NEUROIMAGING NELLE EPILESSIE

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Protocollo RM

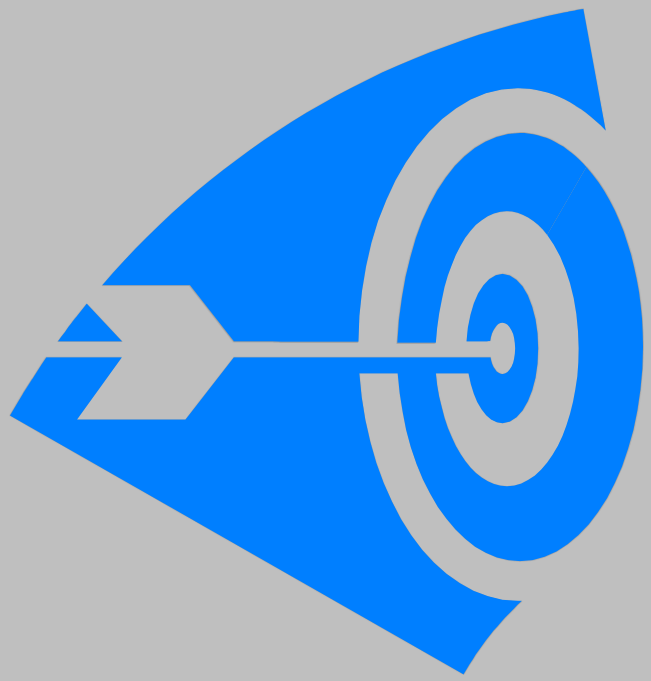
- Assiali T2 5 mm
- Assiali T2 FLAIR 5 mm
- Coronali T2 3 mm
- Coronali T1 IR 3 mm
- Coronali T2 FLAIR 3 mm
- Sagittali T2 FLAIR 3 mm

In caso di sospetto danno della BEE

- Assiali &/o coronali

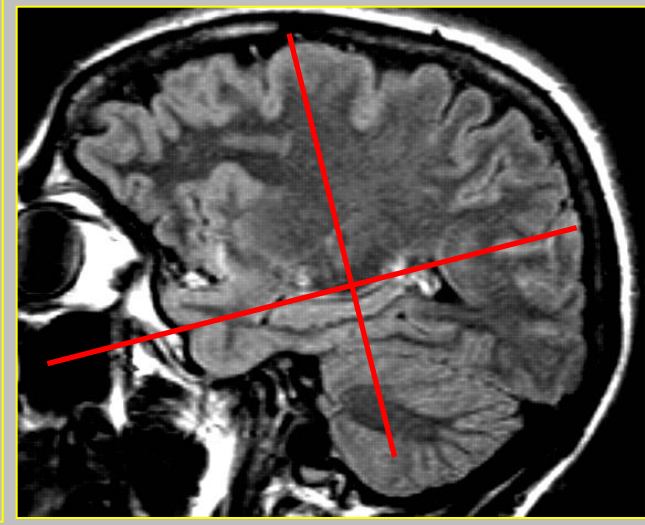
  SE-T1WI senza e con contrasto 5 mm

Esame RM basato sui dati elettroclinici



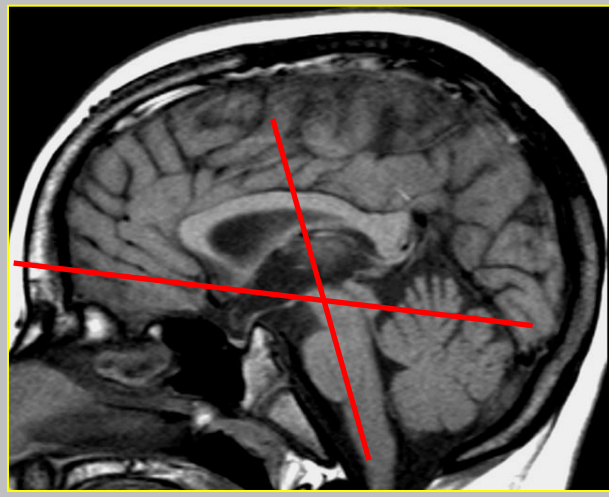
Epilessia temporale

- *Scansioni coronali ortogonali all' asse maggiore dell'ippocampo*
- *Scansioni assiali parallele all' asse maggiore dell'ippocampo*



Epilessia extra-temporale

- *Scansioni assiali parallele alla linea bicommissurale*
- *Scansioni coronali parallele al pavimento del IV° ventricolo*



Risultati neuropatologici su 1079 pz operati per epilessia parziale farmaco-resistente

• MSC (Malformazioni dello sviluppo corticale)	354	(33%)
• **Tumori**	**333**	**(31%)**
• MTS + MSC	152	(14%)
• Criptogenetici	100	(9%)
• MTS isolata	85	(8%)
• Esiti cicatriziali	55	(5%)

Anomalie di sviluppo corticale

1. **Aumentata e abnorme proliferazione**

   megalencefalia e emimegalencefalia

   Abnorme proliferazione

   *Non-neoplastica: DCF di Taylor con BC, Sclerosi tuberosa

   *Neoplastica: DNET, ganglioglioma, gangliocitoma

2. **Migrazione:** Agyria/pachigyria-band spectrum, lissencefalie tipo 1 e 2 e altri tipi, eterotopie nodulari, singoli neuroni eterotopici nella sostanza bianca

3. **Organizzazione:** Polimicrogiria/schizencefalia, DCF senza BC (cito- e architetturali)

R.I.Kuznieky, A.J.Barkovich – Brain & Development 2001

Malformations of Cortical Development

Barkovich A.J. et al – Brain 2012

1. Abnormal neuronal and glial proliferation or apoptosis

A- Congenital microcephaly (MIC)

B- Megalencephaly (MEG)

C- Cortical disgenesis with abnormal cells proliferation

* Non-neoplastic: tuberous sclerosis, FCD IIa & IIb

* Neoplastic: DNET, ganglioglioma, gangliocytoma

2. Abnormal neuronal migration

A- neuroependymal abnormalities: periventricular heterotopia

B- generalized abnormal transmantle migration: LIS classic & other types

C- late radial or tangential transmantle migration: subcortical heterotopia

3. Abnormal postmigrational development

A- Polymicrogyria / schizencephaly

B- Cortical dysgenesis secondary to inborn errors of metabolism

C- FCD type I, FCD type III (a,b,c,d)

Focal Cortical Dysplasia (FCD) Taylor et al. 1971

Focal dysplasia of the cerebral cortex in epilepsy

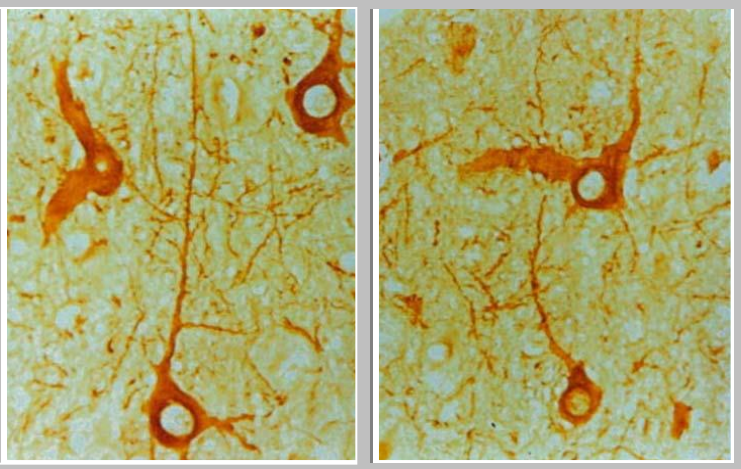
Taylor DC, Falconer MA, Bruton CJ, Corsellis JAN

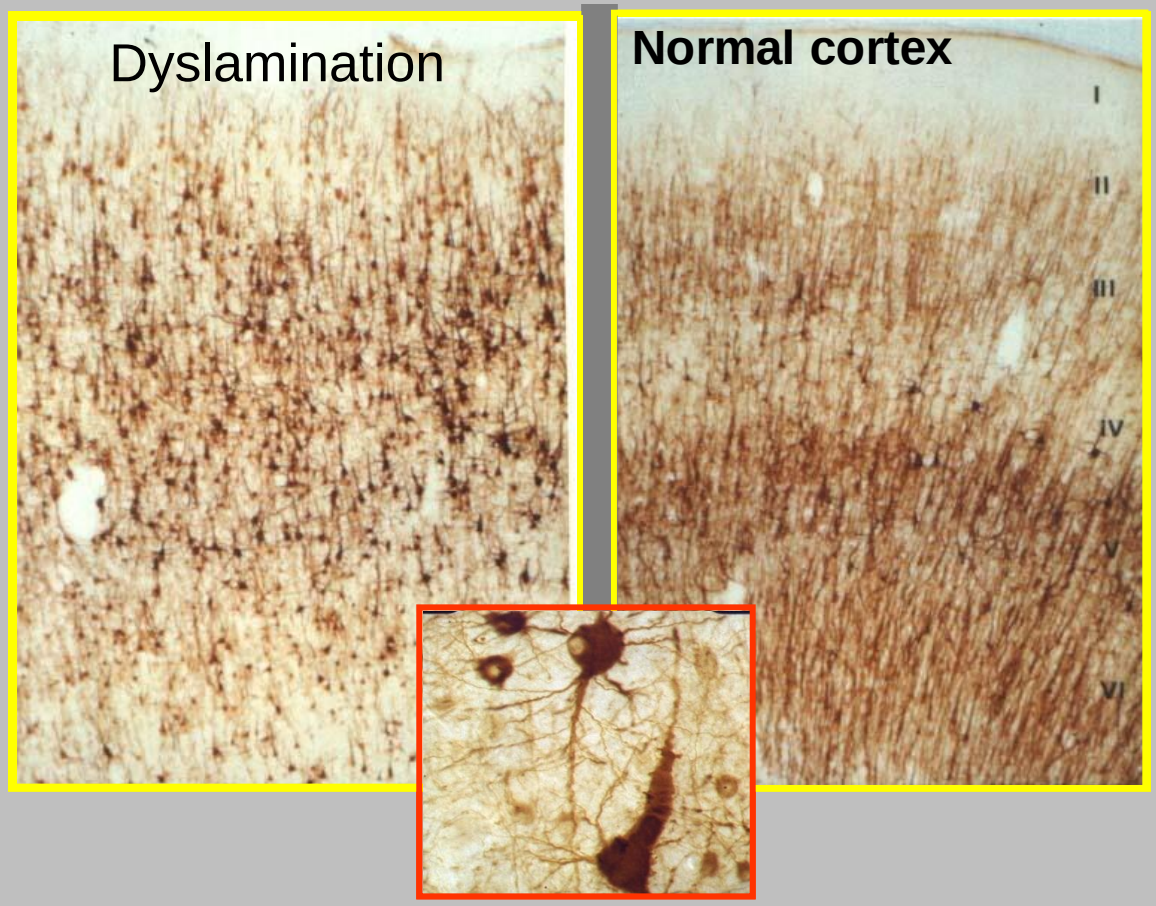
J. Neurol. Nurosurg. Psychiat. 1971; 34:369-87

FCD Type II

- disarray of cortical lamination
- dysmorphic neurons
- +/- balloon cells (+ in Type II b)

Dysmorphic neurons





Balloon cells ⇨ type II b

Brain (2002), **125**, 1719–1732

Focal cortical dysplasia: neuropathological subtypes, EEG, neuroimaging and surgical outcome

L. Tassi,[1] N. Colombo,[2] R. Garbelli,[4] S. Francione,[1] G. Lo Russo,[1] R. Mai,[1] F. Cardinale,[1] M. Cossu,[1] A. Ferrario,[4] C. Galli,[3] M. Bramerio,[3] A. Citterio[2] and R. Spreafico[4]

Type I

- **Architectural Dysplasia** → **Type I A**
- **Cytoarchitectural Dysplasia** → **Type I B**

Type II

- **Taylor's Dysplasia without balloon cells** → **Type II A**
- **Taylor's Dysplasia with balloon cells** → **Type II B**

Terminology and classification of the cortical dysplasias

A. Palmini, MD, PhD; I. Najm, MD; G. Avanzini, MD; T. Babb, PhD; R. Guerrini, MD; N. Foldvary-Schaefer, DO; G. Jackson, MD; H.O. Lüders, MD, PhD; R. Prayson, MD, PhD; R. Spreafico, MD, PhD; and H.V. Vinters, MD

Neurology - 2004

Classificazione istologica delle DCF

Tassi, Colombo, Spreafico et al. Brain 2002 Aug: 125 (Pt 8): 1719-32 / Colombo, Tassi, Spreafico et al. AJNR 24:724-733, April 2003

- **Architetturale (AD)** **(I A)** — **neuroni eterotopici nella SB**
  lieve dislaminazione corticale
- **Citoarchitetturale (CD)** **(I B)** — **neuroni eterotopici nella SB**
  > dislaminazione corticale
  neuroni giganti
- **DCF di Taylor senza BC** **(IIA)** — **neuroni eterotopici nella SB**
  severa dislaminazione corticale
  neuroni giganti e dismorfici
- **DCF di Taylor con BC** **(IIB)** — **neuroni etetrotopici nella SB**
  severa dislaminazione corticale
  neuroni giganti e dismorfici
  cellule balloniformi

The histopathological spectrum of Focal Cortical Dysplasias : a revised classification system of the International League against Epilepsy

I. Blumcke et al. Epilepsia 1-17, 2010

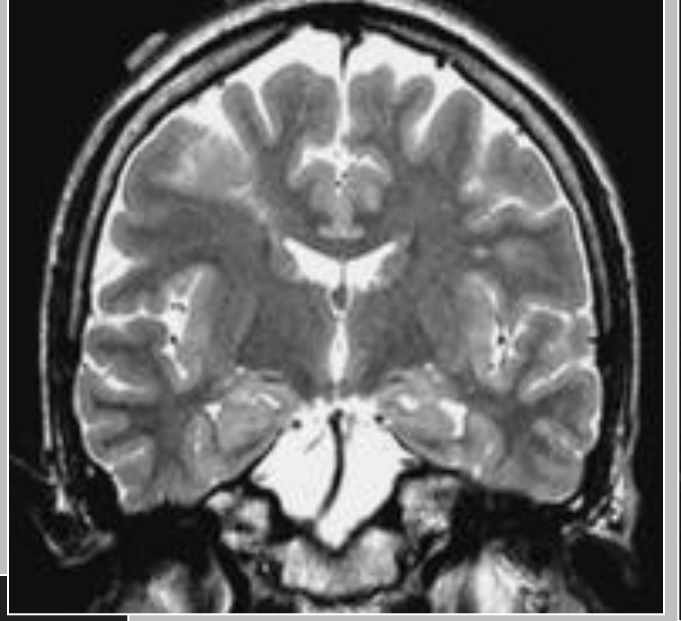
FCD Type I (isolated)	Focal Cortical Dysplasia with abnormal radial cortical lamination **(FCD Ia)**	Focal Cortical Dysplasia with abnormal tangential cortical lamination **(FCD Ib)**	Focal Cortical Dysplasia with abnormal radial and tangential cortical lamination **(FCD Ic)**	
FCD Type II (isolated)	Focal Cortical Dysplasia with dysmorphic neurons **(FCD IIa)**		Focal Cortical Dysplasia with dysmorphic neurons and balloon cells **(FCD IIb)**	
FCD Type III (associated with principal lesion)	Cortical layer abnormalities in the temporal lobe associated with hippocampal sclerosis **(FCD IIIa)**	Cortical layer abnormalities adjacent to a glial or glio-neuronal tumor **(FCD IIIb)**	Cortical layer abnormalities adjacent to vascular malformation **(FCD IIIc)**	Cortical layer abnormalities adjacent to any other lesion acquired during early life, e.g., trauma, porencephaly, encephalitis **(FCD IIId)**

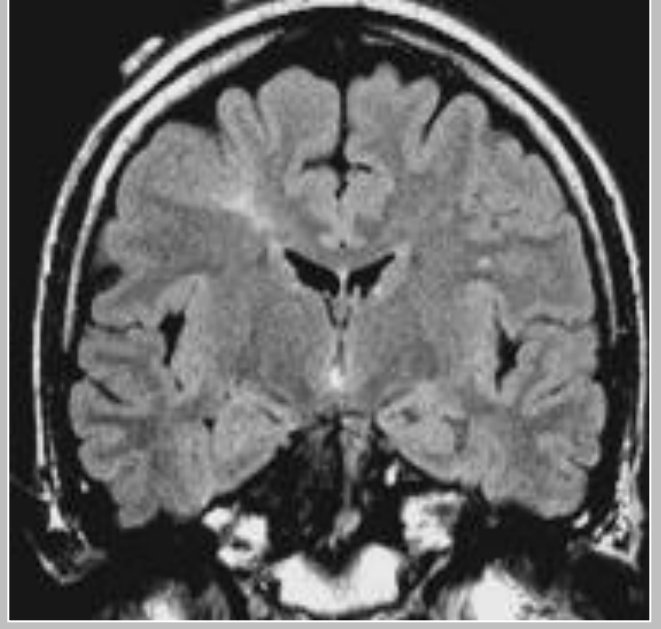
DCF tipo II (Taylor) - Aspetti RM

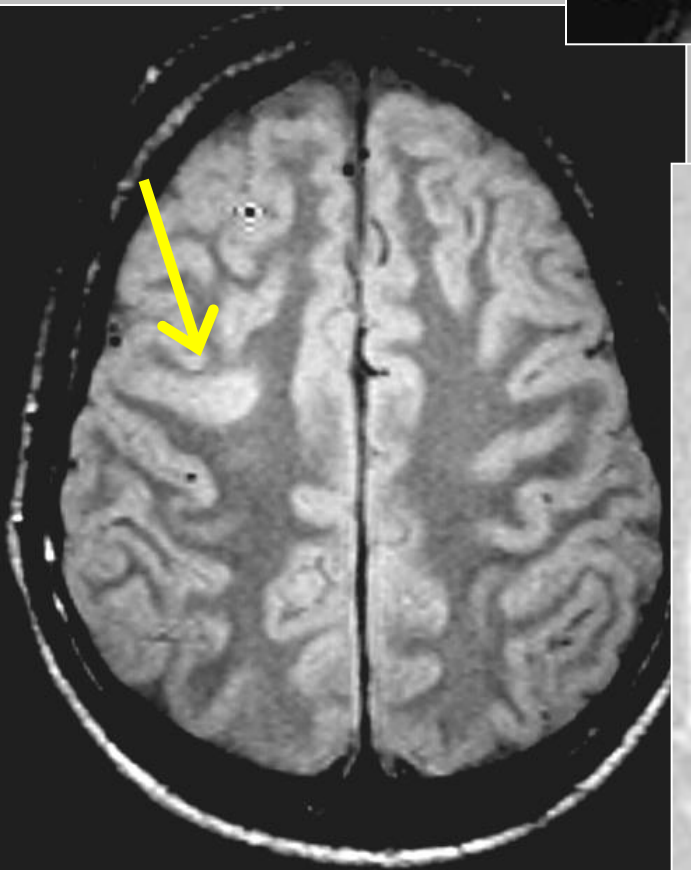
- Aumentato spessore della corteccia (pseudoispessimento)
- Maldefinizione (“blurring”) della giunzione SB/SG, (in alcuni casi il limite è netto)
- Ipersegnale in T2 della SB sottocorticale esteso radialmente al ventricolo (*transmantle sign*) peculiare delle FCD tipo II, più frequente nelle IIB
- Ipersegnale della corteccia in T2
- Girazione corticale dismorfica
- No edema, calcificazioni, effetto massa

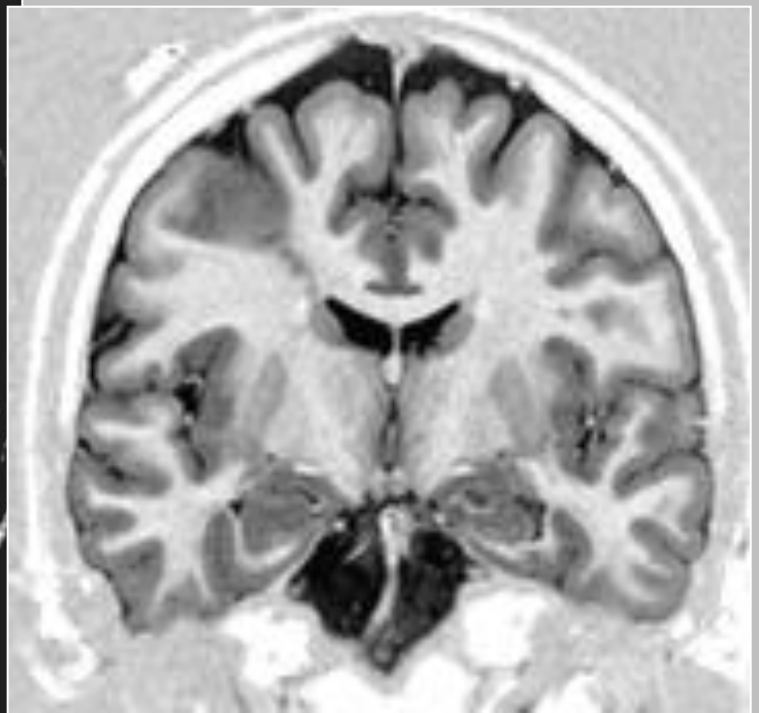
✓*(Colombo et al. Neuroradiology 2012).*

FCD type IIb (BC +)



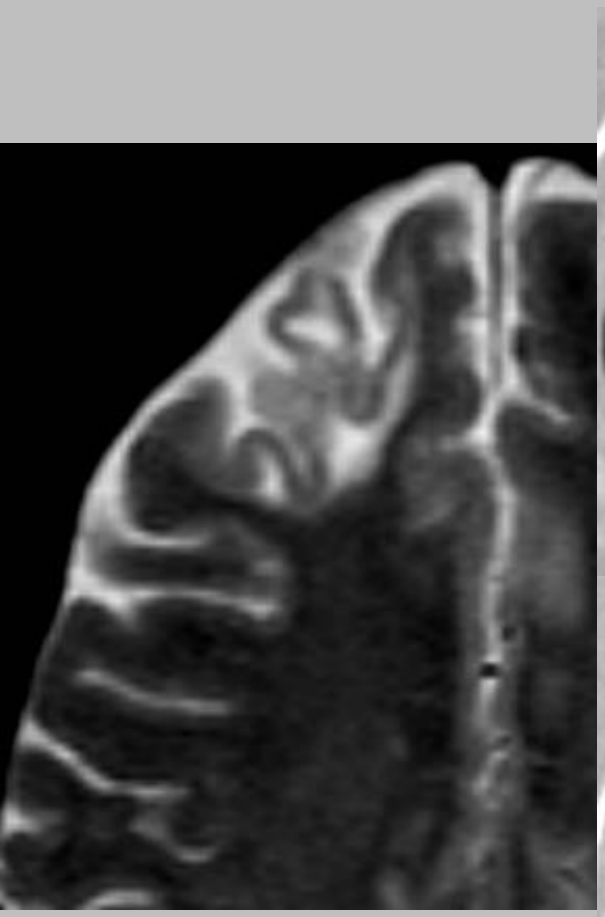


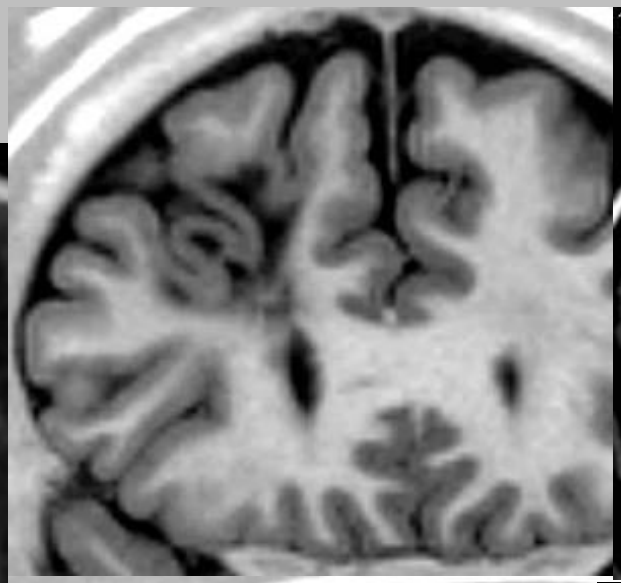


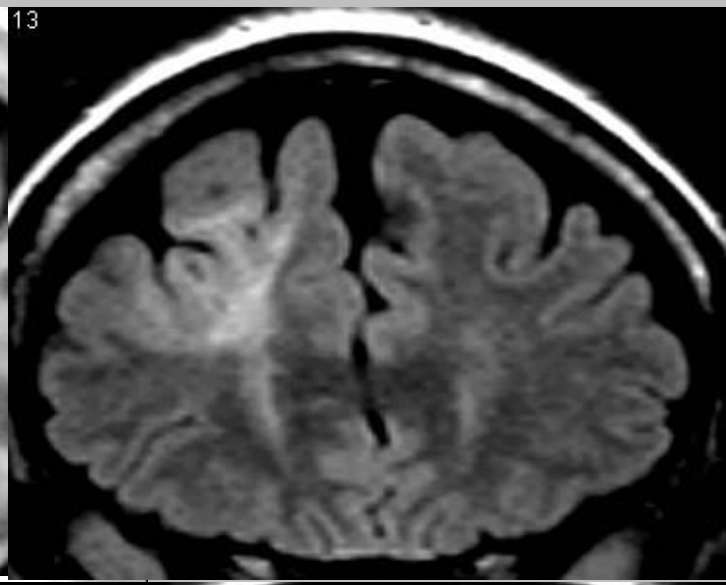


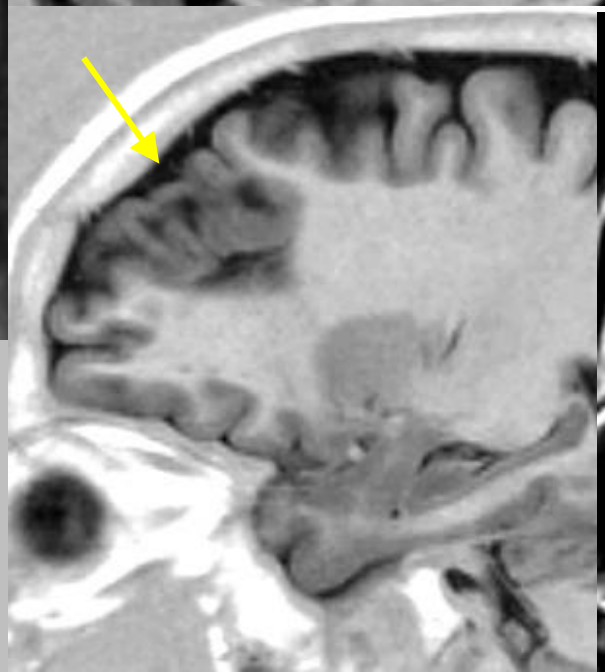
Z.M.
R. Fr. Epilepsy
Onset: 7 yrs
Surgery: 16 yrs
Sz freq./m: 20
Outcome: I a

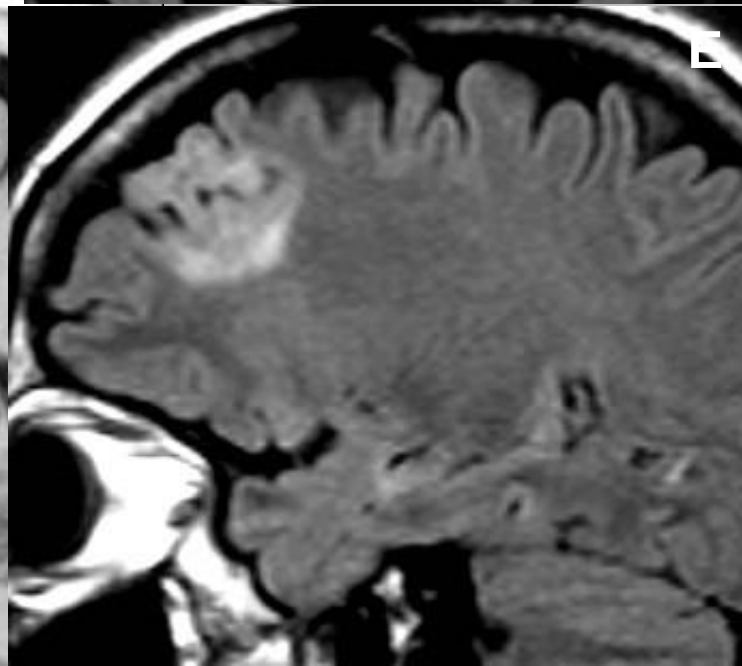
FCD type IIb (BC +)









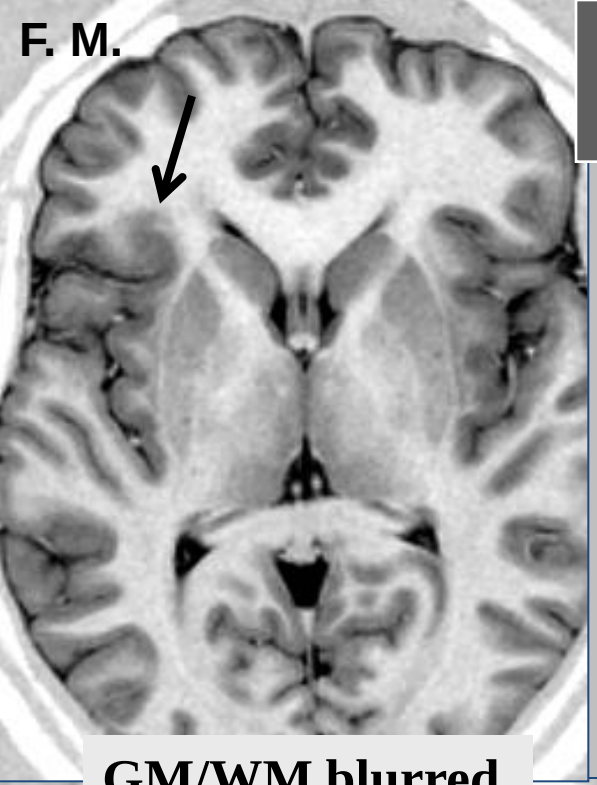


S.S.

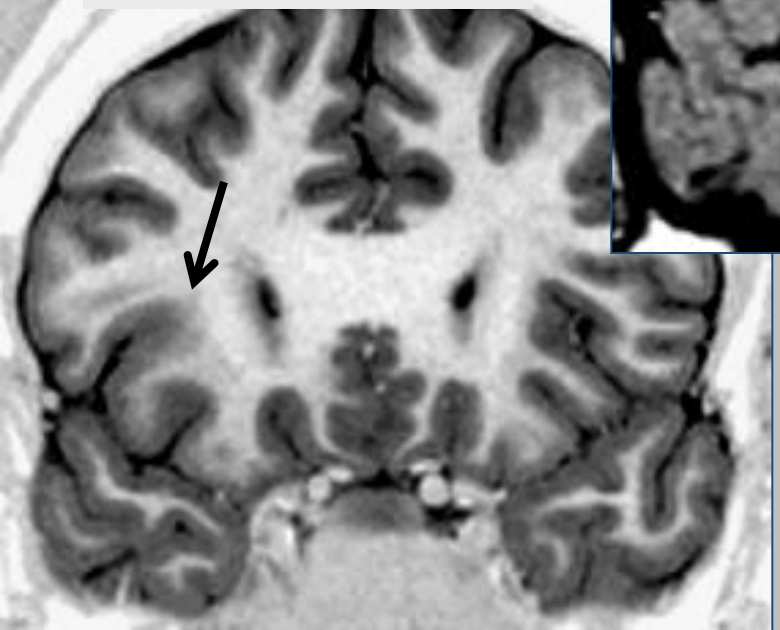
F. M.

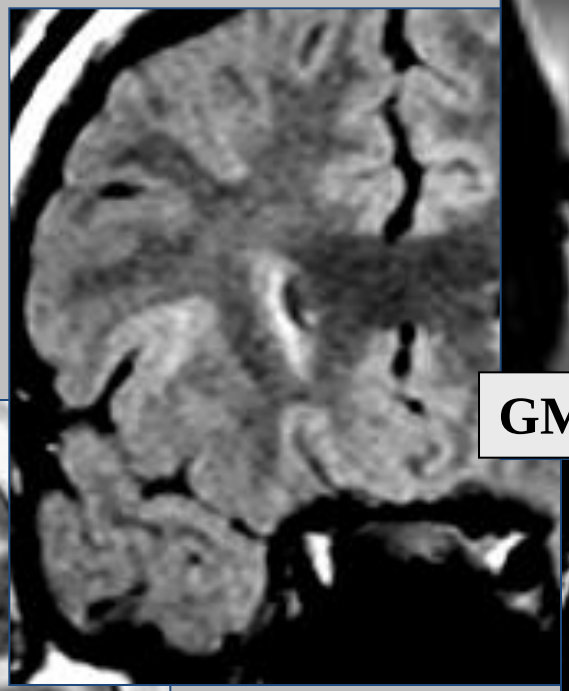
FCD type IIb (BC +)

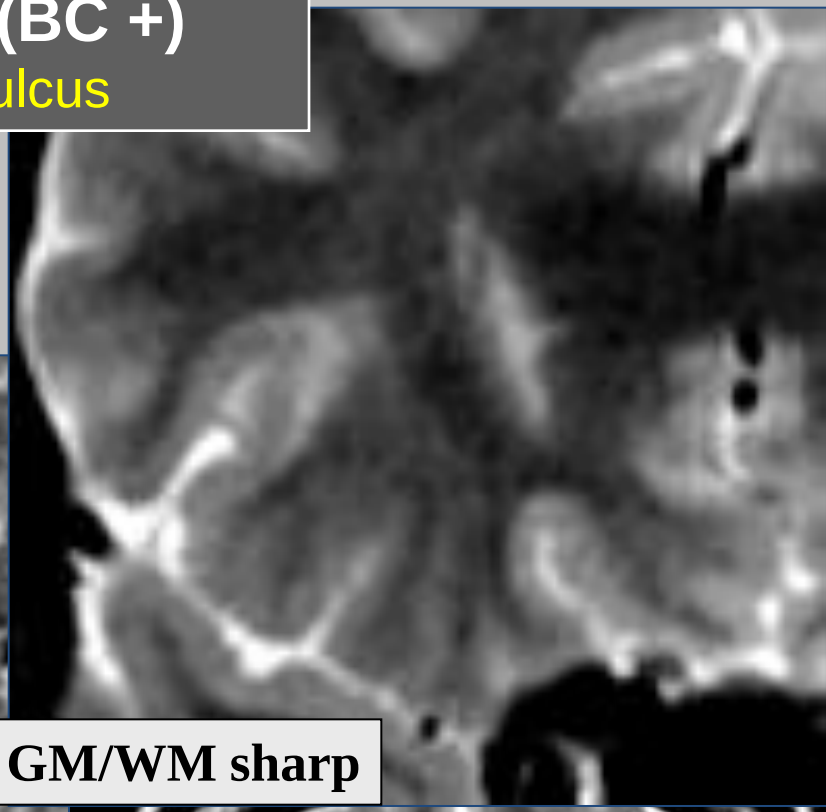
deep in the sulcus



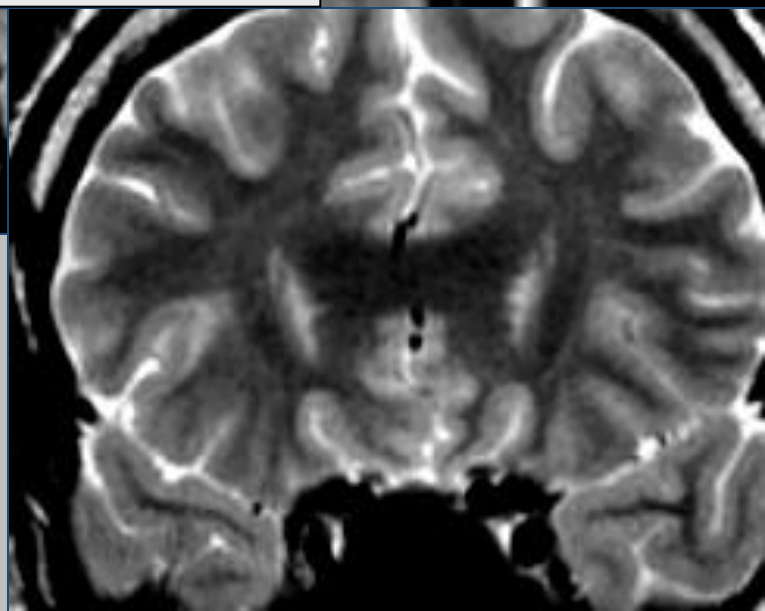
GM/WM blurred.





