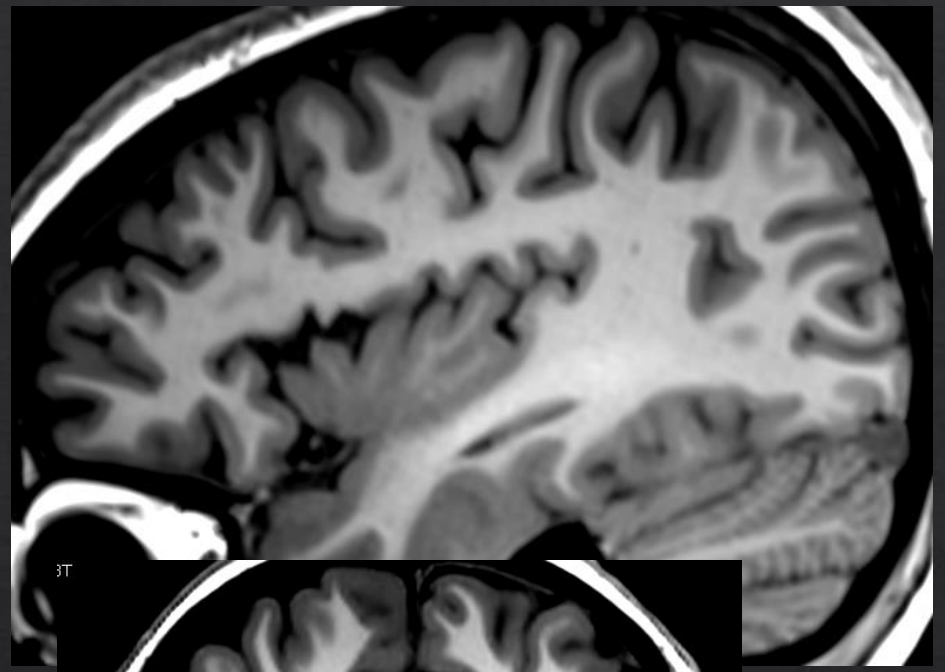
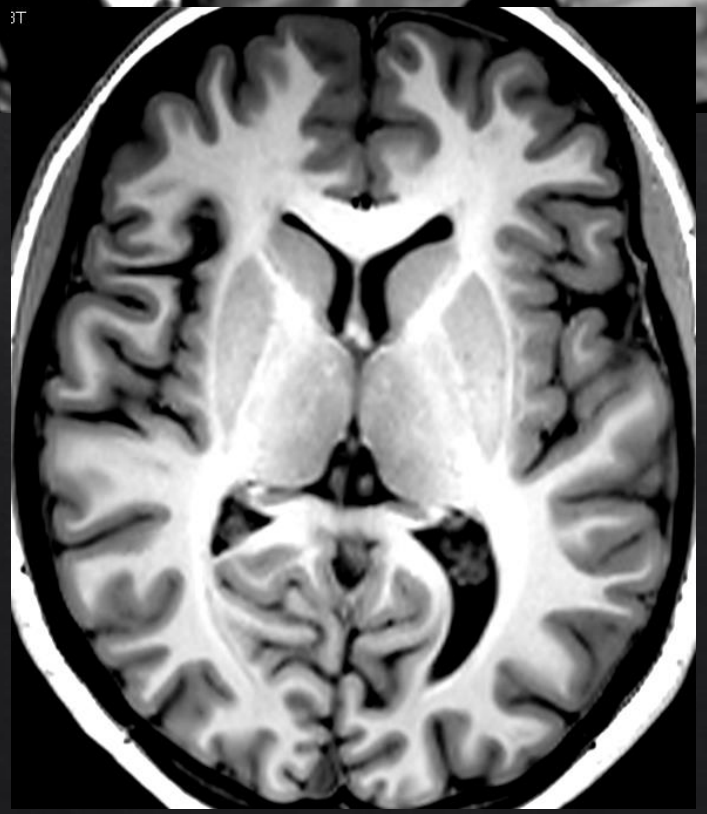
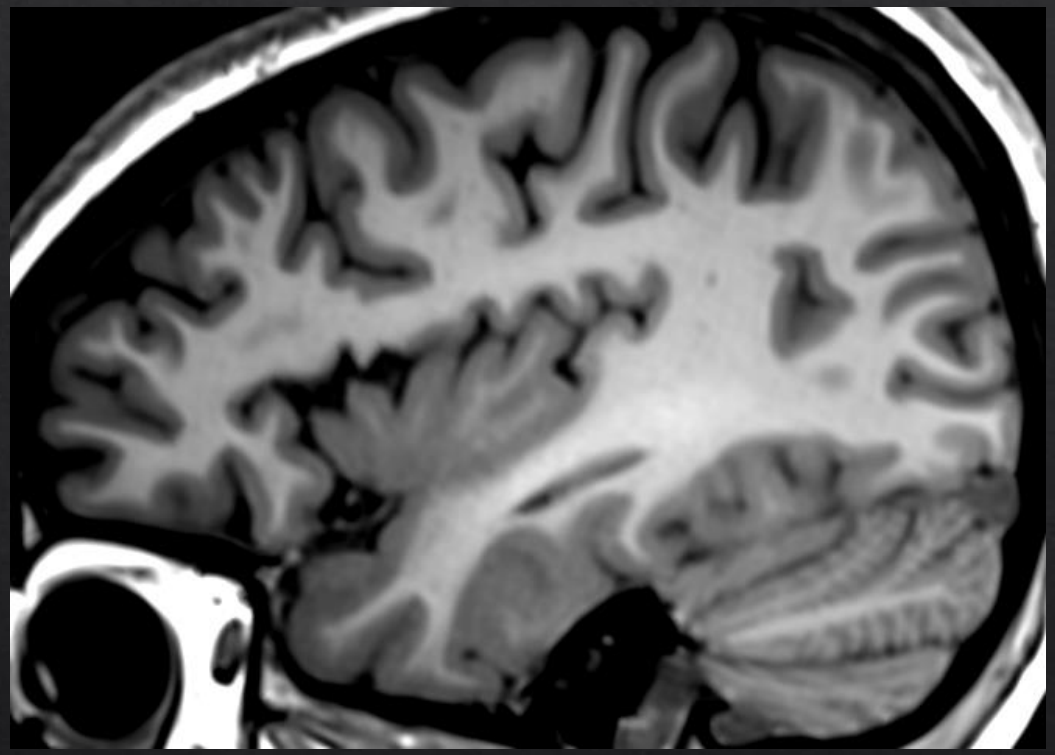


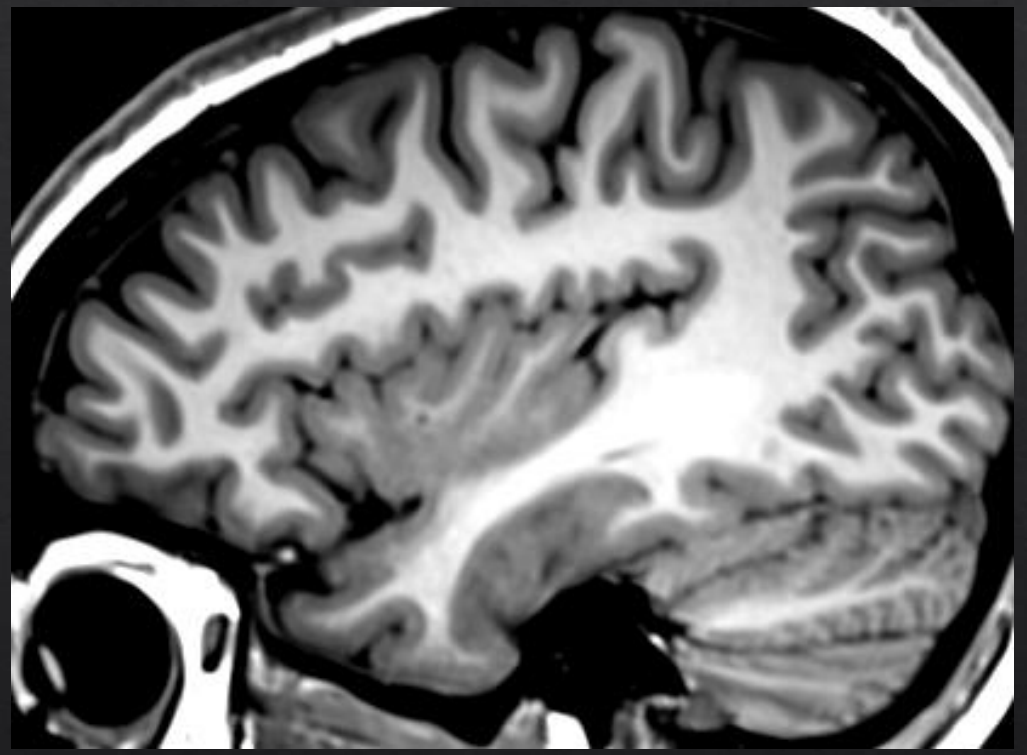




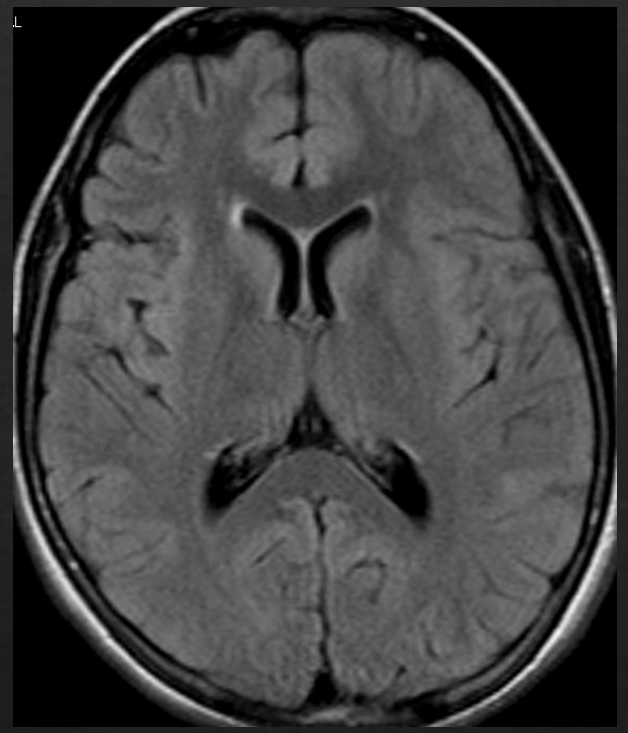


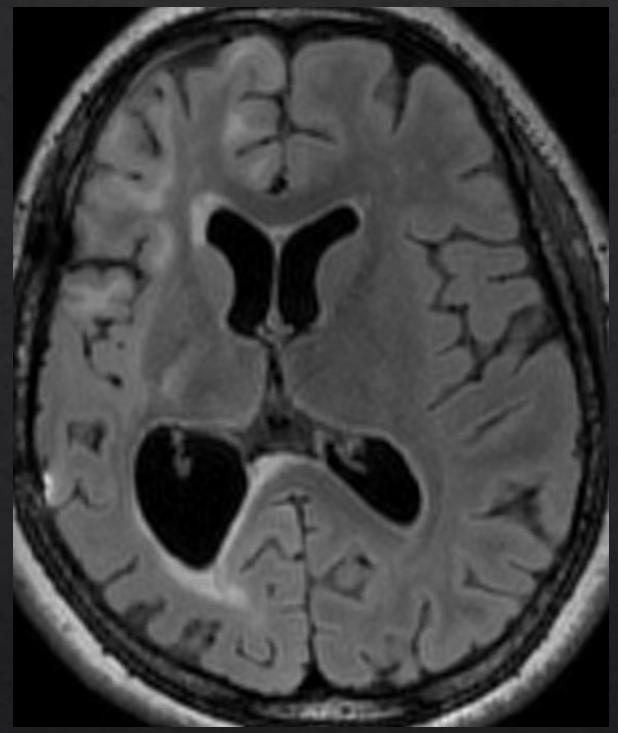


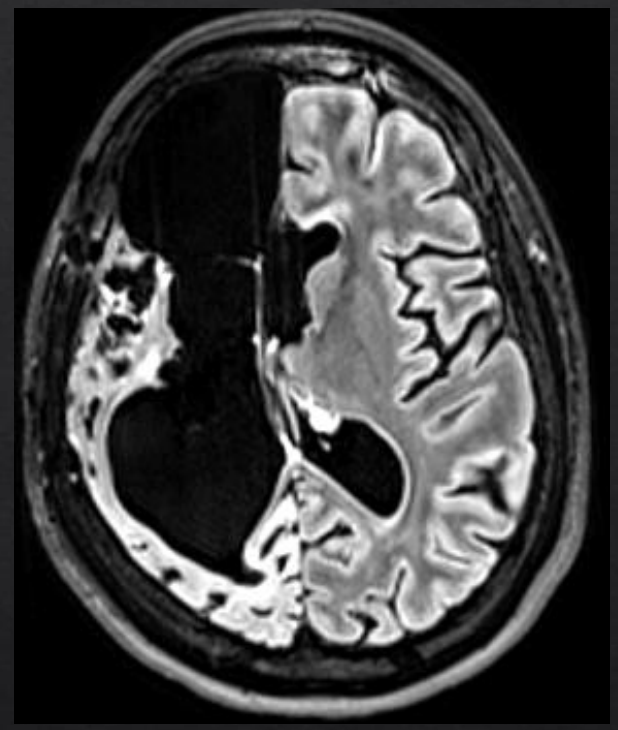




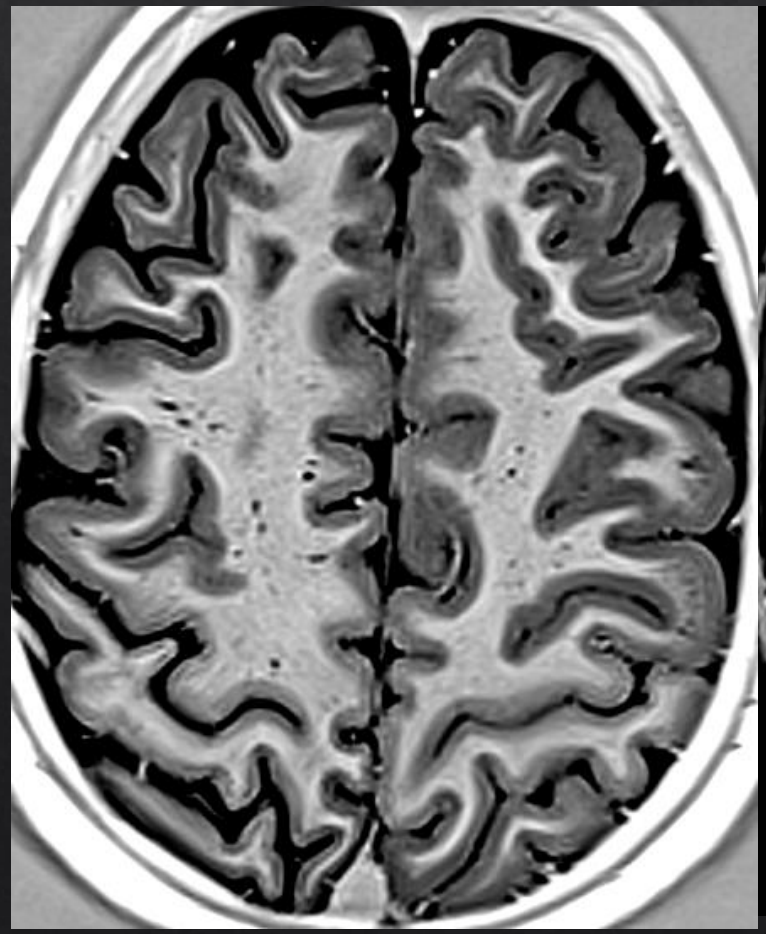
# Lesión periventricular: “Cup” frontal

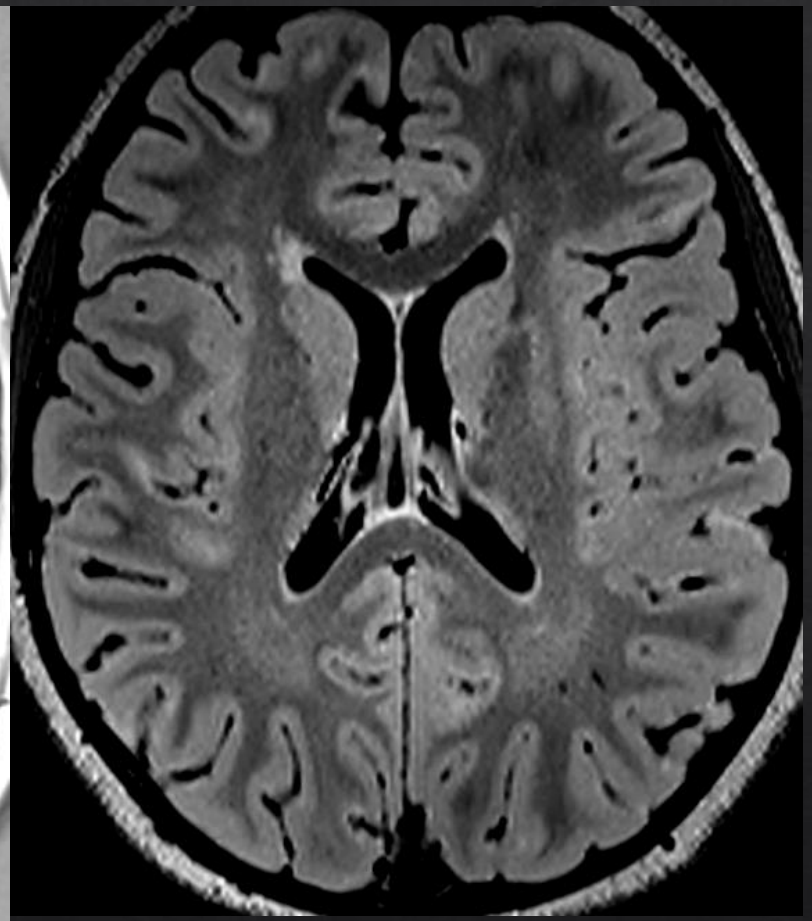


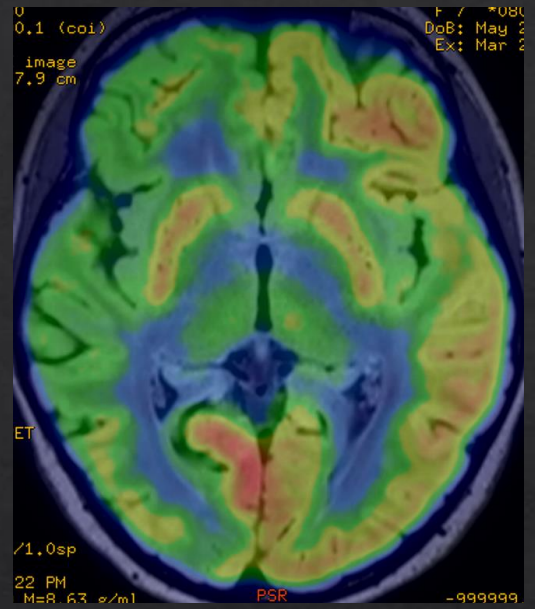