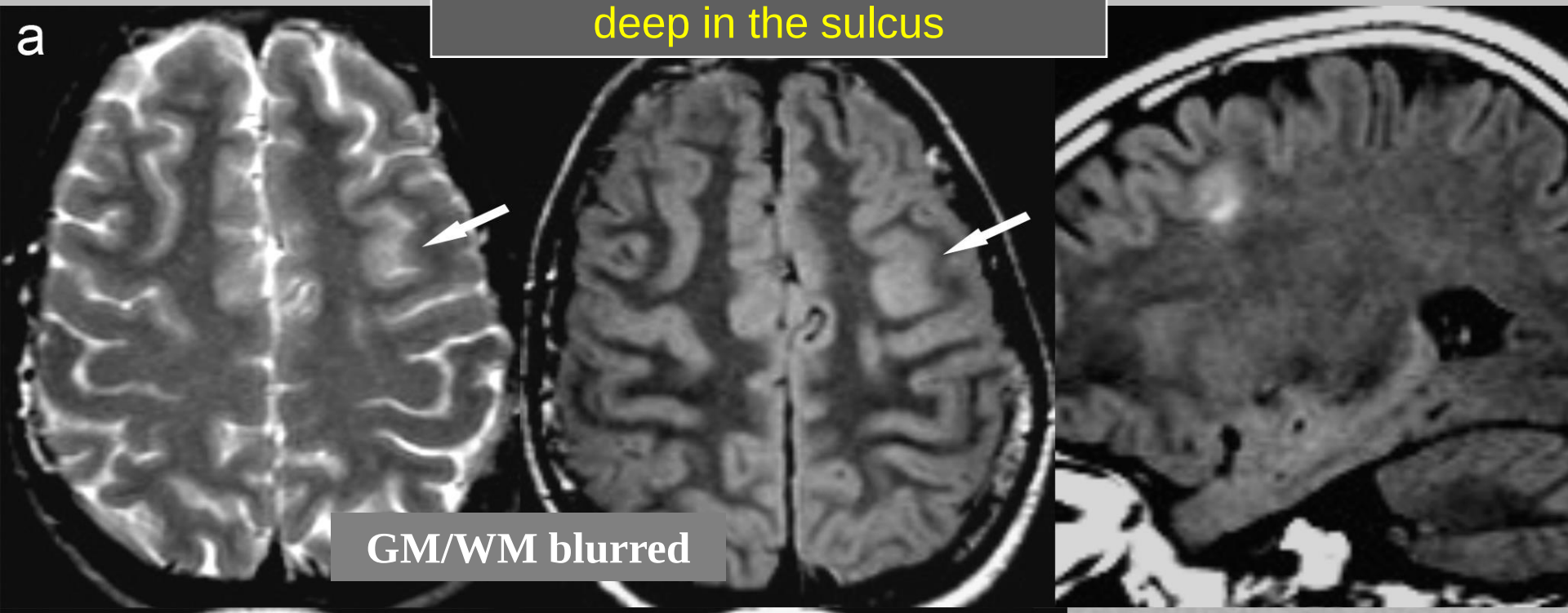


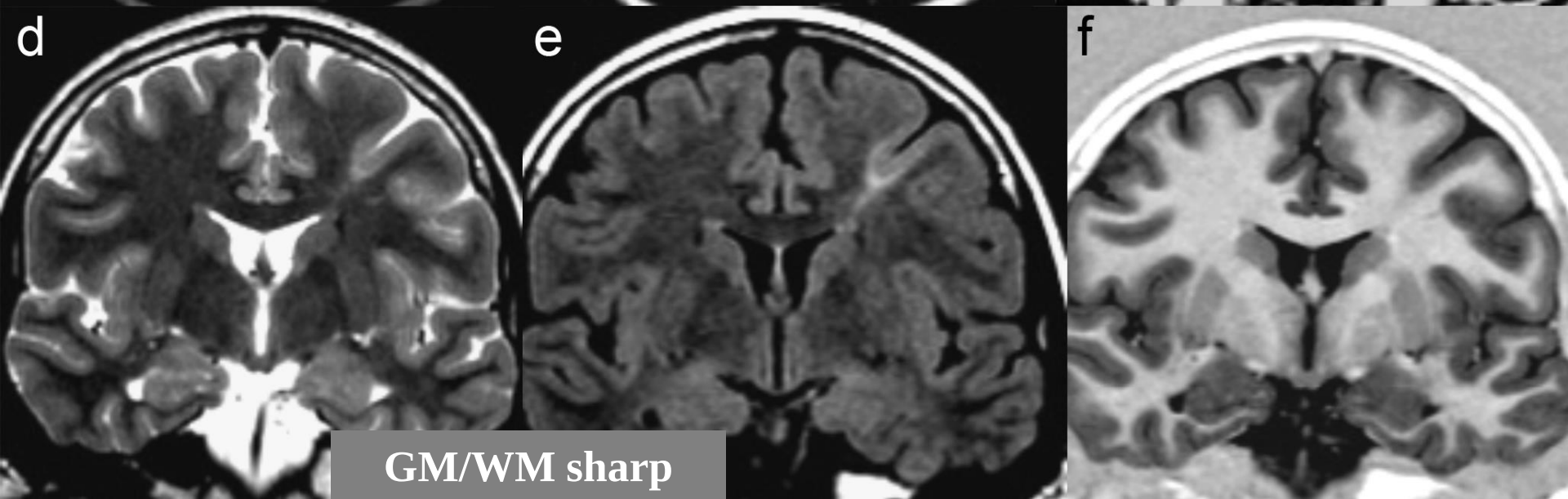
GM/WM sharp

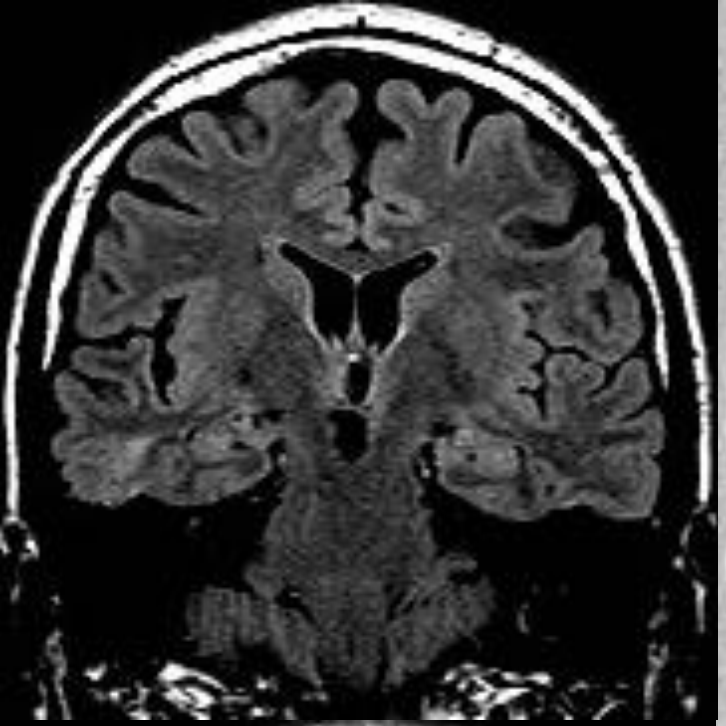


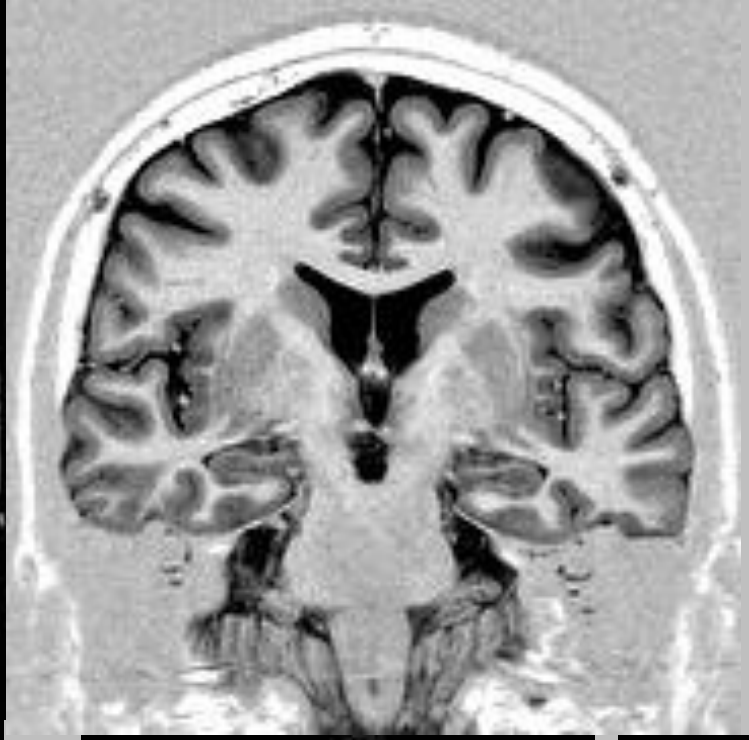
FCD type IIb (BC +)

deep in the sulcus

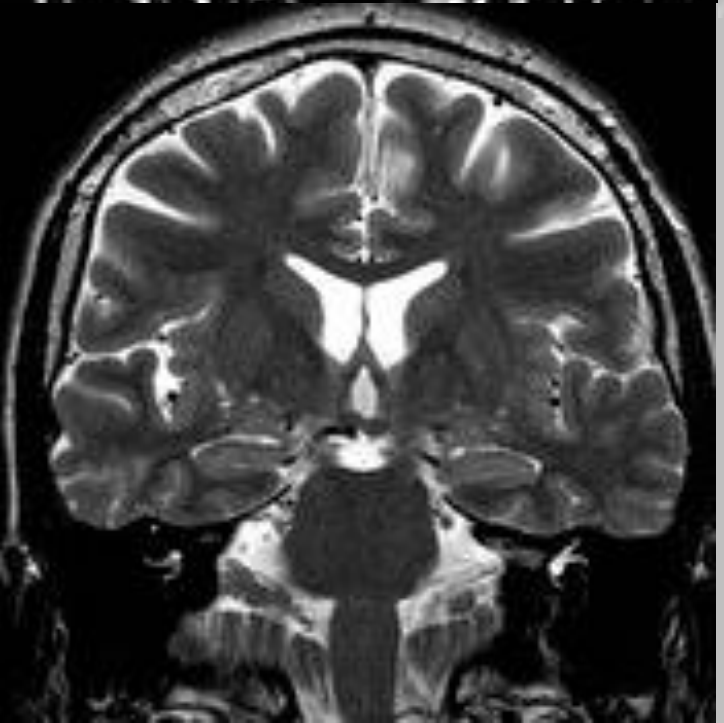


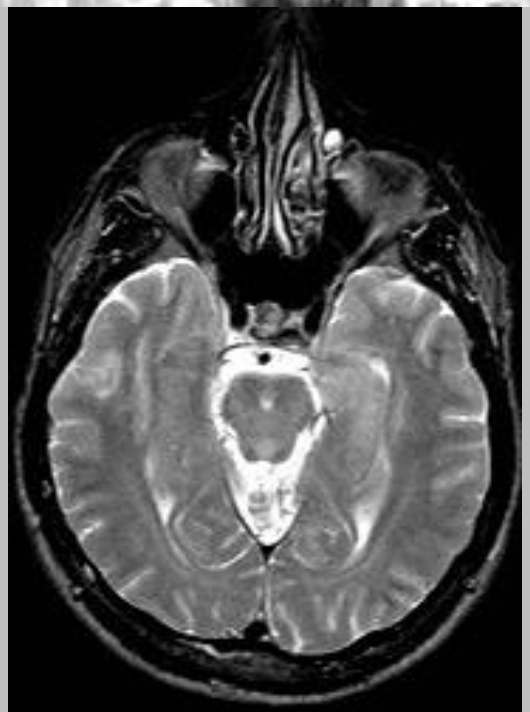


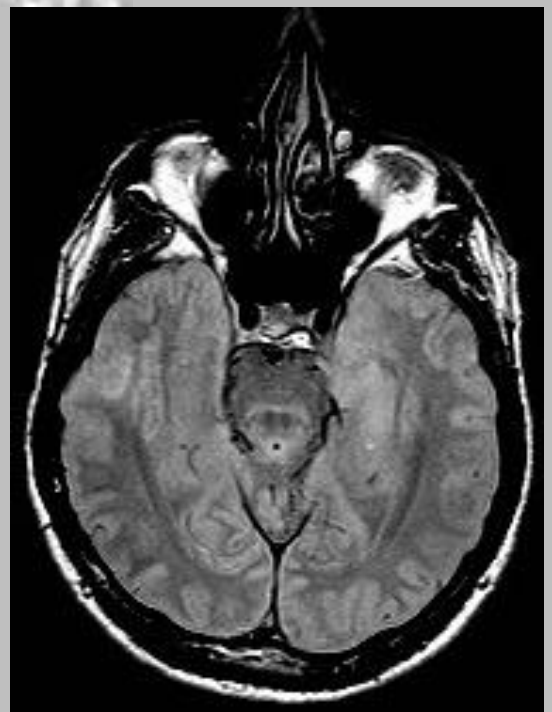




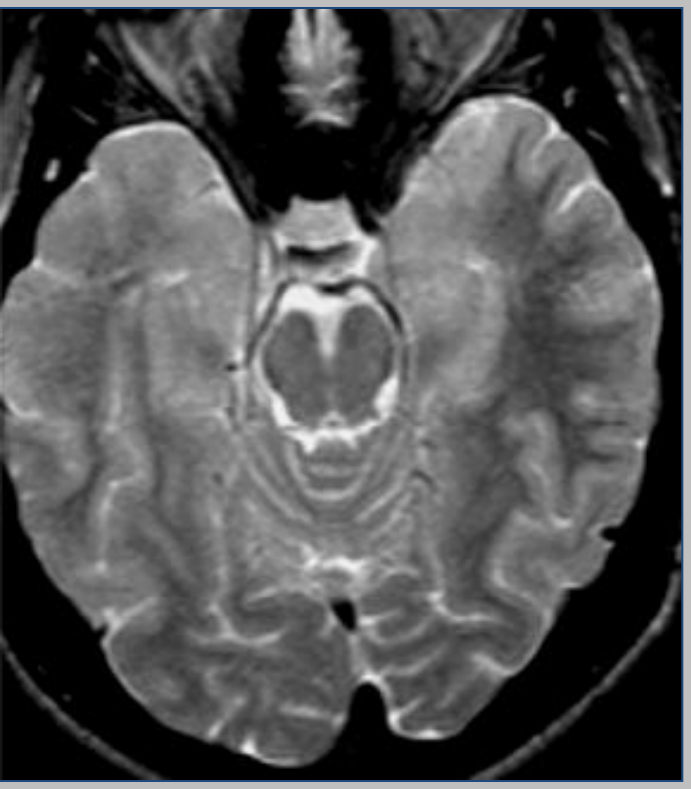
DCF tipo Taylor IIb con BC

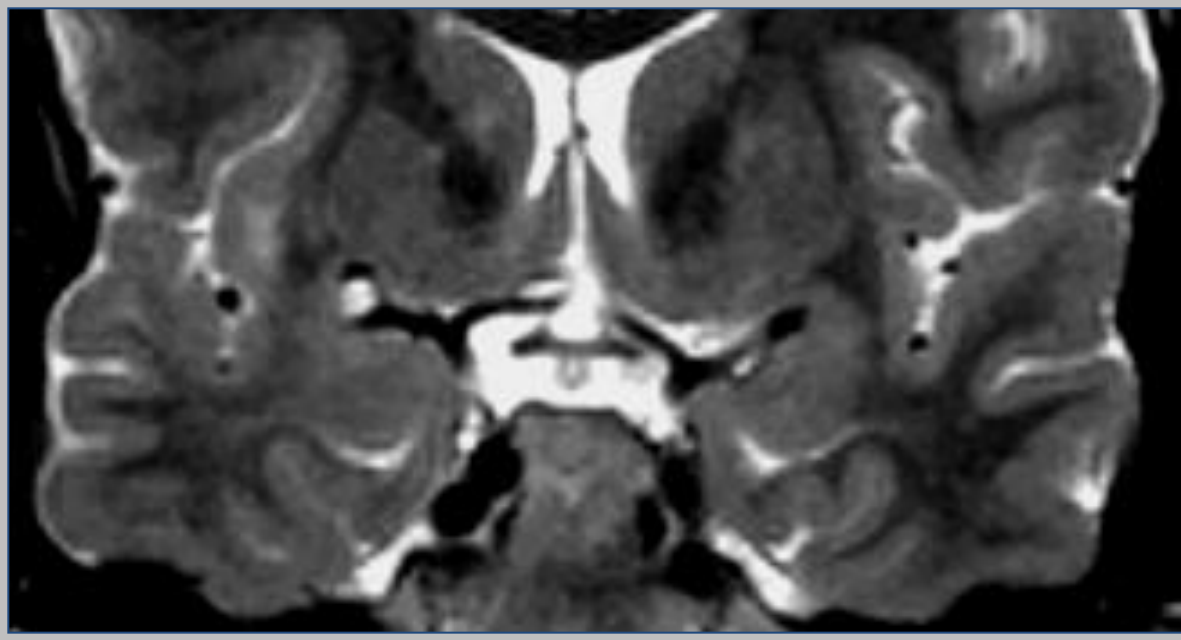


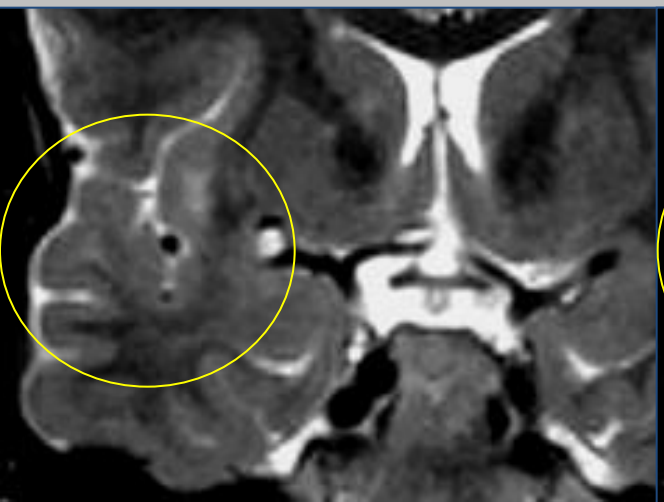


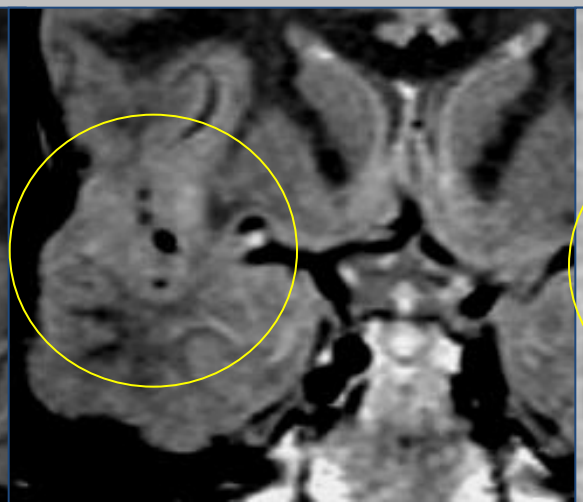


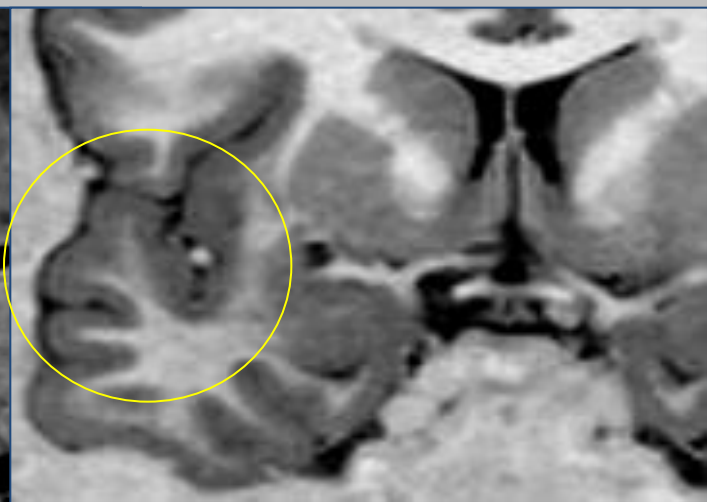
FCD type IIa (BC -)



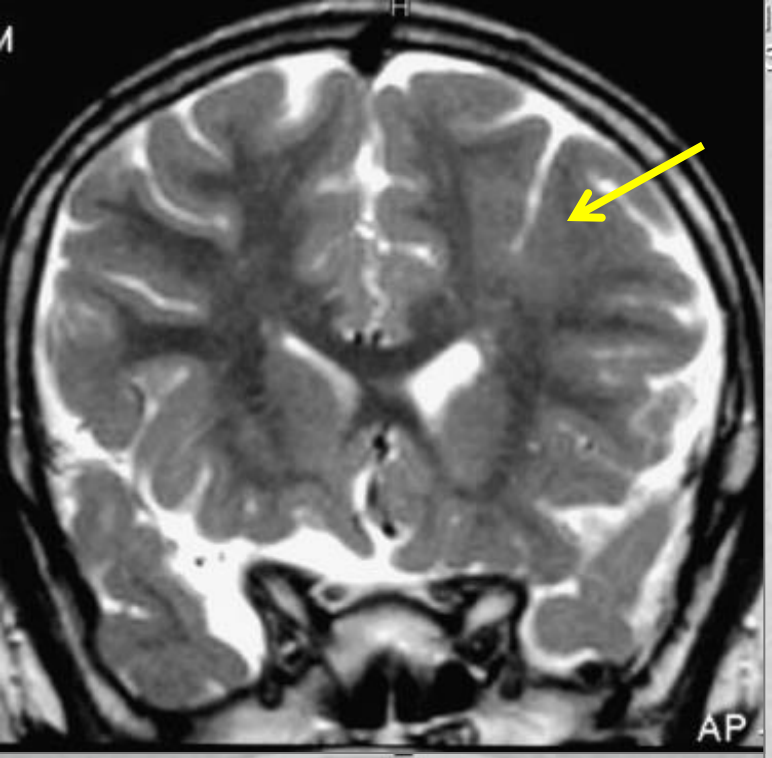


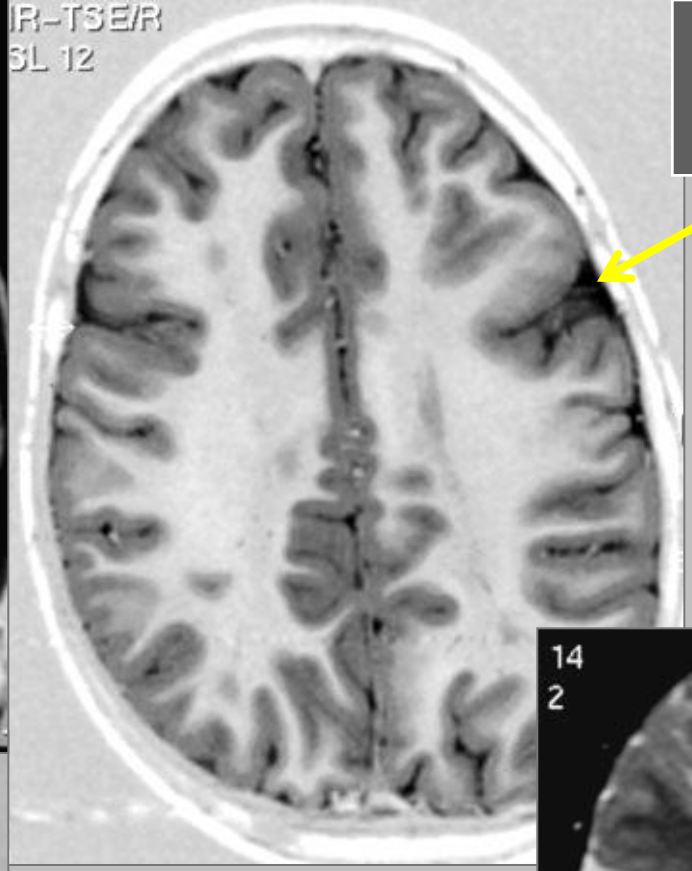


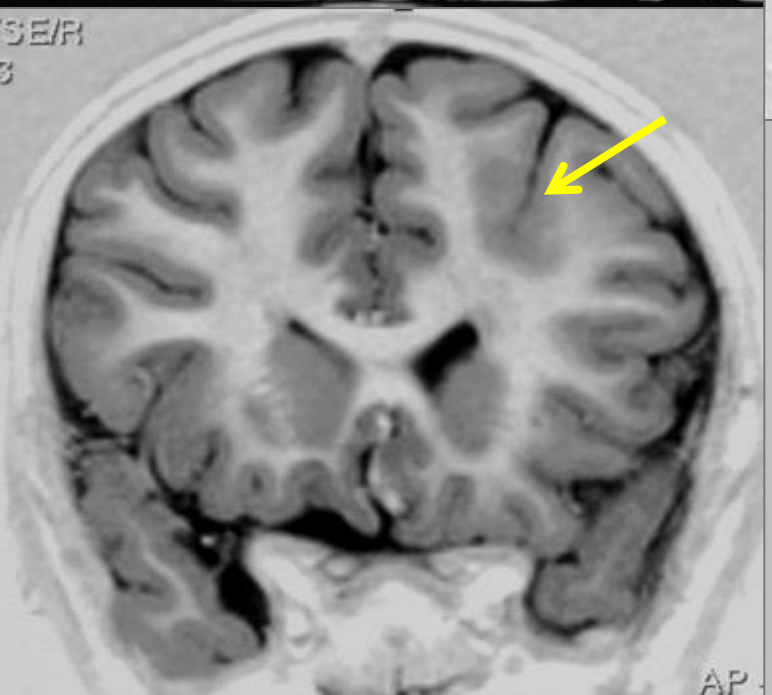


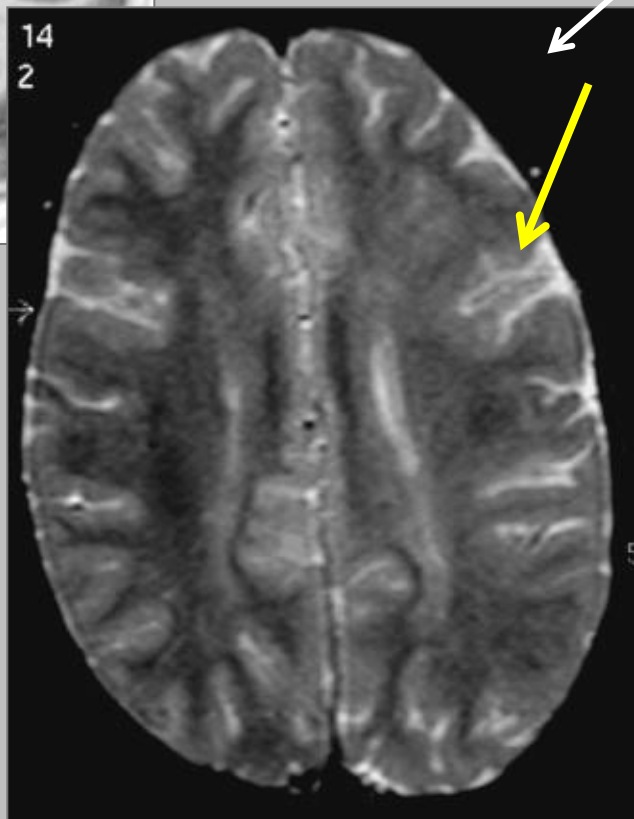


FCD type IIa (BC -)





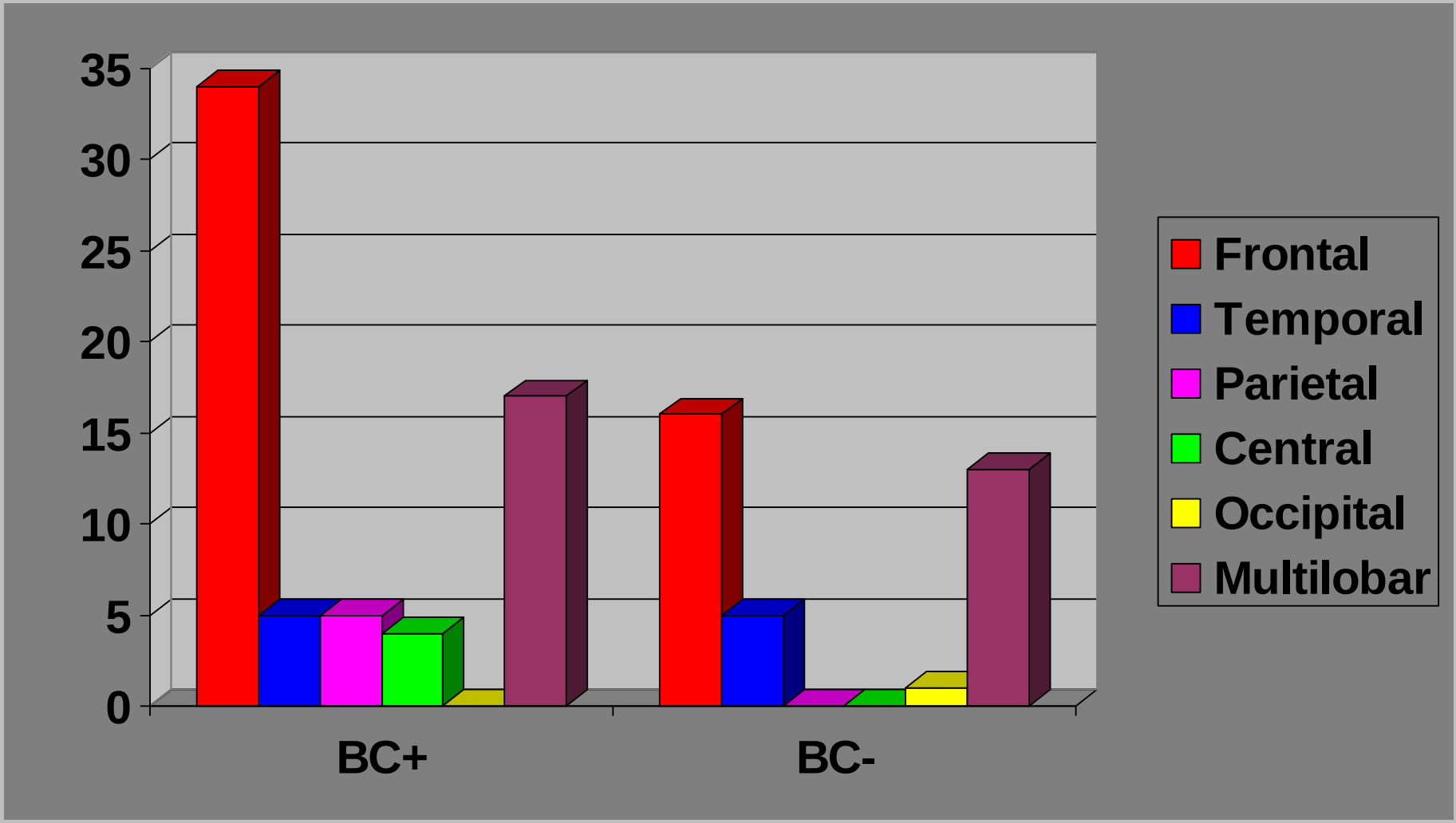




M.R.

Lobe of intervention

118 pts with FCD Type II



118 FCD Type II.

Comparison of histopathology and retrospective MRI diagnosis

Histopathology	MRI abnormal	MRI normal
FCD type IIa (n= 37)	19 (51%)	18 (49%)
FCD type IIb (n=81)	74 (91%)	7 (9%)
Total 118	93 (79%)	25 (21%)

118 FCD Type II.

Patients characteristics and surgical outcome.

Histology	M/F	Seizure outcome (Engel Class I) after 2 years from intervention
FCD IIa (n=37)	22/15	25 (68%)
FCD IIb (n=81)	38/43	74 (91%)
Total (n=118)	60/58	99 (84%)

Epilepsia, **(*):1–17, 2010
doi: 10.1111/j.1528-1167.2010.02777.x

SPECIAL REPORT

The clinicopathologic spectrum of focal cortical dysplasias: A consensus classification proposed by an ad hoc Task Force of the ILAE Diagnostic Methods Commission[1]

*[2]Ingmar Blümcke, †Maria Thom, ‡Eleonora Aronica, §Dawna D. Armstrong, ¶Harry V. Vinters, #Andre Palmini, **Thomas S. Jacques, ††Giuliano Avanzini, ‡‡A. James Barkovich, §§Giorgio Battaglia, ¶¶Albert Becker, ##Carlos Cepeda, ***[3]Fernando Cendes, †††Nadia Colombo, ‡‡‡Peter Crino, §§§J. Helen Cross, ¶¶¶Olivier Delalande, ###François Dubeau, ****John Duncan, ††††Renzo Guerrini, ‡‡‡‡Philippe Kahane, §§§§Gary Mathern, ¶¶¶¶Imad Najm, ####Çiğdem Özkara, *****Charles Raybaud, †††††Alfonso Represa, ‡‡‡‡‡Steven N. Roper, §§§§§Noriko Salamon, ¶¶¶¶¶Andreas Schulze-Bonhage, #####Laura Tassi, ******Annamaria Vezzani, and ††Roberto Spreafico

Table 1. The three-tiered ILAE classification system of focal cortical dysplasia (FCD) distinguishes isolated forms (FCD Types I and II) from those associated with another principal lesion (FCD Type III).

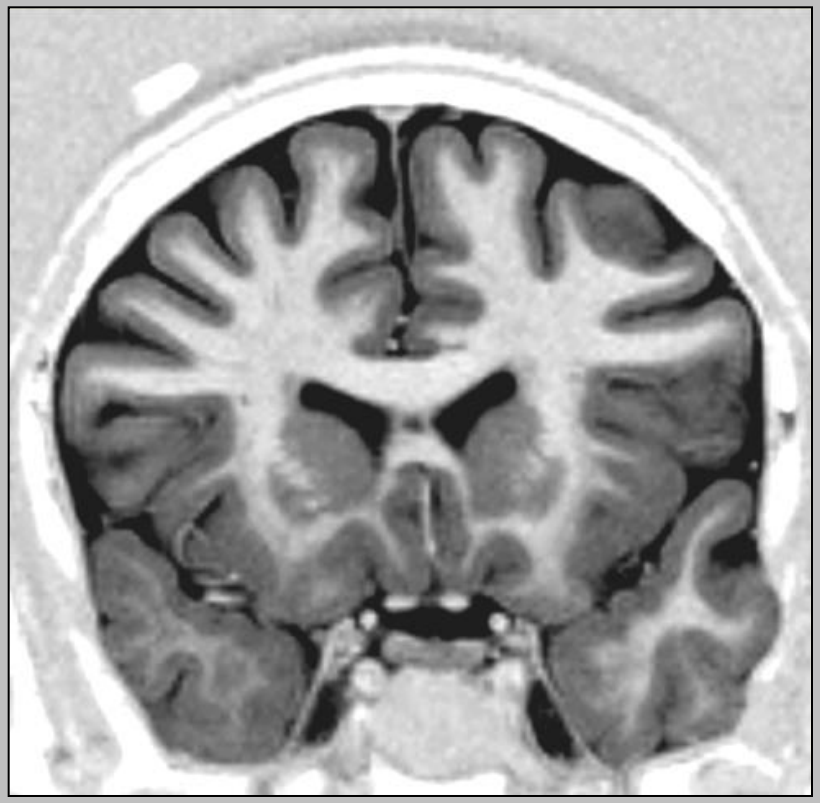
FCD Type I (isolated)	Focal cortical dysplasia with abnormal radial cortical lamination (FCD Type Ia)	Focal cortical dysplasia with abnormal tangential cortical lamination (FCD Type Ib)	Focal cortical dysplasia with abnormal radial and tangential cortical lamination (FCD Type Ic)	
FCD Type II (isolated)	Focal cortical dysplasia with dysmorphic neurons (FCD Type IIa)		Focal cortical dysplasia with dysmorphic neurons and balloon cells (FCD Type IIb)	
FCD Type III (associated with principal lesion)	Cortical lamination abnormalities in the temporal lobe associated with hippocampal sclerosis (FCD Type IIIa)	Cortical lamination abnormalities adjacent to a glial or glioneuronal tumor (FCD Type IIIb)	Cortical lamination abnormalities adjacent to vascular malformation (FCD Type IIIc)	Cortical lamination abnormalities adjacent to any other lesion acquired during early life, e.g., trauma, ischemic injury, encephalitis (FCD Type IIId)

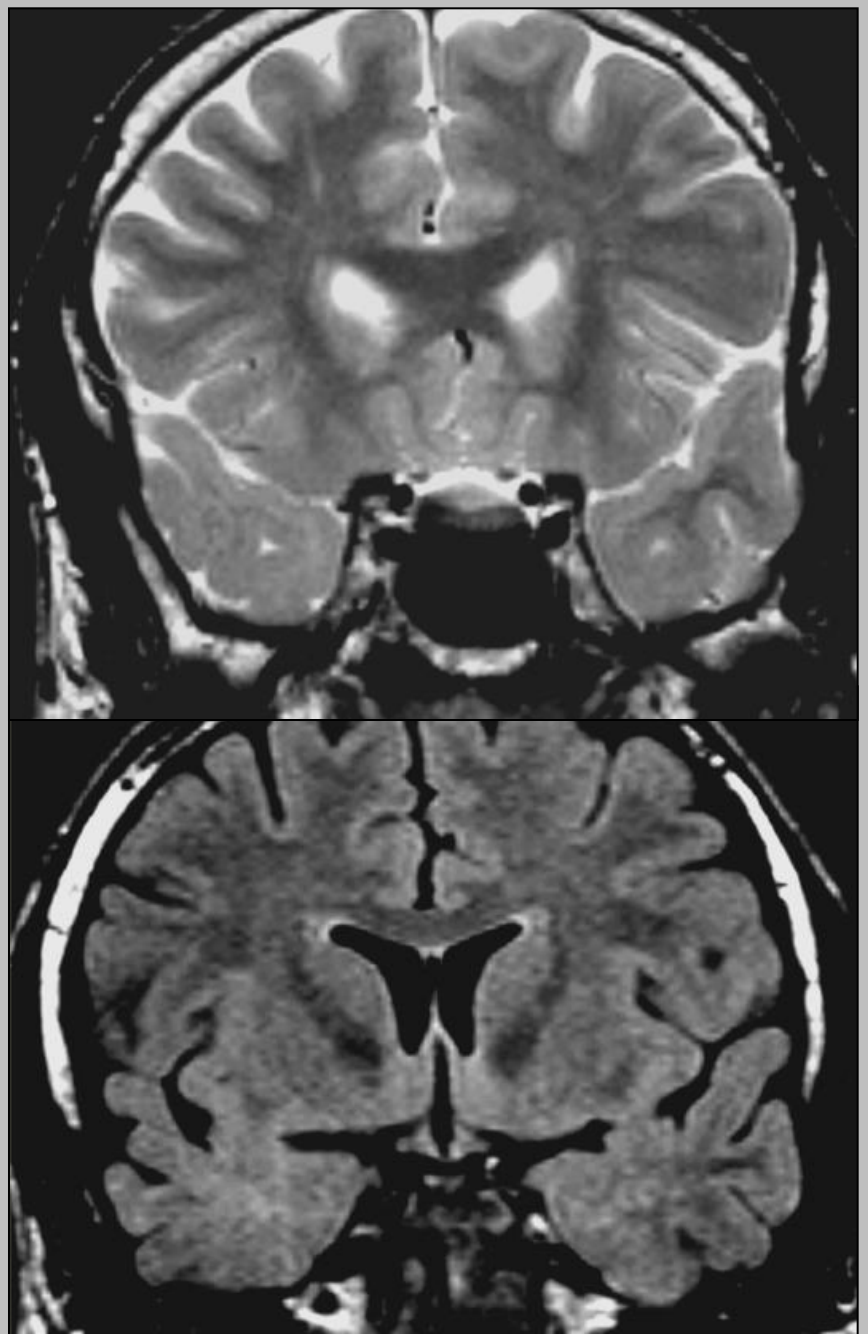
FCD Type III (not otherwise specified, NOS): if clinically/radiologically suspected principal lesion is not available for microscopic inspection.
Please note that the rare association between FCD Types IIa and IIb with hippocampal sclerosis, tumors, or vascular malformations should not be classified as FCD Type III variant.

FCD Tipo I - Aspetti RM

- Ipoplasia lobare o sublobare
- Ridotto volume della SB
- Blurring della giunzione bianca/grigia
- Iperintensità della SB in T2WI
- Mild subcortical WM hypointensity on T1WI
- Anomalie della girazione corticale
- Incremento dello spessore corticale

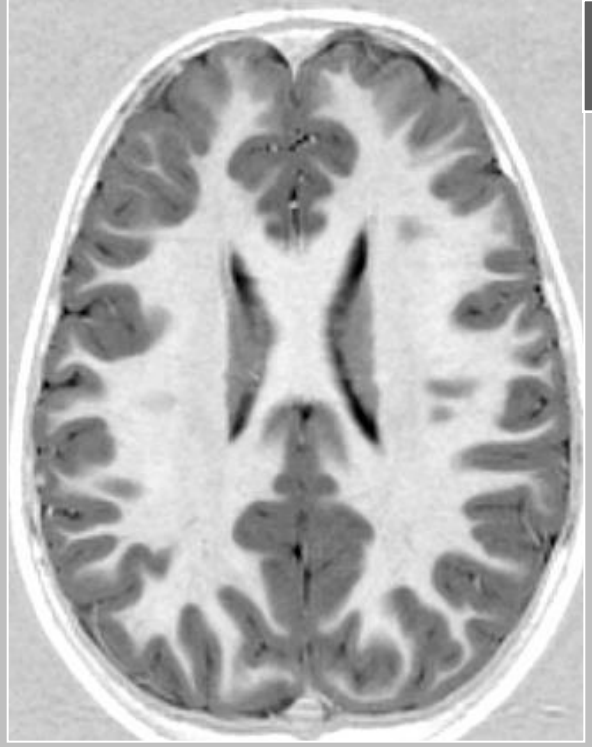
Isolated FCD Type I (no HS)

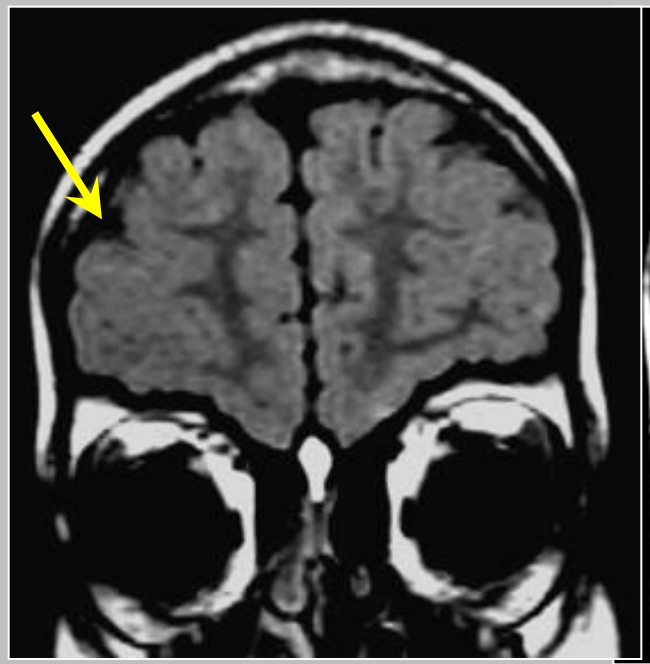


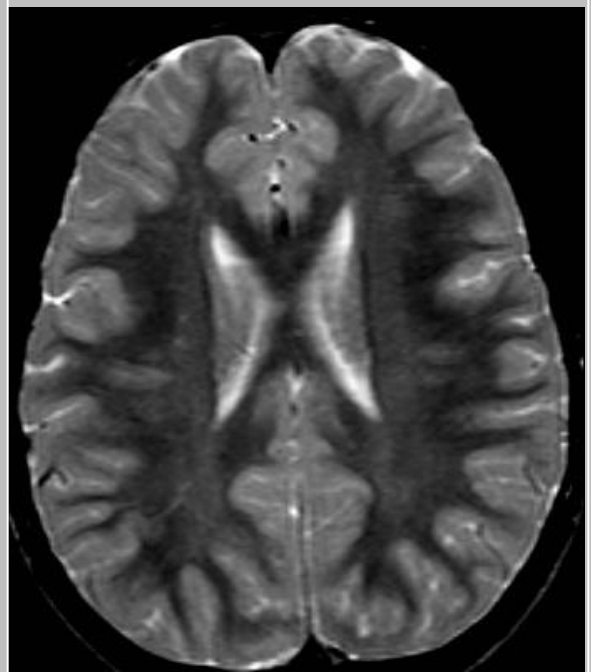


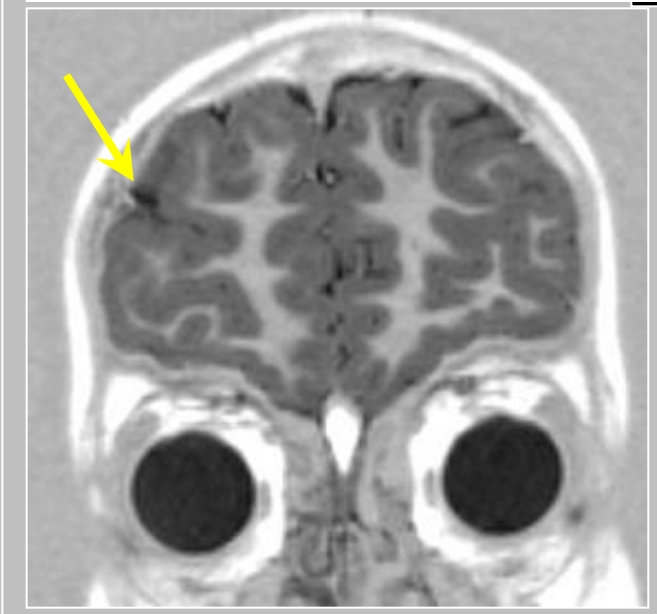
FCD Type I

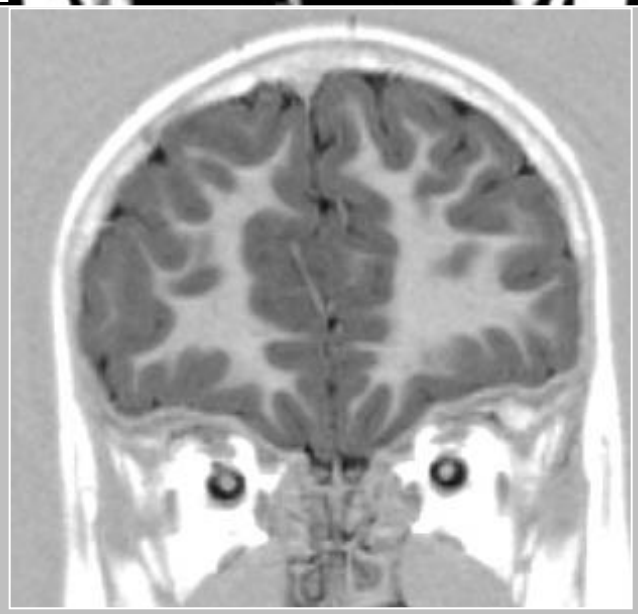
Fav. G.
R. F epilepsy
Sz freq/m: 120
Outcome I a