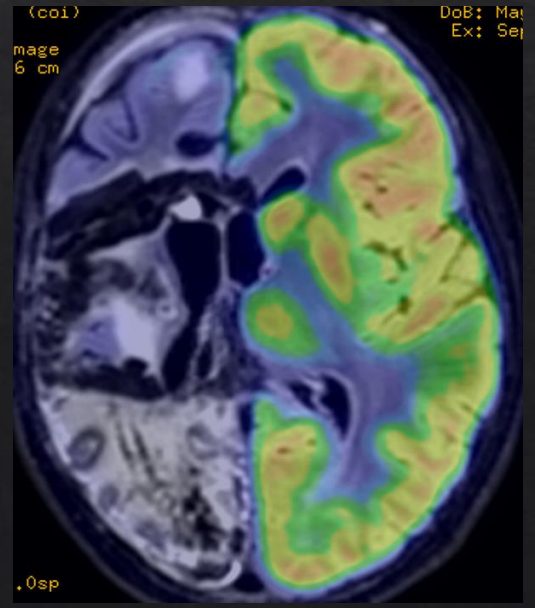


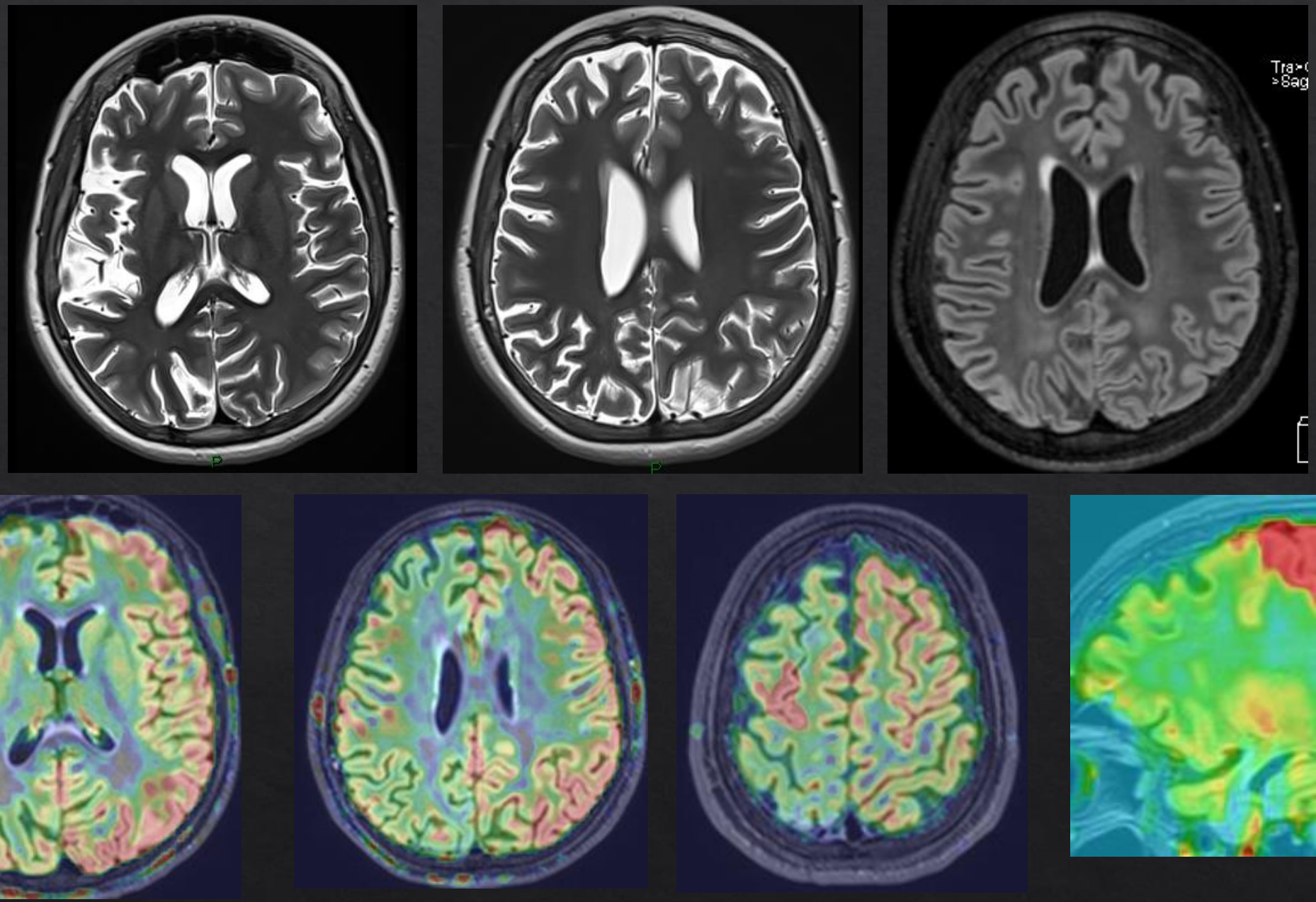
# Atrofia y lesiones de SB





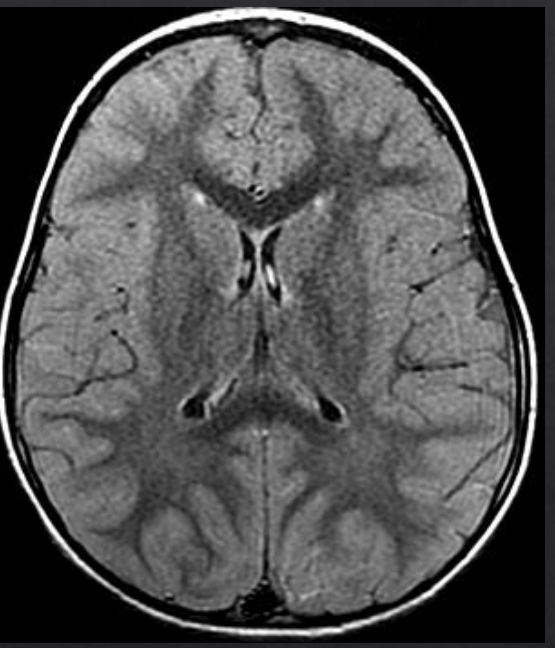


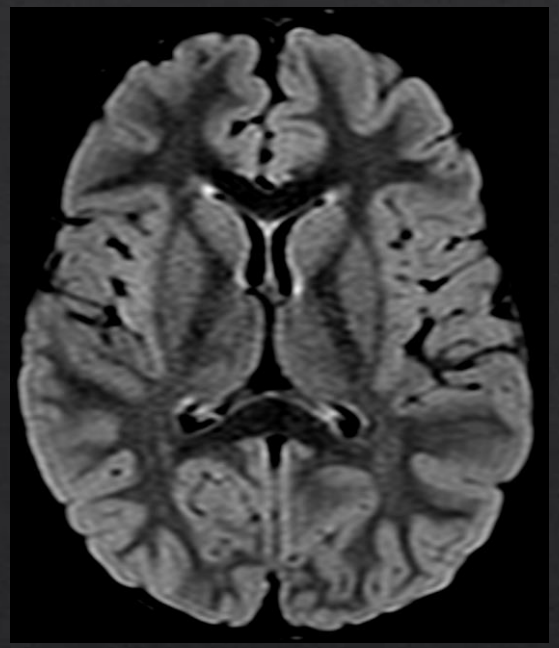



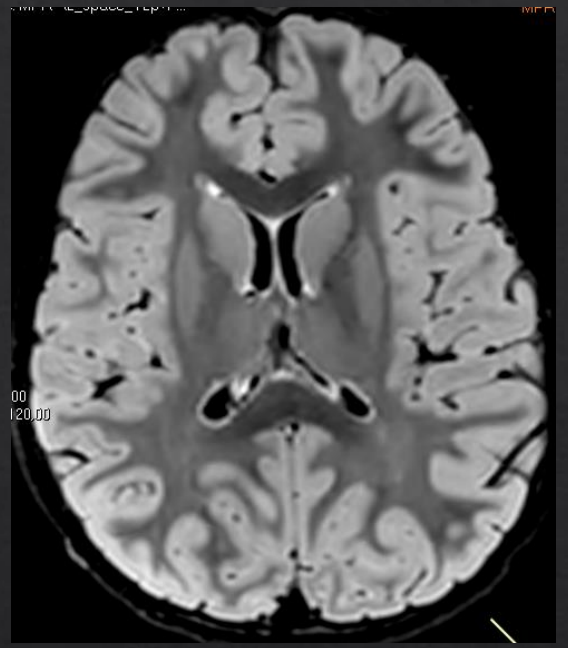


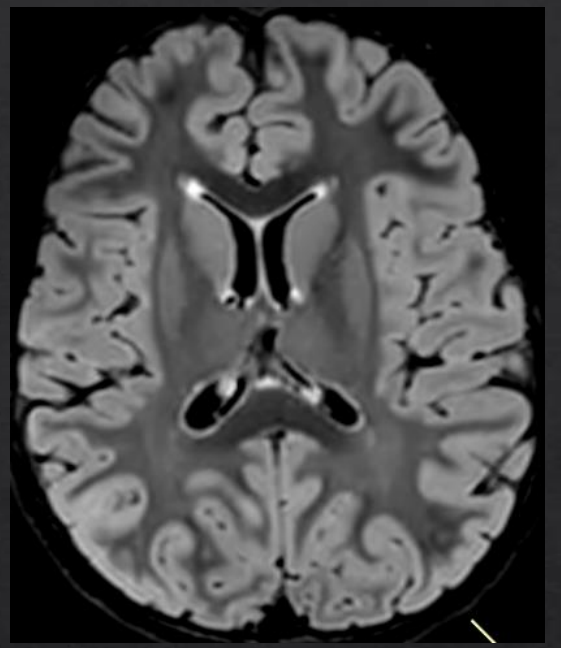
Atrofia bilateral, hipoperfusión asimétrica con hiperperfusion frontal