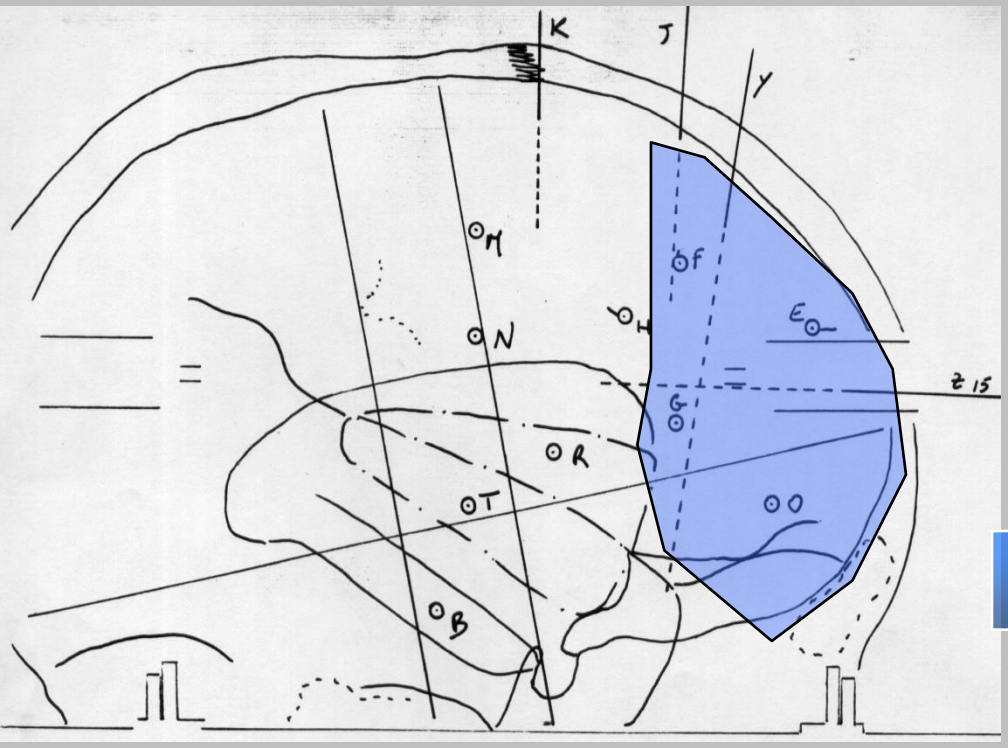








Schema LL



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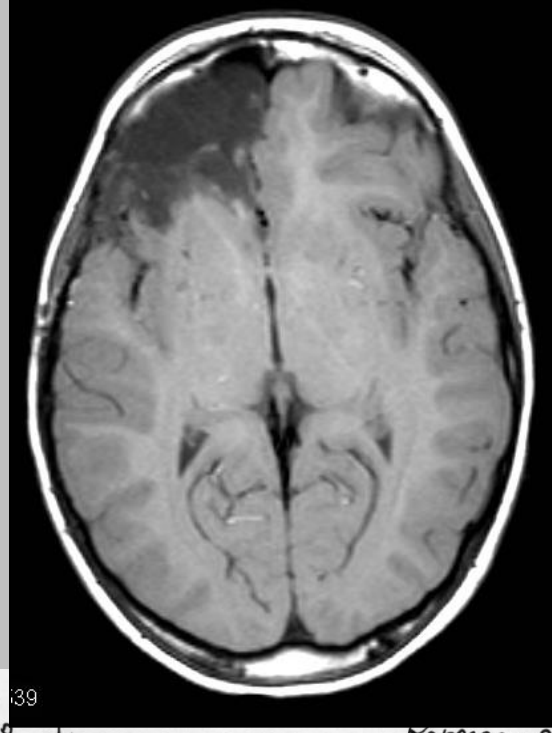
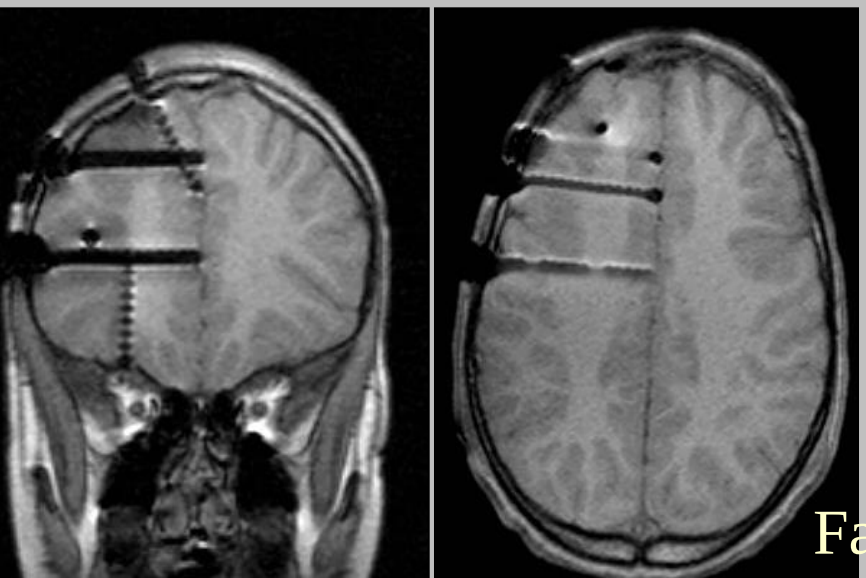
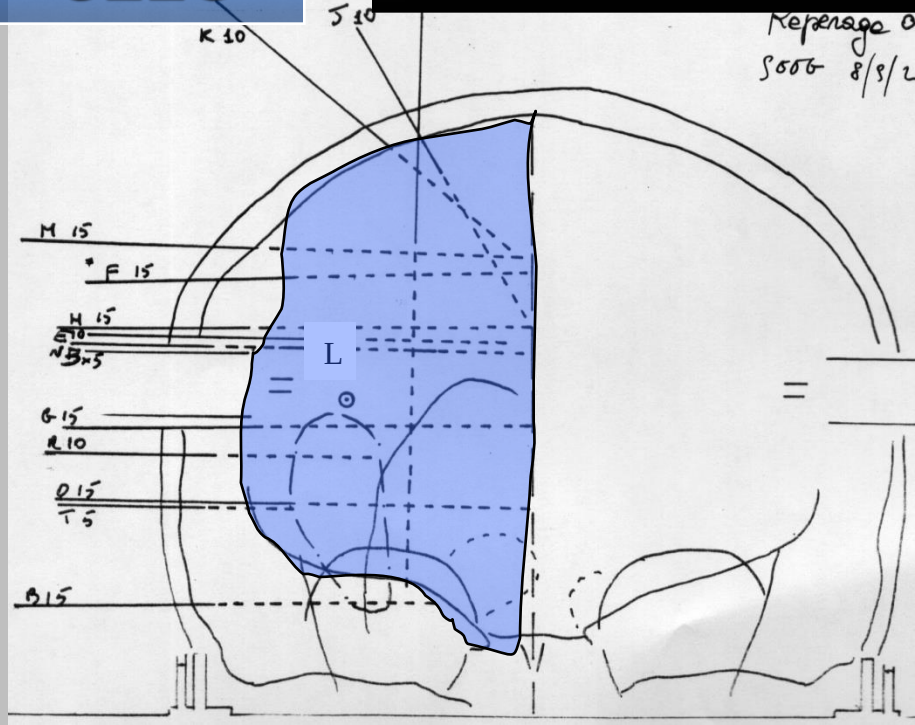


SEEG





Fav.G.

Epilepsia, **(*):1–17, 2010
doi: 10.1111/j.1528-1167.2010.02777.x

SPECIAL REPORT

The clinicopathologic spectrum of focal cortical dysplasias: A consensus classification proposed by an ad hoc Task Force of the ILAE Diagnostic Methods Commission[1]

*[2]Ingmar Blümcke, †Maria Thom, ‡Eleonora Aronica, §Dawna D. Armstrong, ¶Harry V. Vinters, #Andre Palmini, **Thomas S. Jacques, ††Giuliano Avanzini, ‡‡A. James Barkovich, §§Giorgio Battaglia, ¶¶Albert Becker, ##Carlos Cepeda, ***[3]Fernando Cendes, †††Nadia Colombo, ‡‡‡Peter Crino, §§§J. Helen Cross, ¶¶¶Olivier Delalande, ###François Dubeau, ****John Duncan, ††††Renzo Guerrini, ‡‡‡‡Philippe Kahane, §§§§Gary Mathern, ¶¶¶¶Imad Najm, ####Çiğdem Özkara, *****Charles Raybaud, †††††Alfonso Represa, ‡‡‡‡‡Steven N. Roper, §§§§§Noriko Salamon, ¶¶¶¶¶Andreas Schulze-Bonhage, #####Laura Tassi, ******Annamaria Vezzani, and ††Roberto Spreafico

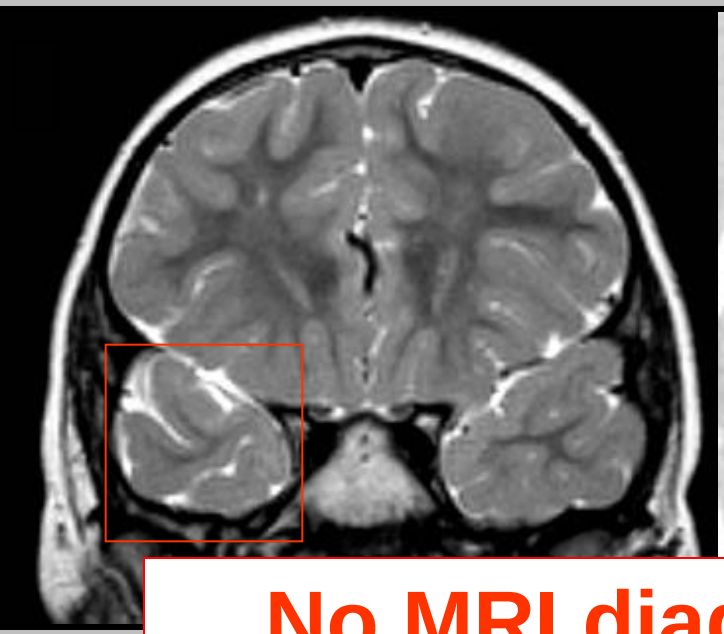
Table 1. The three-tiered ILAE classification system of focal cortical dysplasia (FCD) distinguishes isolated forms (FCD Types I and II) from those associated with another principal lesion (FCD Type III).

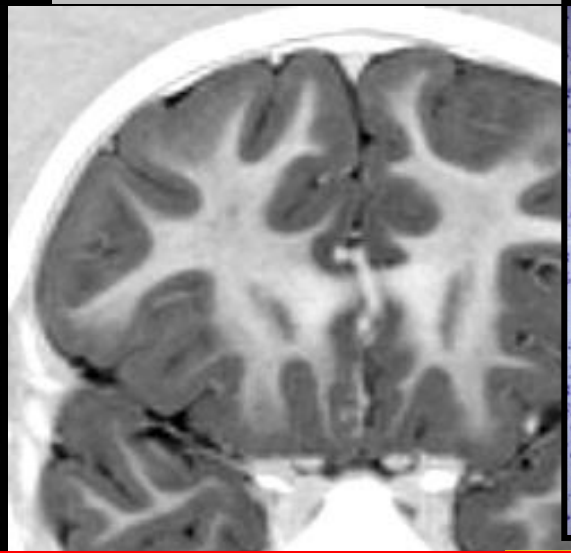
FCD Type I (isolated)	Focal cortical dysplasia with abnormal radial cortical lamination (FCD Type Ia)	Focal cortical dysplasia with abnormal tangential cortical lamination (FCD Type Ib)	Focal cortical dysplasia with abnormal radial and tangential cortical lamination (FCD Type Ic)	
FCD Type II (isolated)	Focal cortical dysplasia with dysmorphic neurons (FCD Type IIa)		Focal cortical dysplasia with dysmorphic neurons and balloon cells (FCD Type IIb)	
FCD Type III (associated with principal lesion)	Cortical lamination abnormalities in the temporal lobe associated with hippocampal sclerosis (FCD Type IIIa)	Cortical lamination abnormalities adjacent to a glial or glioneuronal tumor (FCD Type IIIb)	Cortical lamination abnormalities adjacent to vascular malformation (FCD Type IIIc)	Cortical lamination abnormalities adjacent to any other lesion acquired during early life, e.g., trauma, ischemic injury, encephalitis (FCD Type IIId)

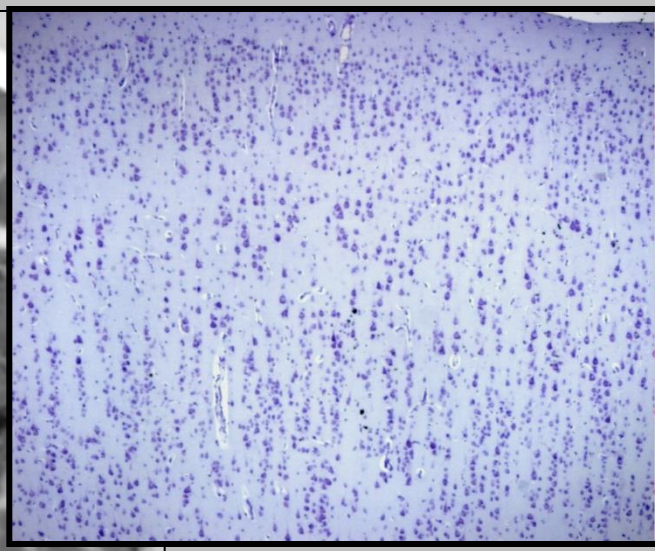
FCD Type III (not otherwise specified, NOS): if clinically/radiologically suspected principal lesion is not available for microscopic inspection.
Please note that the rare association between FCD Types IIa and IIb with hippocampal sclerosis, tumors, or vascular malformations should not be classified as FCD Type III variant.

Neuropathology: cortical layer abnormalities (FCD Type I) + HS → FCD Type IIIa

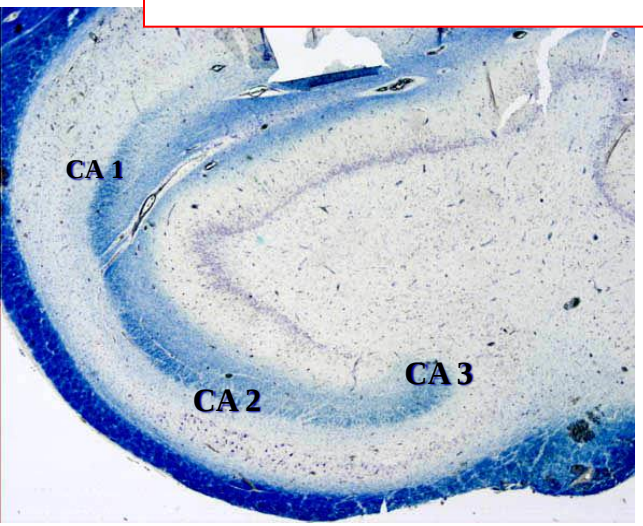
H. NIGUARDA

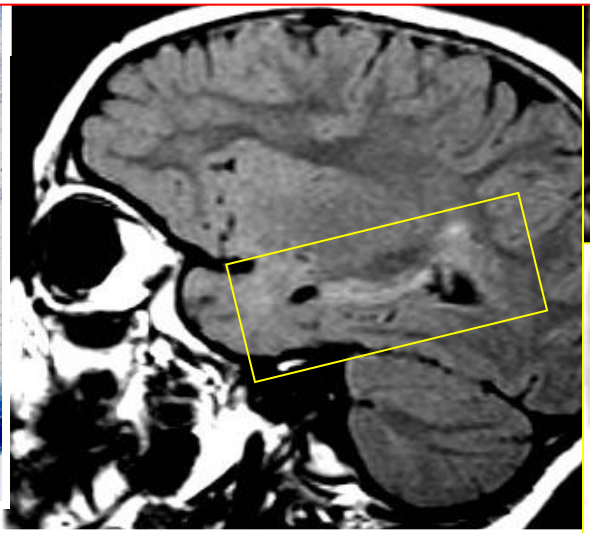


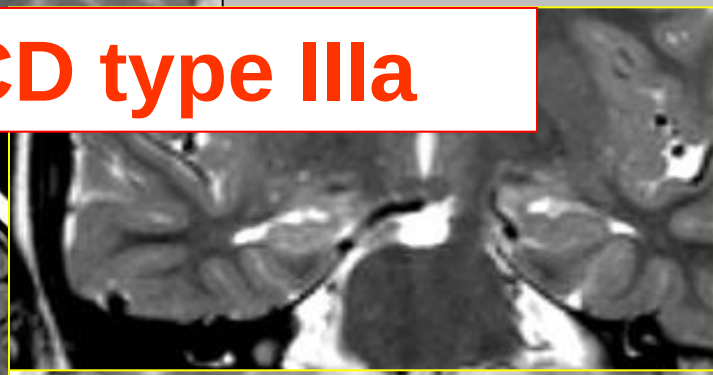


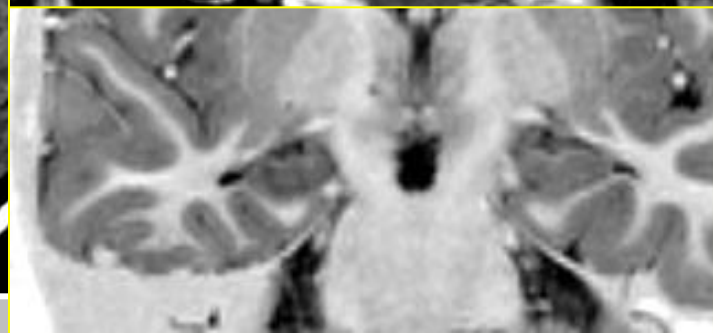


No MRI diagnosis of FCD type IIIa

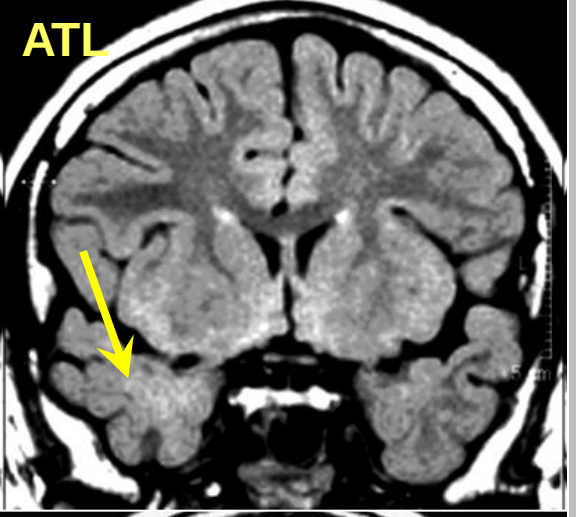


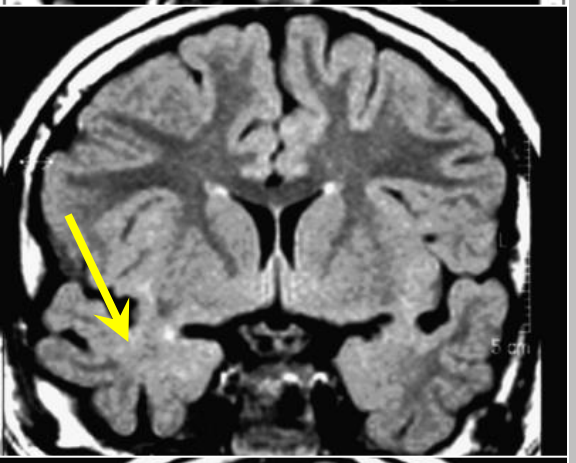


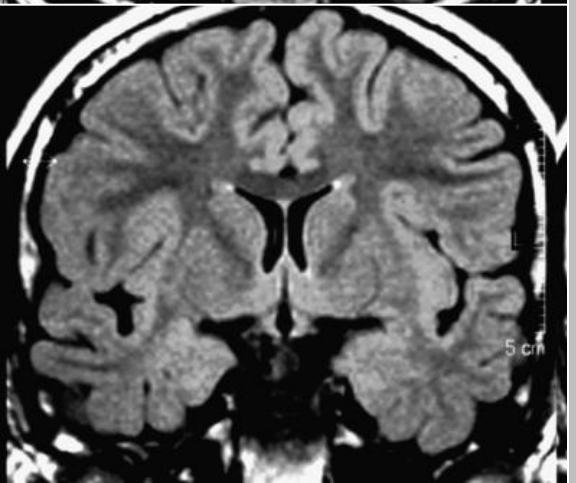




R.R.





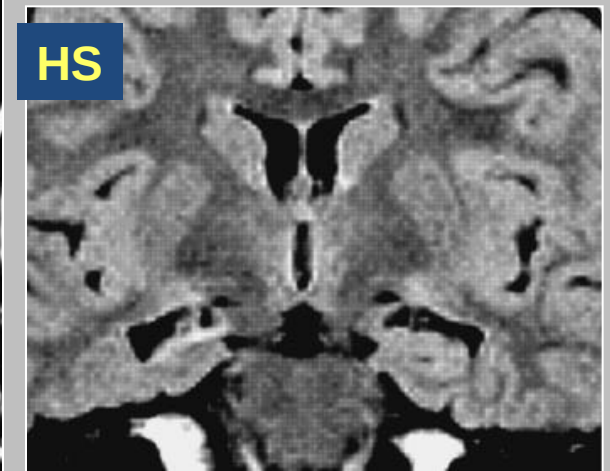


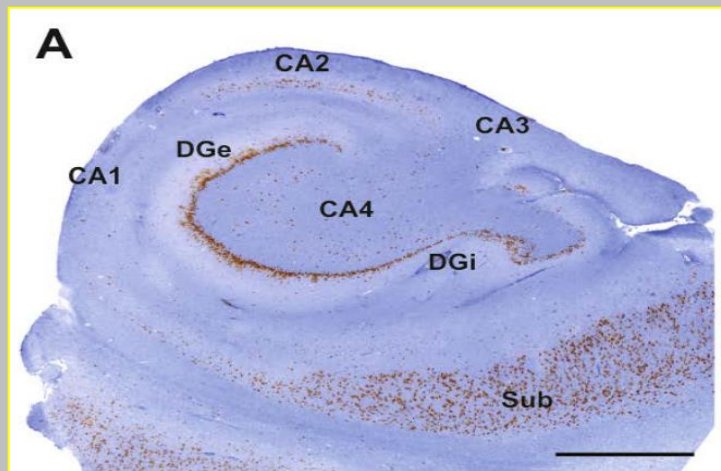
MR - *HS + TP abnormalities*

- Ipsilateral temporo-polar atrophy +/-
- hyperintense signal on T2WI of the WM +/-
- blurring of the GM/WM junction

there is no consensus regarding the genesis of temporo-polar abnormalities

- **gliosis ?**
- **developmental cortical abnormalities (FCD I – mMCD) ?**
- **loss of myelin ?**
- **non-specific increase in water content ?**
- **vascular/metabolic alteration ?**





Temporal Pole
GM / WM blurring

Brain Advance Access published June 22, 2012

doi:10.1093/brain/aws149 Brain 2012: Page 1 of 13 | 1

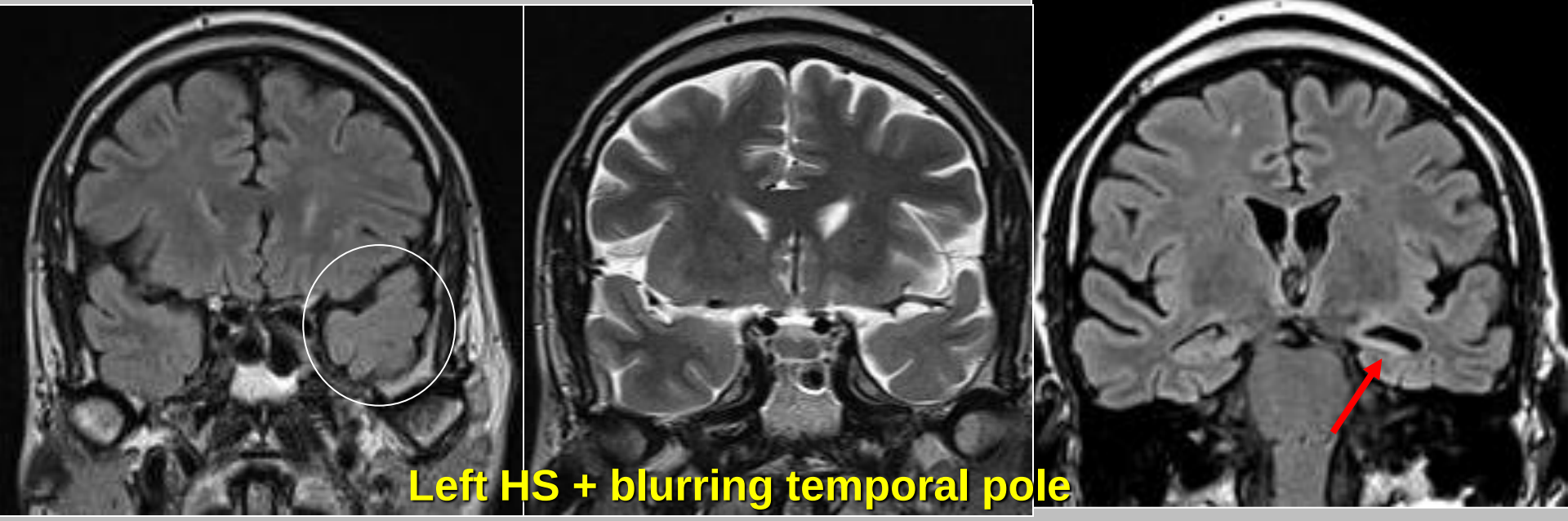
BRAIN
A JOURNAL OF NEUROLOGY

Blurring in patients with temporal lobe epilepsy: clinical, high-field imaging and ultrastructural study

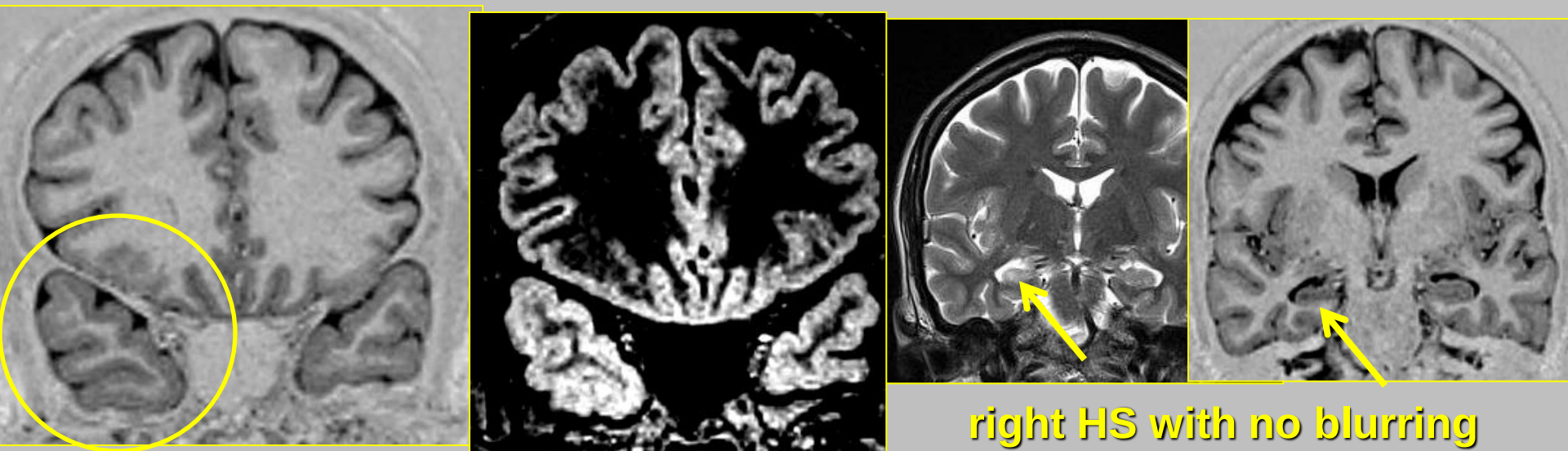
Rita Garbelli,[1] Gloria Milesi,[1] Valentina Medici,[1] Flavio Villani,[1] Giuseppe Didato,[1] Francesco Deleo,[1] Ludovico D'Incerti,[2] Michela Morbin,[3] Giulia Mazzoleni,[3] Anna Rita Giovagnoli,[4] Annalisa Parente,[4] Ileana Zucca,[4] Alfonso Mastropietro[4,5] and Roberto Spreafico[1]

Pts population: 32 epileptic patients operated on for TLE
with radiological evidence and neuropathologic proven HS

Group 1: 18 pts blurring
Group 2: 14 no- blurring

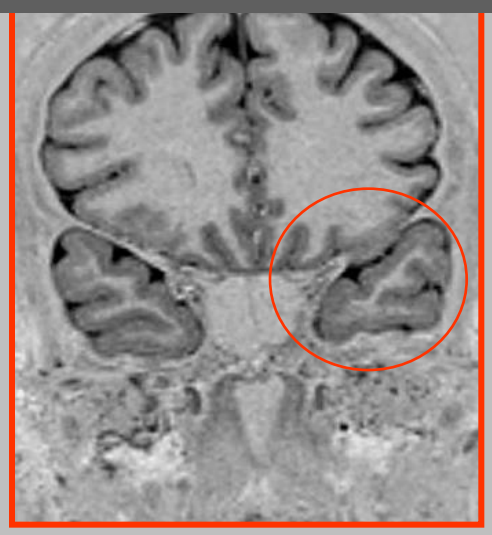


Left HS + blurring temporal pole

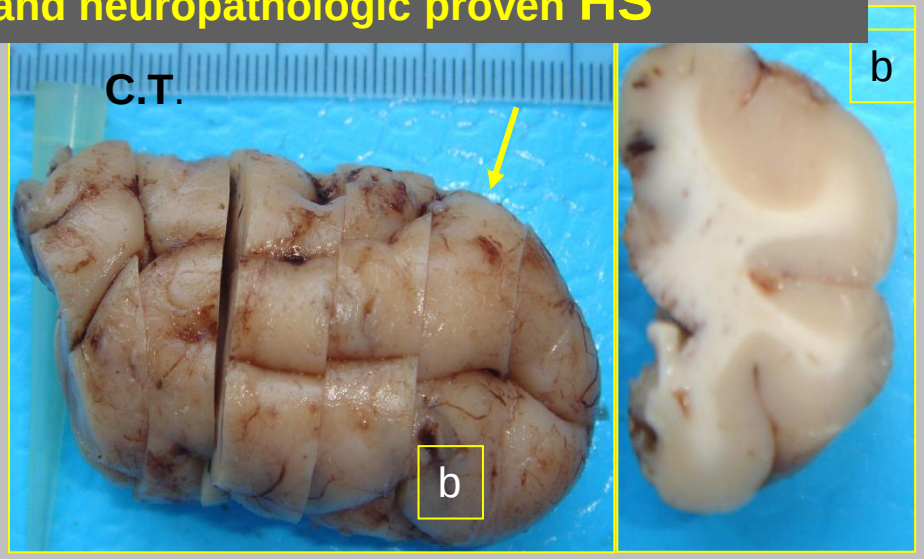


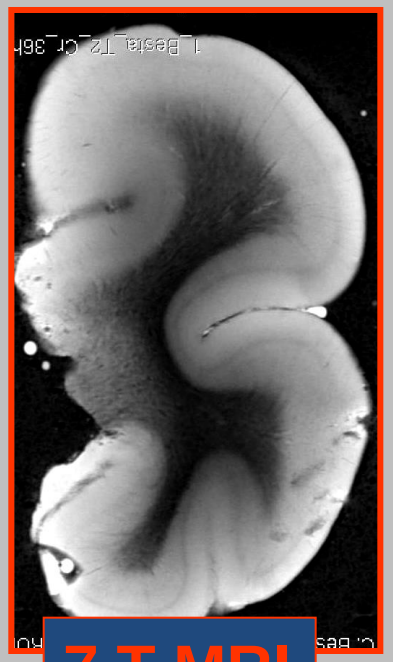
right HS with no blurring

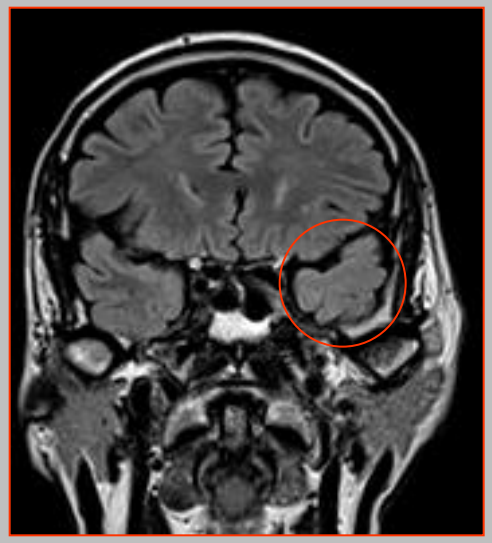
Pts population: 32 epileptic patients operated on for TLE
with radiological evidence and neuropathologic proven HS

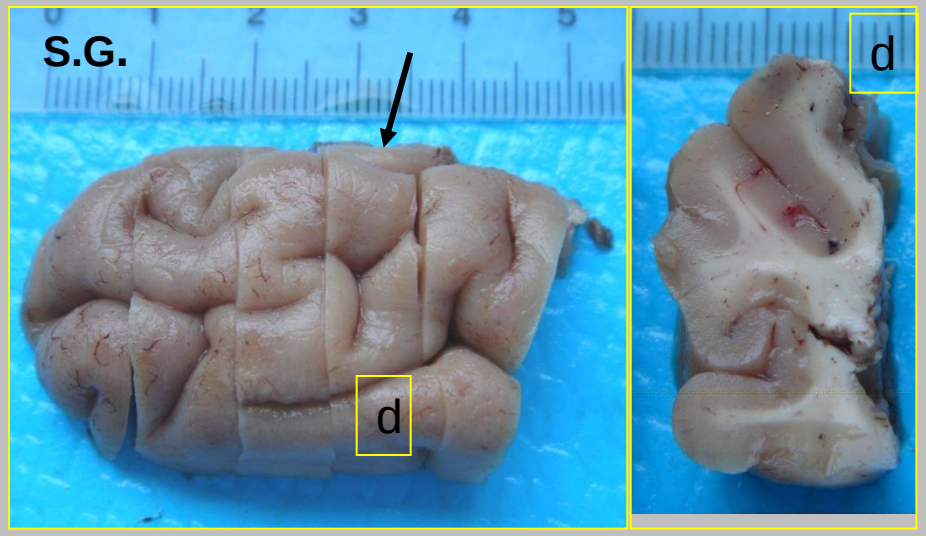


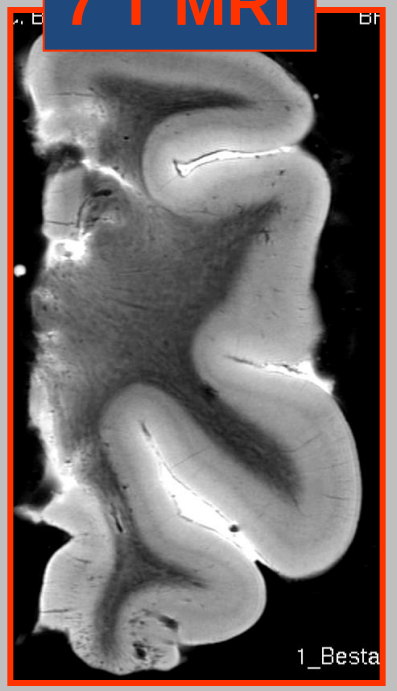
1.5 T MRI





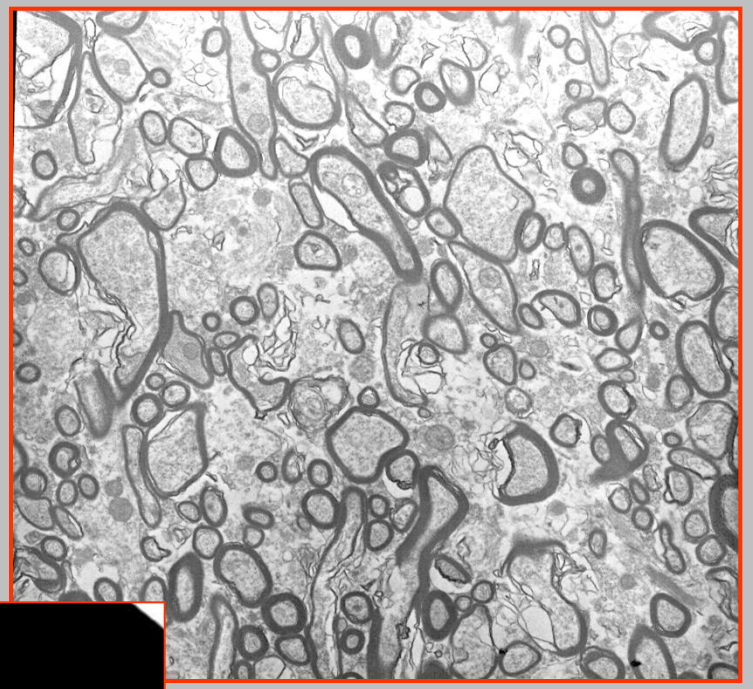


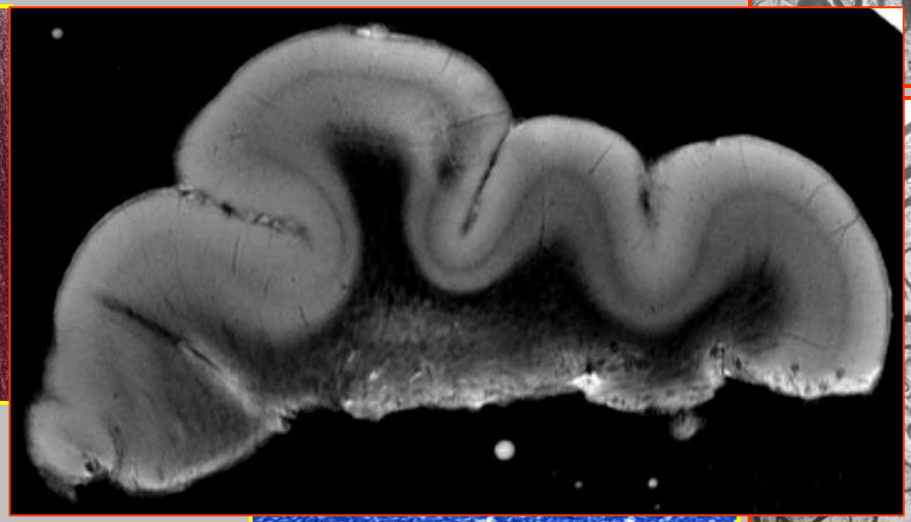


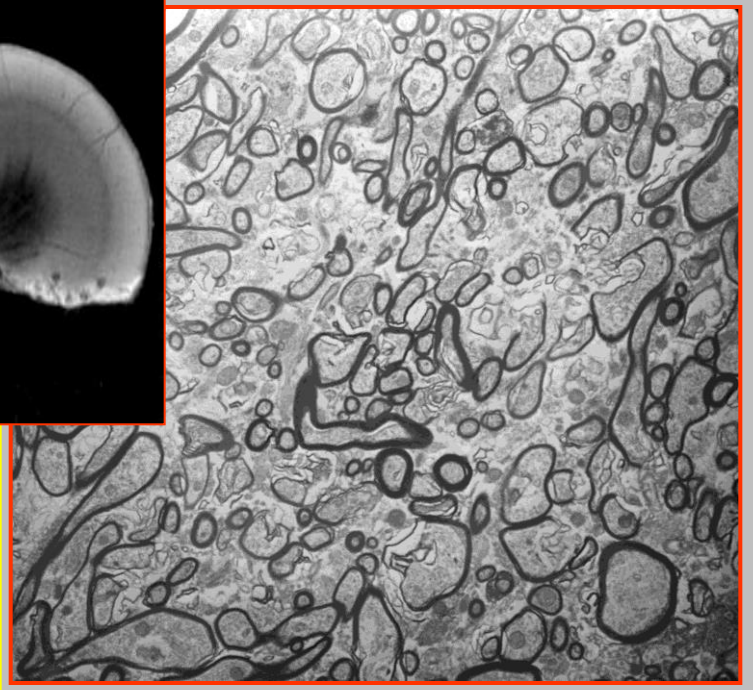


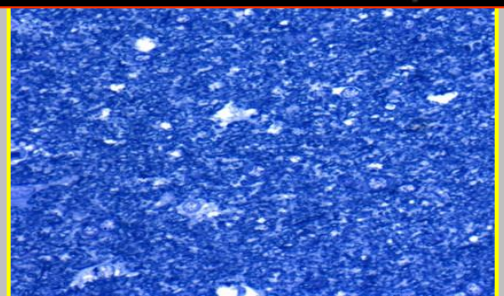
courtesy of R. Garbelli & R.Spreafico,
Neurological Institute "C.Besta" Milano

NO BLURRING D.F.





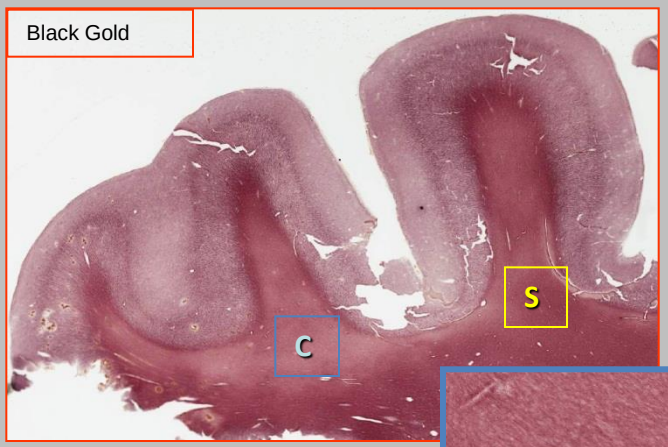




courtesy of
R. Garbelli & R.Spreafico,
Neurological Institute
"C.Besta" Milano

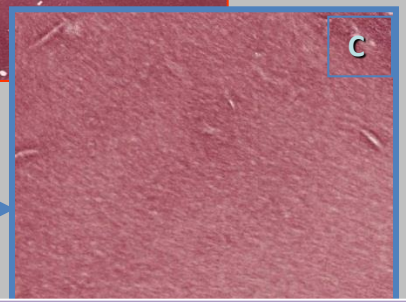
BLURRING - V.