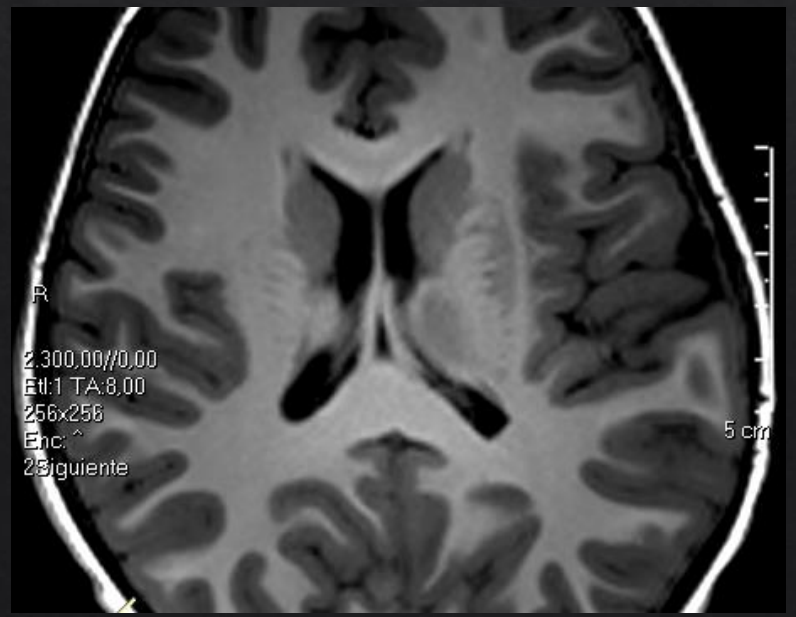
# CUP que aparece

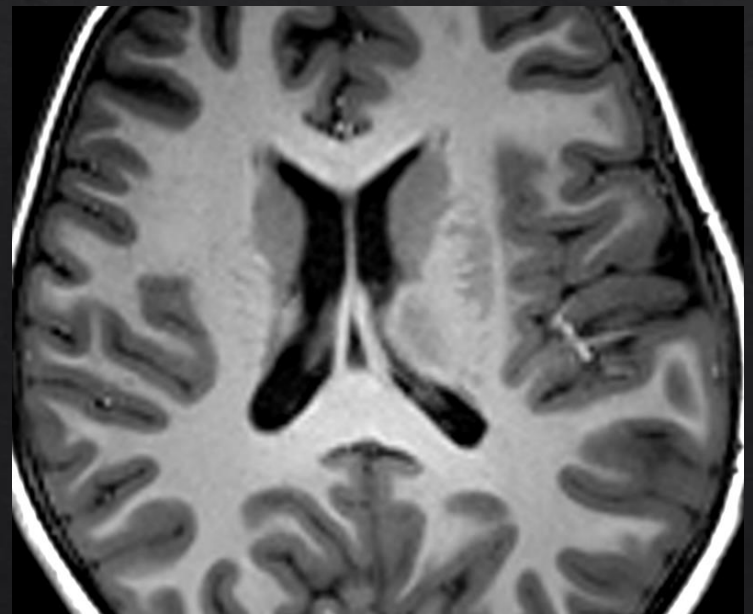


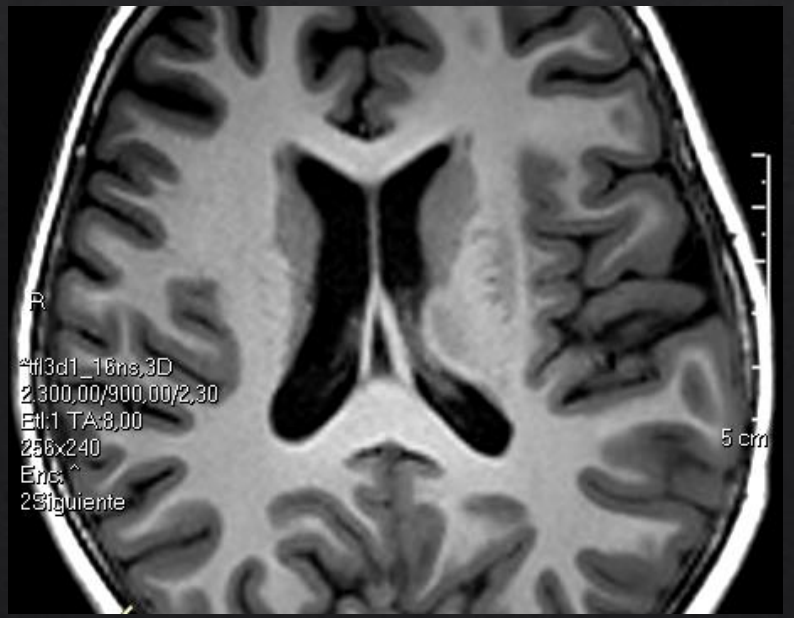










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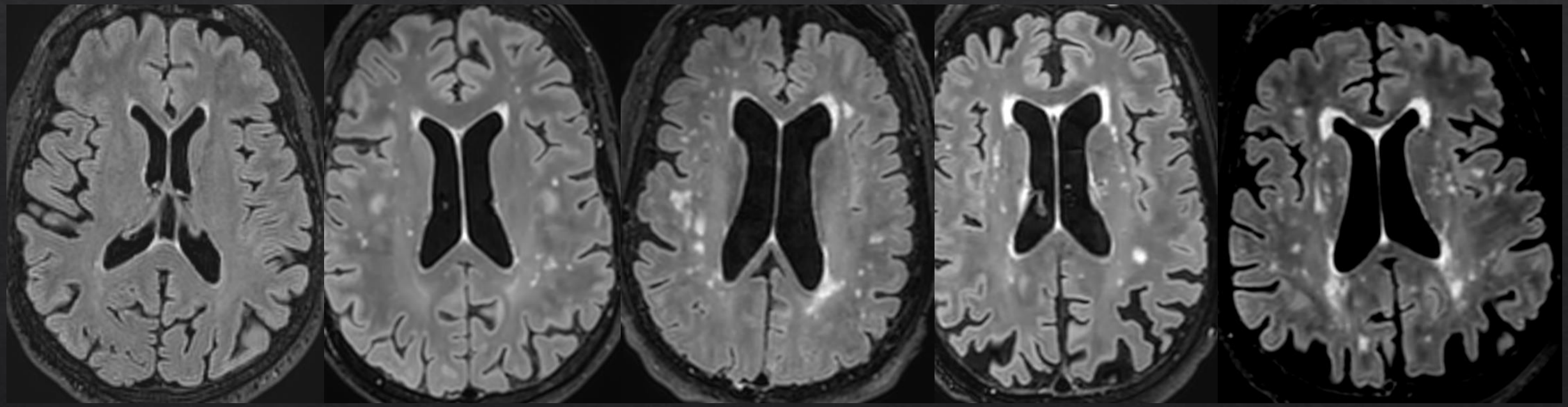
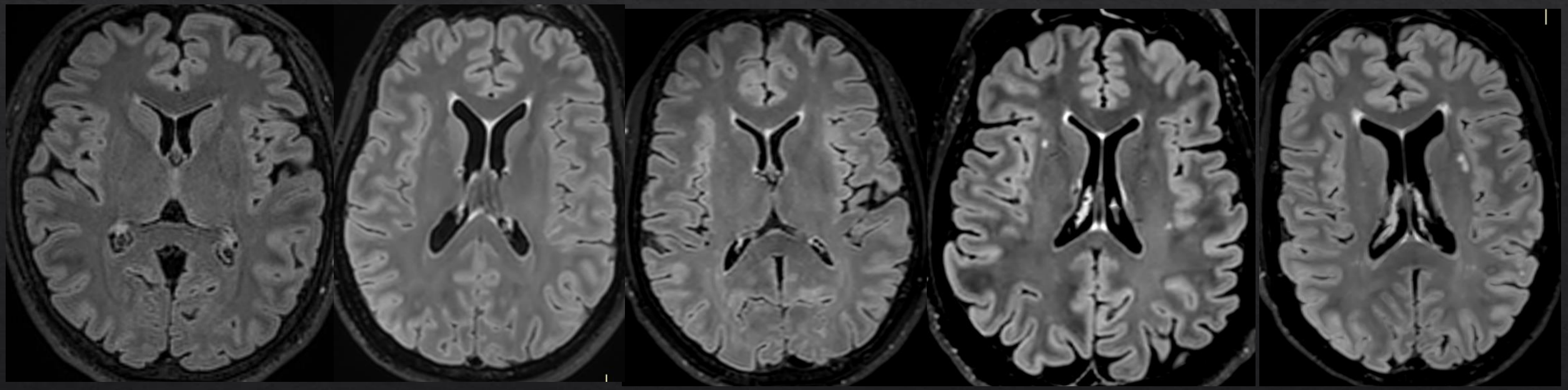
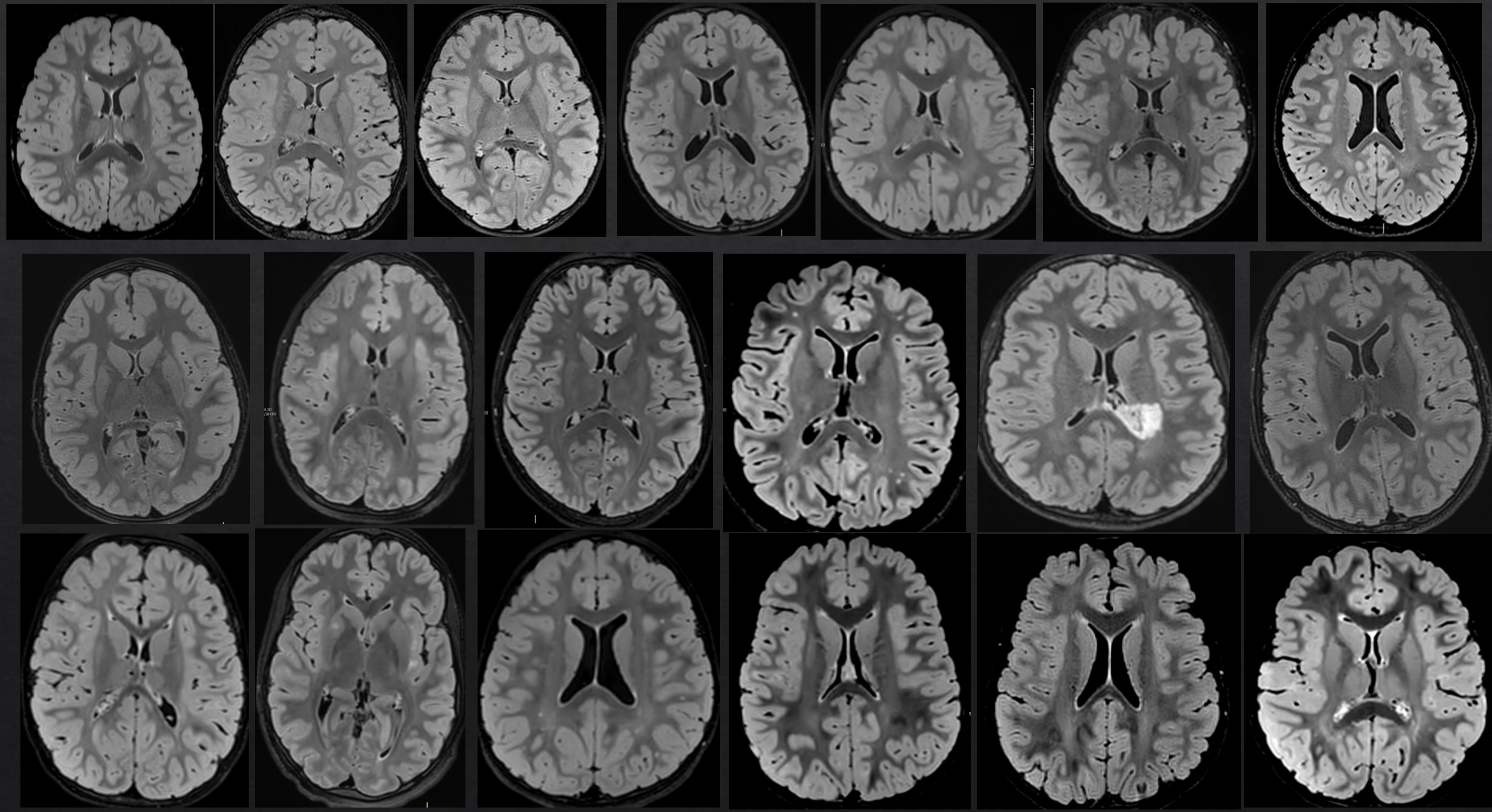