Black Gold

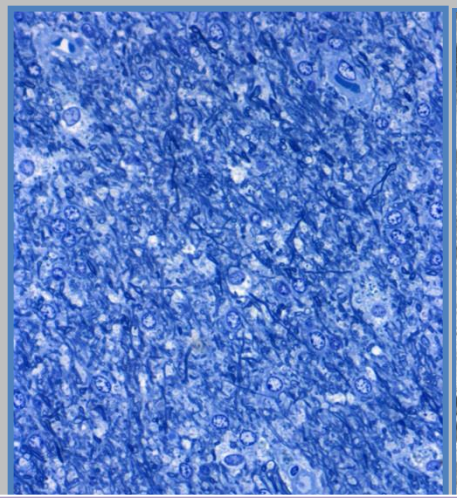


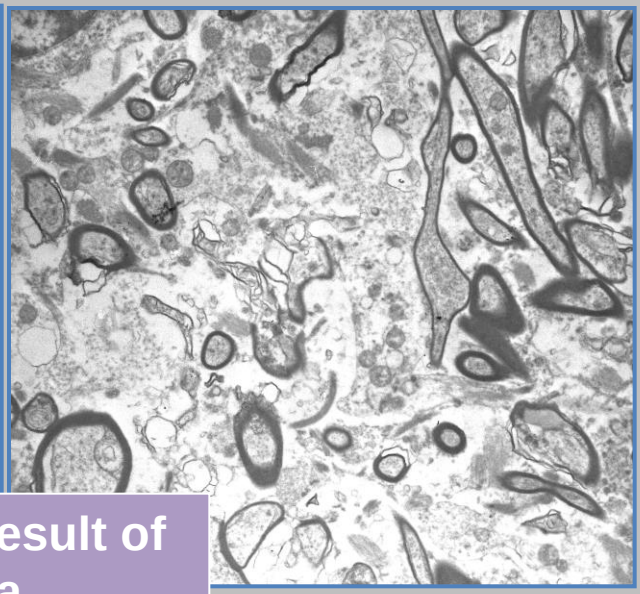
Luxol fast blue

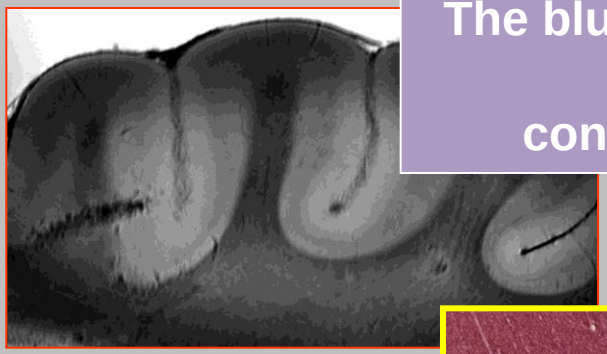
EM



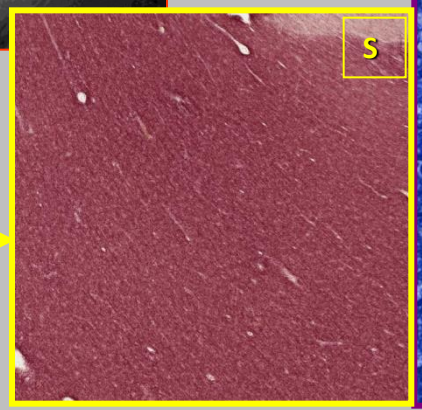
pale zone →



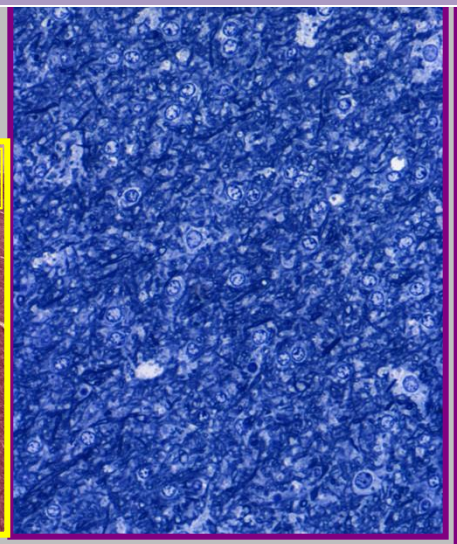


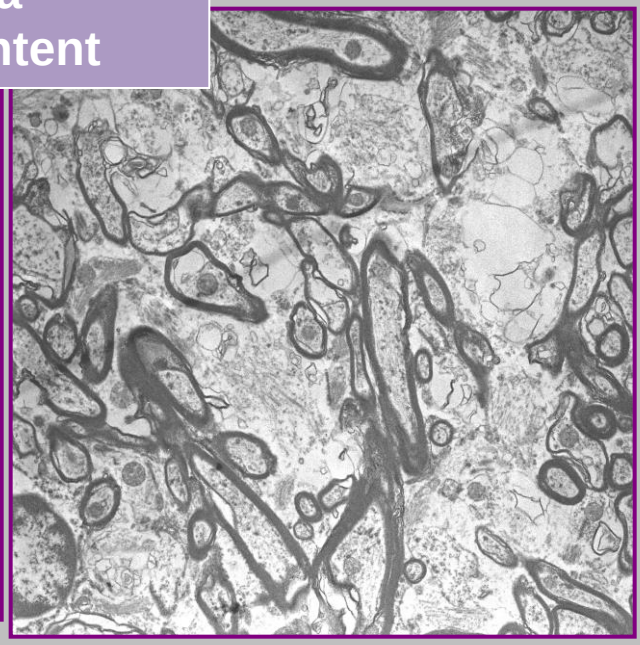


The blurring detected by MRI is the result of fibers degeneration and to a consequent increase in water content



dark zone →





Temporal Pole neocortex

Pts population: 32 epileptic patients operated on for TLE with radiological evidence and neuropathologic proven HS

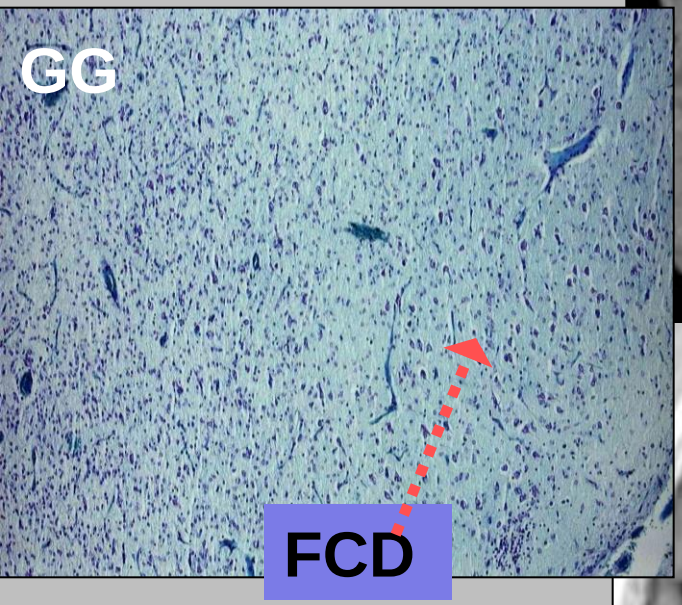
		Temporal cortex histopathology	
	Patients	Cortical dyslamination (FCD type IIIa)	Normal cortex
Group 1 HS + blurring	18	15 (83%)	3
Group 2 HS - No blurring	14	13 (92%)	1

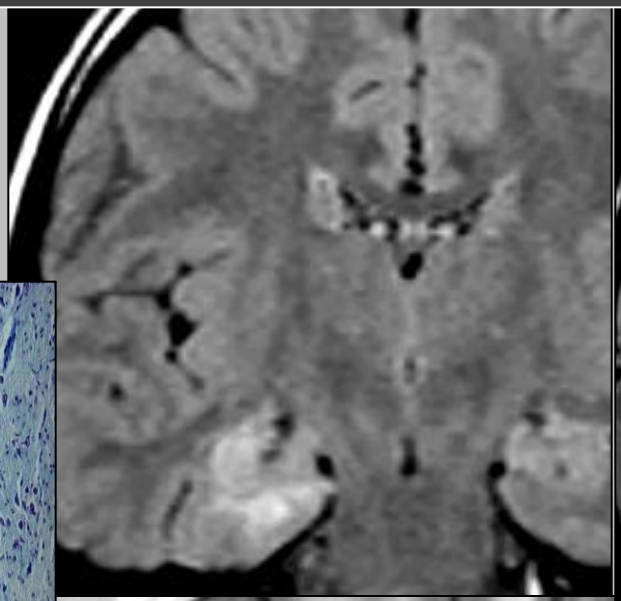
Epilessia del lobo temporale diagnosi RM

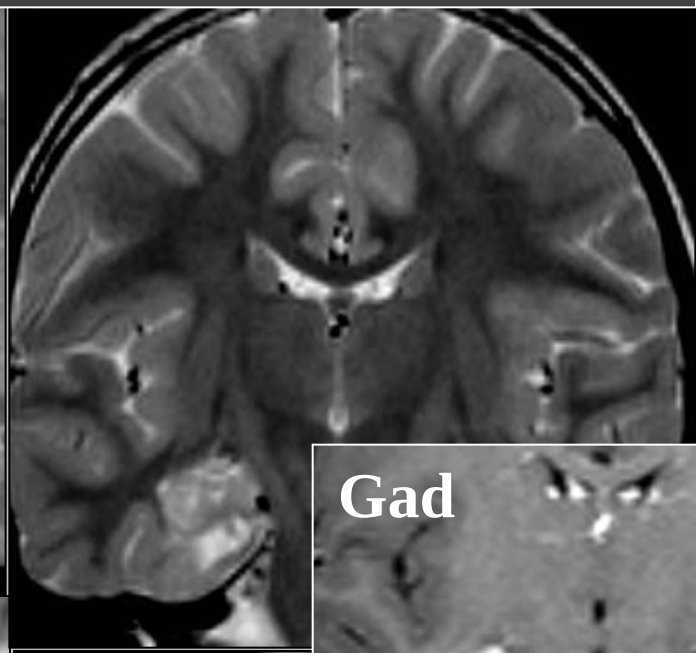
RM	Neuropatologia
1- HS isolata	
2- HS + alterazione SB ➢ Ipsilaterale all'atrofia temporo-polare +/- ➢ Iperintensità in T2WI sostanza bianca +/- ➢ blurring giunzione bianca/grigia	FCD type IIIa Corteccia normale / gliosi
3- HS + seconda lesione ⟹	Dual Pathology

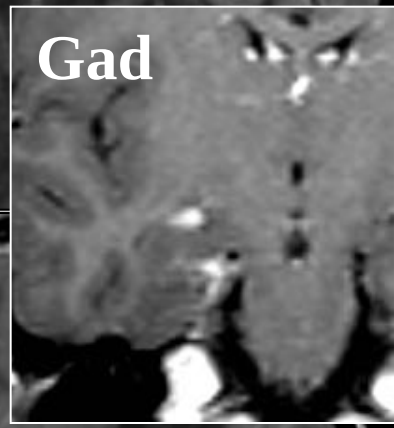
Non esistono criteri diagnostici basati sulla RM in grado di formulare ipotesi circa l'esistenza o meno di una FCD III

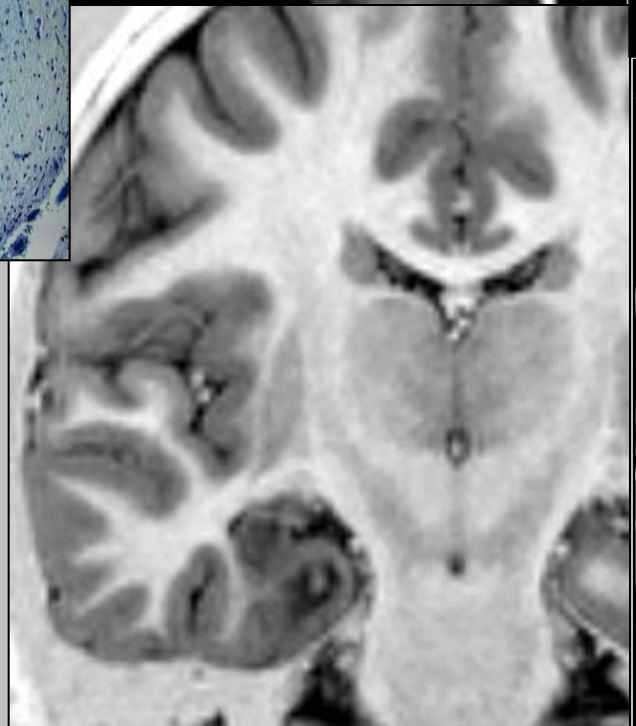
Neuropathology: FCD Type I + Ganglioglioma = FCD Type III b



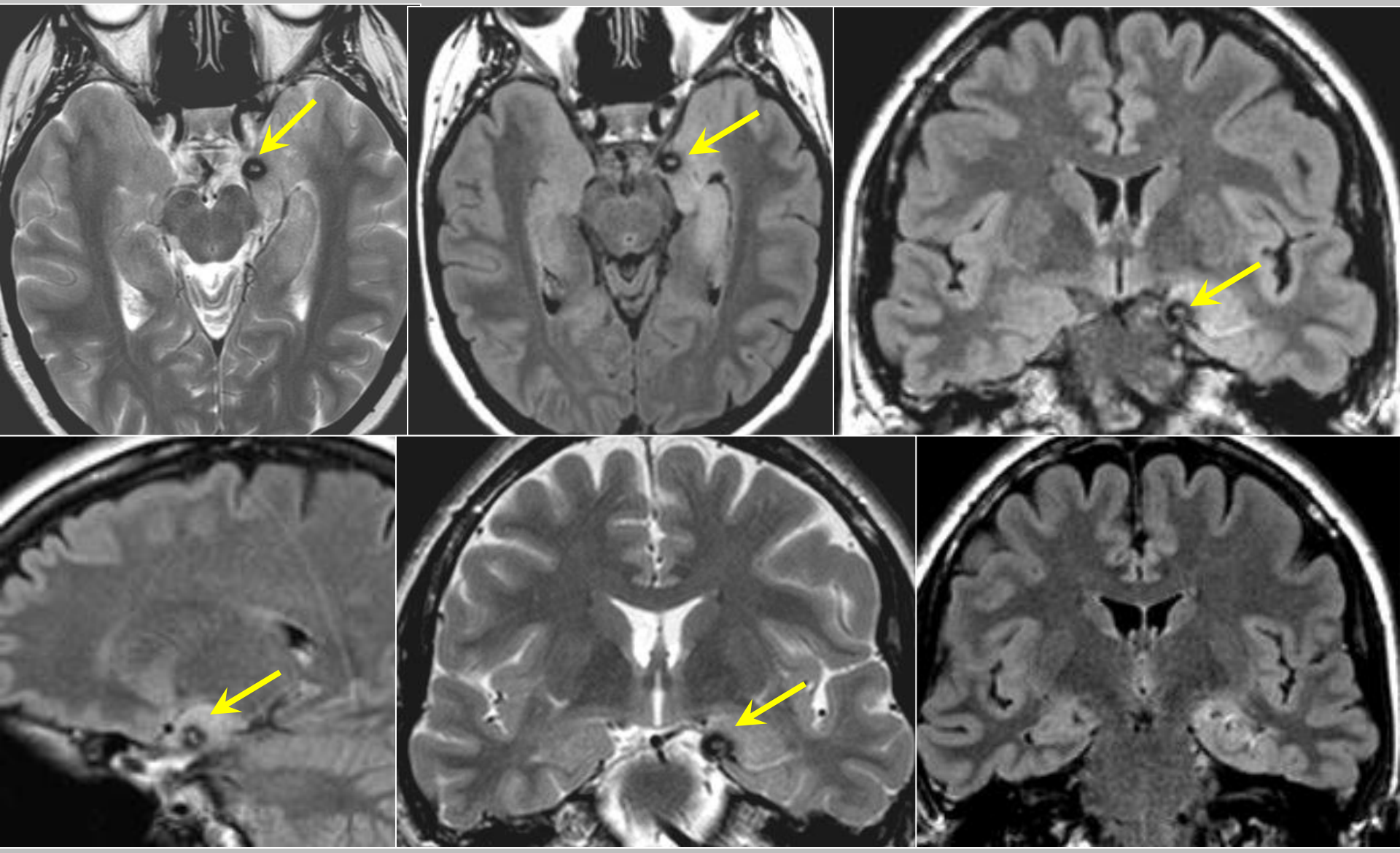




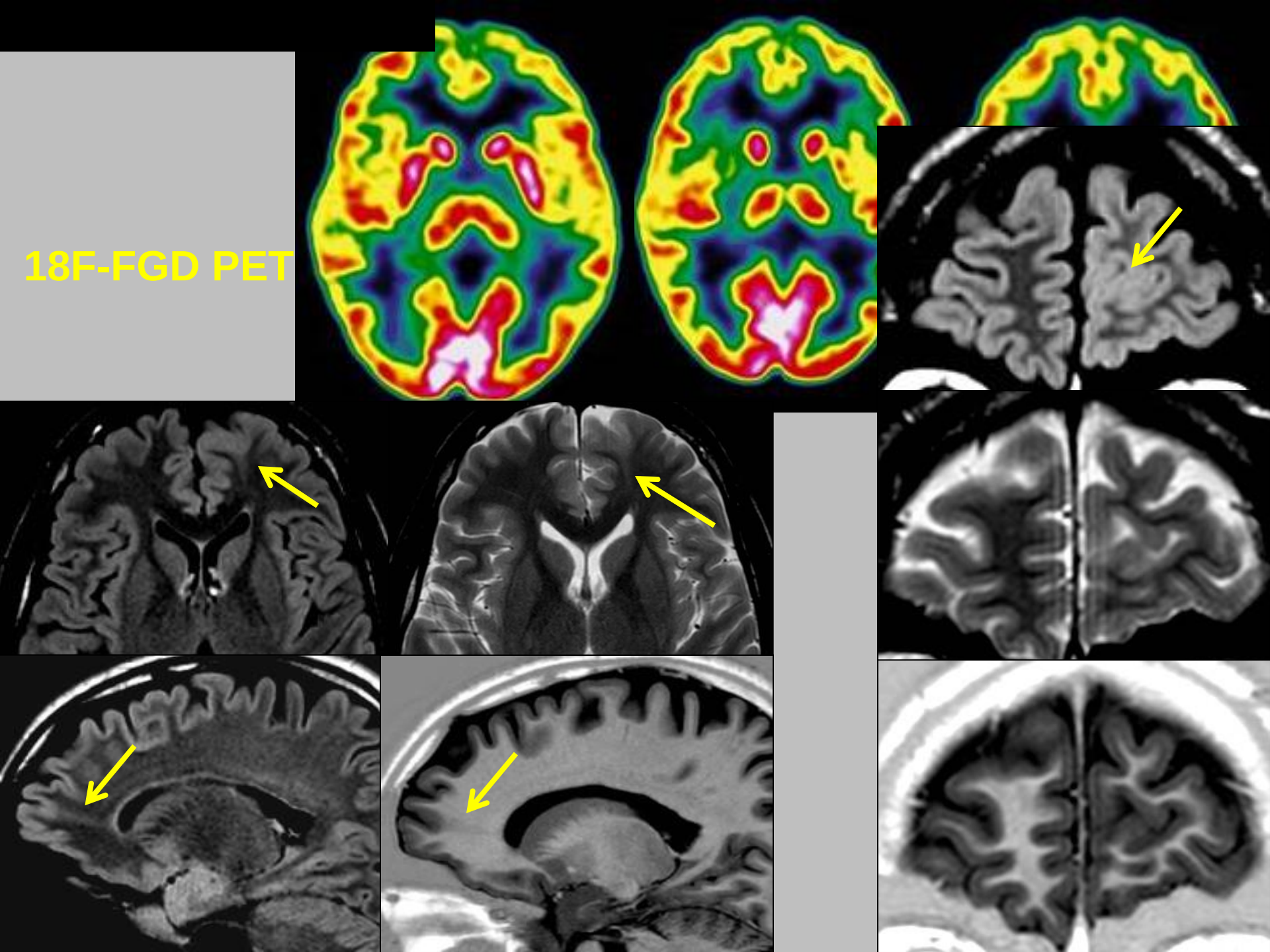


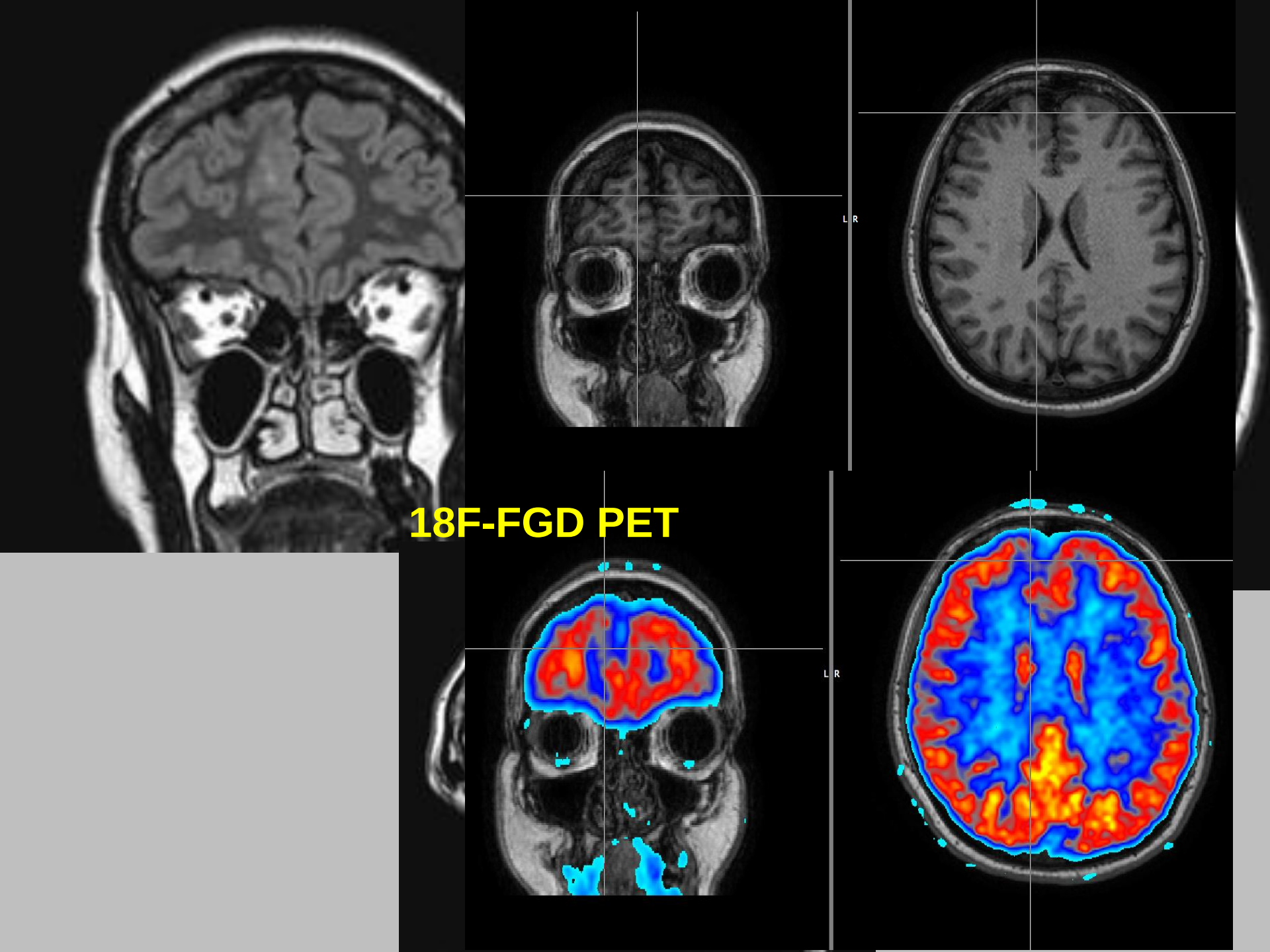


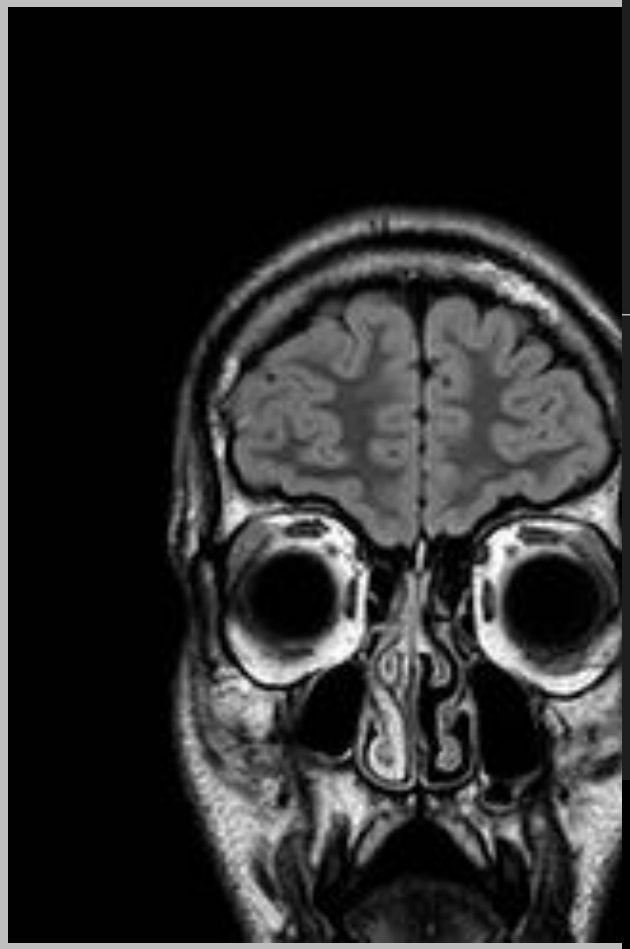
Neuropathology: FCD Type I + cavernoma = FCD Type III c

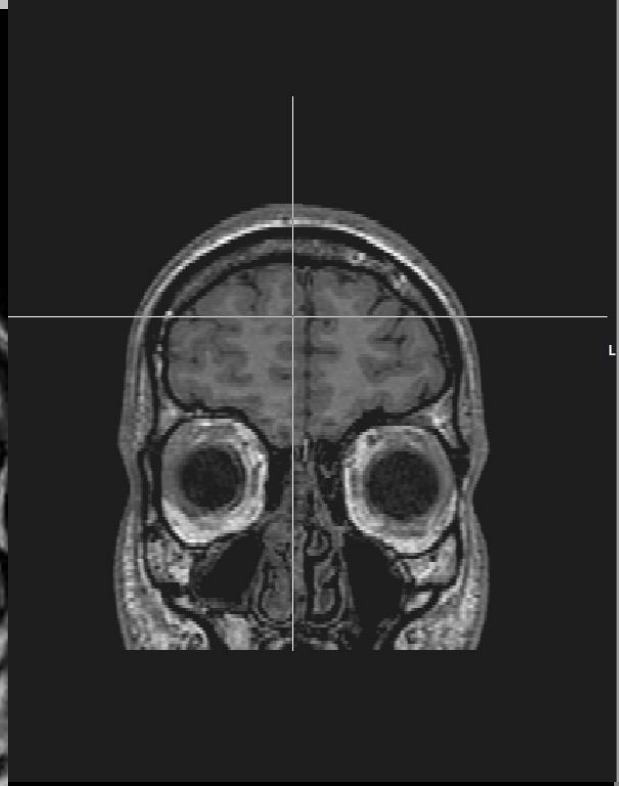


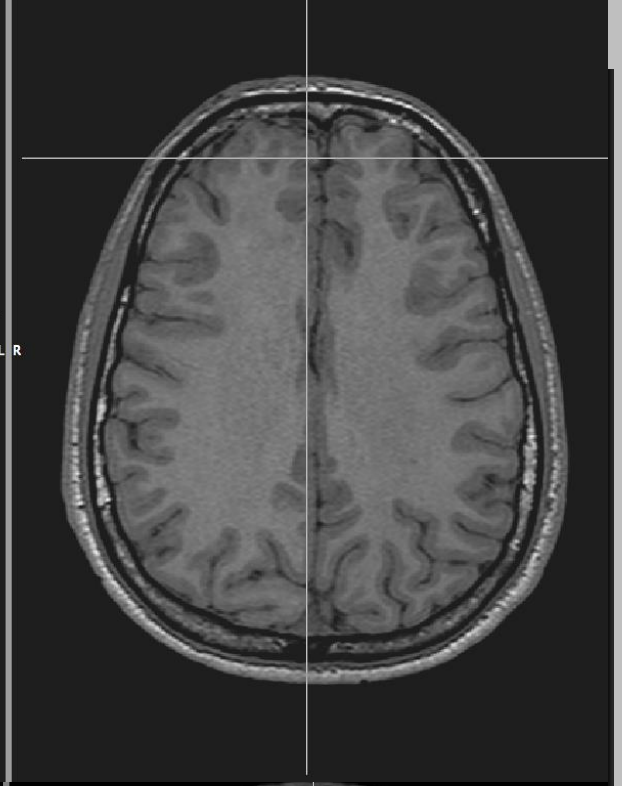
18F-FGD PET

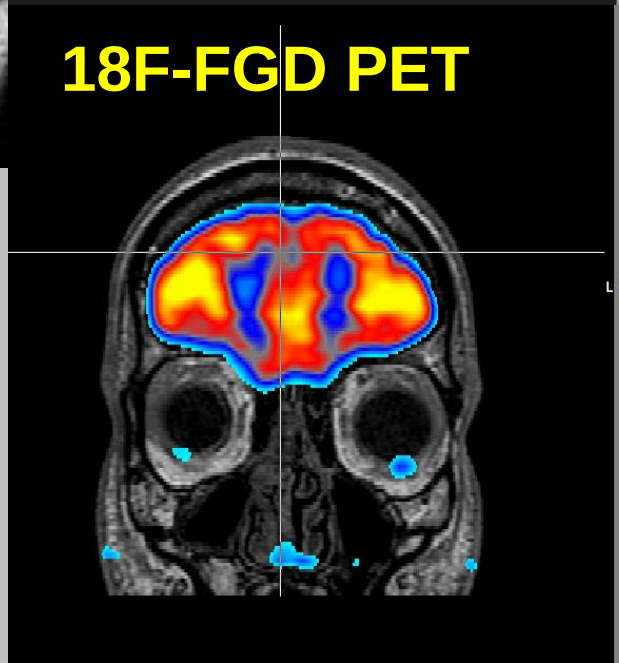


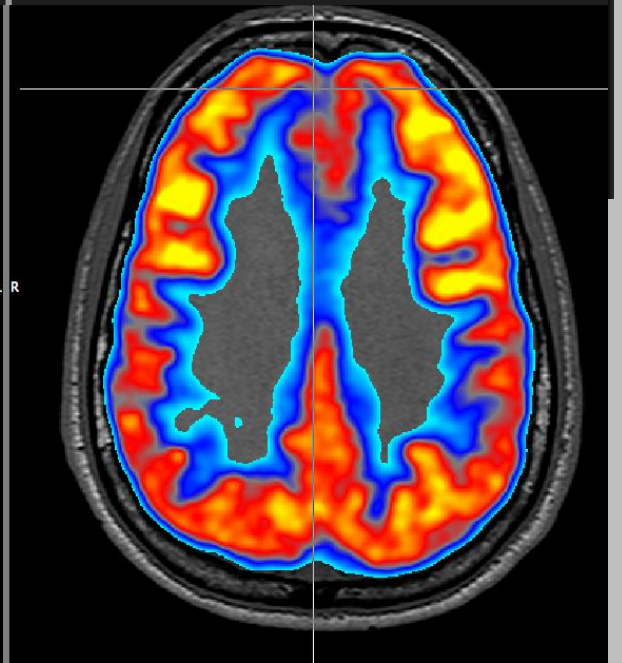

18F-FGD PET





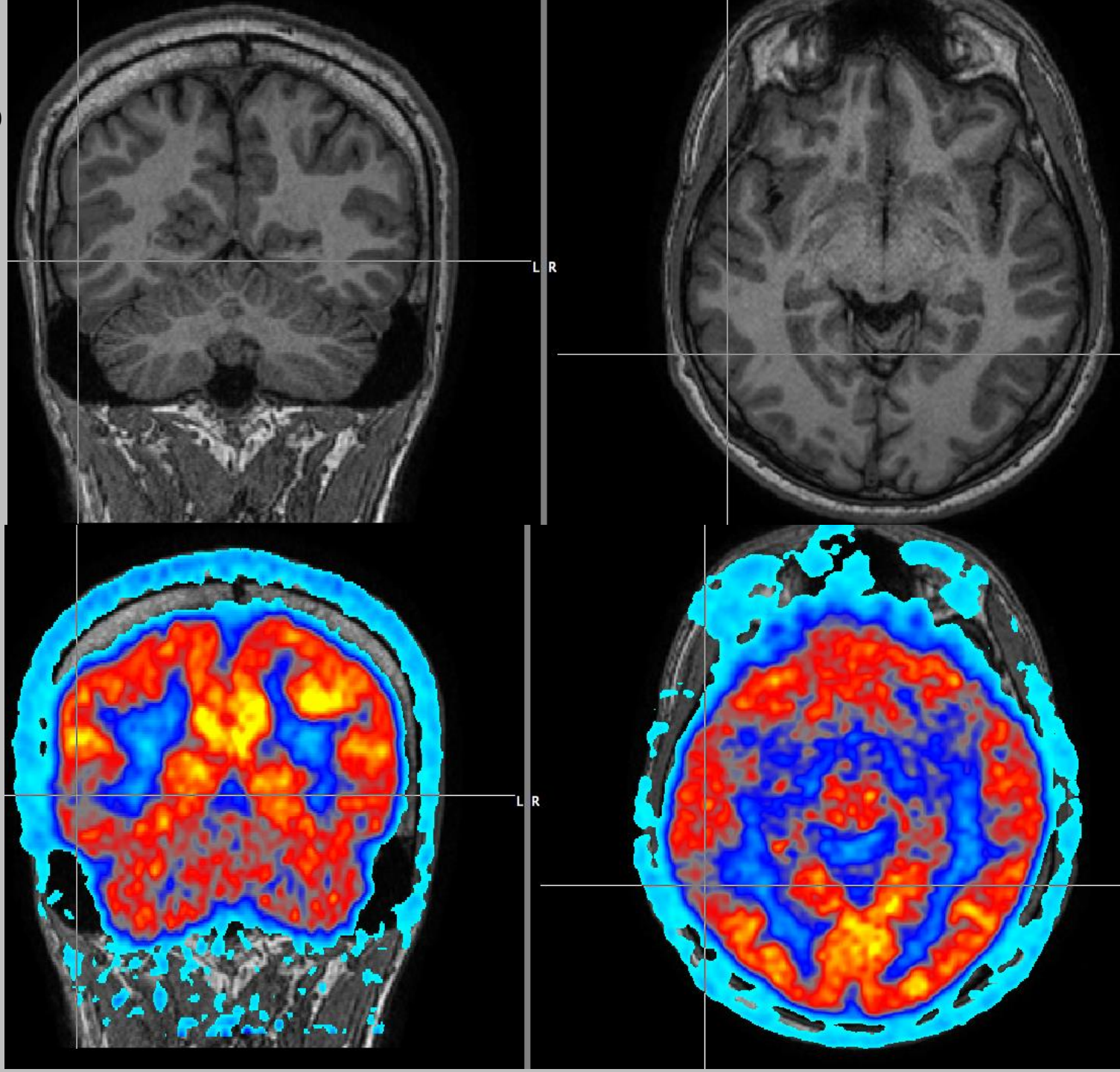


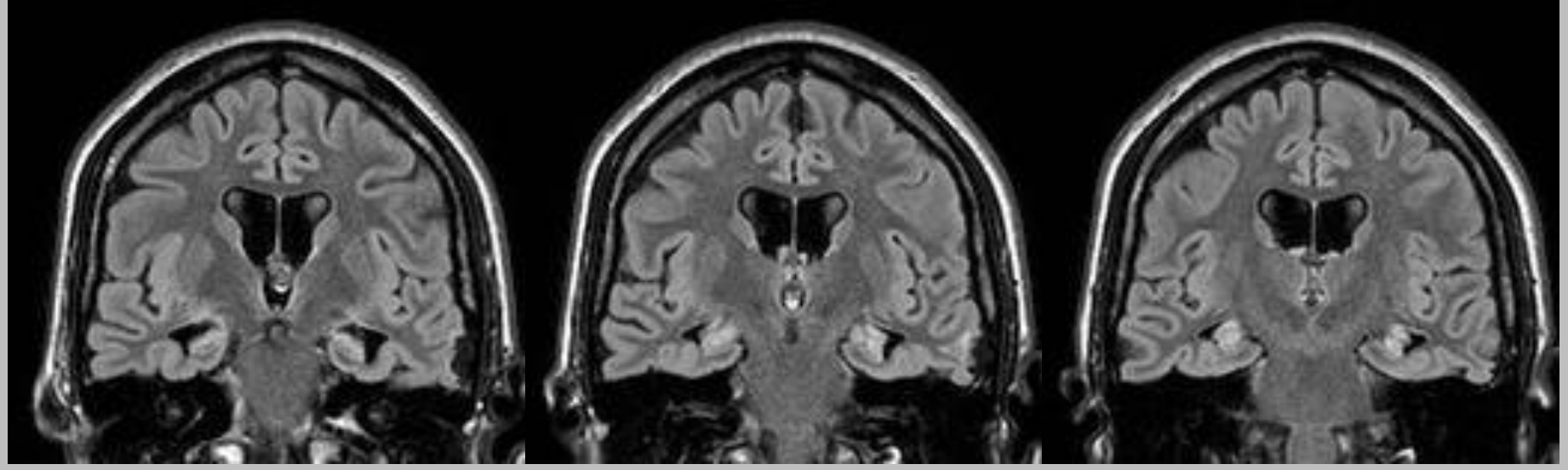




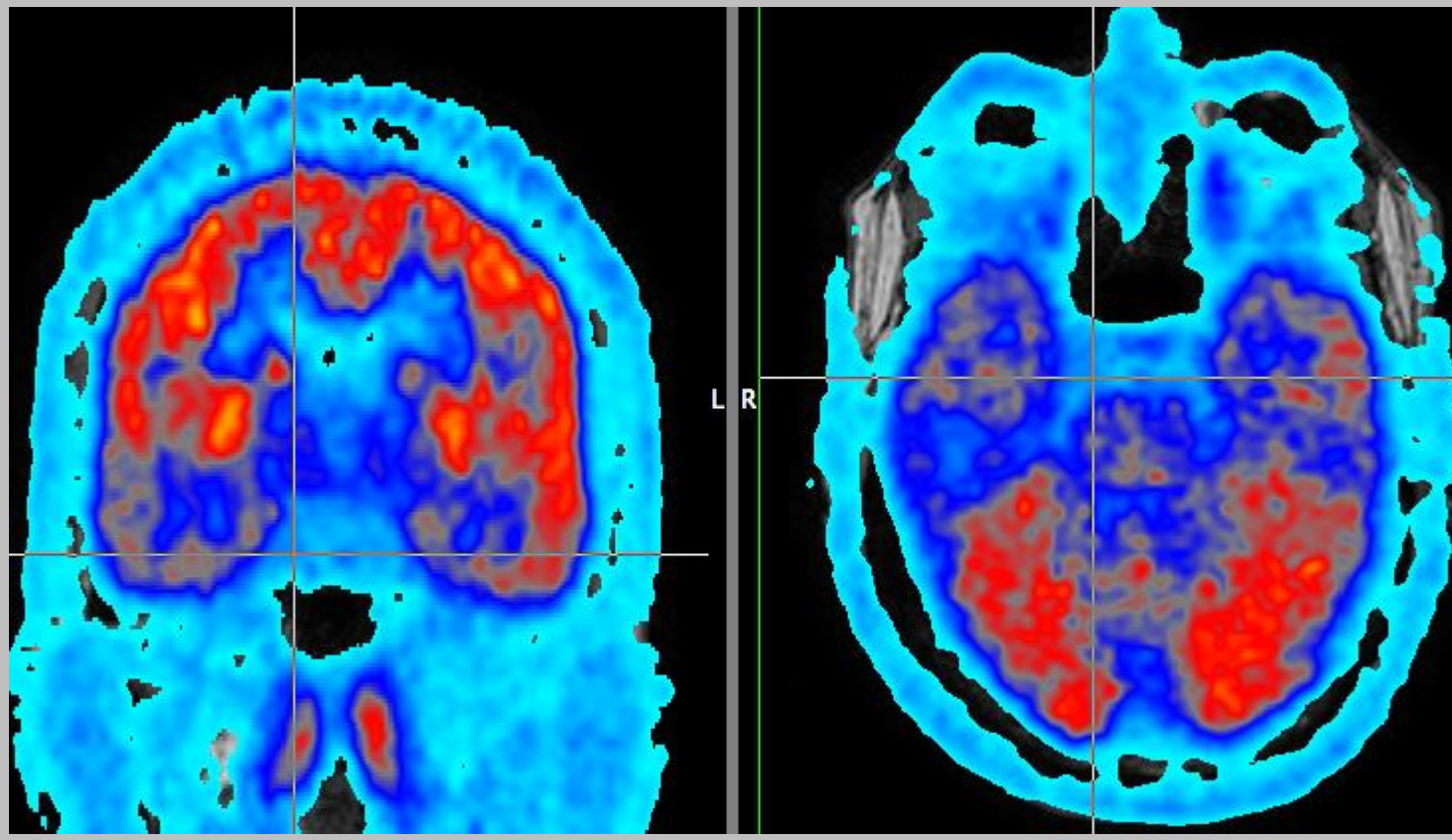
M.A.
Crisi parziali frontali dx
RM negativa
SEEG: focolaio frontale dx
PET ipometabolismo front. dx
Intervento, istologia Taylor IIb.
Classe Ia Engels a 2 aa dalla chirurgia

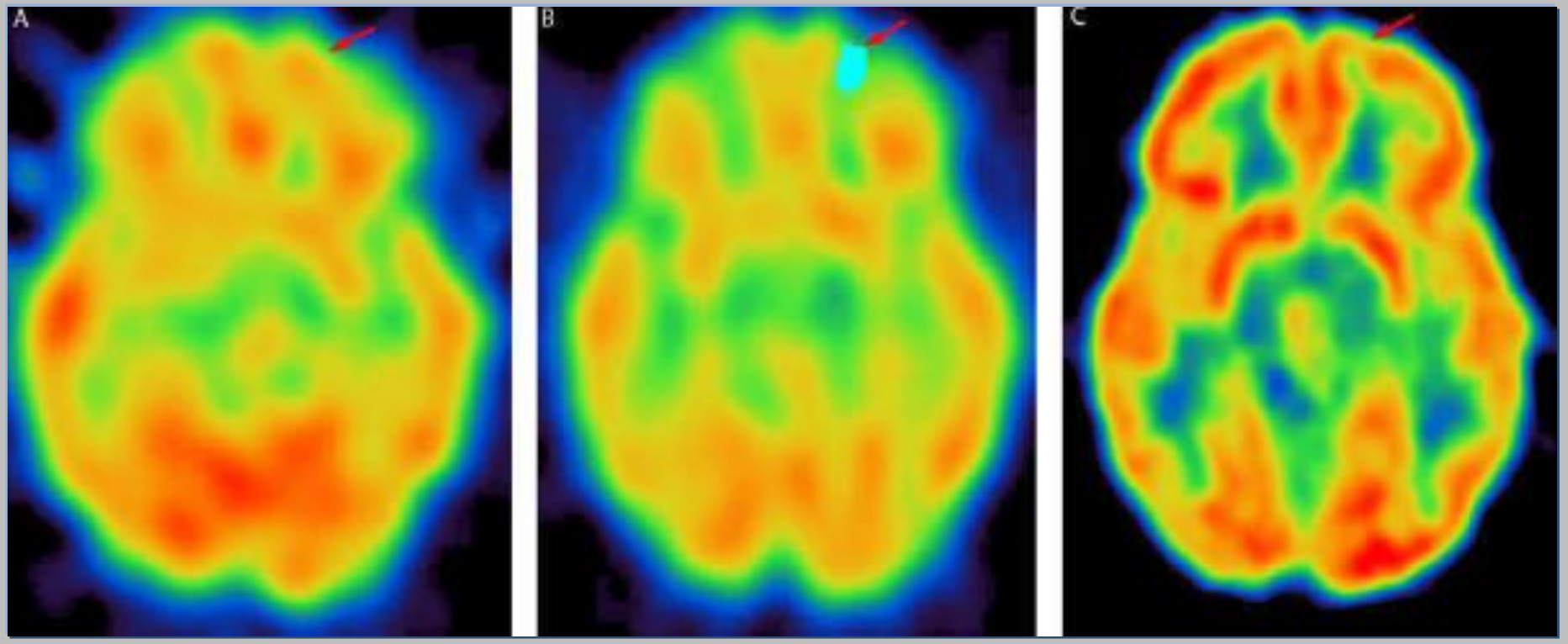
AM crisi ad esordio TO dx
VEEG: focolaio TO dx
RM: anomala girazione TO dx
PET: focolaio ipo-Metabolico TO dx
Intervento
Istologia: Taylor IIa
A 2 aa dopo int. netta riduzione delle crisi (solo notturne)





E. B.
Epilessia Temporale
RM: sclerosi Ippocampale bilaterale
PET DFG: Ipometabolismo temporale bilaterale più marcato a dx
Intervento in T dx
Classe I di Engels dopo 2 aa





Interictal Tc-ECD SPECT ictal Tc-ECD SPECT FDG-PET

PET and SPECT in epilepsy: A critical review

C. la Fougère*, A. Rominger, S. Förster, J. Geisler, P. Bartenstein

Department of Nuclear Medicine, Ludwig Maximilian University of Munich, Marchioninistrasse 15, D-81377 Munich, Germany

Epilepsy & Behavior 15 (2009) 50–55

SCLEROSI TUBEROSA

transmissione autosomica dominante

mutazioni e polimorfismi dei geni:

TSC1 (amartina) sul Ch 9q34

TSC2 (tuberina) sul Ch16p 13.3

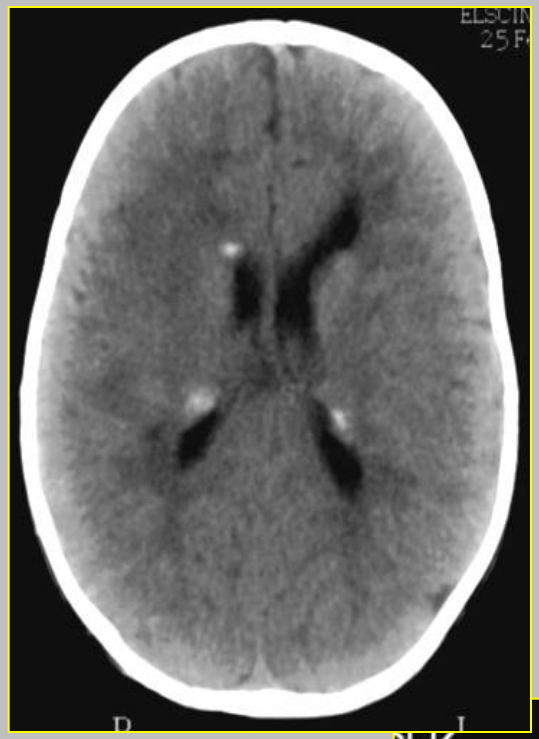
con funzione di:

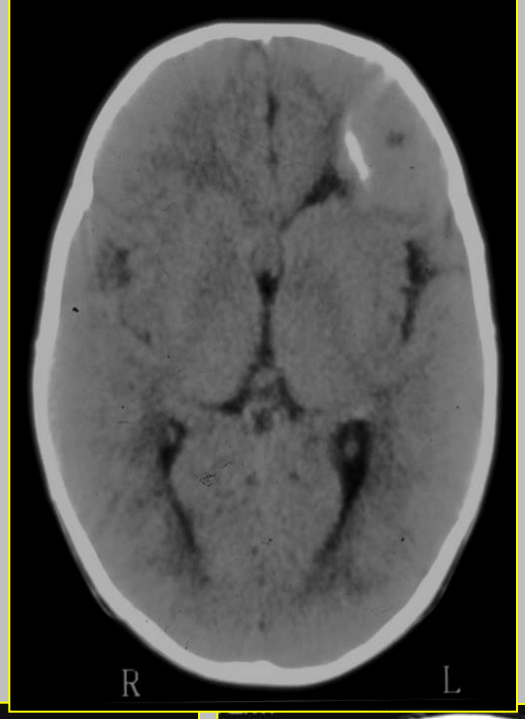
- **regolazione della crescita e proliferazione cellulare**
- **onco-soppressione**

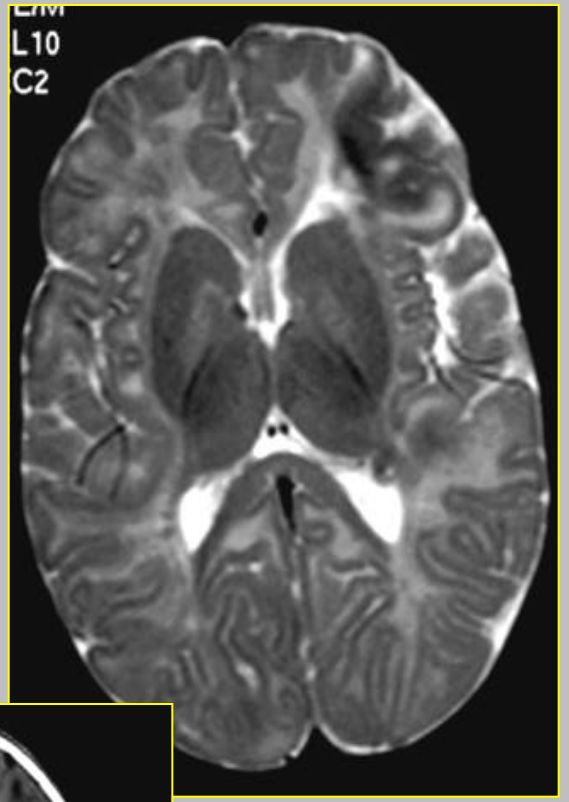
- **Forme TSC2-correlate si manifestano con fenotipi più severi**

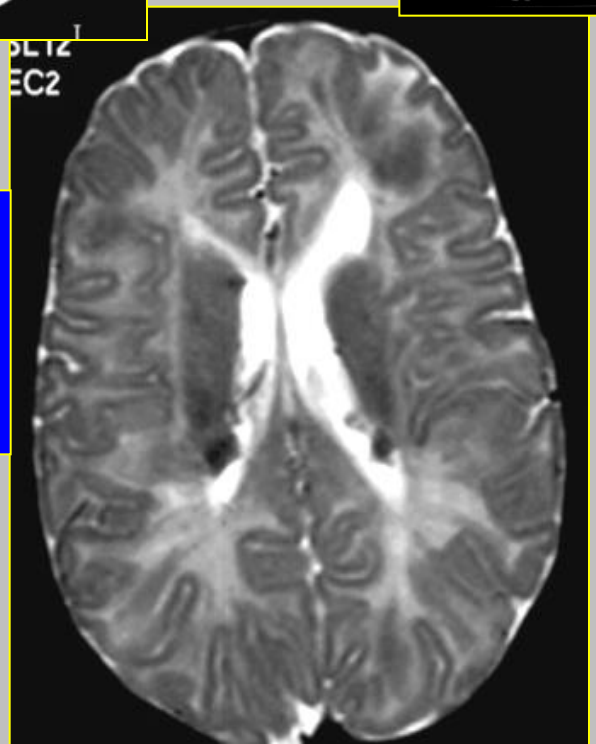
SCLEROSI TUBEROSA – Aspetti RM

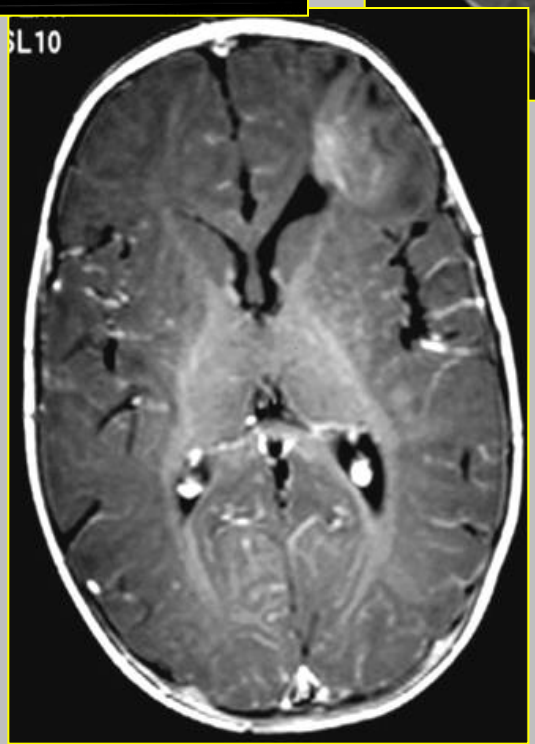
- **Noduli calcifici periventricolari**
- **Tuberi cortico-sottocorticali, spesso calcifici**
- **Enhancement raro**
- **Blurring della giunzione SB/SG**
- **Amartomi multipli della SB**
- **Astrocitoma gigantocellulare intraventricolare**





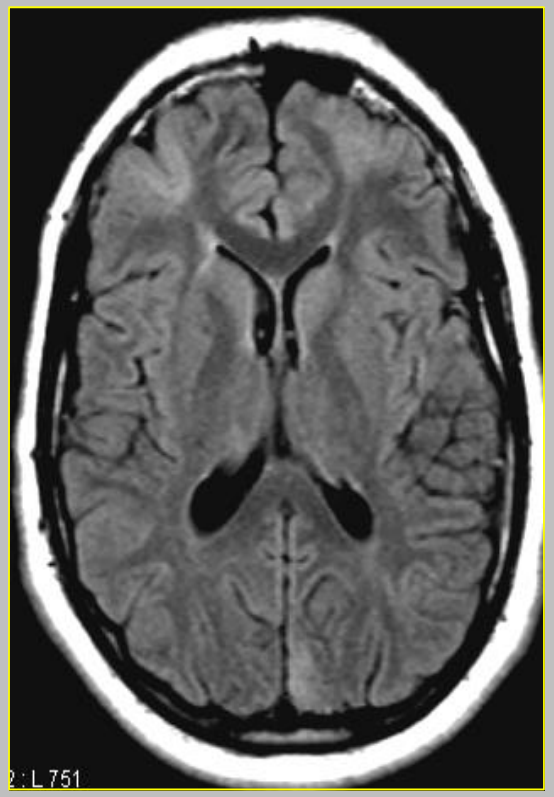


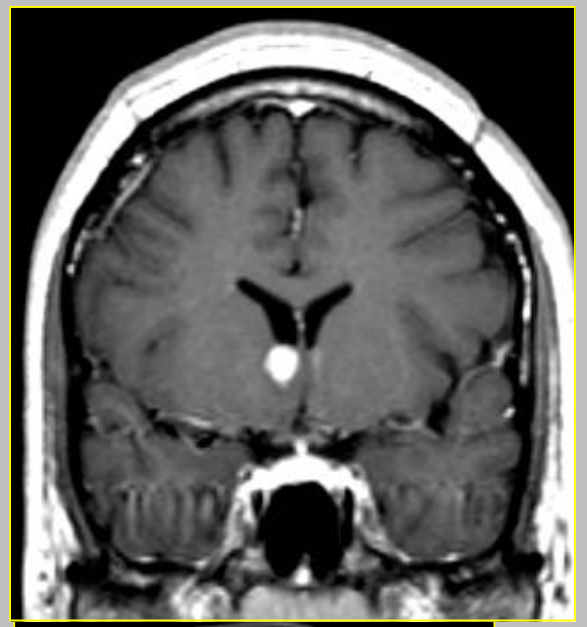


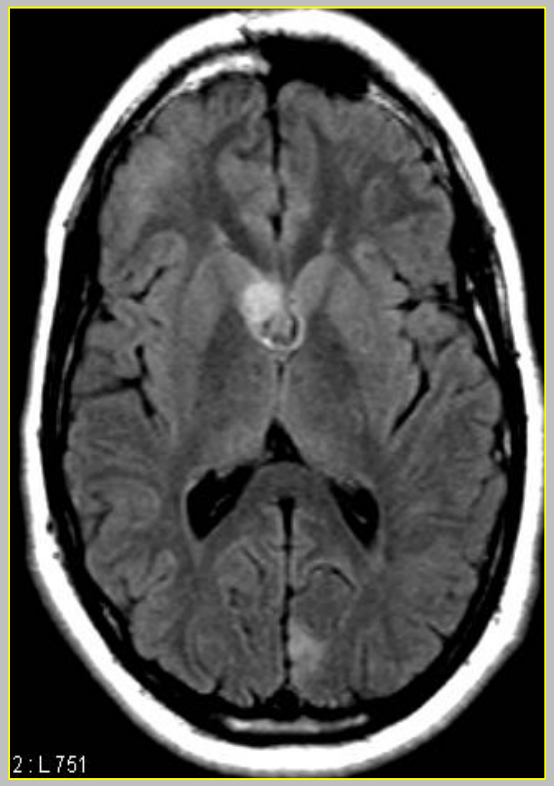


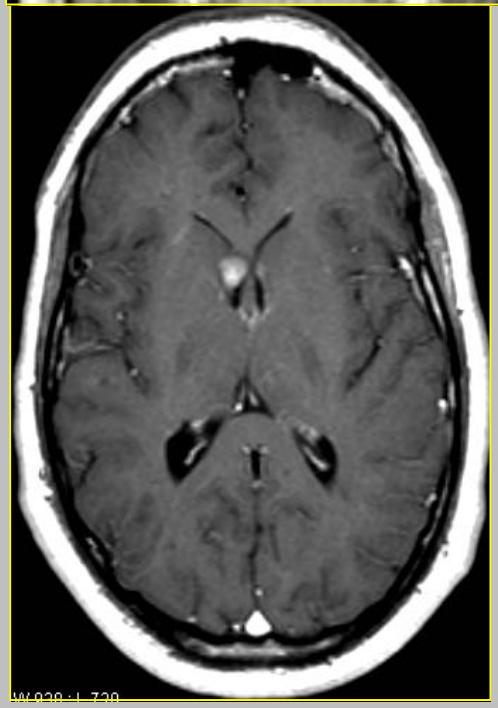
SCLEROSI TUBEROSA

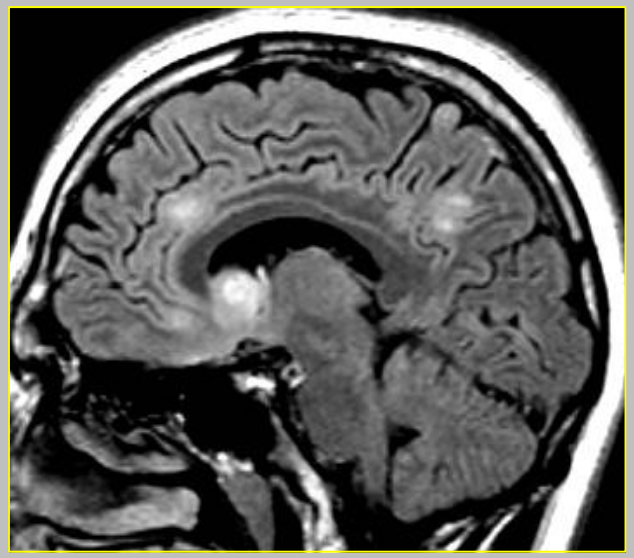
SCLEROSI TUBEROSA
ASTROCITOMA
GIGANTOCELLULARE



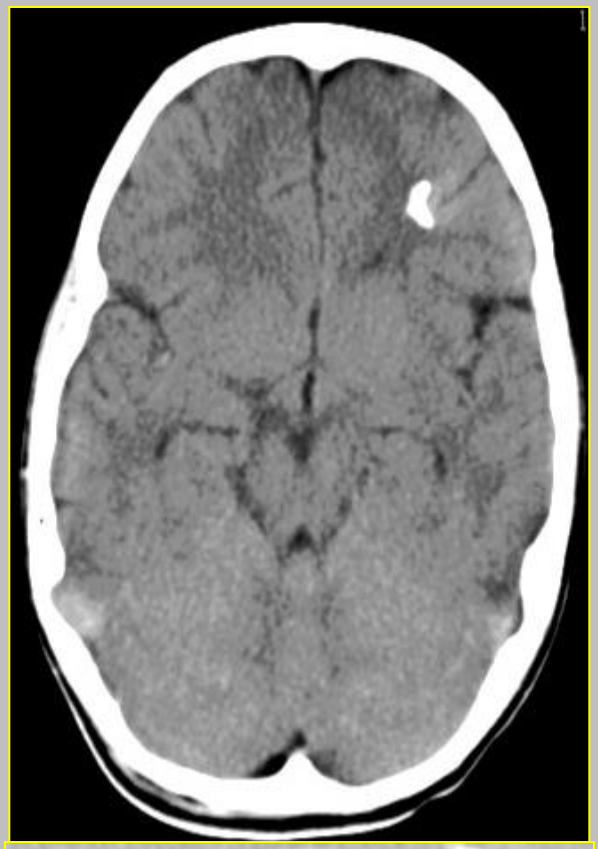


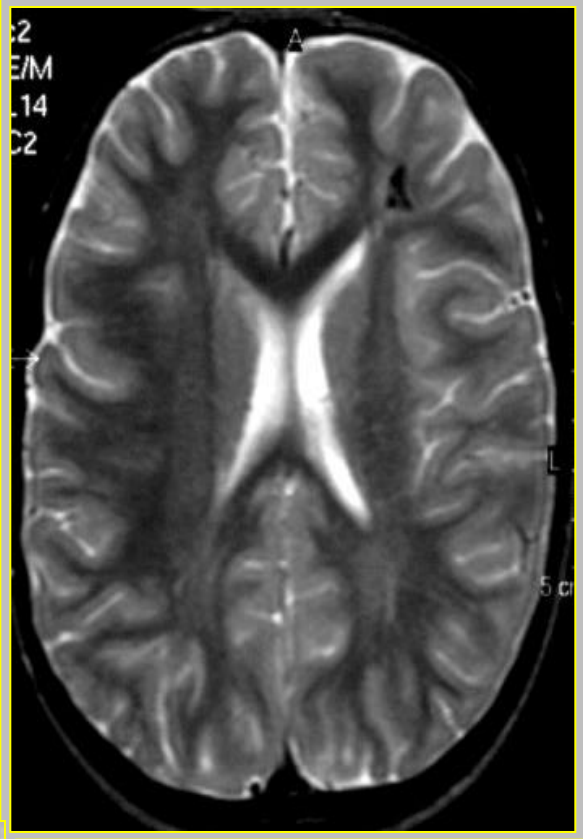


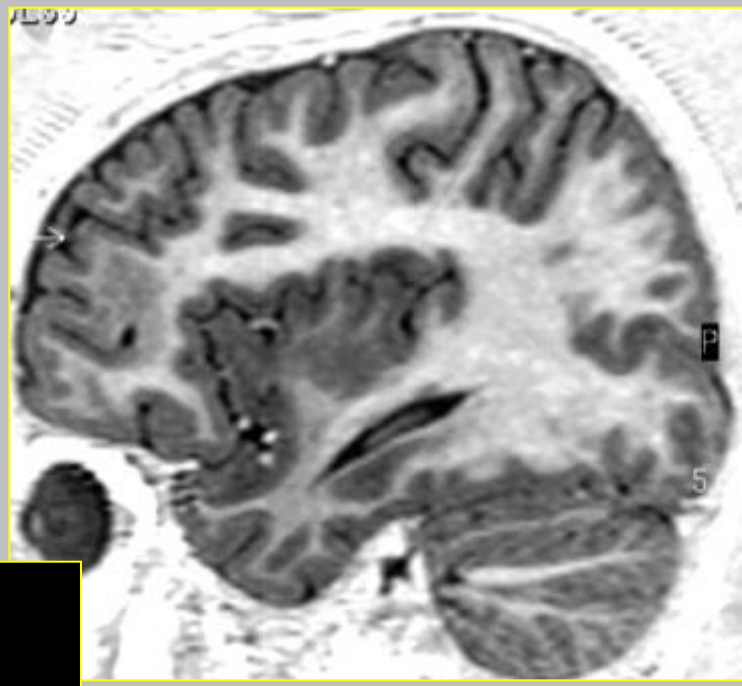


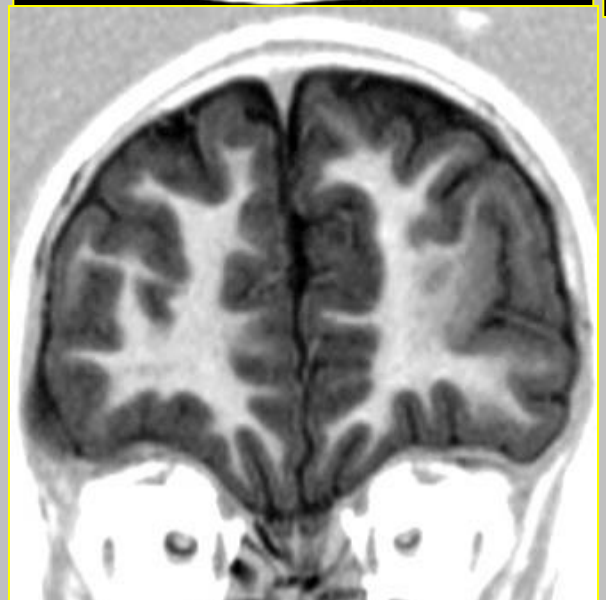


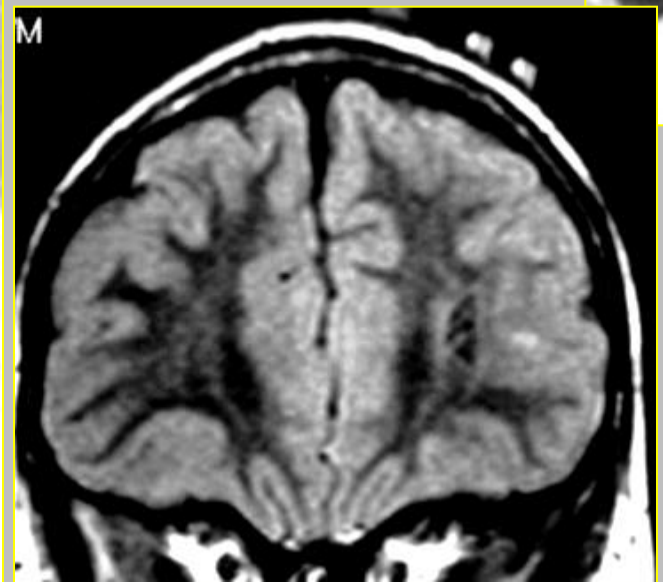
SCLEROSI TUBEROSA











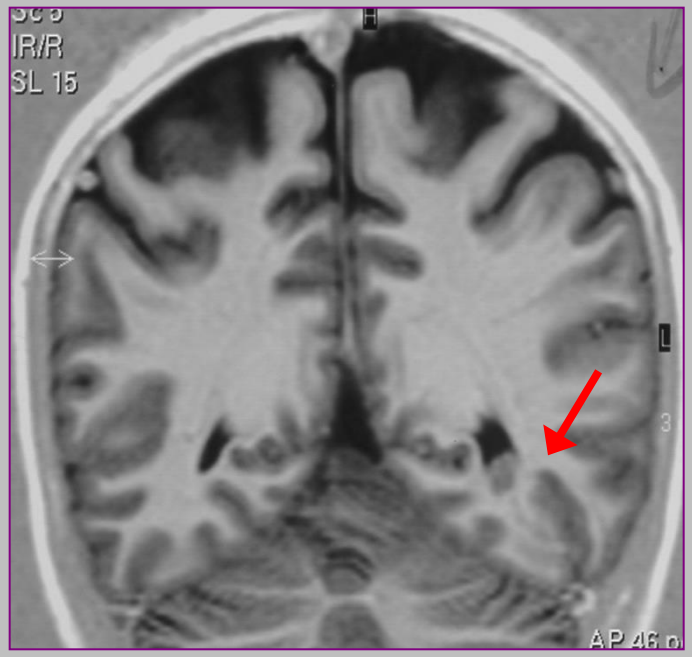
ETEROTOPIE

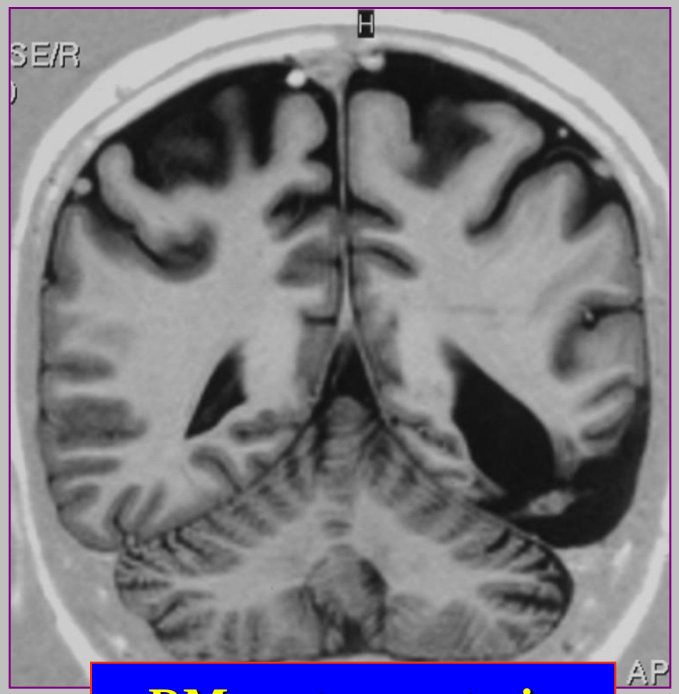
- **Nodulare subependimale -periventricolare**
  - **Unilaterale (> destra)**
  - **Bilaterale (BPNH) trasmissione x-linked dominante**
    - **si manifesta nelle femmine**
    - **letalità prenatale nei maschi**

→ **Gene: Filamina 1, FLN1, sul Ch Xq28**

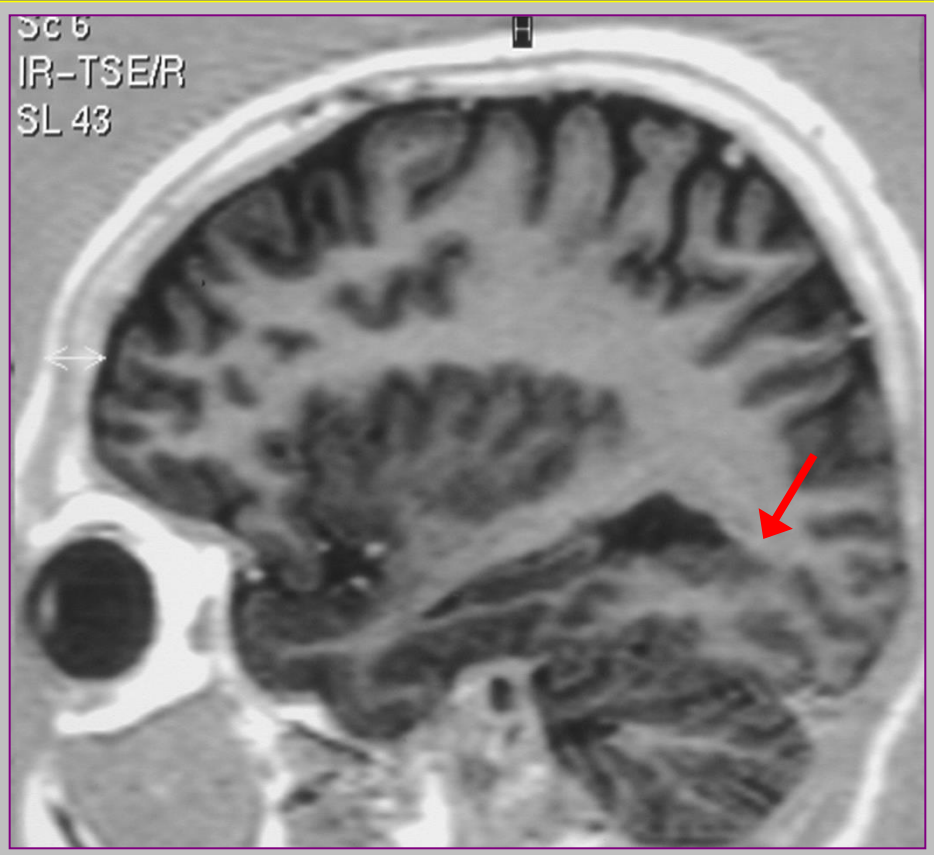
- **Nodulare sottocorticale**
- **A banda sottocorticale o “doppia corteccia” (SBH) (Agyria-pachigyria-band spectrum)**

Eterotopia nodulare periventricolare *monolaterale*

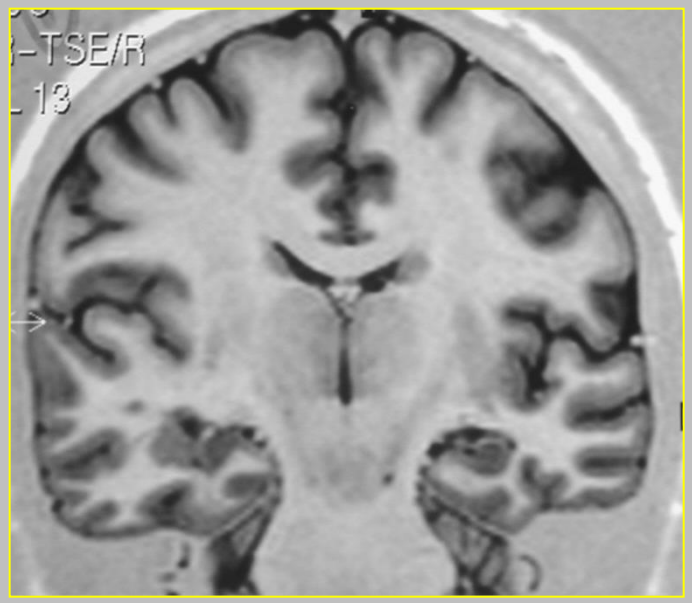


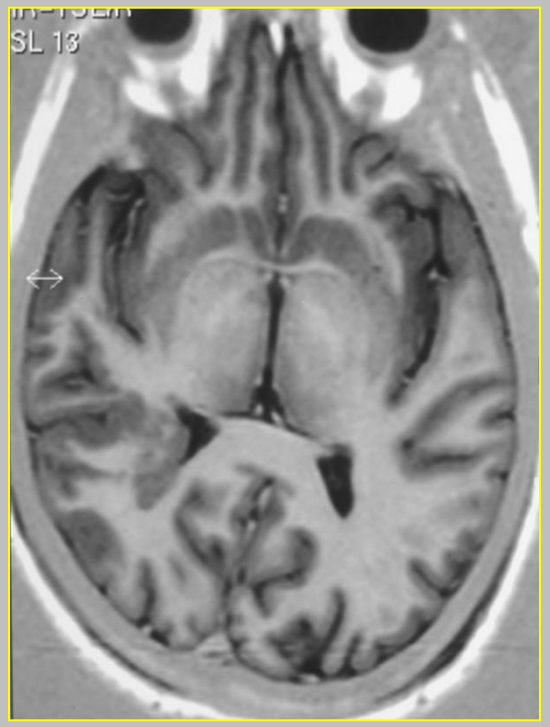


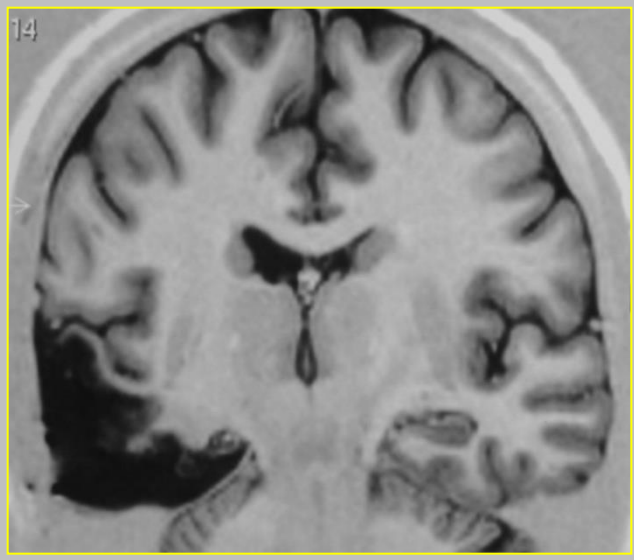
RM post-operatoria

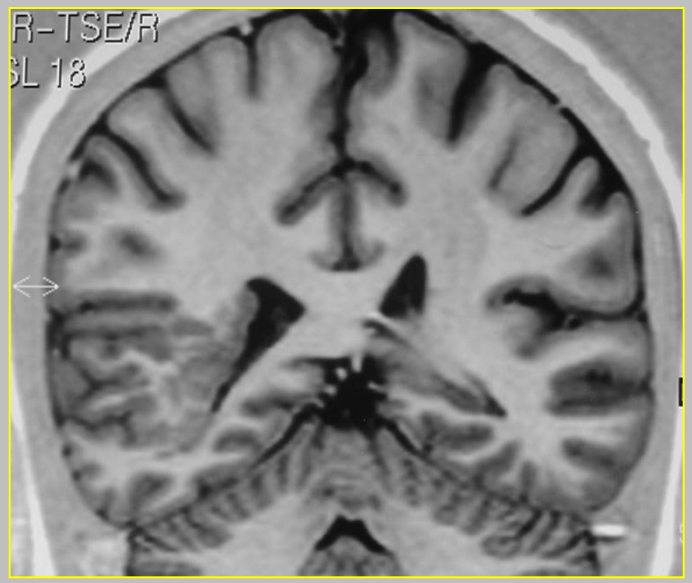


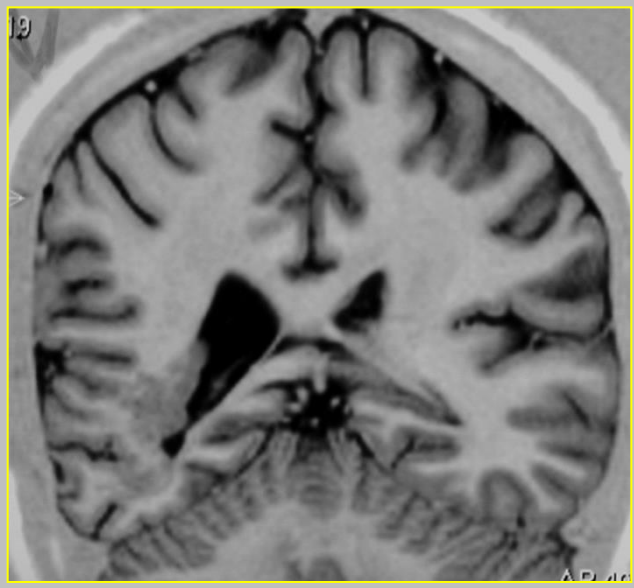
Eterotopia nodulare periventricolare-sottocorticale monolaterale











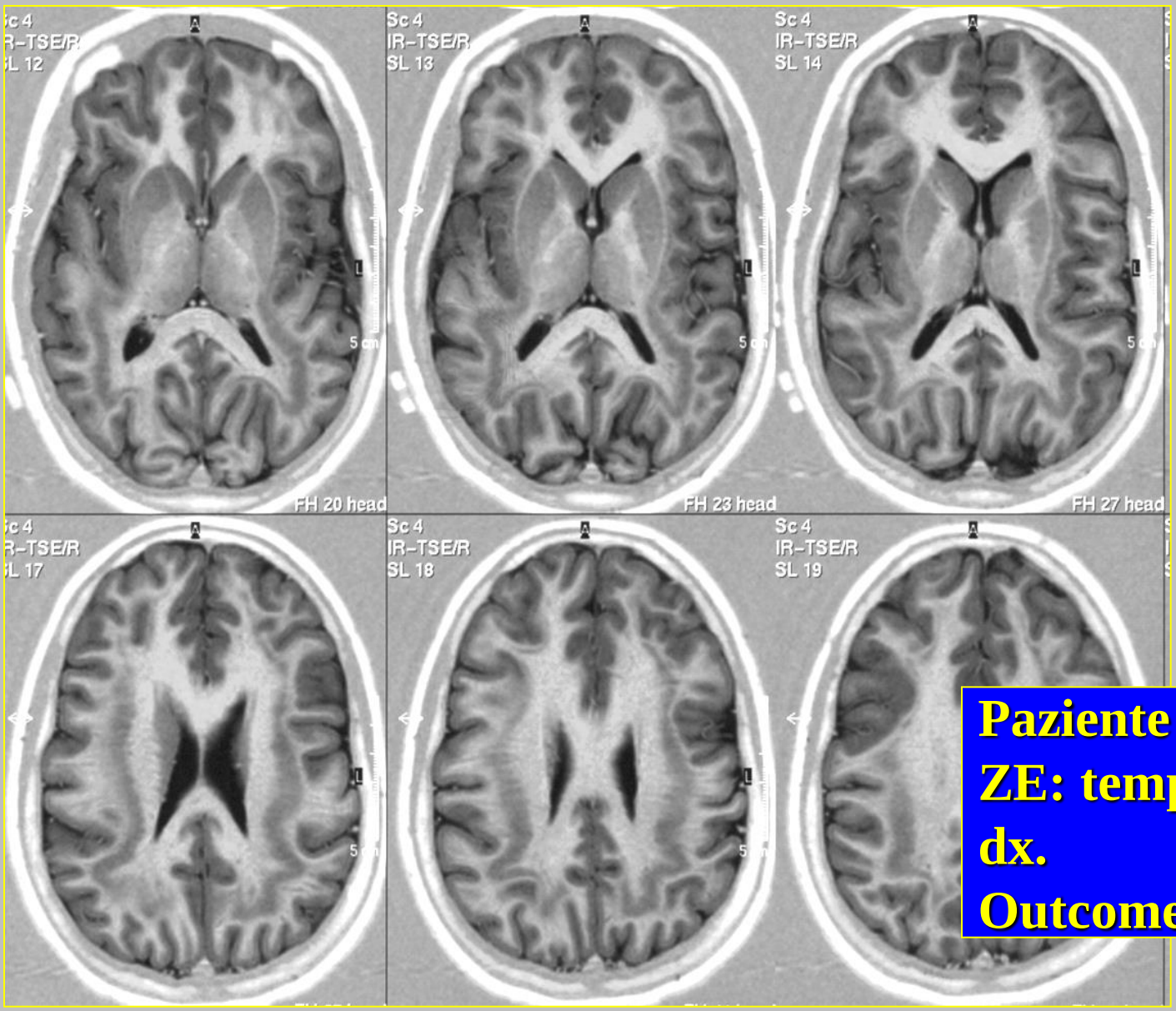
RM postoperatoria
Pz. libero da crisi a 3 aa dall'intervento

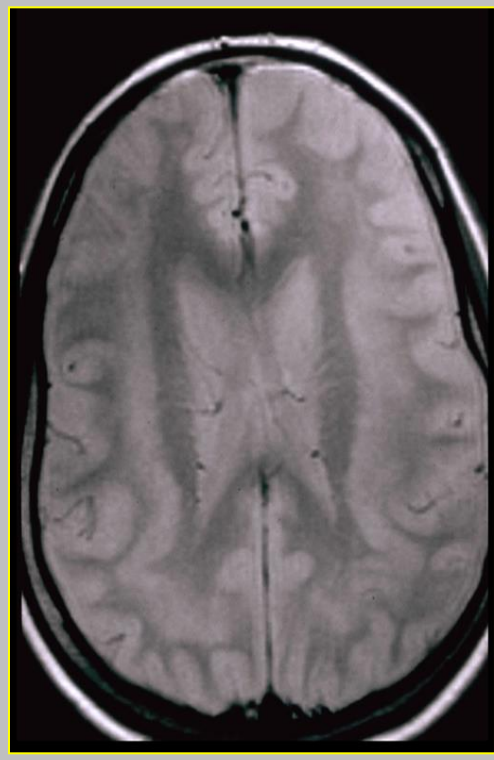
Agiria-pachigiria-band spectrum

- **SBH –lissencefalia trasmissione X-linked dominante**
  - ***"doppia corteccia"* (SBH) nelle femmine**
  - ***Lissencefalia classica* nei maschi**