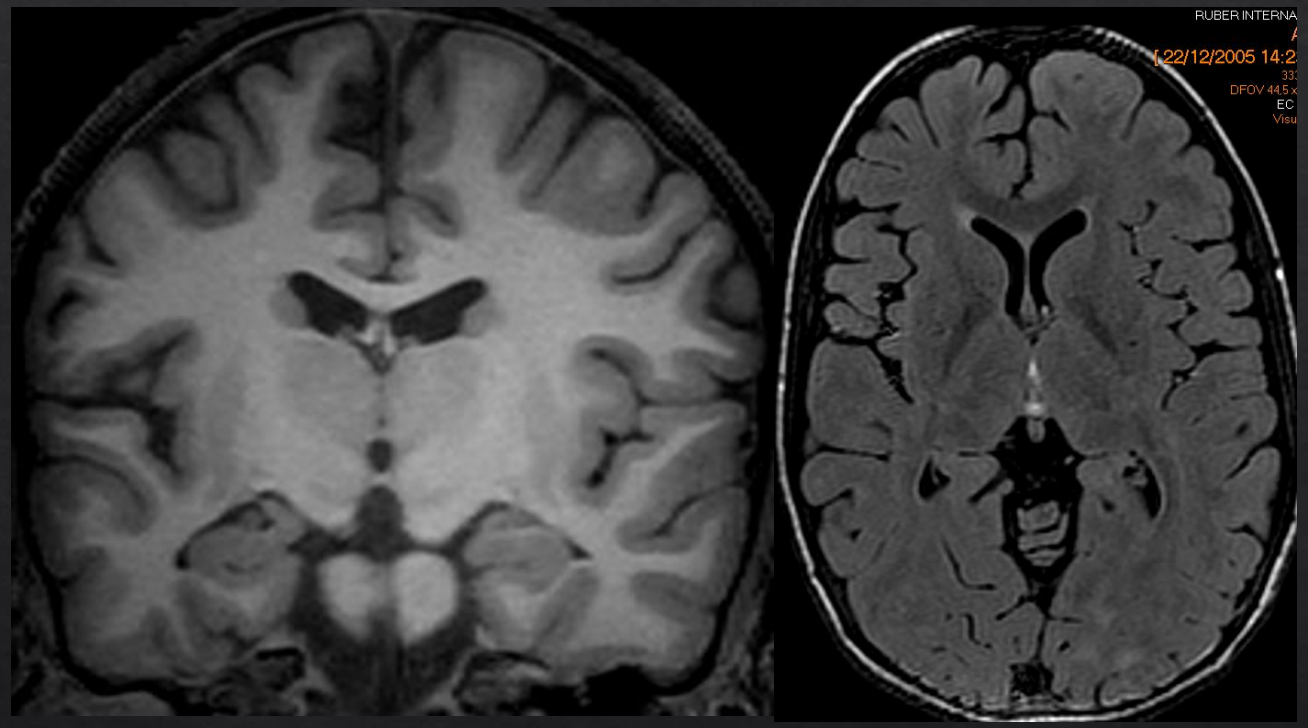
# Cup 5ª década

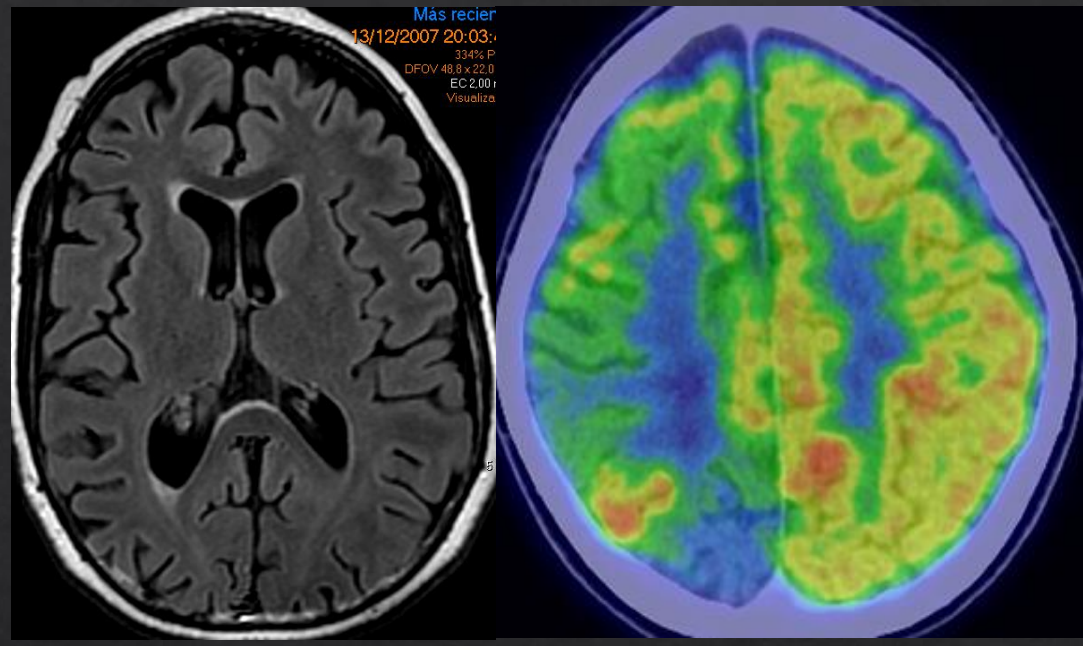


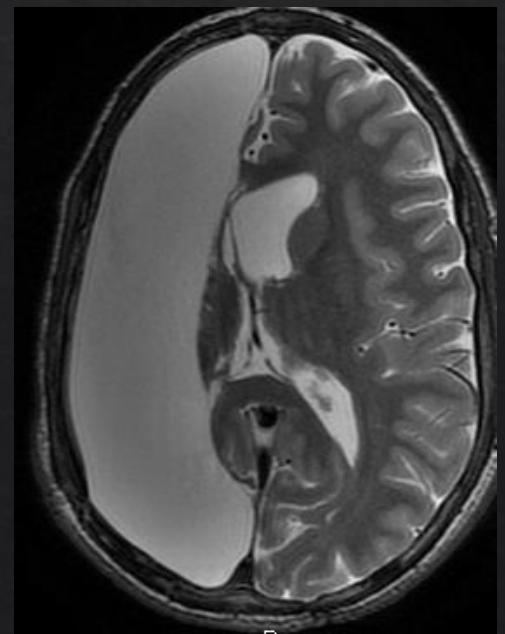
# Cup 1ª década



# Atrofia y alteración de señal?

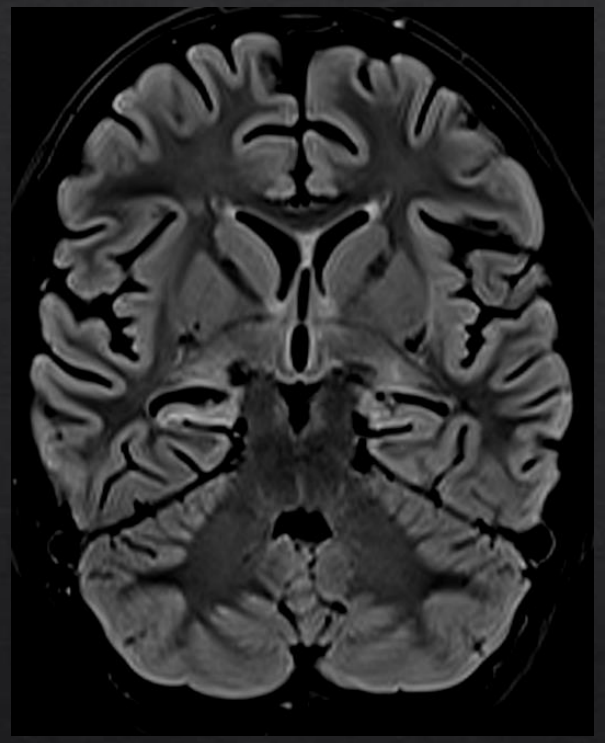


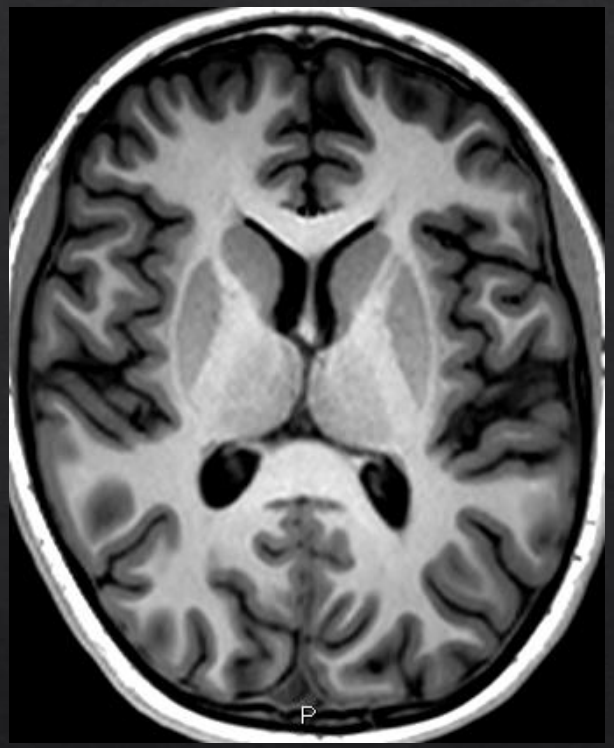



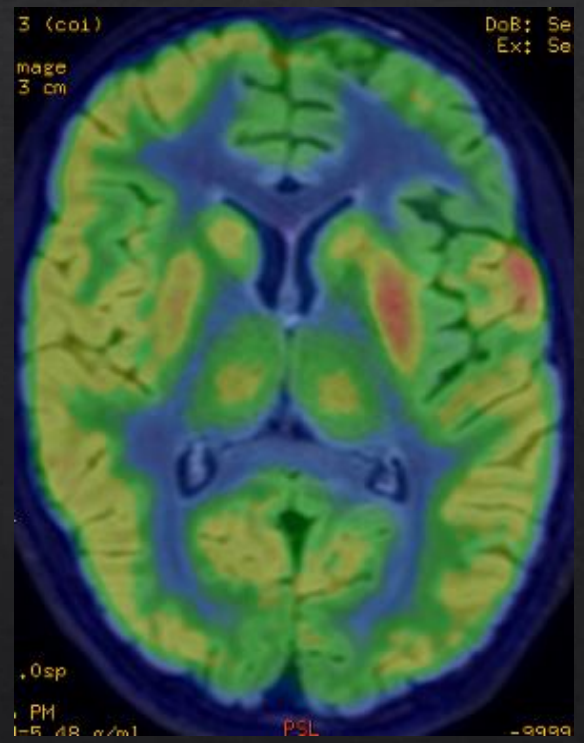



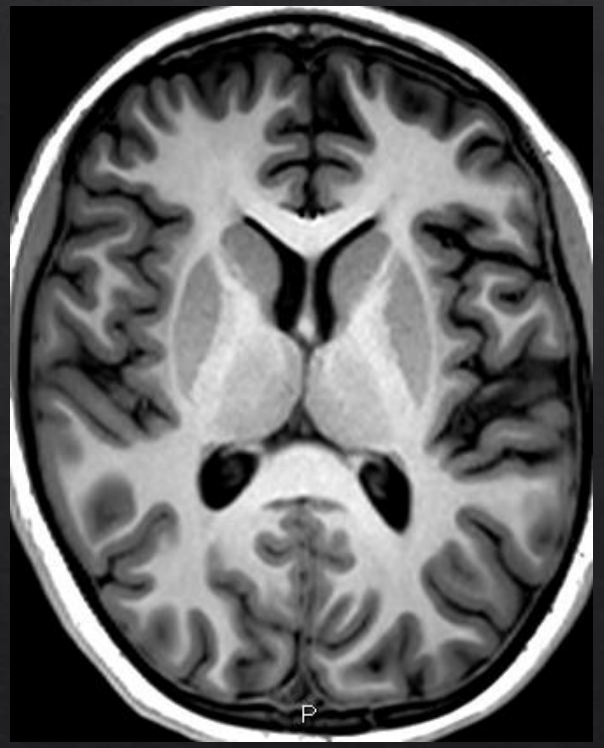
11 años , CPC, déficit motor,  EEG característico, no deterioro cognitivo
Hallazgos “no concluyentes”, se recomienda control

# Signos sutiles

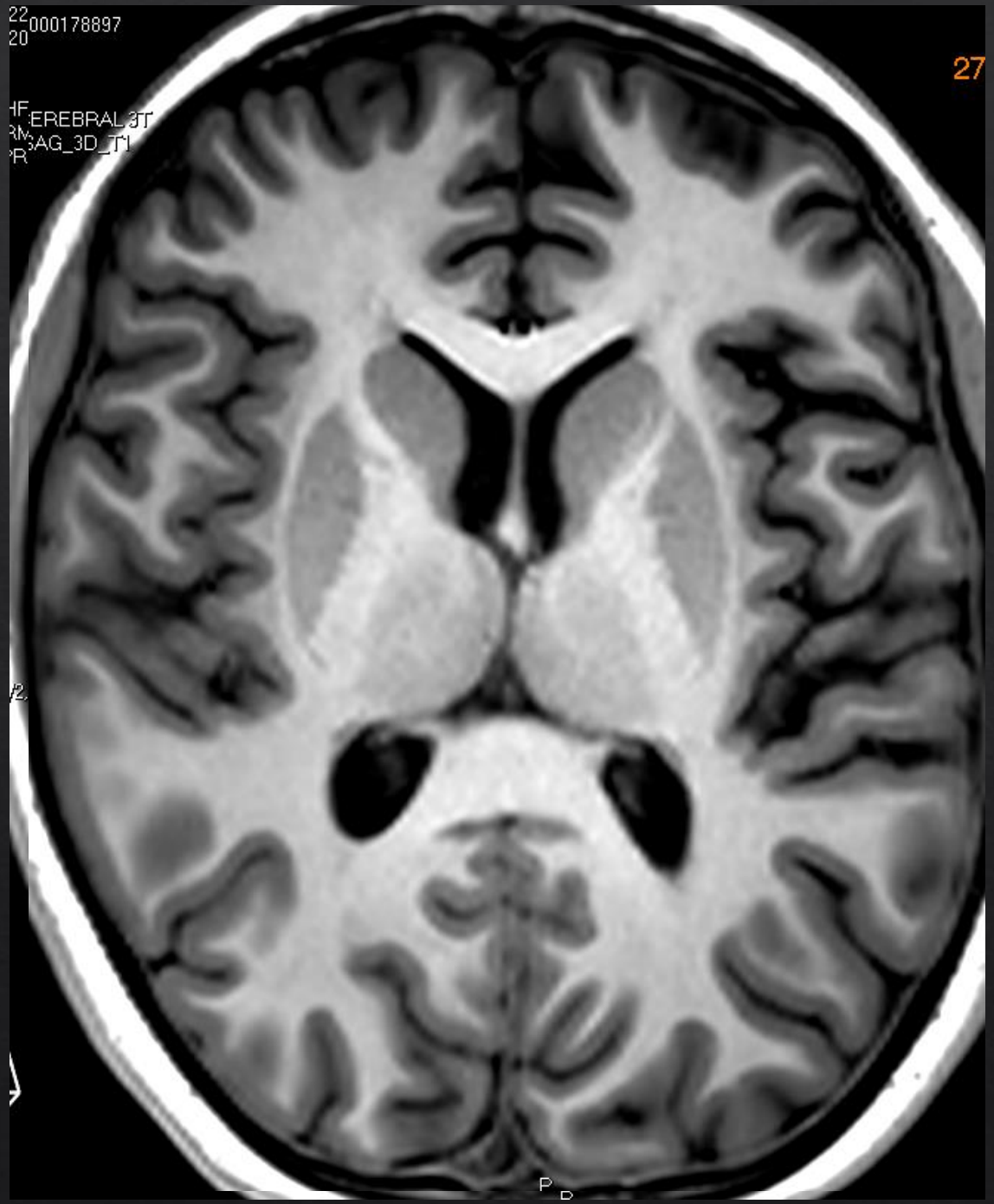


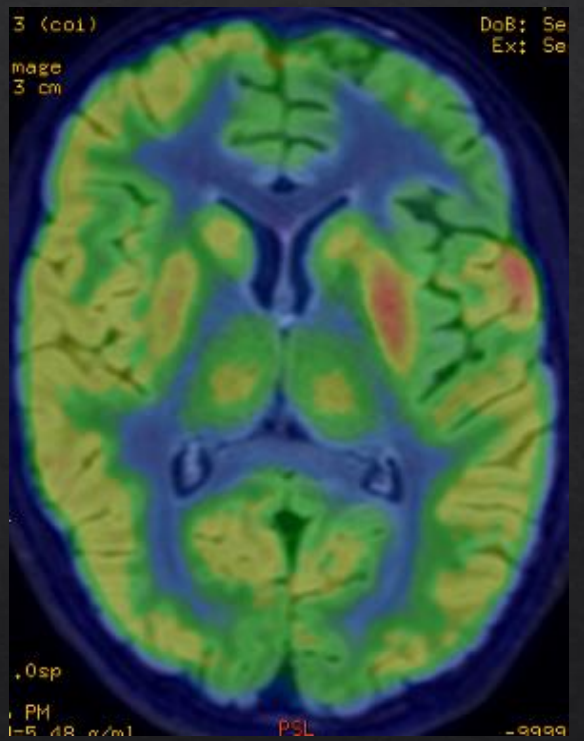




6 años, CPC, déficit motor, EEG positivo, ETM bilat: encefalitis límbica?
Halazgos no concluyentes? Control PRECOZ


000178897
27
EREBRAL 3T
AG_3D_T1


3 (col)
DoB: Se
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mage
3 cm
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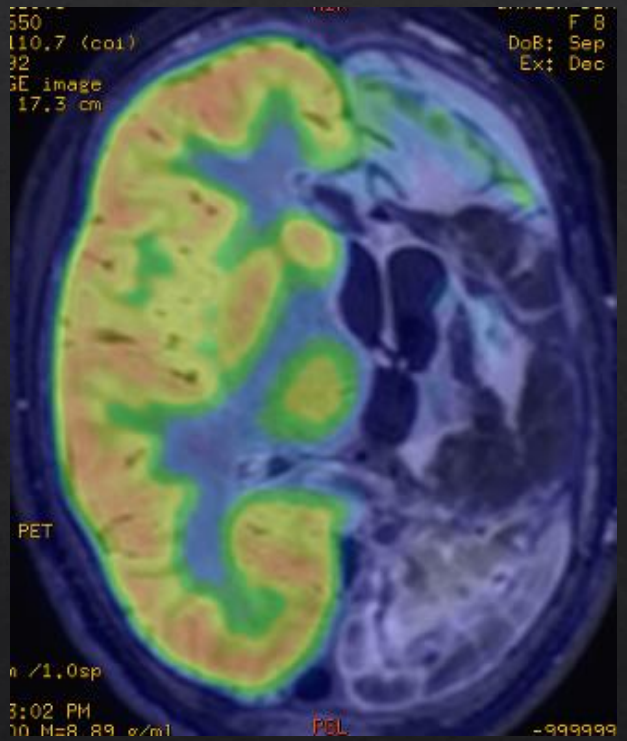
PM
PSL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17.3 cm
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PET
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3:02 PM
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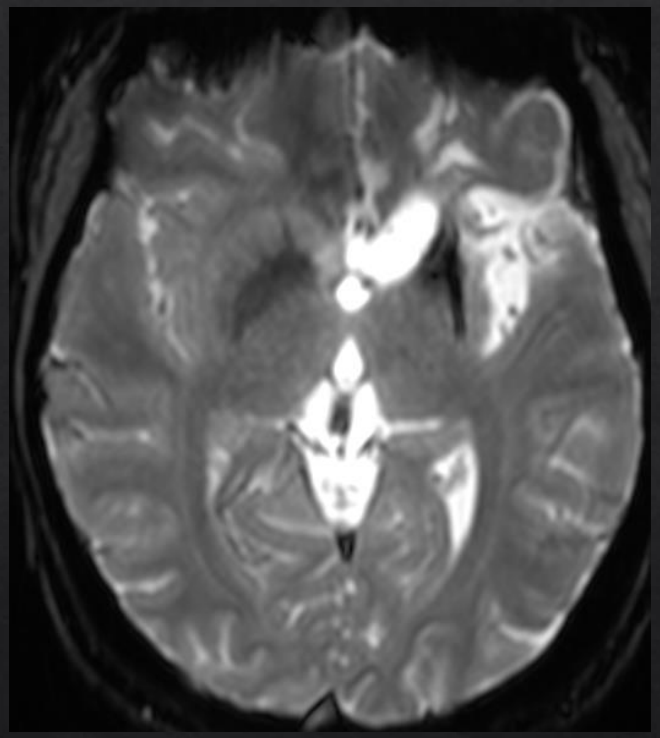
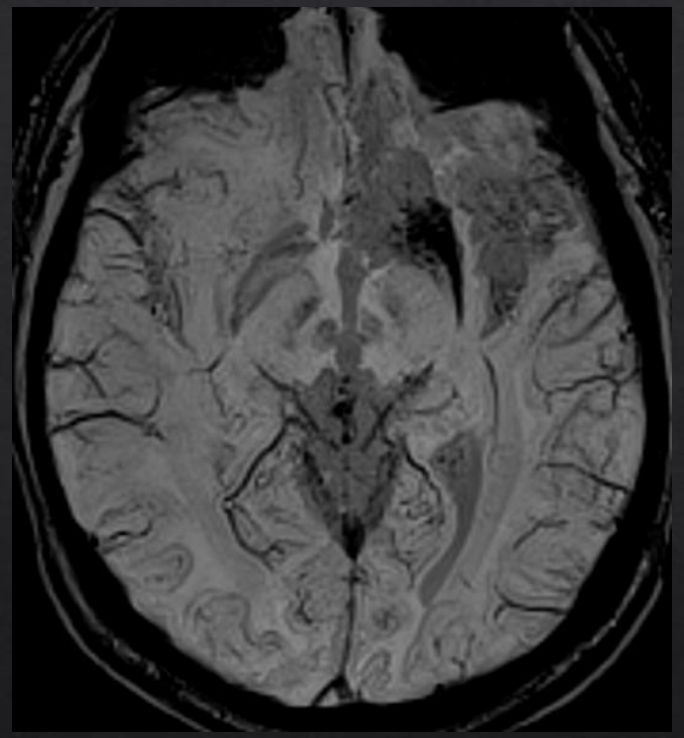
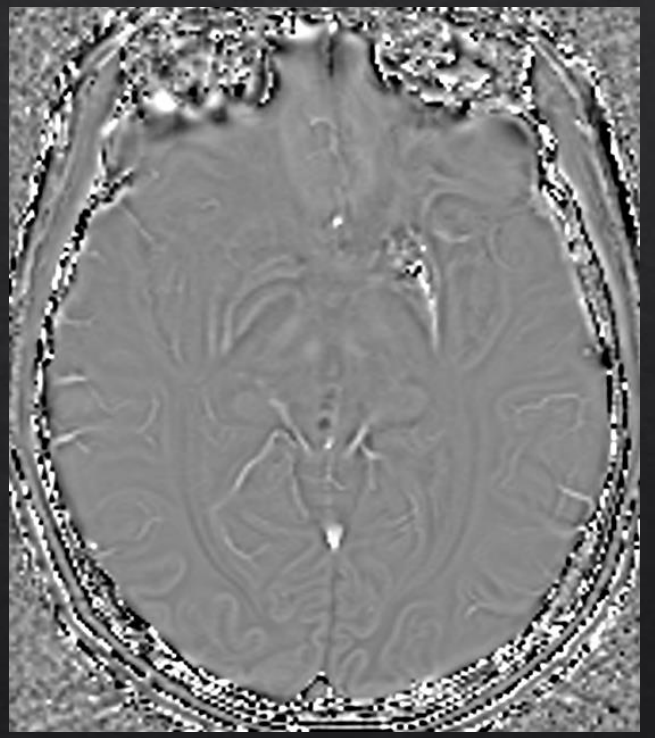
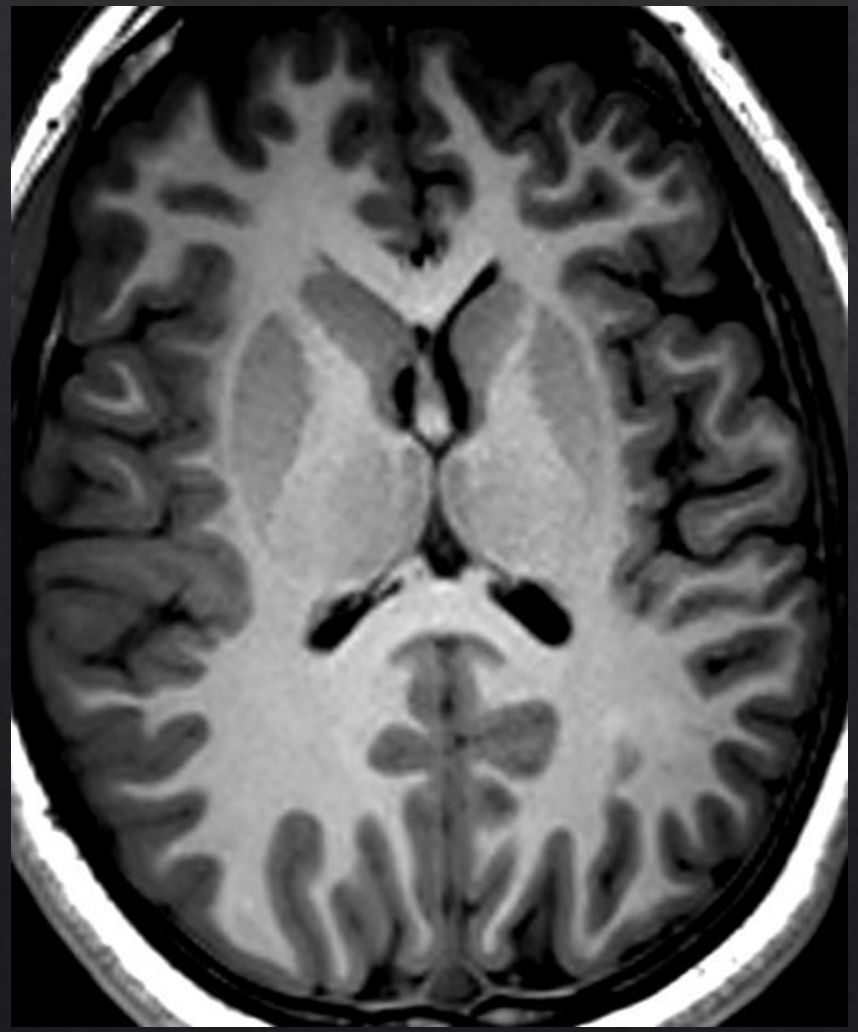
# Depósito de hierro en N. Lenticular