Gene: doublecortin, DCX, sul Ch Xq22.3-q23

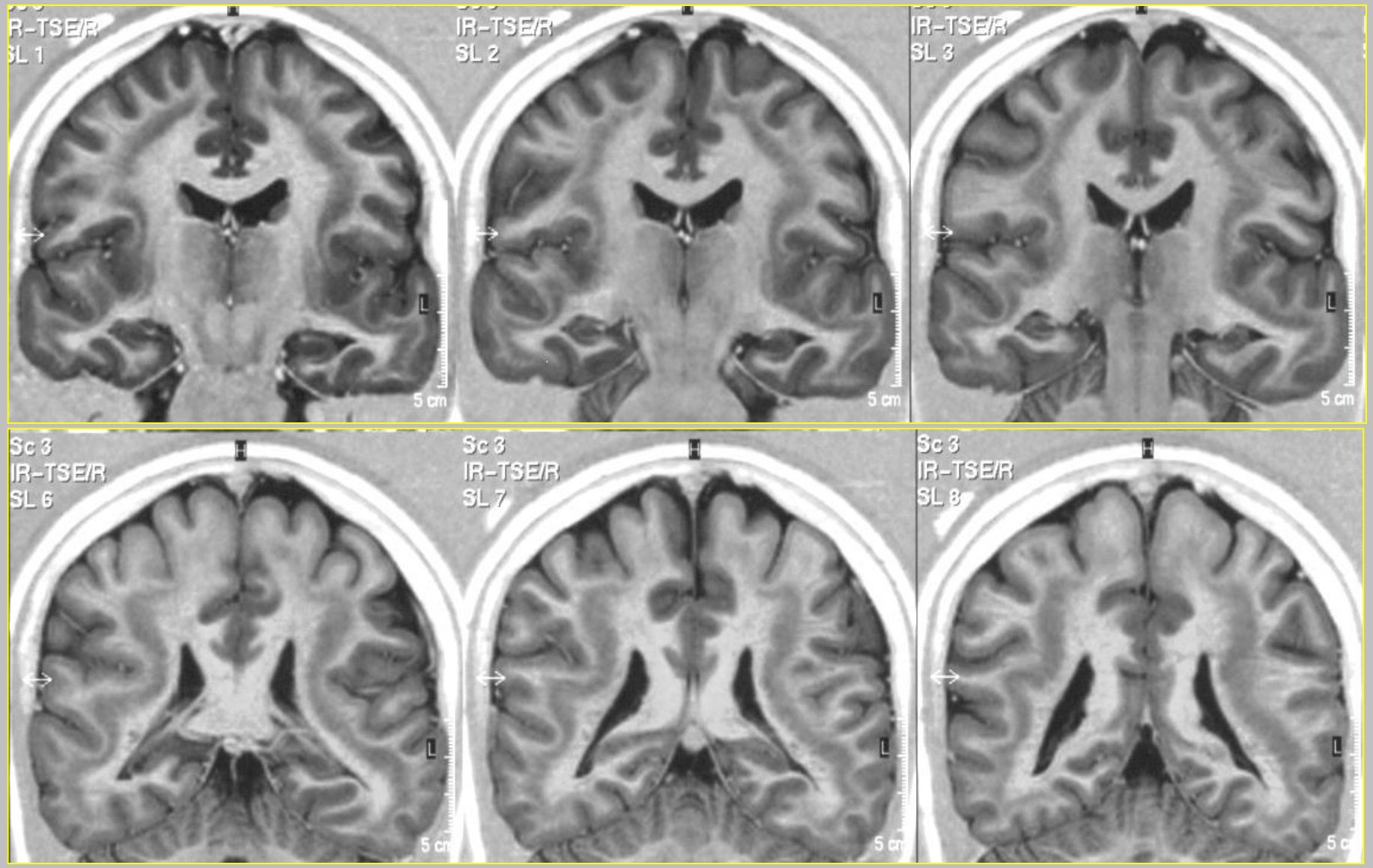
Eterotopia a Banda - Doppia Corteccia





Paziente femmina operata
ZE: temporo-occipitale dx.
Outcome 4 aa: classe 1b

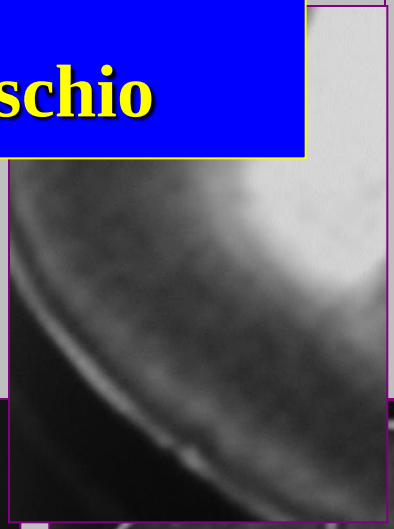
Eterotopia a Banda - Doppia Corteccia

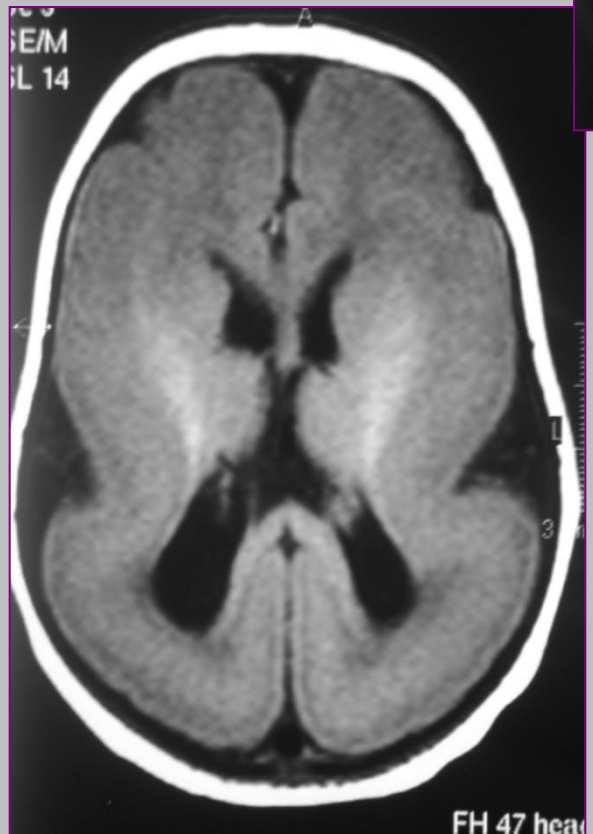


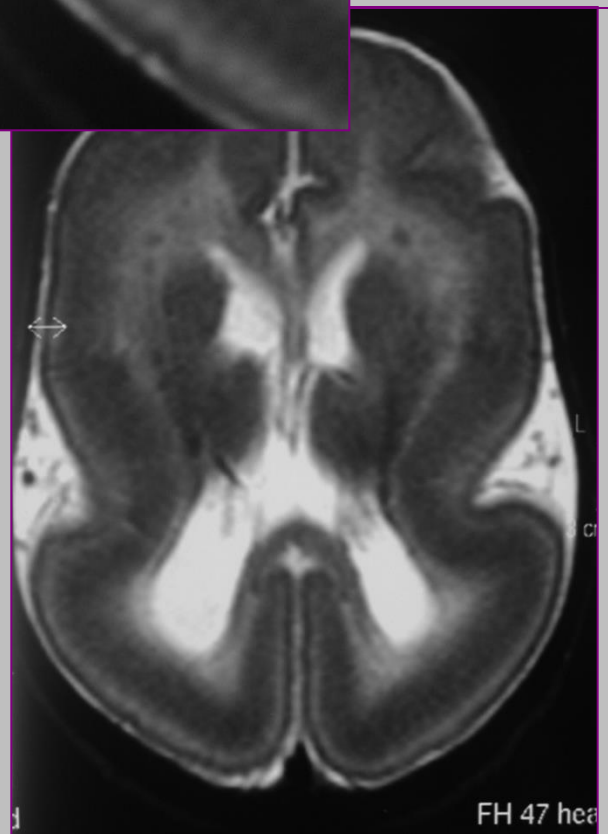
LISSENCEFALIA
classica
Paziente maschio

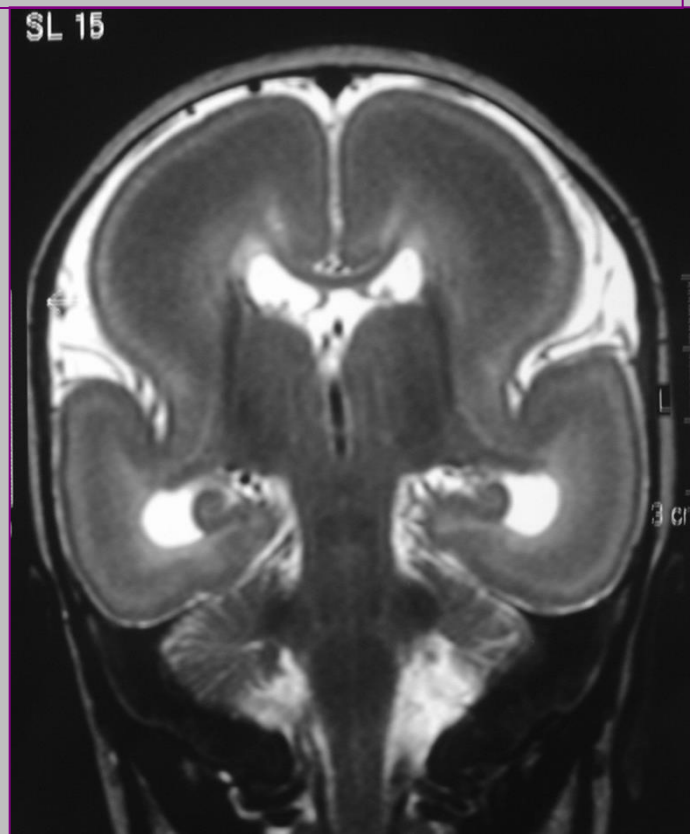
- cervello liscio con perdita totale della solcazione
- corteccia spessa (> 8 mm)
- giunzione bianca/grigia rettilinea
- ipersegnale lineare intracorticale in T2

  (strato di cellule sparse)
- microcefalia



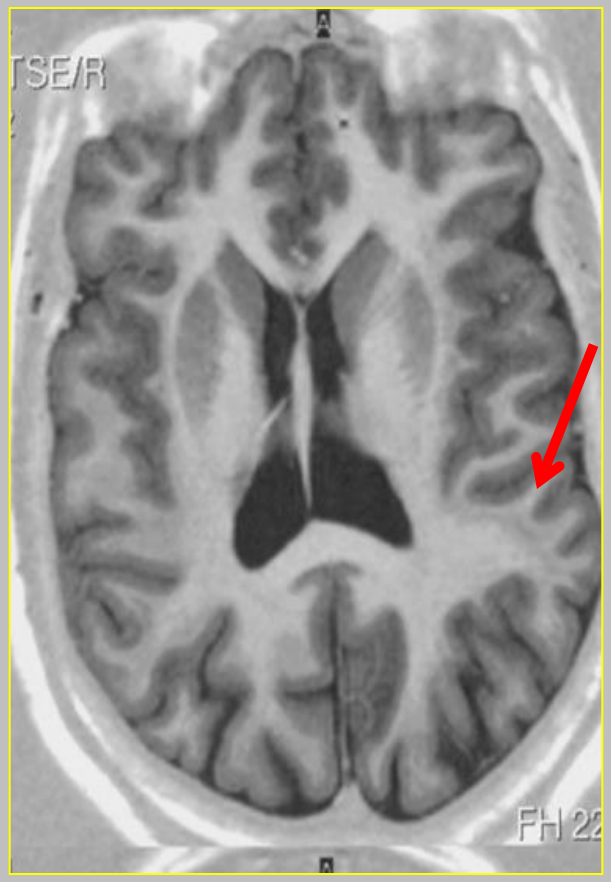


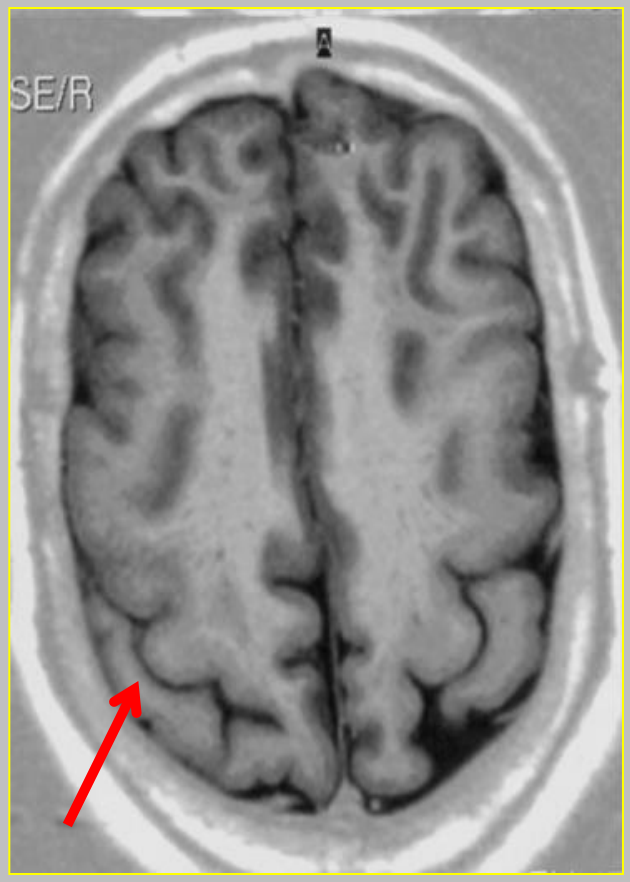


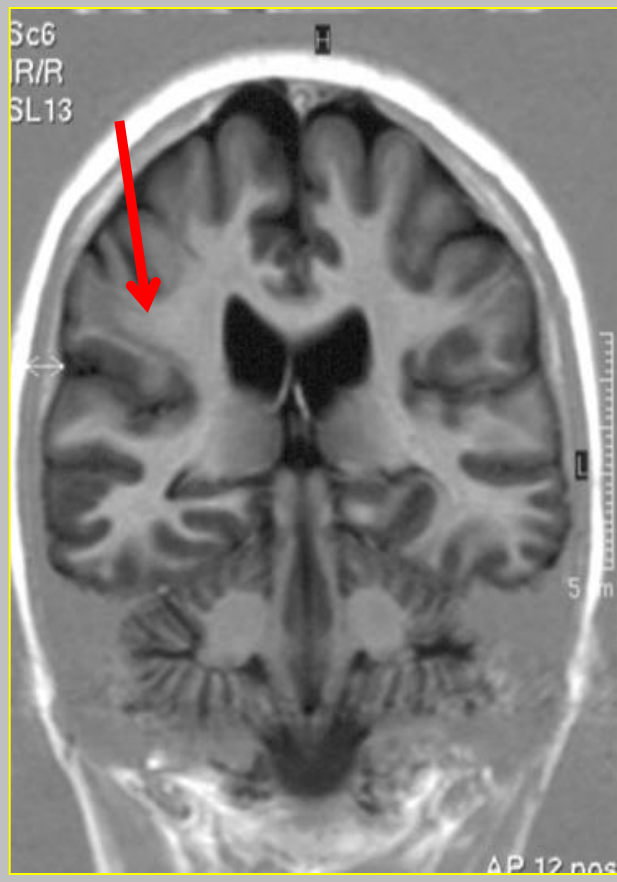


PACHIGIRIA + DOPPIA CORTECCIA INCOMPLETA

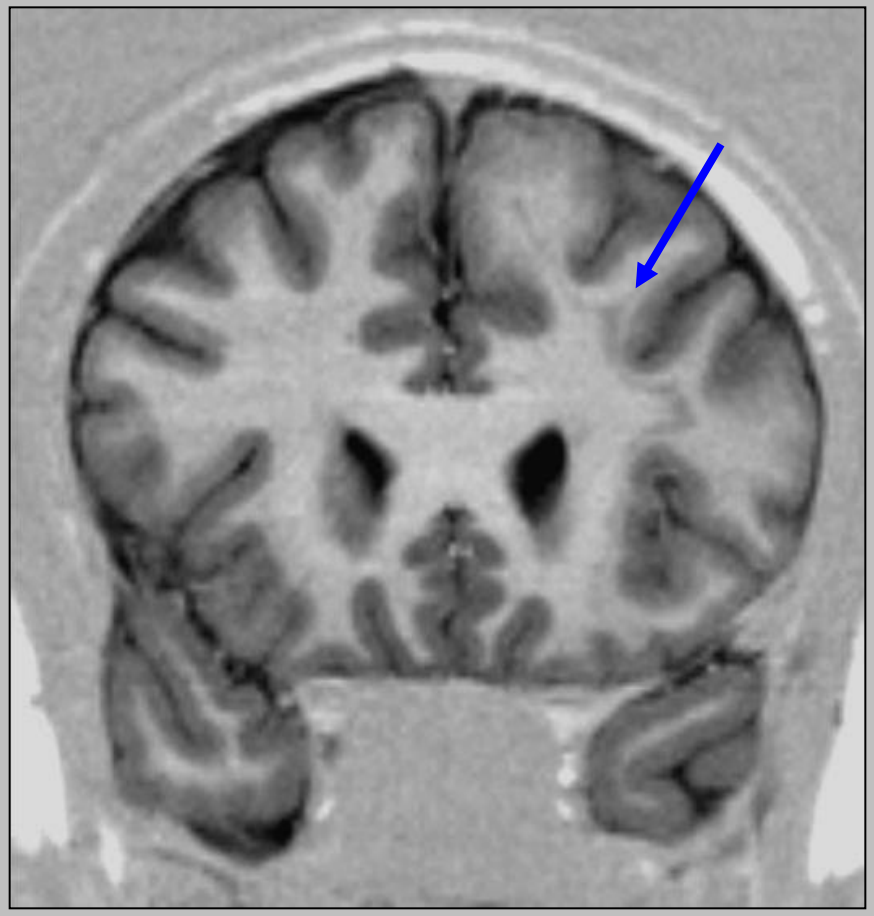
Paziente femmina

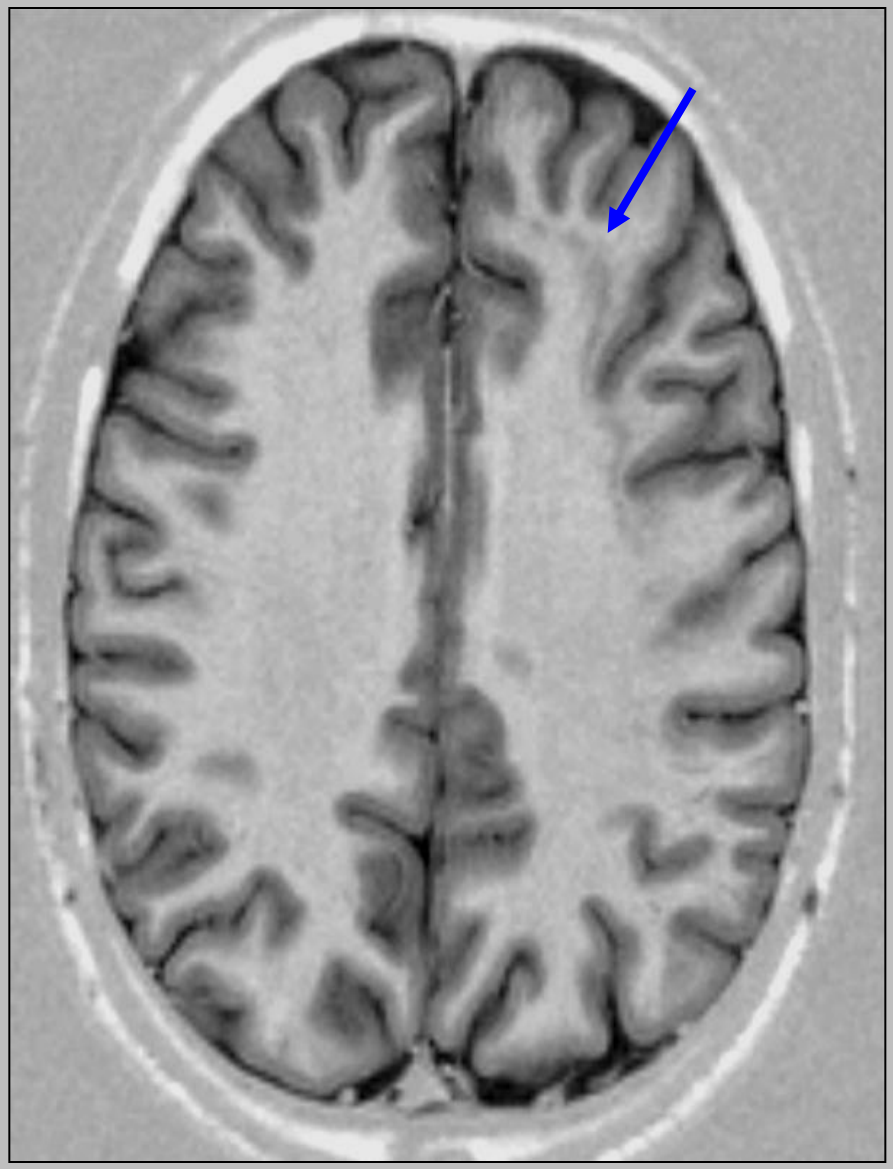






DOPPIA CORTECCIA INCOMPLETA
Paziente maschio



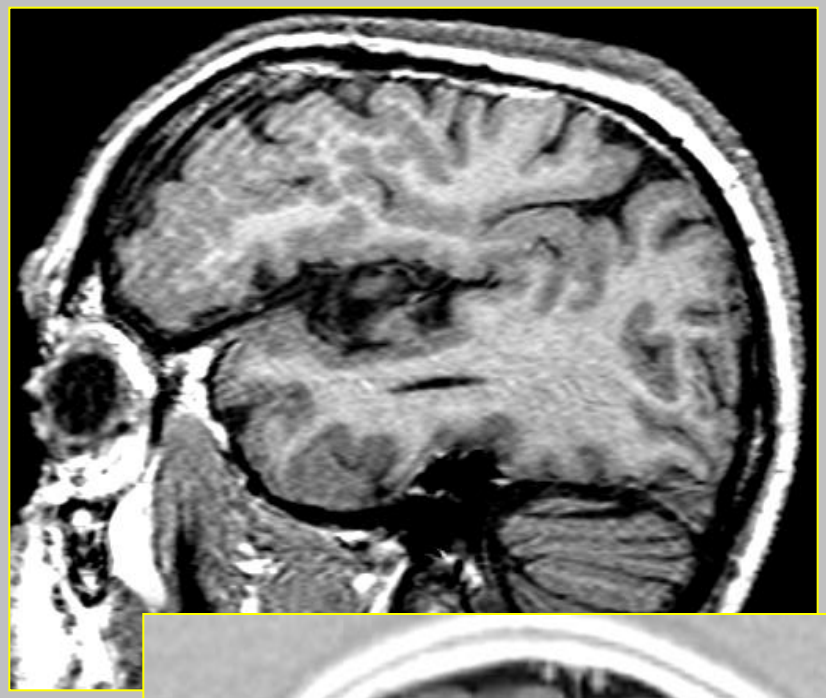


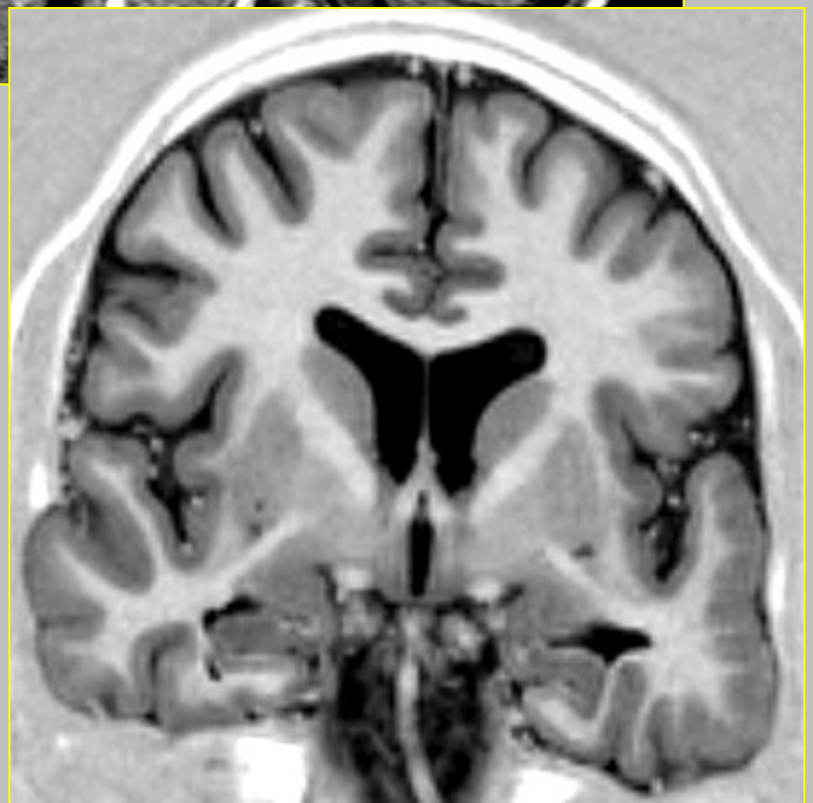
Mutazioni sia DCX che LIS1
Brain 2002,125: 2507-2522

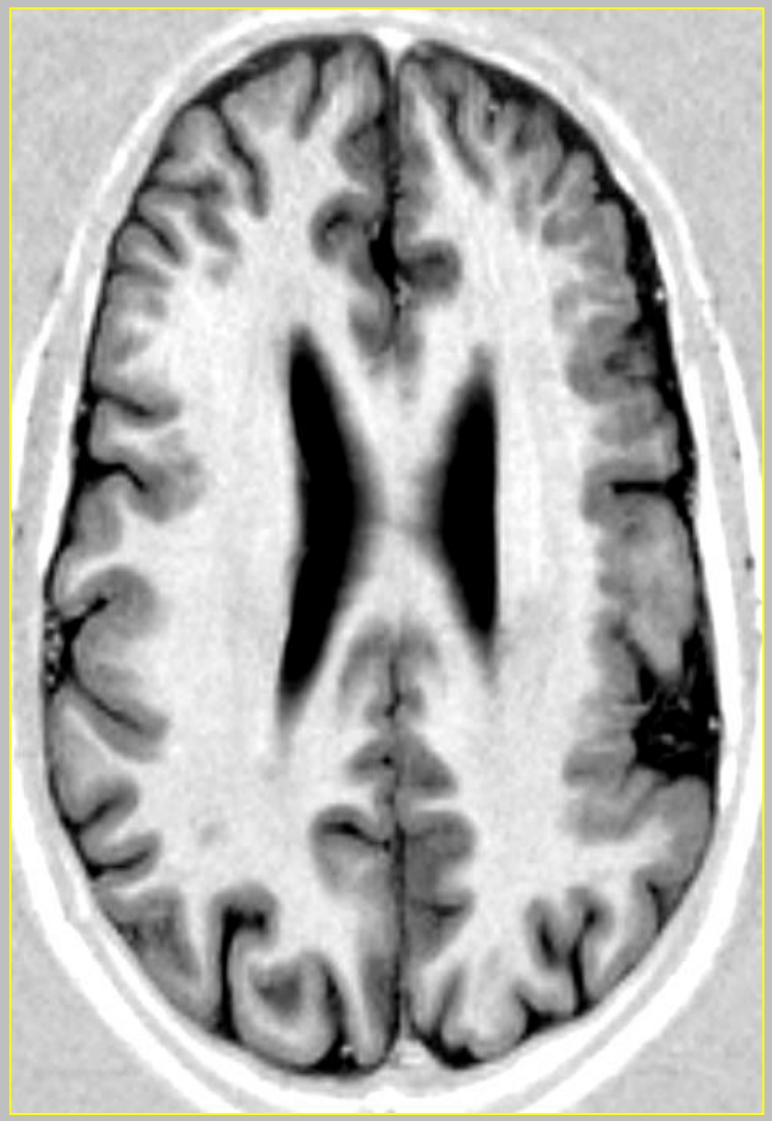
POLIMICROGIRIA

- Corteccia pachigirica
- Corteccia microgirica, normale o assottigliata
- Superficie corticale bozzoluta
- Giunzione SG/SB netta, seghettata o irregolare
- Ridotta SB sottocorticale
- Ripiegamento corticale
- Allargamento degli spazi subaracnoidei sovrastanti
- Anomalie venose associate

Polimicrogiria







TUMORI EPILETTOGENI

- Esordio clinico precoce
- Generalmente di basso grado ed a lenta crescita

TUMORI EPILETTOGENI

Serie

- 1079 pazienti operati per epilessia parziale farmaco-resistente presso il Centro per la Chirurgia dell'Epilessia dell'Ospedale Niguarda di Milano dal 1995 al 2011

Tumori (n°333)

ISOLATI

- Neoplasie 232 (69,6%)
- Amartomi 8 (2.4%)

- Neoplasie o amartomi

associati a MSC (displasie corticali) 93 (28%)

WHO classification

Istotipi
tumori (n°333)

• Ganglioglioma (GG)	130 (39%)
• Tumore Disembroplastico Neuroepiteliale (DNET)	86 (26%)
• Amartoma	46 (14%)
• Oligodendroglioma	25 (7%)
• Astrocitoma Pilocitico	16 (5%)
• Xantoastrocitoma Pleomorfo (PXA)	14 (4%)
• Astrocitoma	10 (3%)
• Dermoide	3 (1%)
• Meningioangiomatosi	3 (1%)

DNET
(Dysembryoplastic Neuroepithelial Tumor)

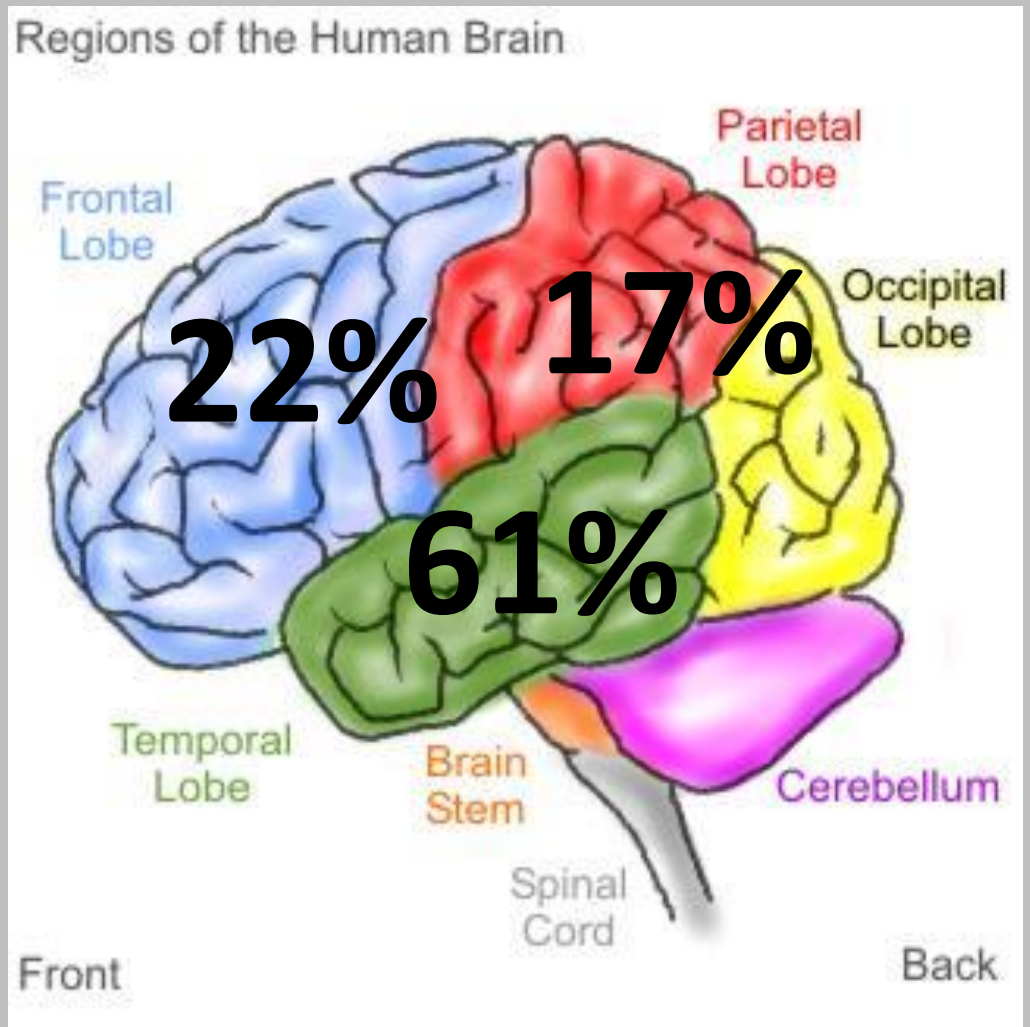
- Neoplasia glio-neuronale benigna (grado I° WHO)
- Usualmente sopratentoriale e con costante interessamento corticale
- Architettura multinodulare con “glioneuronal element” (fascicoli di assoni adesi a cellule simil-oligodendrogliali ed a neuroni dispersi in una matrice mixoide)
- Non atipie citologiche
- Possibile associazione con displasia corticale.

DNET (86 casi)

Temporali: 52 (61%)

Anteriori: 19 (22%)

Posteriori: 15 (17%)



DNET (86 casi)

- Sesso: 30 femmine 56 maschi
- Lato: 44 dx 42 sx

- Età d'esordio delle crisi
  0-10 aa: 49 (57%)
  10-18 aa: 24 (28%)
  > 18 aa: 13 (15%)

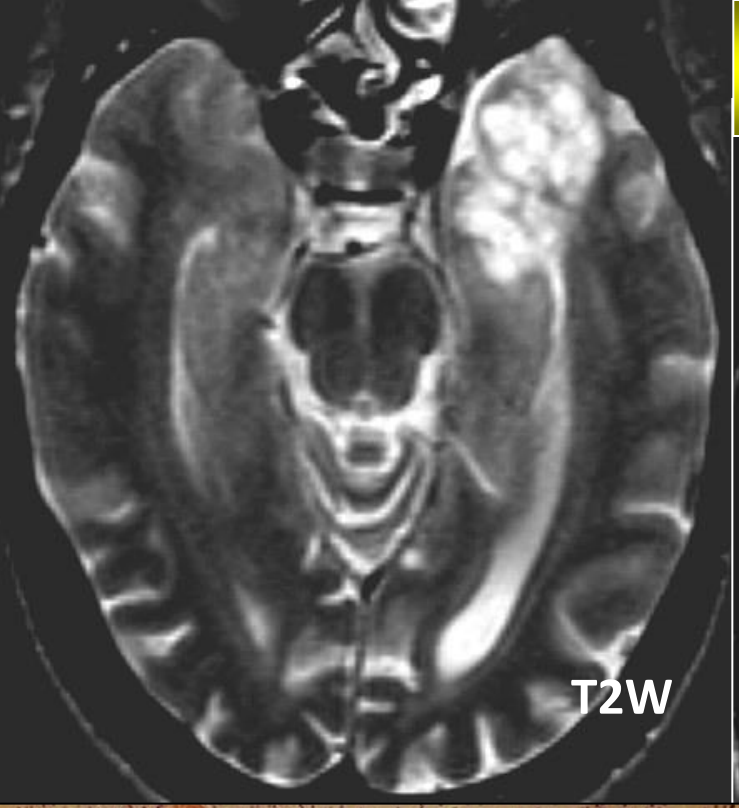
Outcome crisi dopo intervento chirurgico

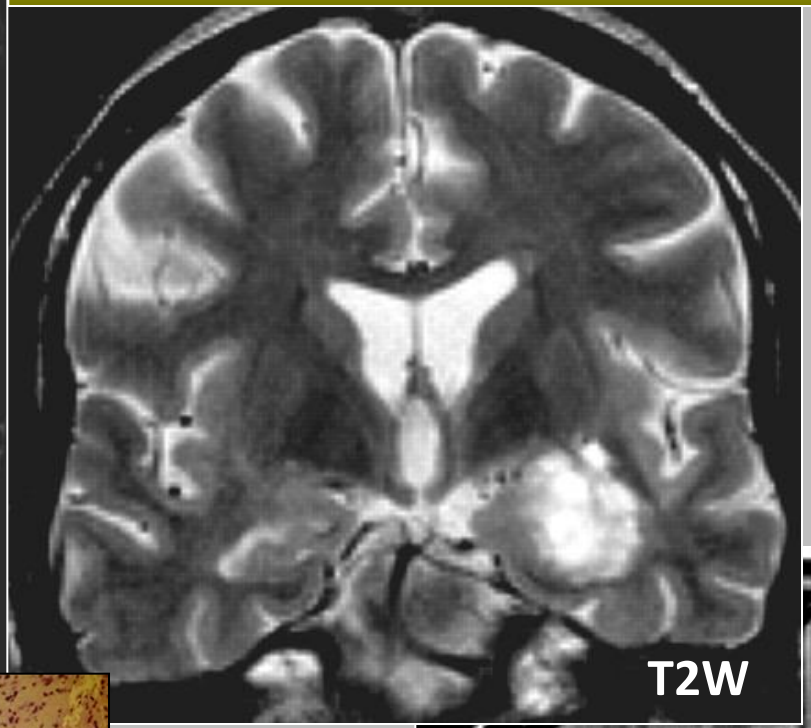
Guariti (classe I di Engel): 69 (80%)
Non guariti: 17 (20%)

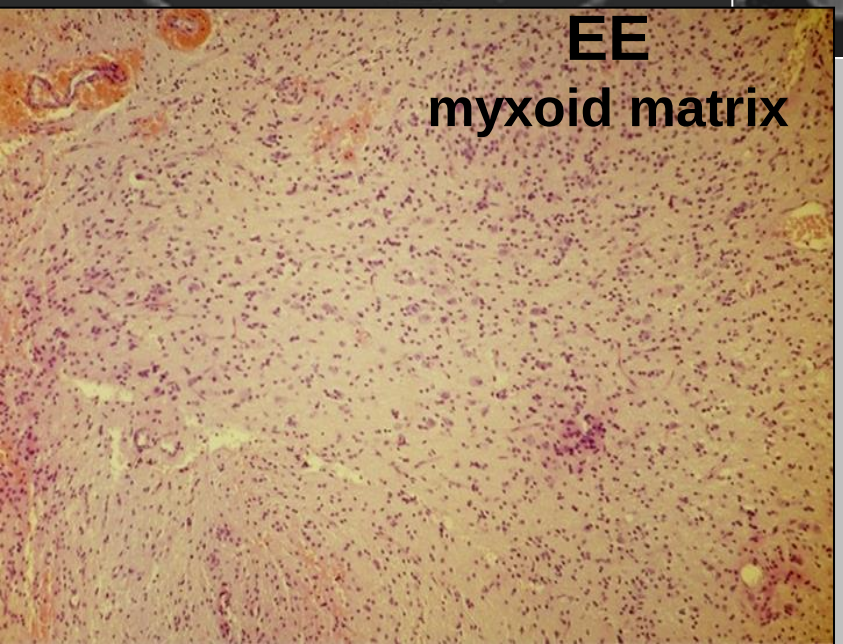
Aspetti neuroradiologici dei DNET

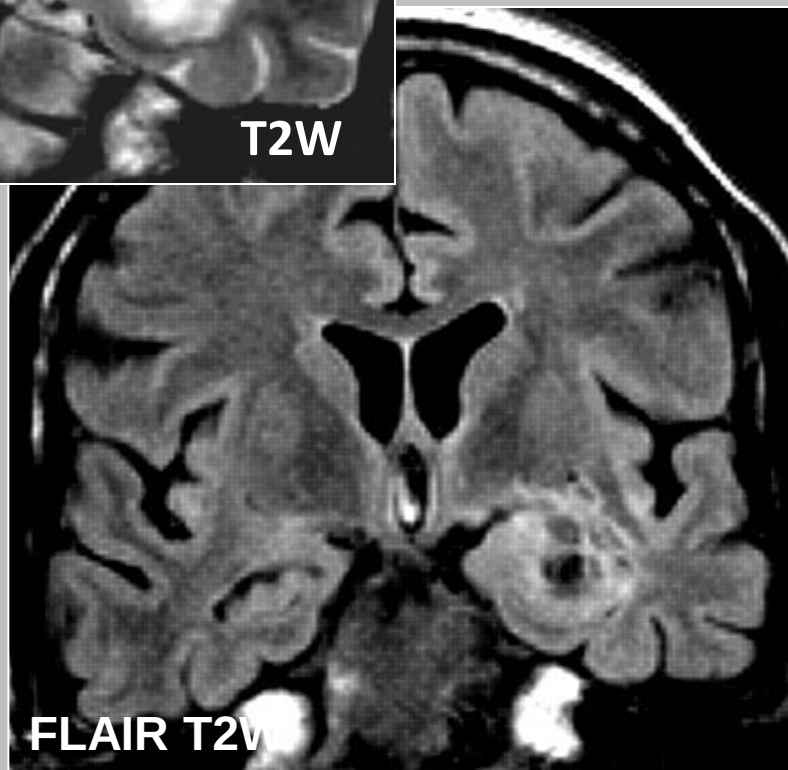
- Piccole dimensioni <3 cm: 47 (55%), 3-6 cm: 32 (37%), >6 cm: 7 (8%)
- Margini netti
- Effetto massa raro 15 casi (17%)
- Aspetto cistico o multicistico
- Componenti solide
- Aspetto giriforme
- Rare calcificazioni 6 casi (7%)
- No edema
- Rimodellamento della teca cranica 12 casi (19%)
- Enhancement raro 18 casi (20%)