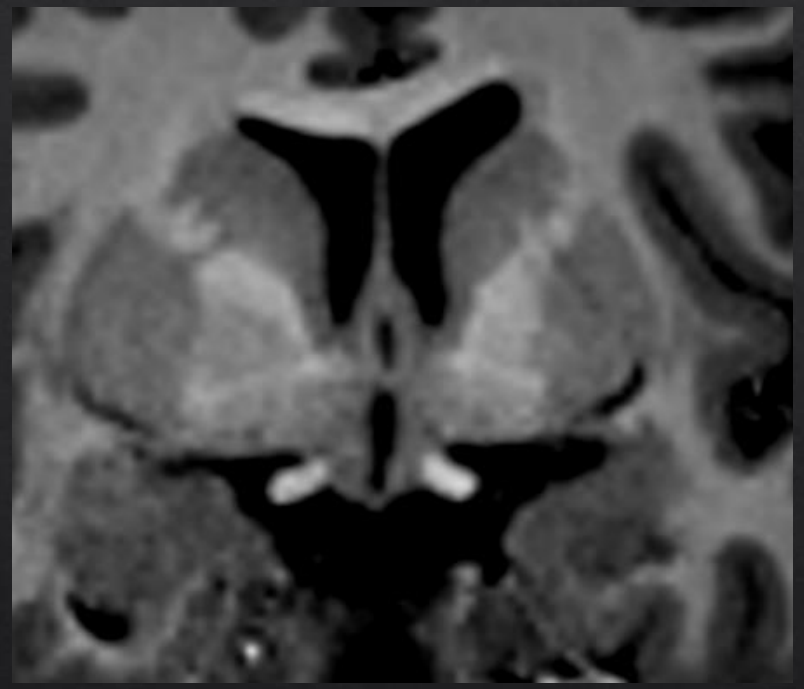




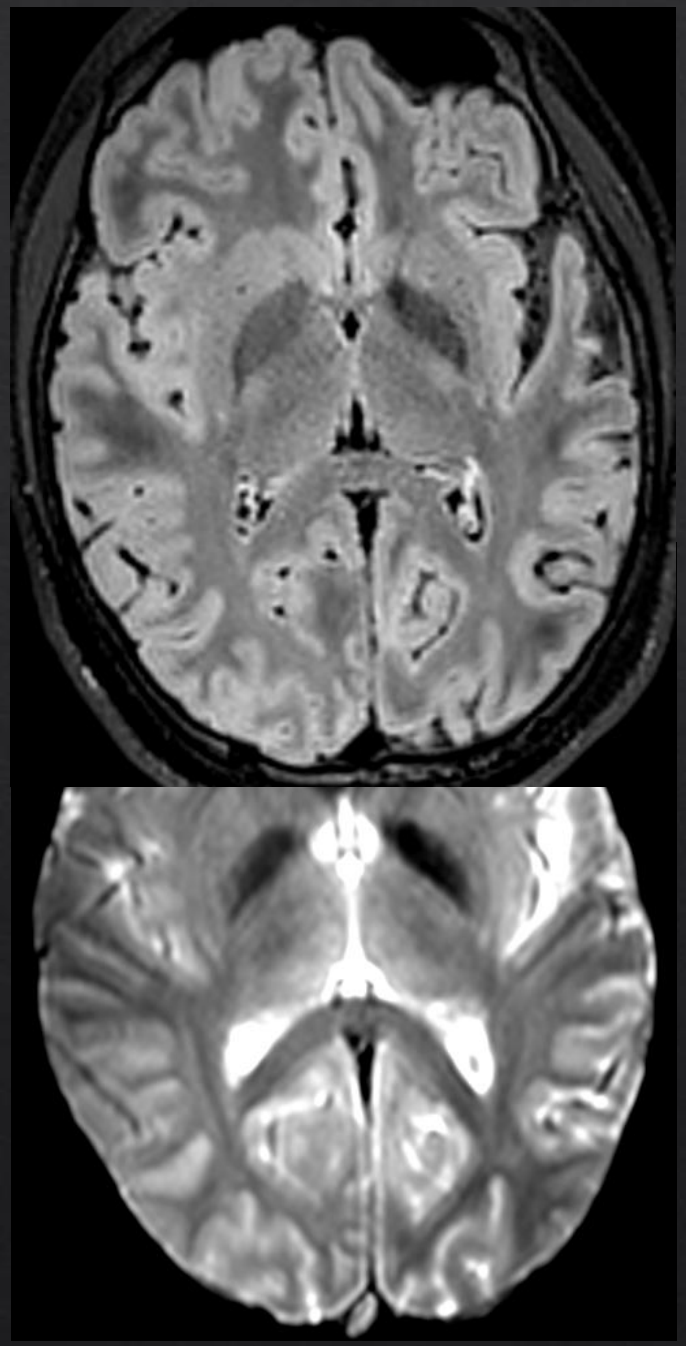
# Lesión del G Pálido



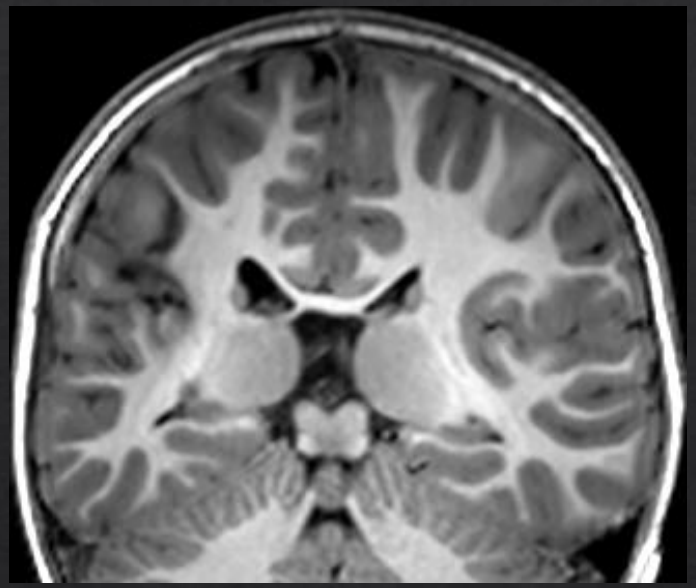
Atrofia fronto-silviana

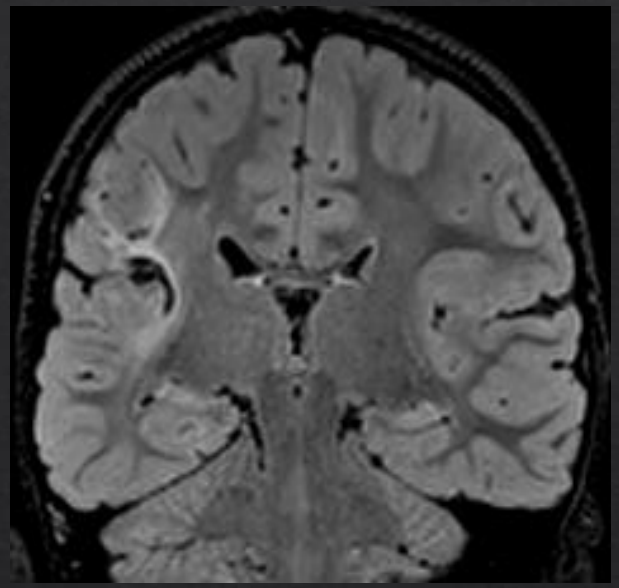


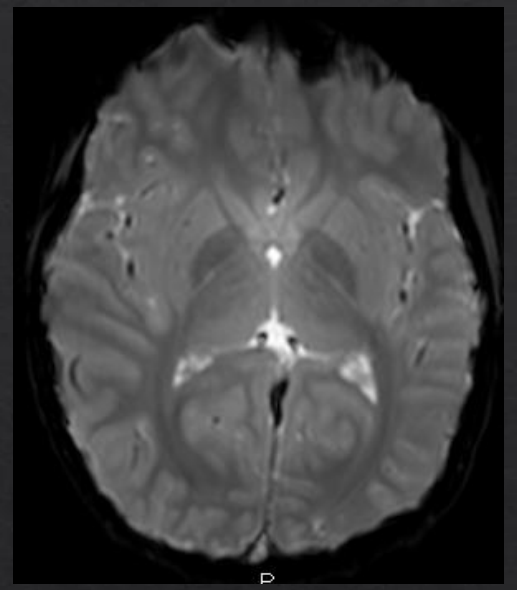
Lesión del G.Pálido sin atrofia del C Estriado