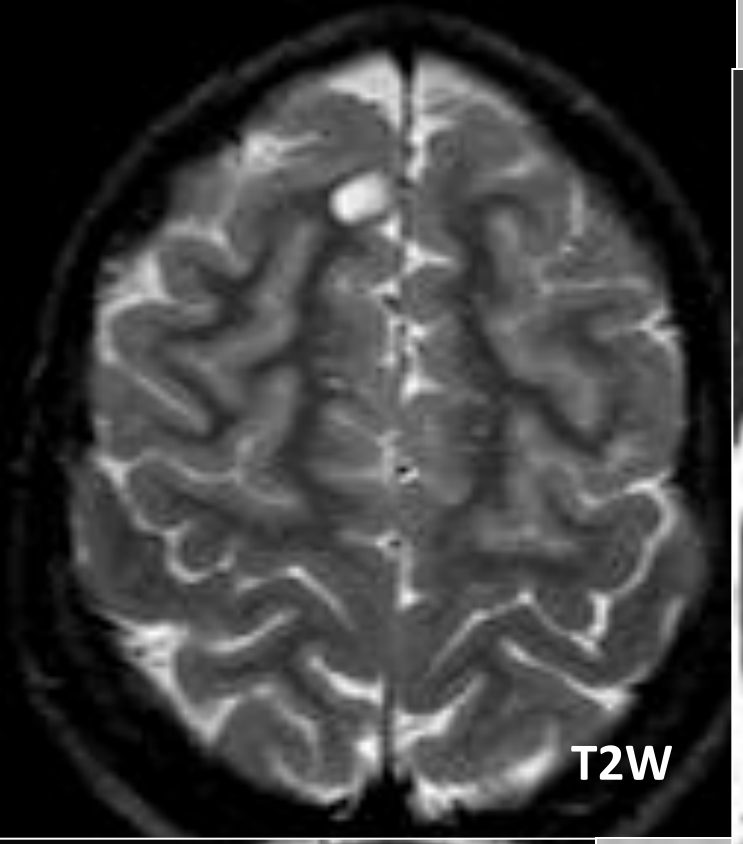
DNET Multicistico

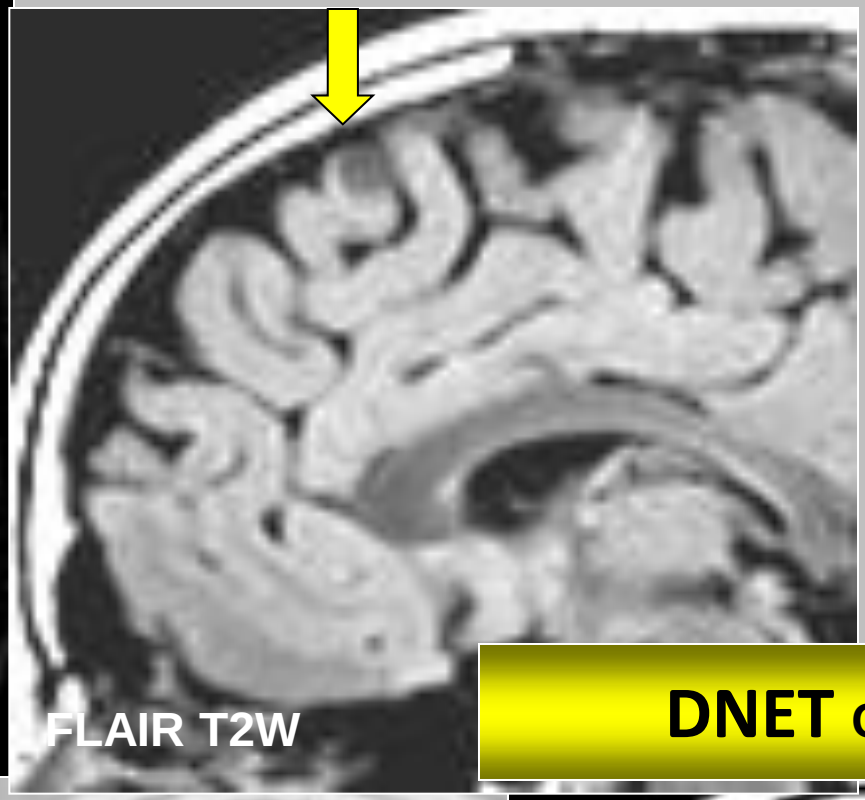




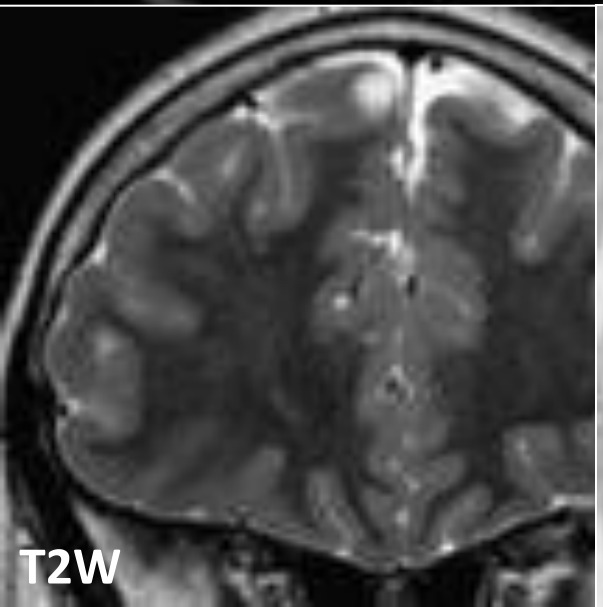


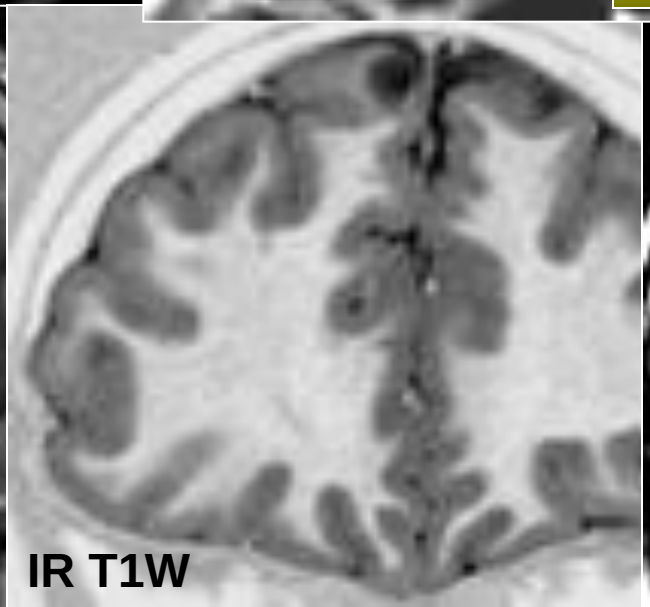


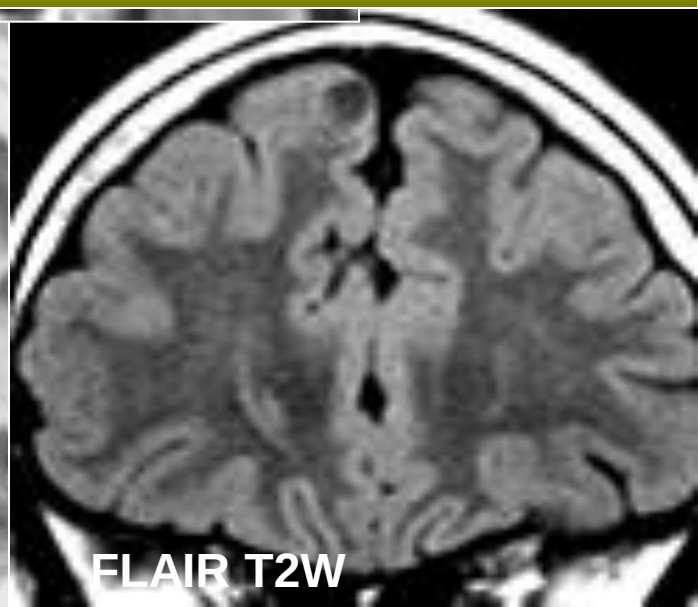




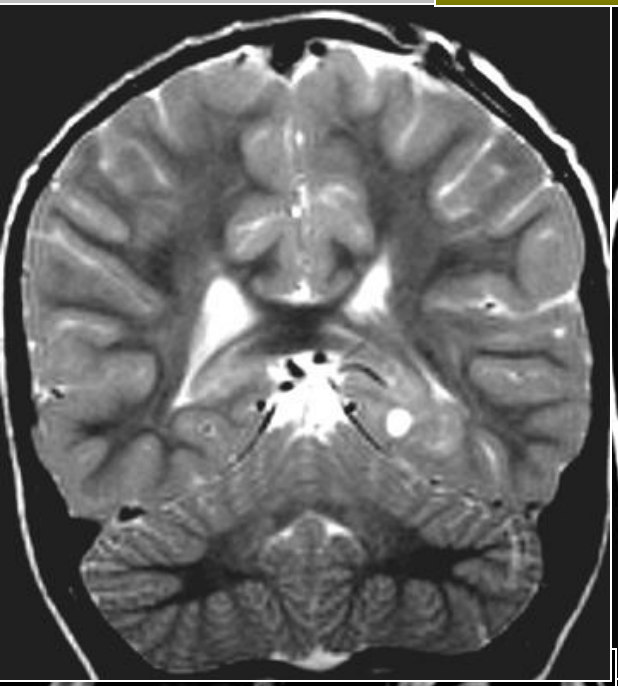
DNET Cistico

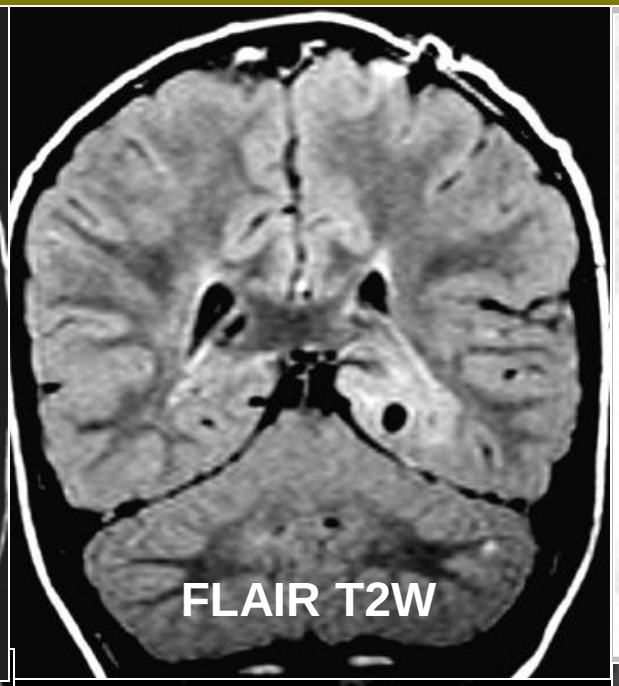


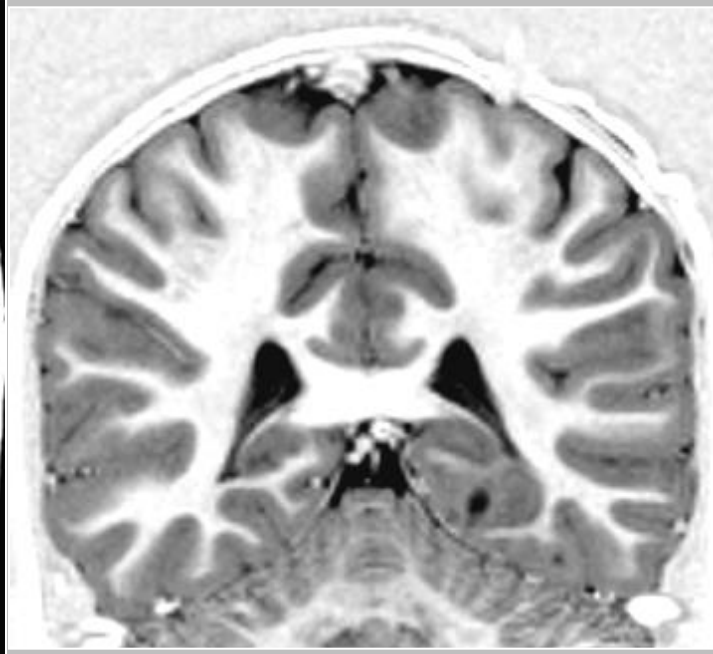


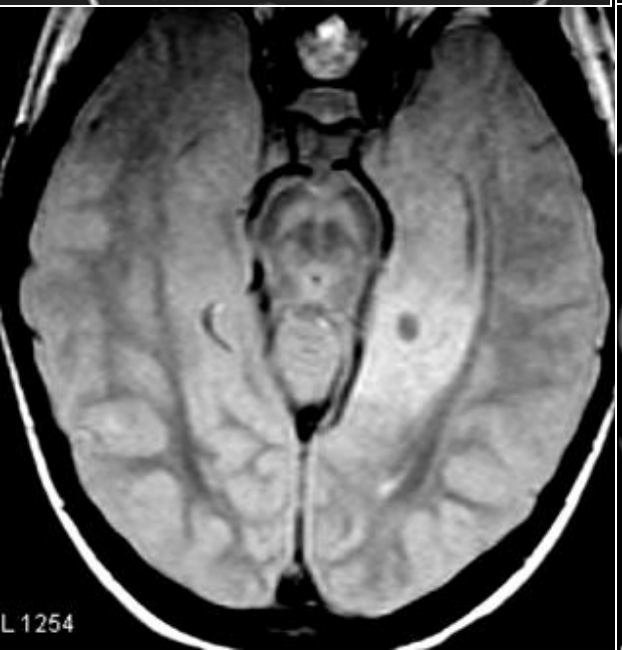


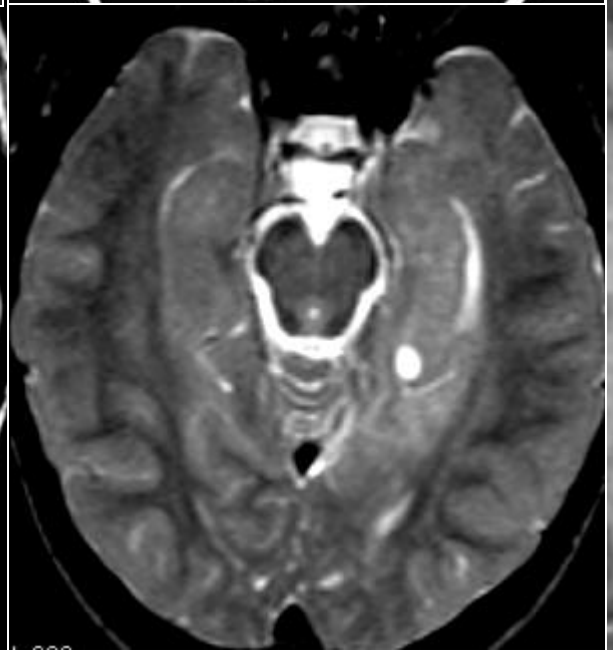
DNET Cistico-solido

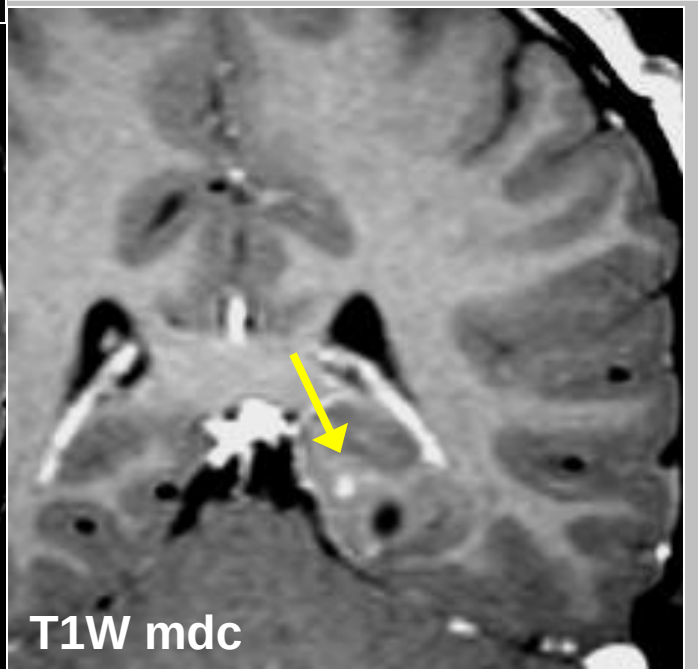




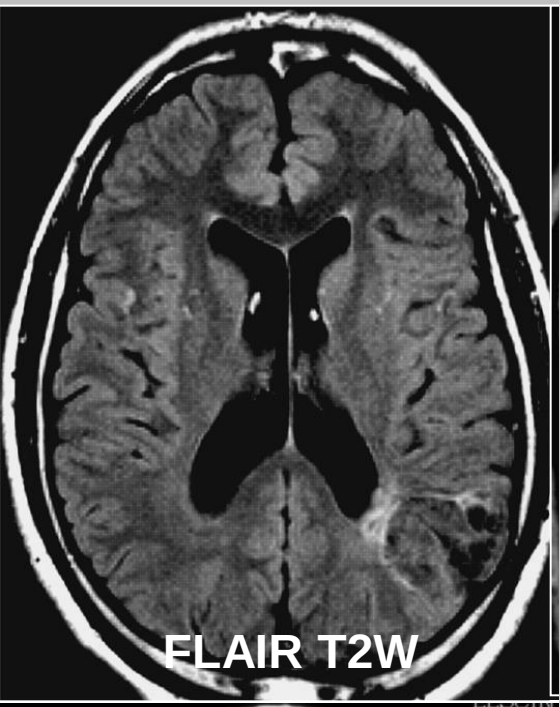


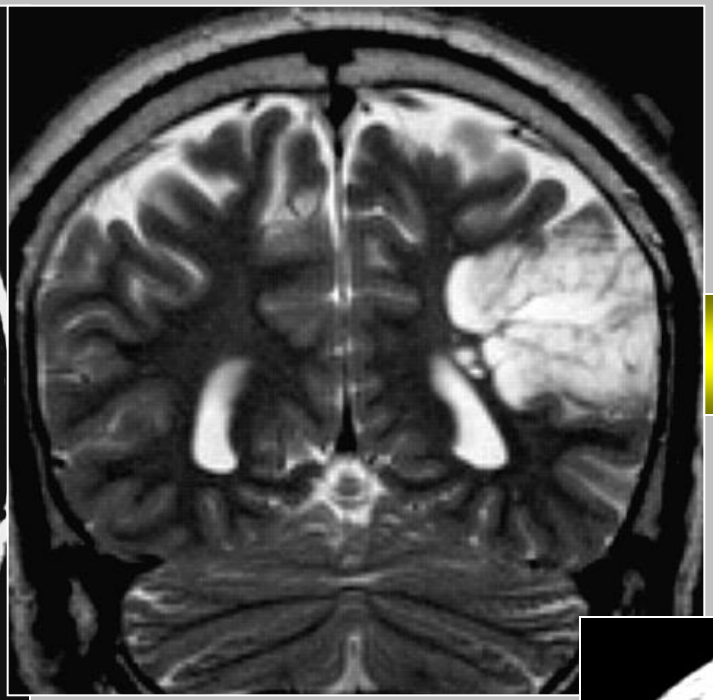


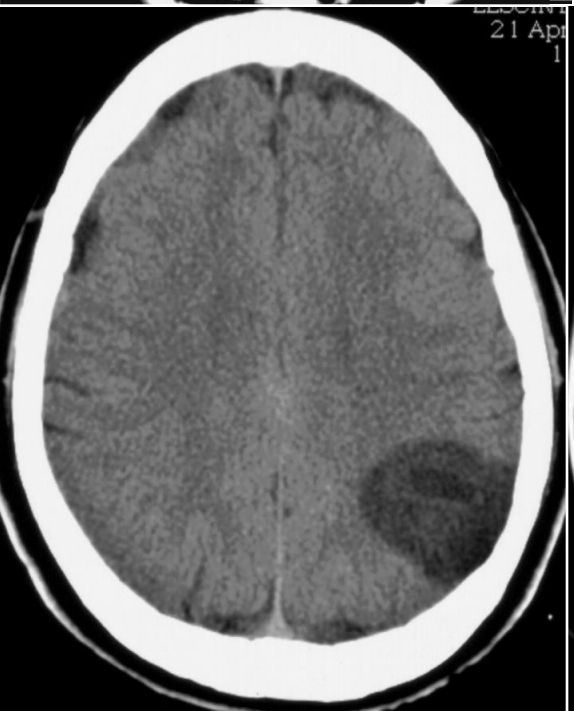


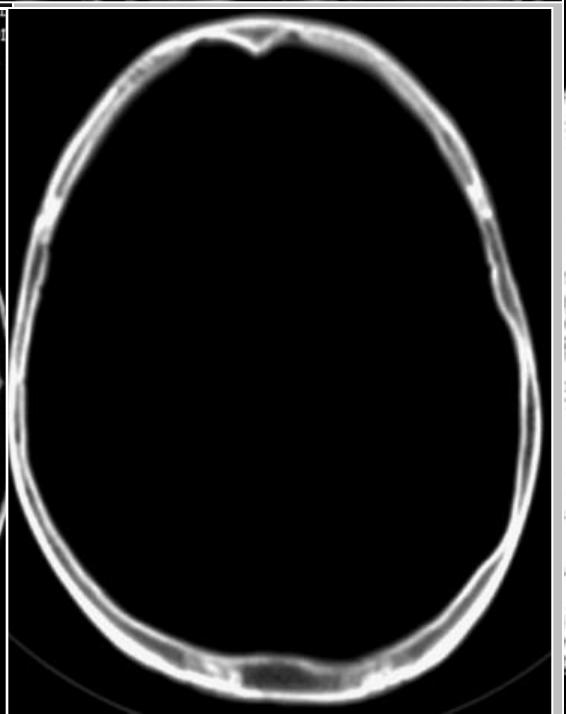


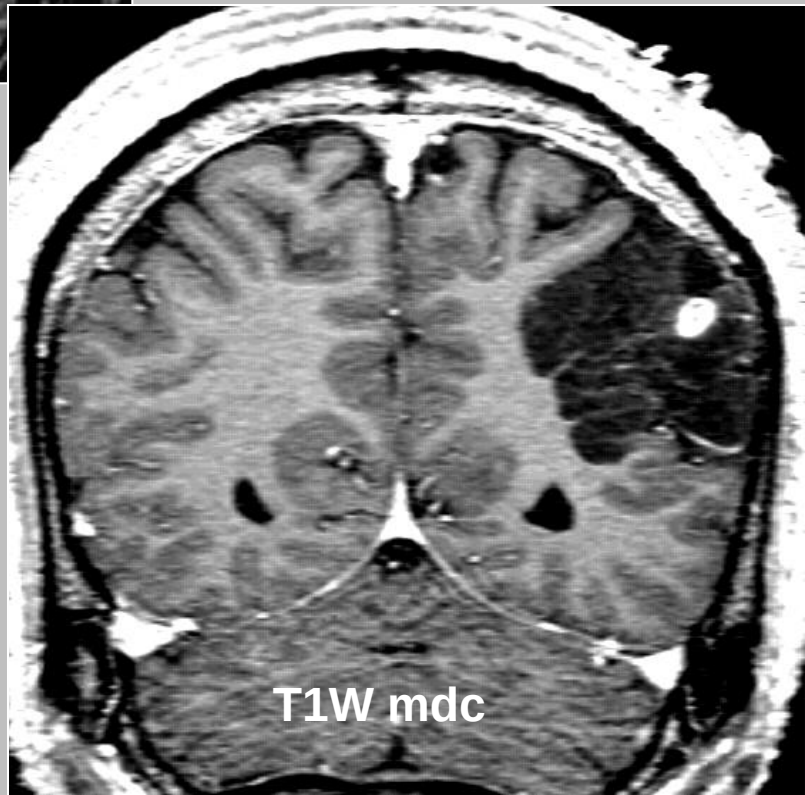
DNET Giriforme



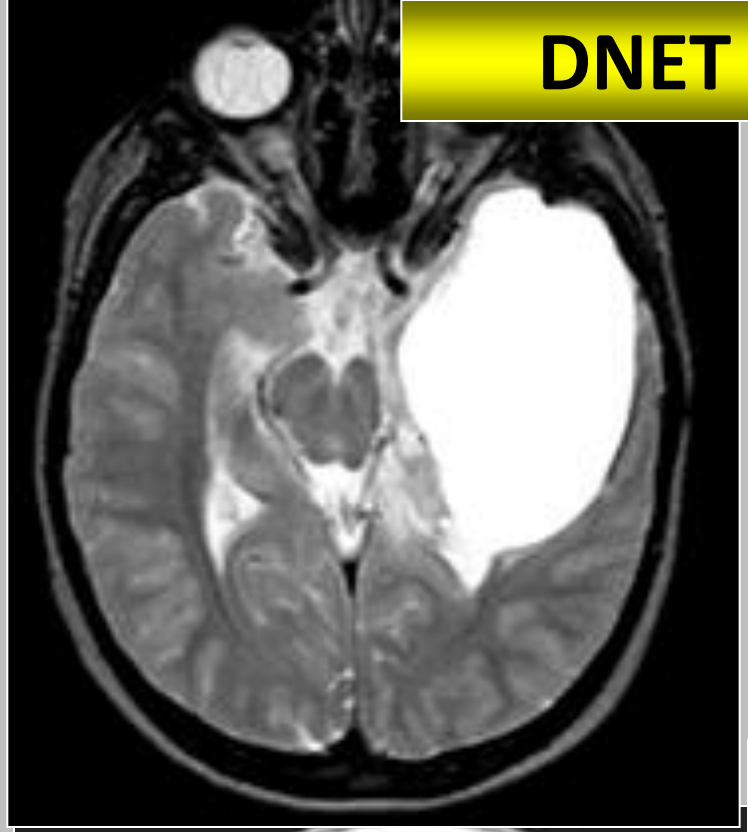


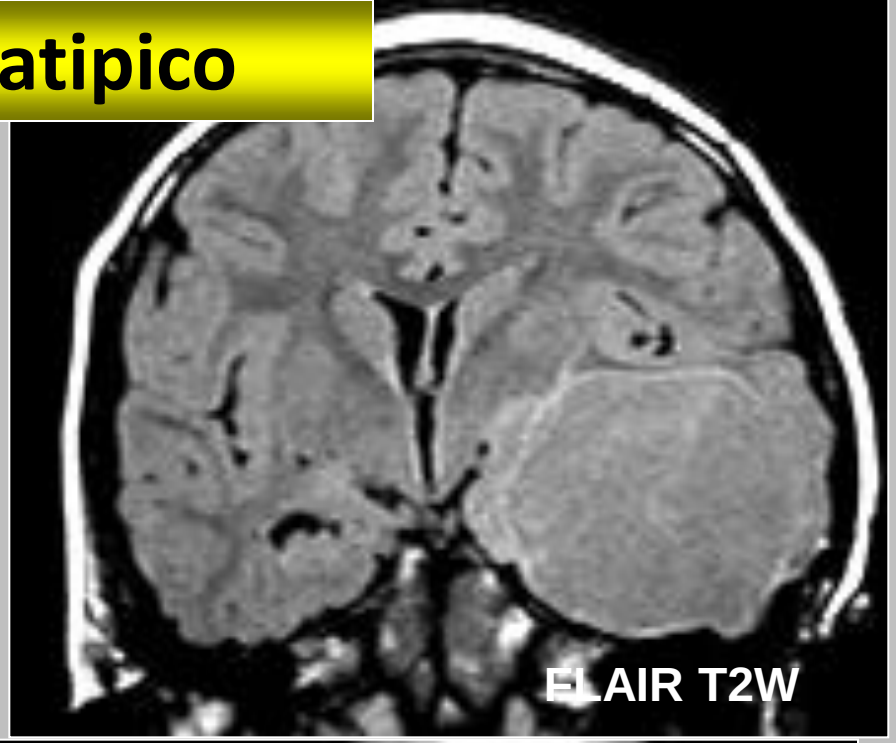


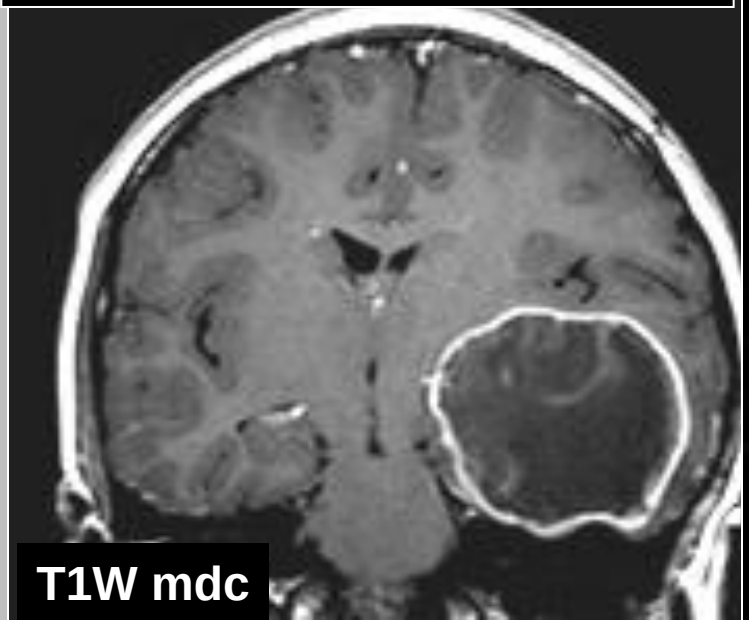


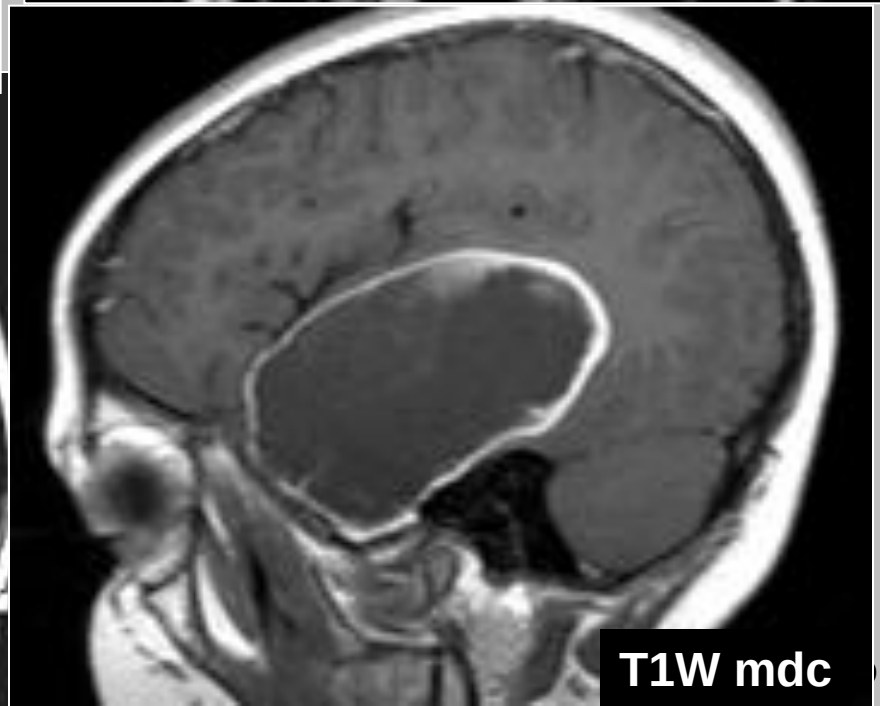


DNET atipico







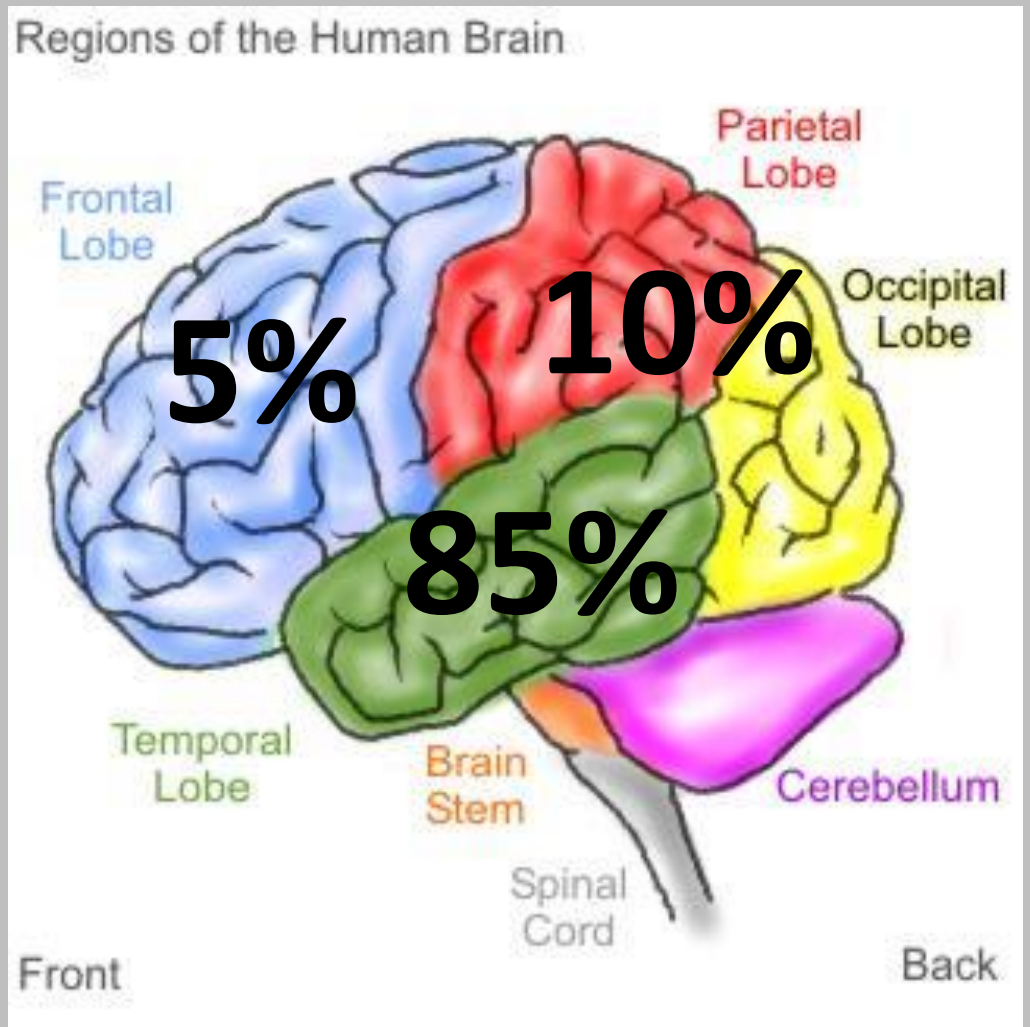


Ganglioglioma

- Tumore neuroepiteliale a lento accrescimento
- Composizione: cellule ganglionari mature e cellule gliali neoplastiche
- Nella maggior parte dei casi Grado I° o II° WHO, possibile trasformazione anaplastica delle componenti gliali
- Possibile associazione con displasie corticali

Gangliogliomi (130 casi)

- Temporali: 111 (85%)
- Posteriori: 12 (10%)
- Anteriori: 7 (5%)



Gangliogliomi (130 casi)

- Sesso: 59 femmine 71 maschi
- Lato: 68 dx 62 sx

- Età d'esordio delle crisi
  0-10 aa: 76 (58%)
  10-18 aa: 41 (32%)
  > 18 aa: 13 (10%)

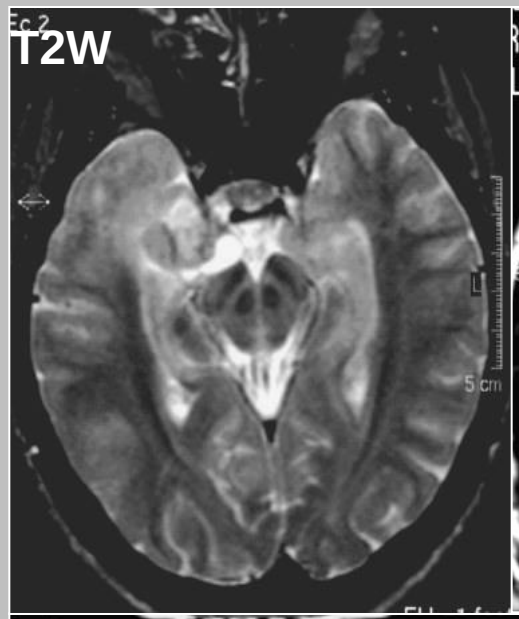
Outcome crisi dopo intervento chirurgico

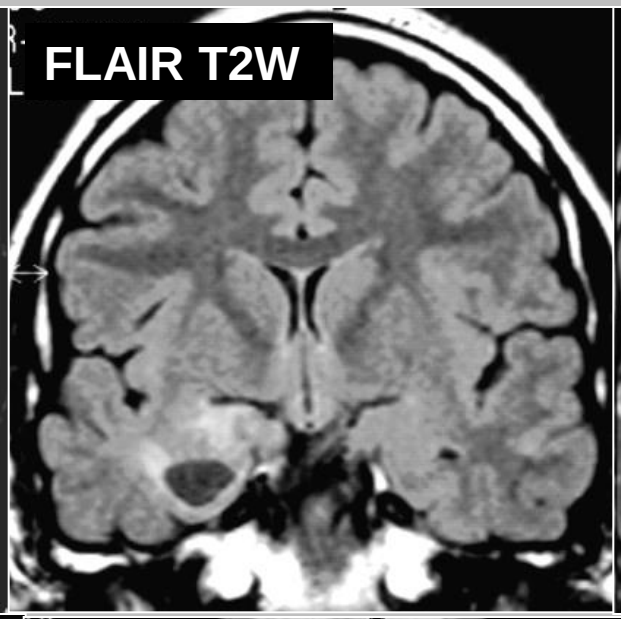
- Guariti (classe I di Engel): 111 (85%)
- Non guariti: 19 (15%)

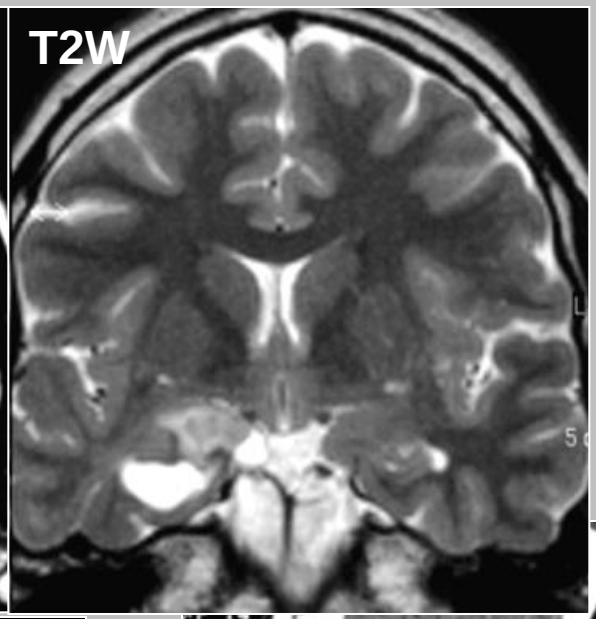
Aspetti neuroradiologici dei GG

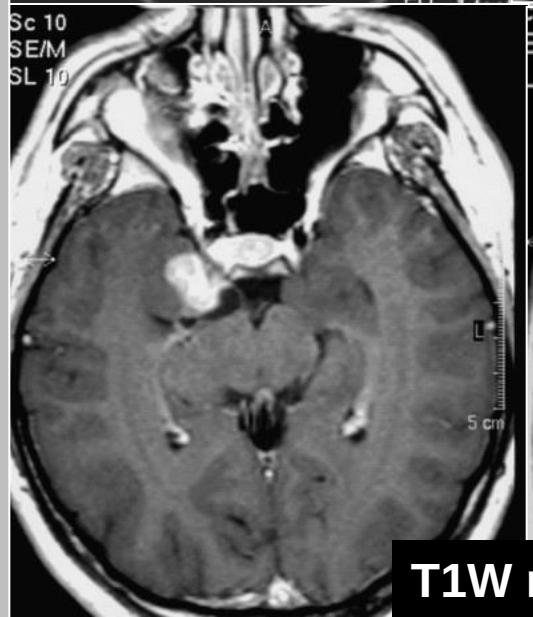
- Dimensioni <3 cm:77 (60%), 3-6 cm: 47 (36%), >6cm:6 (4%)
- Margini netti
- Raro effetto massa 22 casi (17%)
- Componenti cistiche
- Componenti solide
- Calcificazioni frequenti 38 casi (39%)
- Raro edema 3 casi (2%)
- Possibile enhancement 52 casi (40%)
- Rimodellamento teca cranica 12 casi (9%)