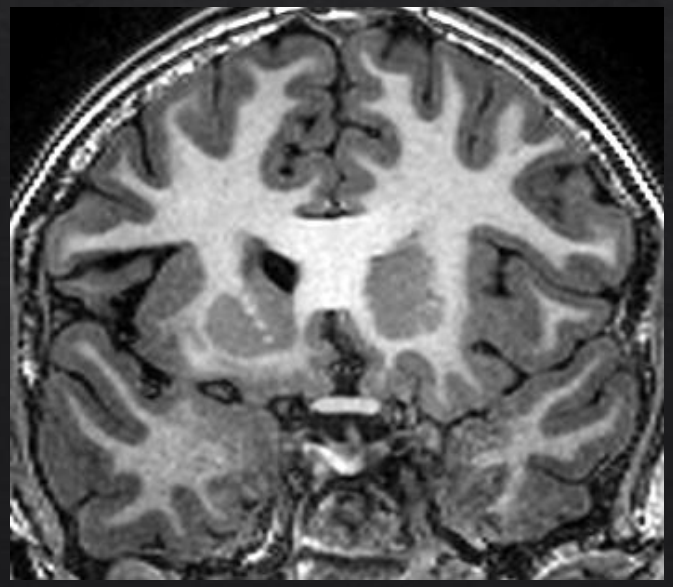


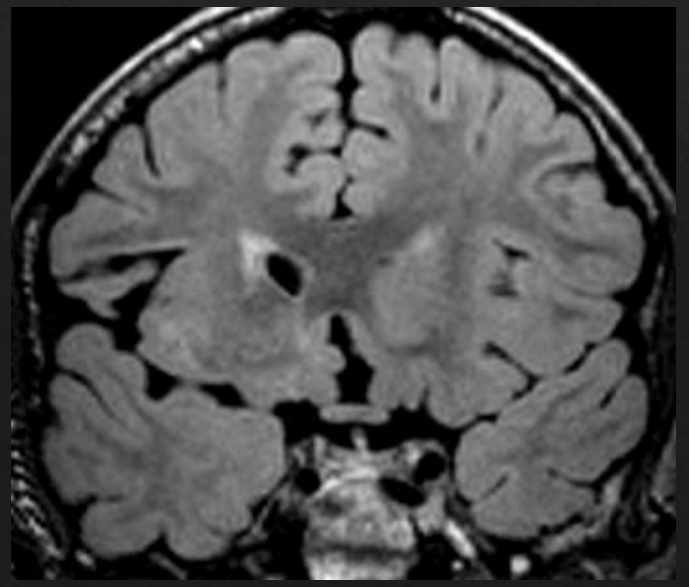
# Infarto vs Rasmussen

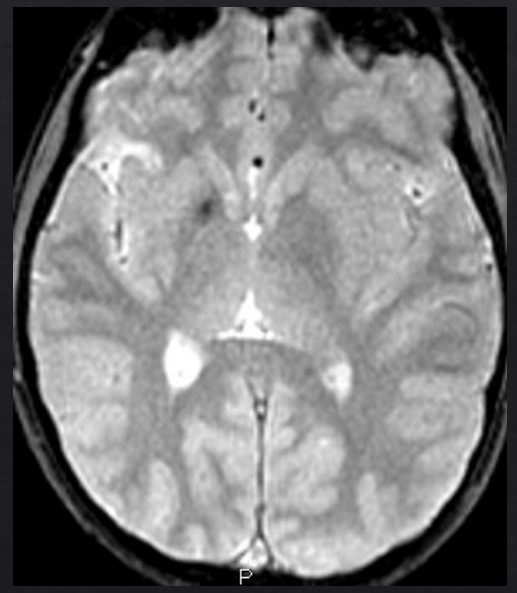




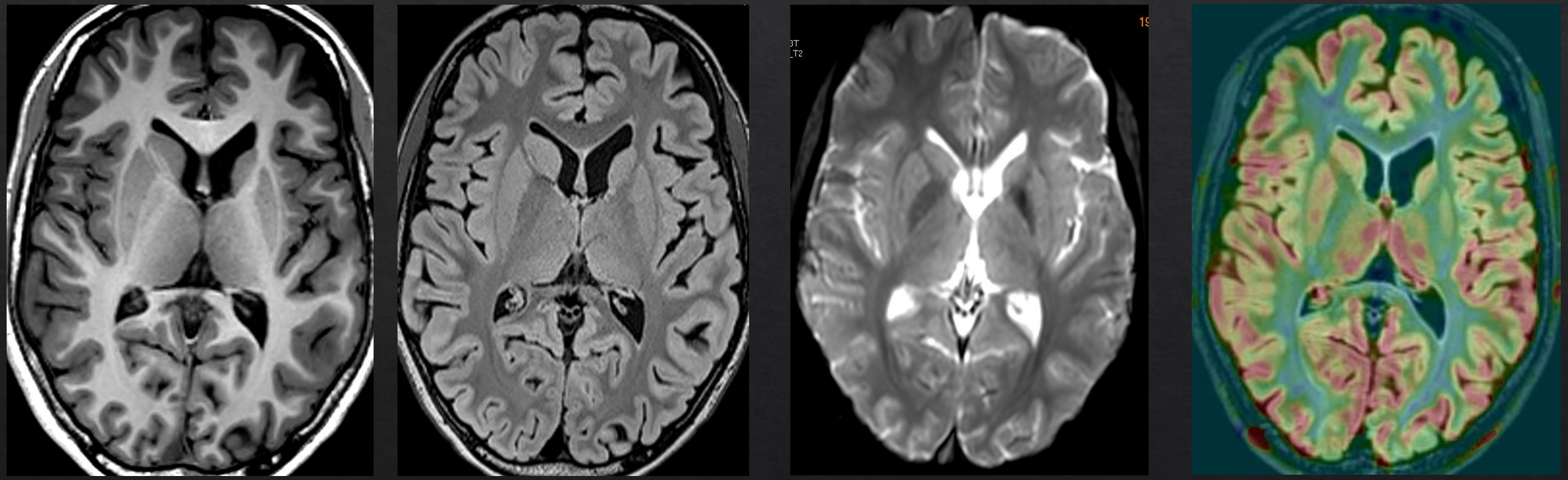




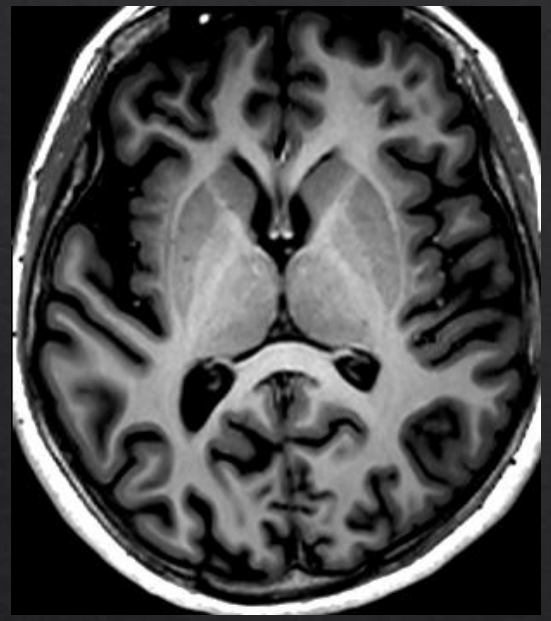
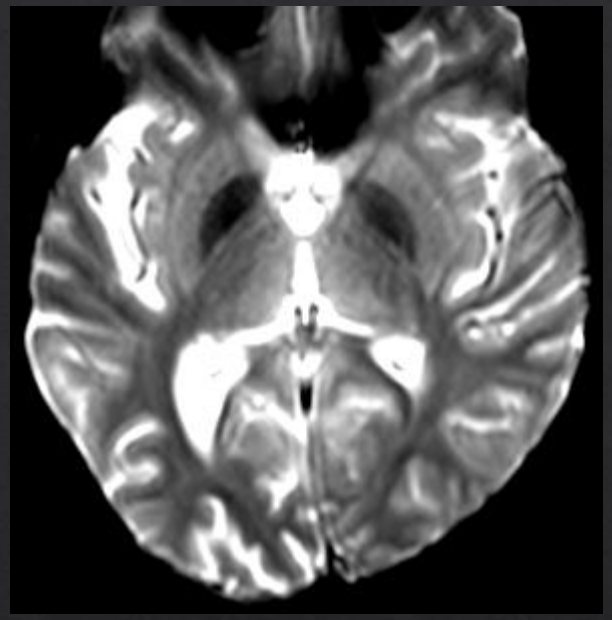
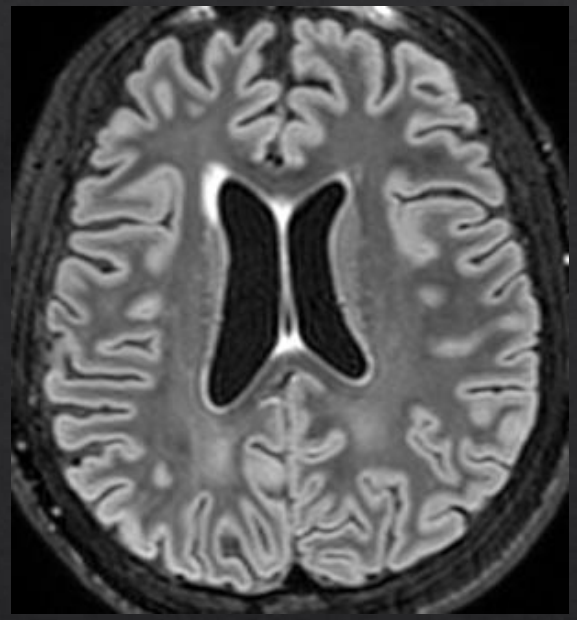
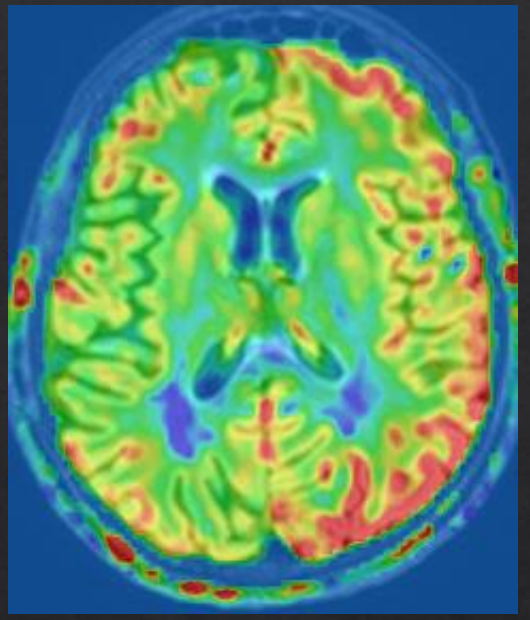
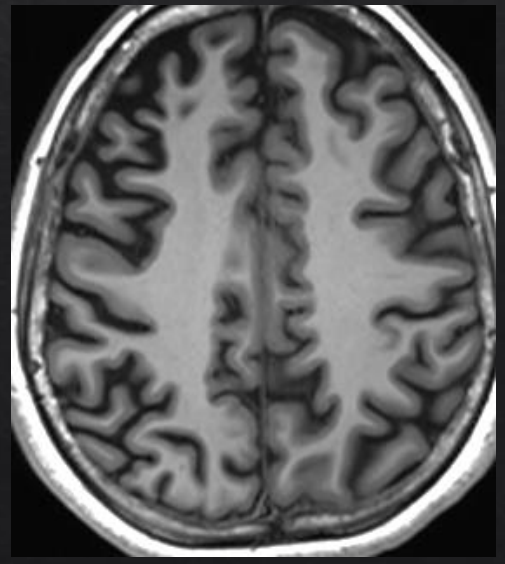
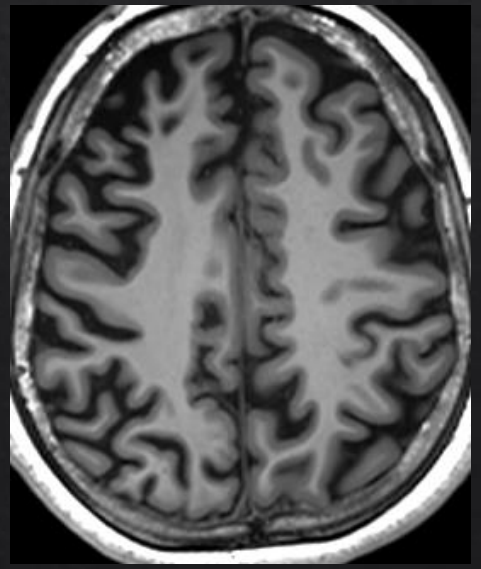
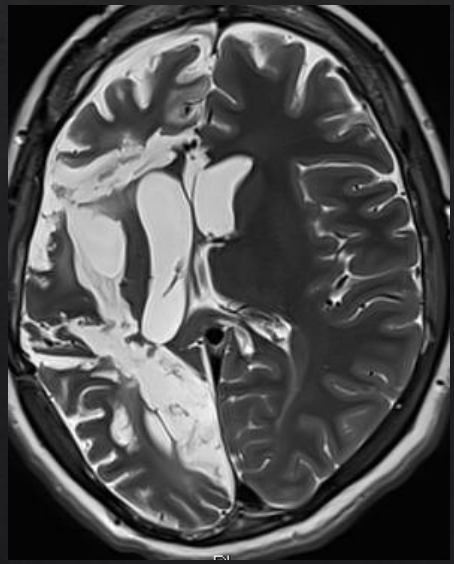




# Solo Estriado



# Atrofia, cup y swi

# Conclusiones

- Síndrome de Rasmussen
  - Amplio espectro de alteraciones
  - VARIABILIDAD:
    - Localización
    - Expresión clínica
    - Imagen
    - Evolución
- Diagnóstico precoz importante
  - Previene deterioro neurológico
  - Mejora resultados
- La combinación de RM de alta resolución (Cup, Pálido) con imagen funcional permite valorar lesiones sutiles y optimiza los resultados