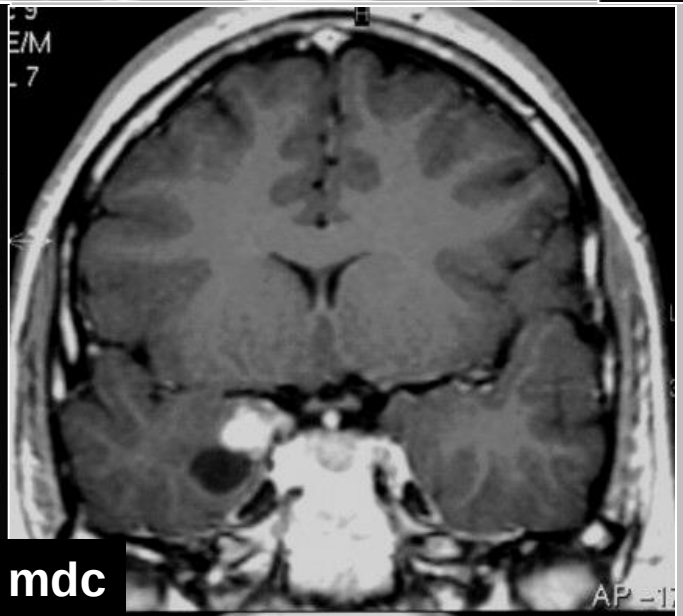
Ganglioglioma

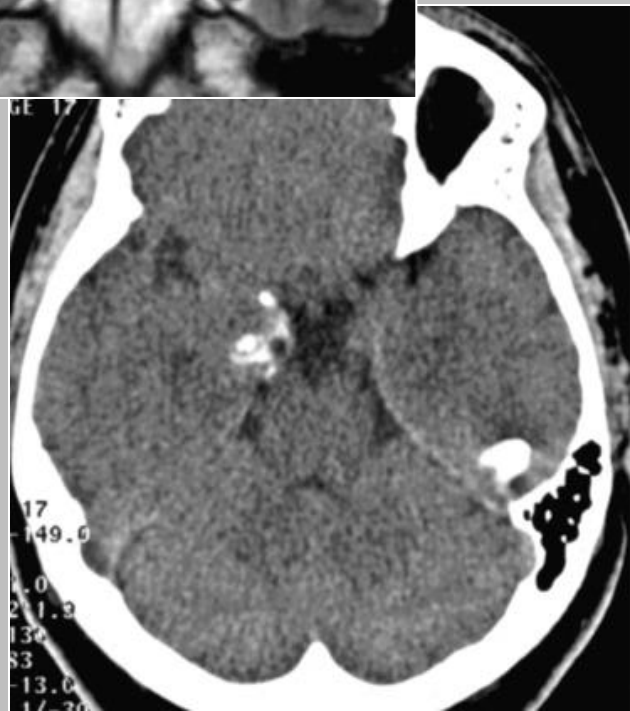




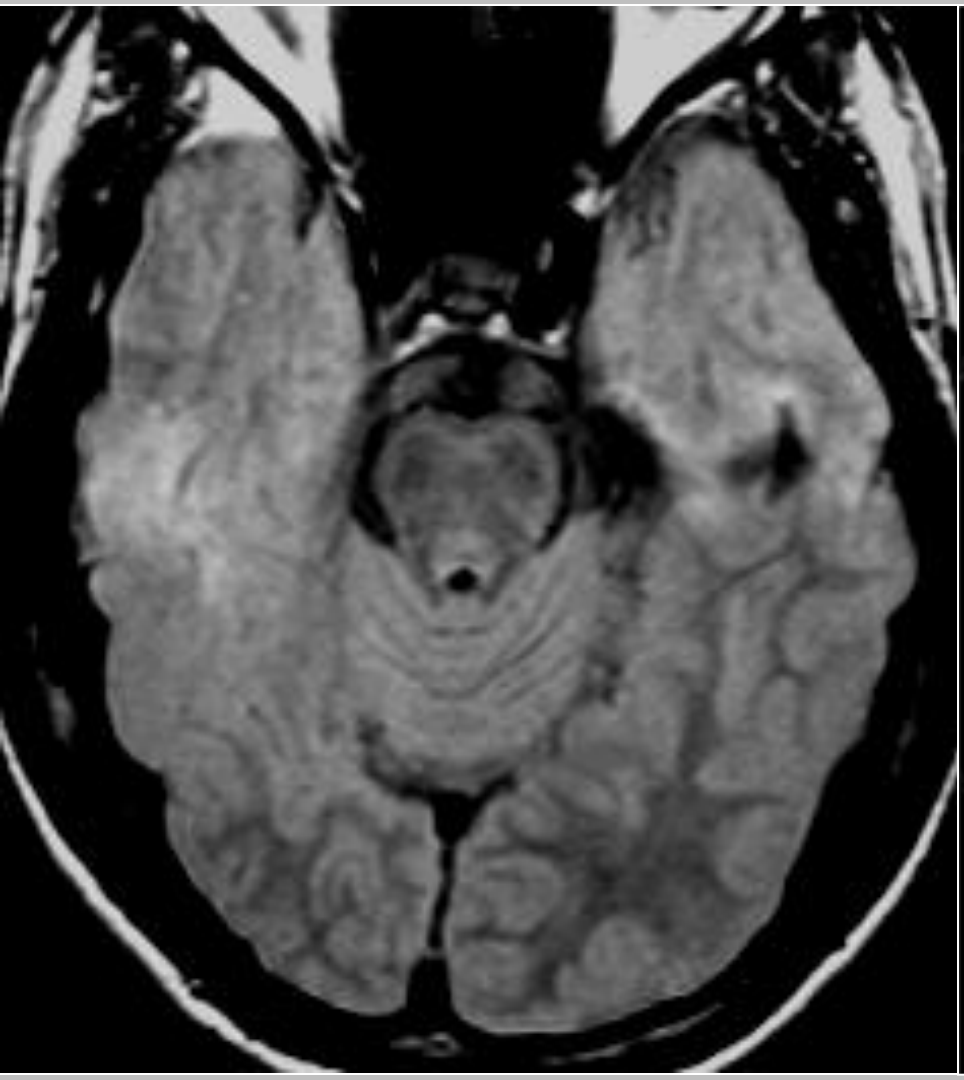


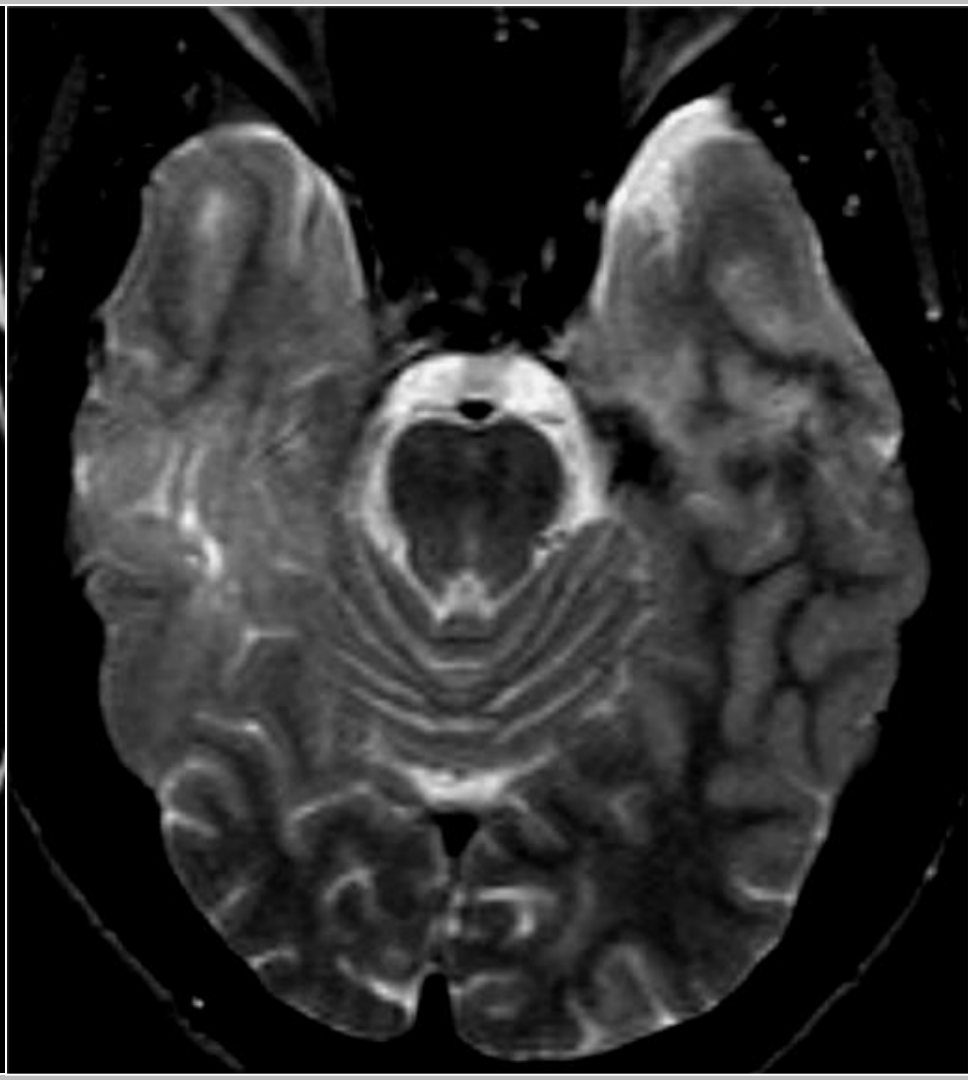






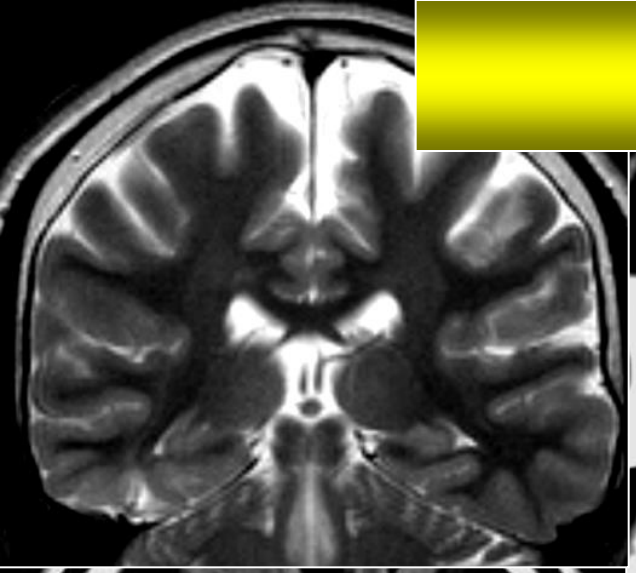
Ganglioglioma

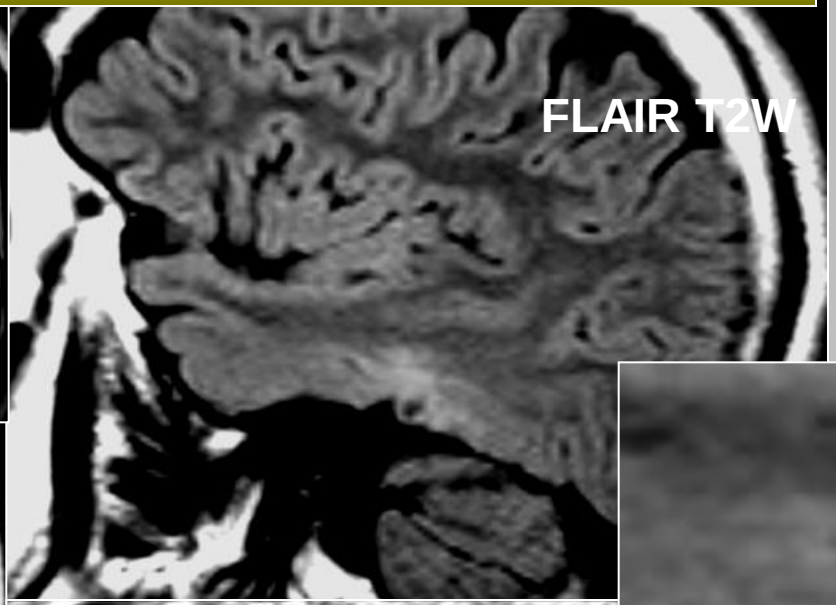


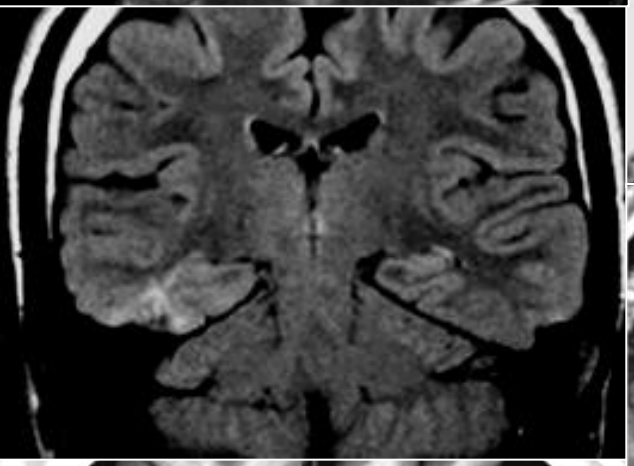


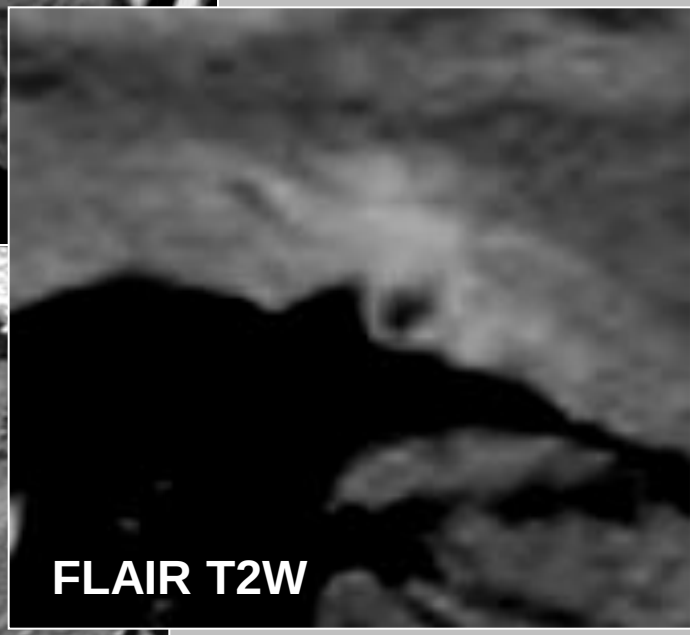
Ganglioglioma

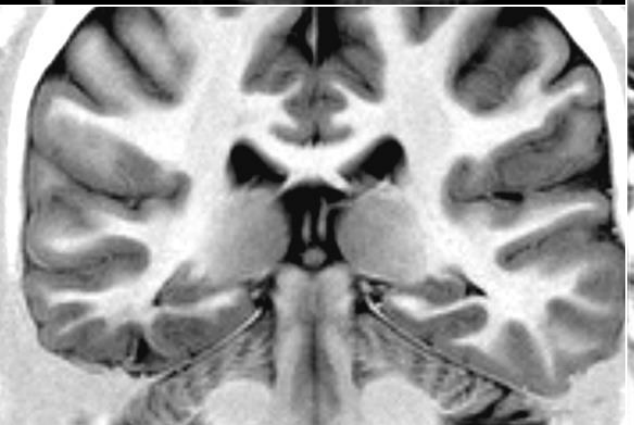
vs DNET

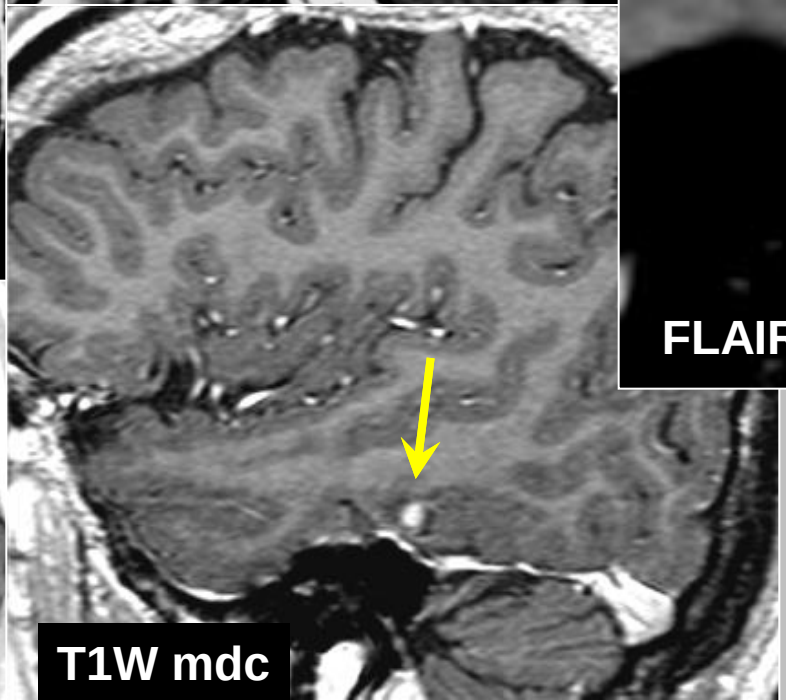






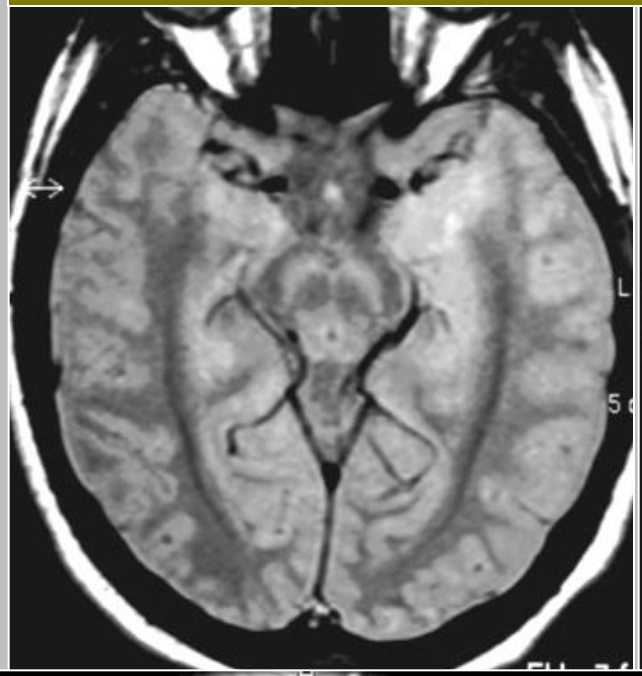


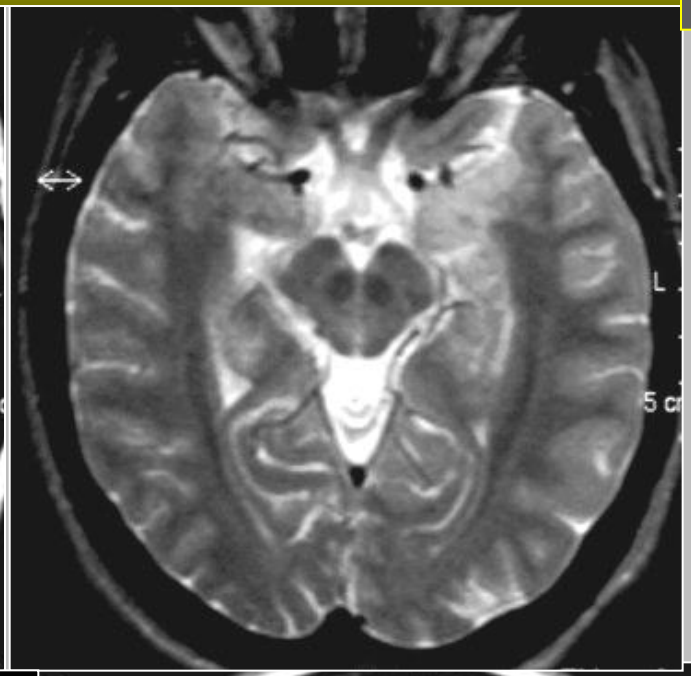


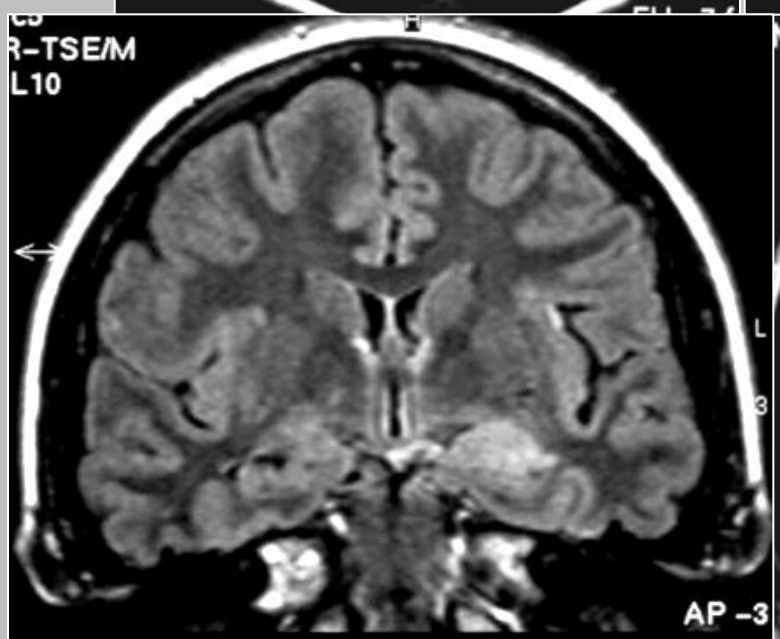


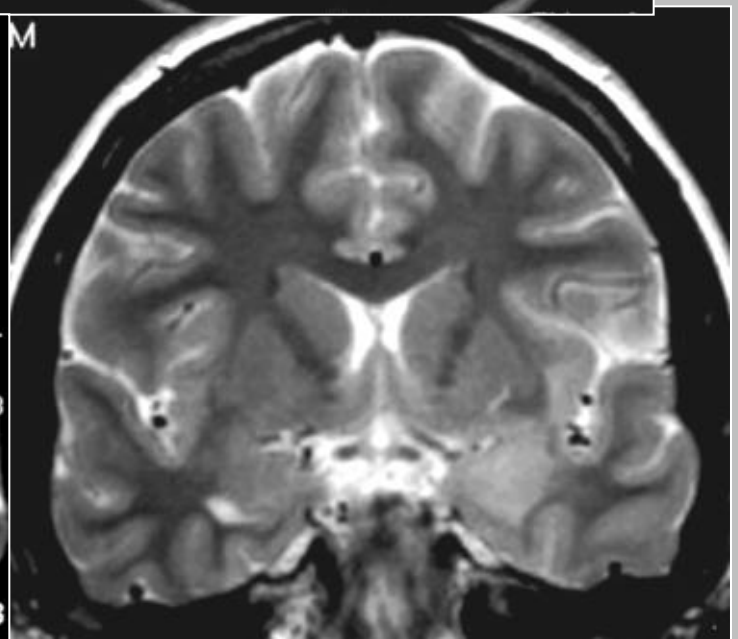
Ganglioglioma

vs tumore gliale









Altri Tumori (n°117)

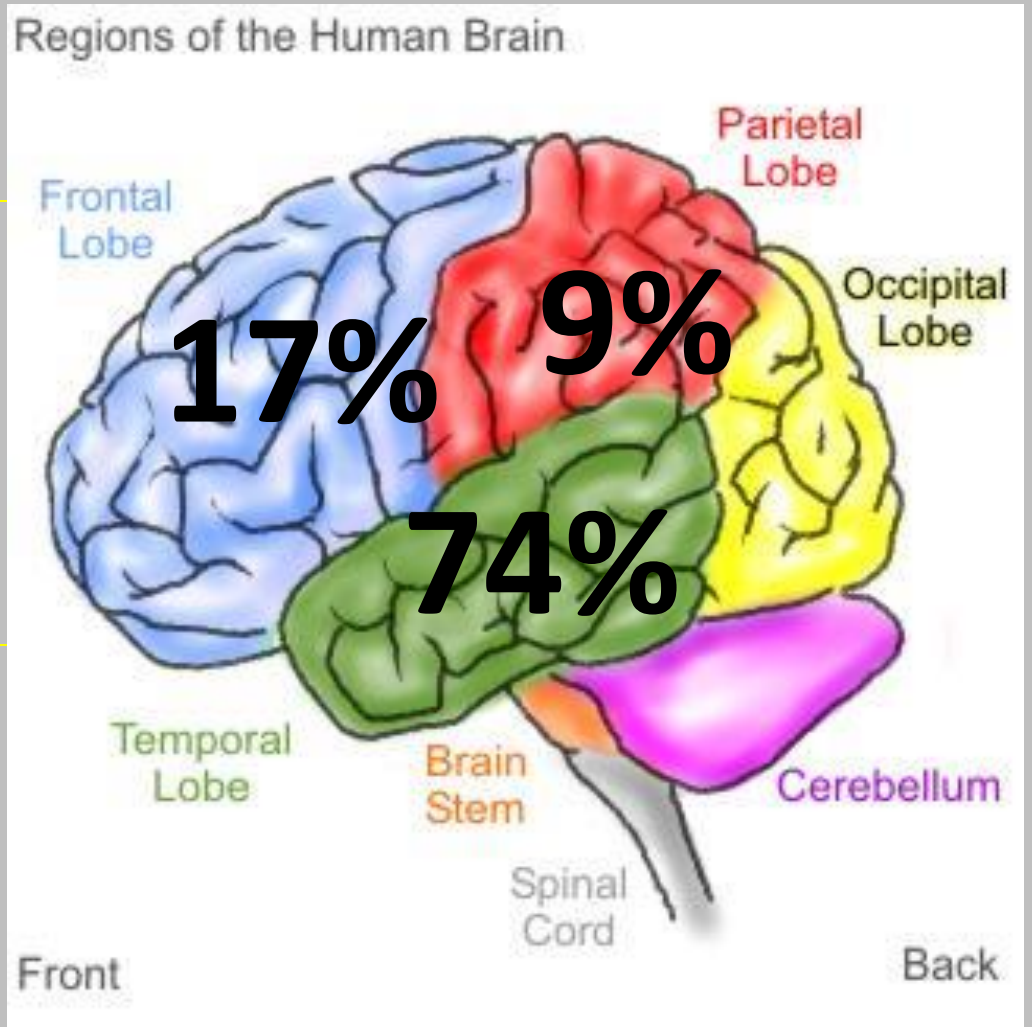
• Amartoma		46
• Oligodendroglioma		25
• Astrocitoma Pilocitico		16
• Xantoastrocitoma Pleomorfo	(PXA)	14
• Astrocitoma		10
• Dermoide		3
• Meningioangiomatosi		3

Altri tumori (117 casi)

Temporali: 87 (74%)

Anteriori: 20 (17%)

Posteriori: 10 (9%)



Altri tumori(n°117)

- Sesso: 52 femmine 65 maschi
- Lato: 56 destro 61 sinistro

- Età d'esordio delle crisi

  0-10 aa: 50 (52%)

  10-18 aa: 31 (25%)

  > 18 aa: 27 (23%)

Outcome crisi dopo intervento chirurgico

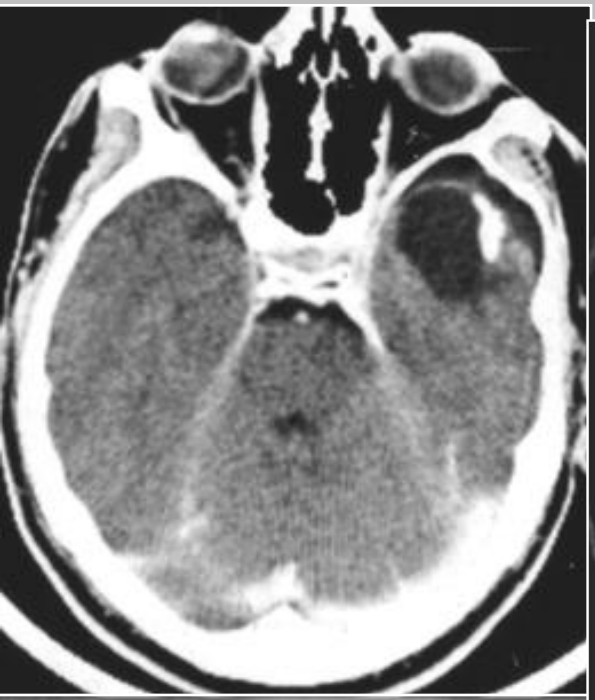
- Guariti (classe I di Engel): 90 (76%)
- Non guariti: 27 (24%)

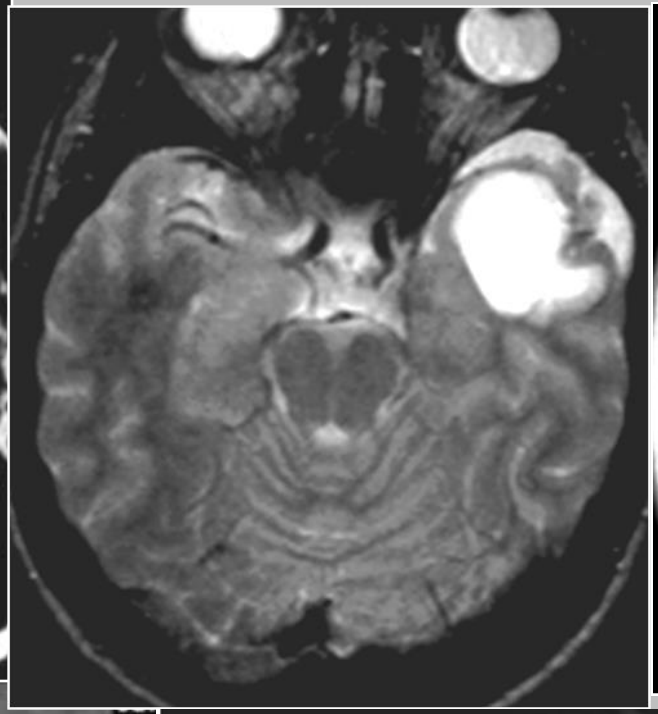
Xantoastrocitoma Pleomorfo (PXA)

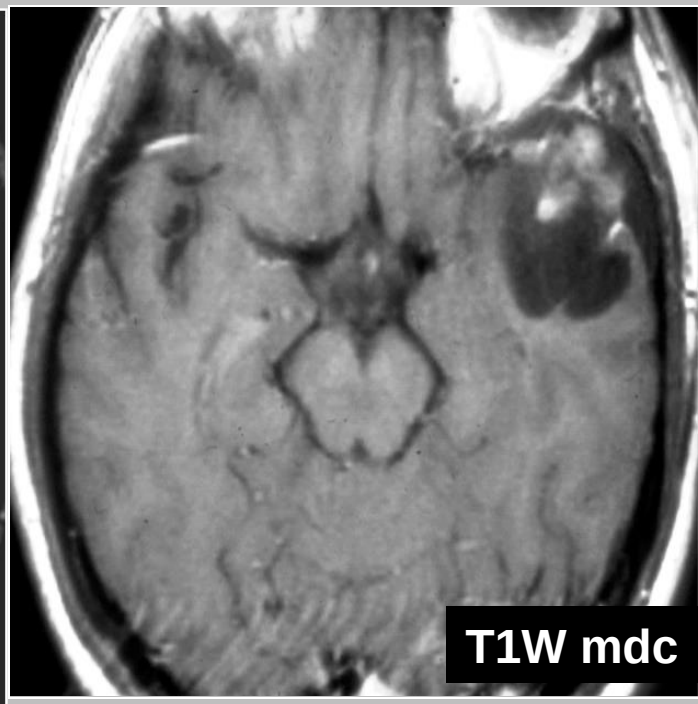
- Neoplasia astrocitaria del bambino e giovane adulto con prognosi relativamente favorevole (WHO grado II°)
- Localizzazione superficiale negli emisferi cerebrali e coinvolgimento delle meningi
- Istologia: cellule pleomorfe e lipidizzate che esprimono GFAP e spesso circondate da una matrice reticolinica

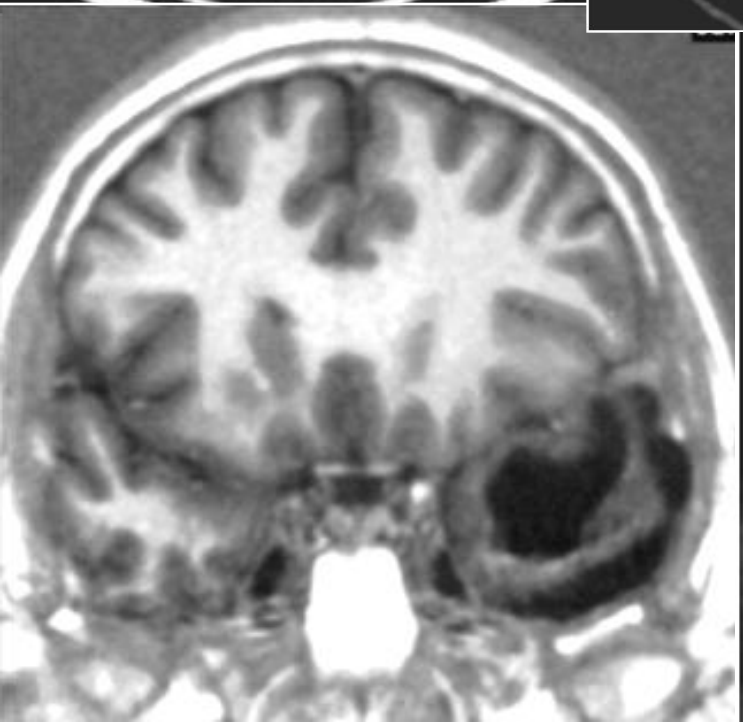
PXA
Aspetti neuroradiologici

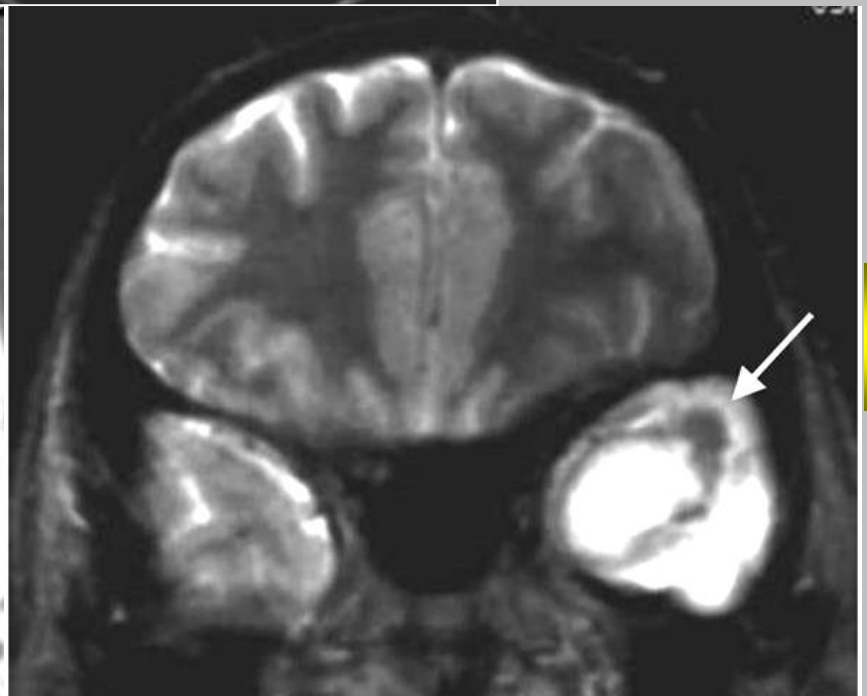
- Localizzazione superficiale con coinvolgimento meningeo
- Intrappolamento e dilatazione spazi subaracnoidei
- Cistico con nodulo murale
- Enhancement del nodulo ed enhancement periferico della parete cistica
- Calcificazioni
- Possibile edema
- Rimodellamento della teca cranica
- Sede: generalmente nel lobo temporale









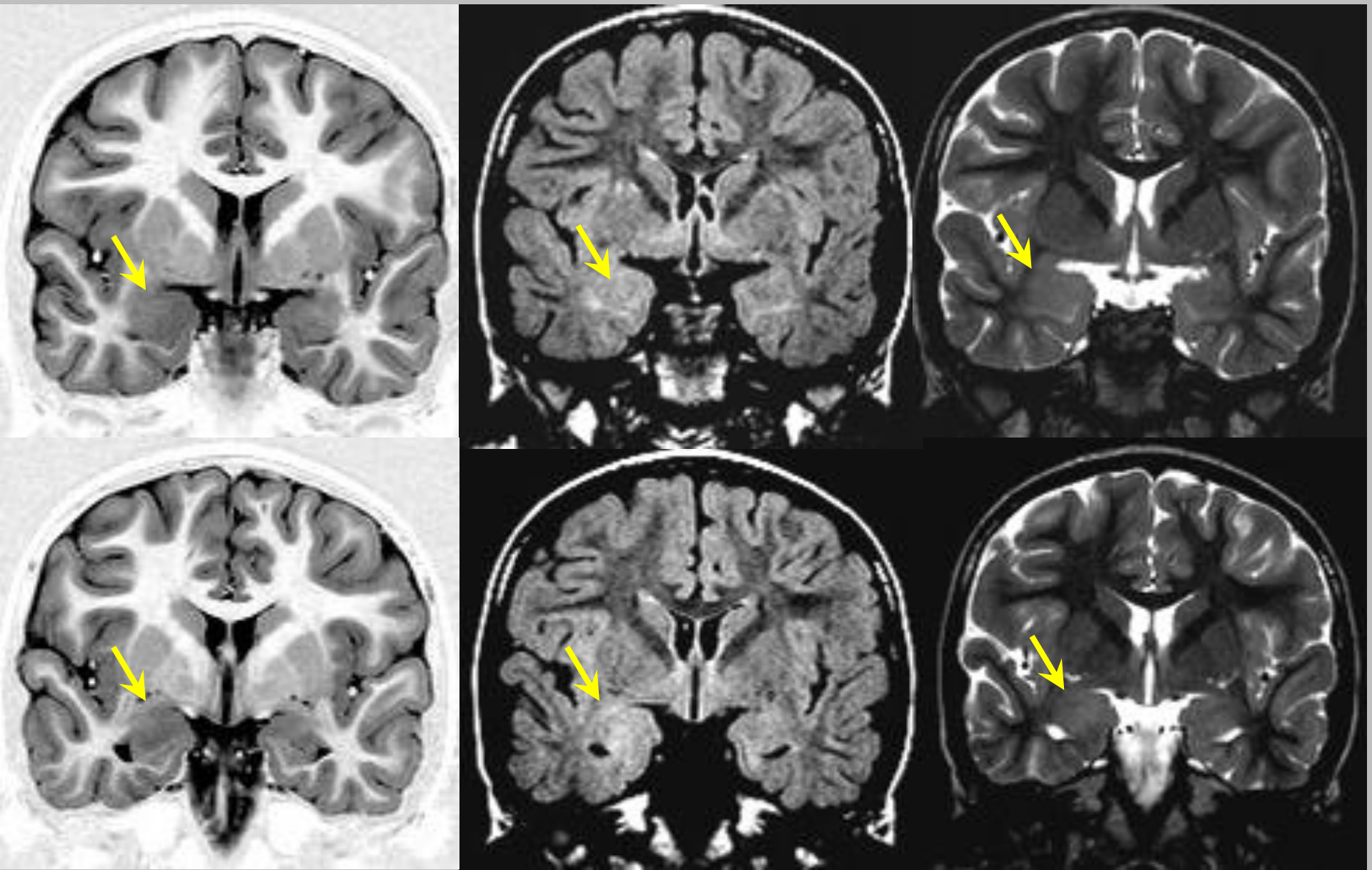


PXA

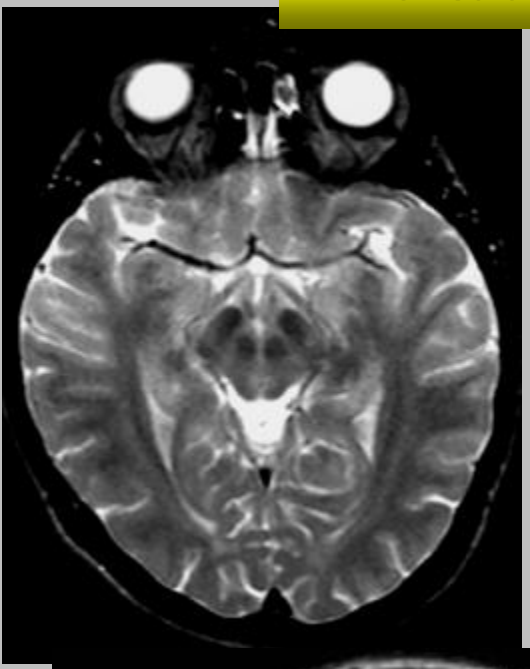
Amartoma

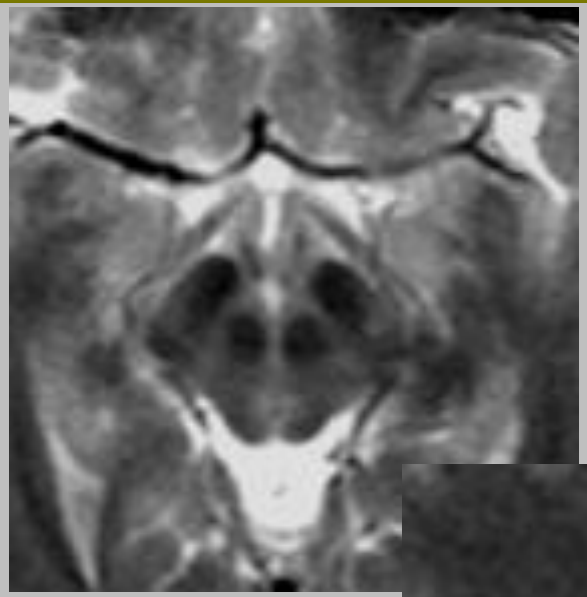
- lesione benigna dovuta ad inusuale proliferazione di cellule gliali e neuronali.
- Composto da cellule disorganizzate ma mature che sono normalmente presenti nel cervello, senza atipie citologiche.

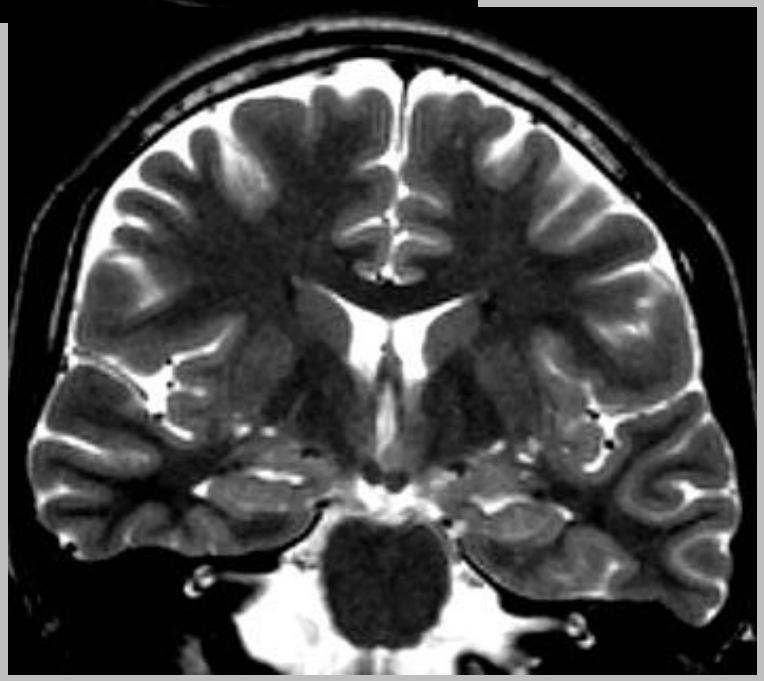
Amartoma

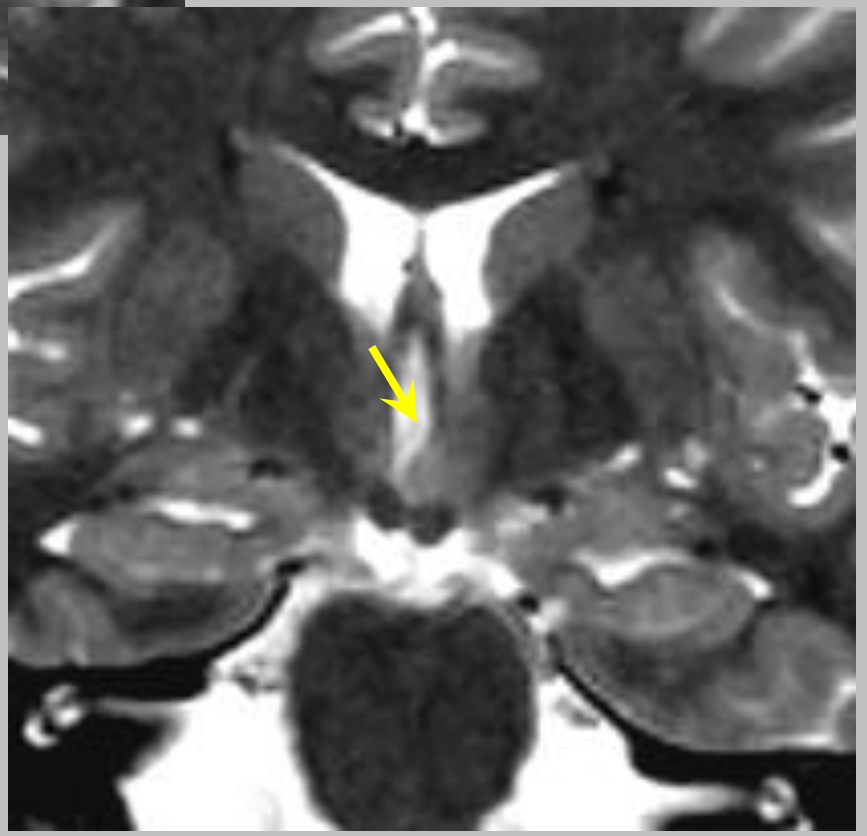


Amartoma ipotalamico



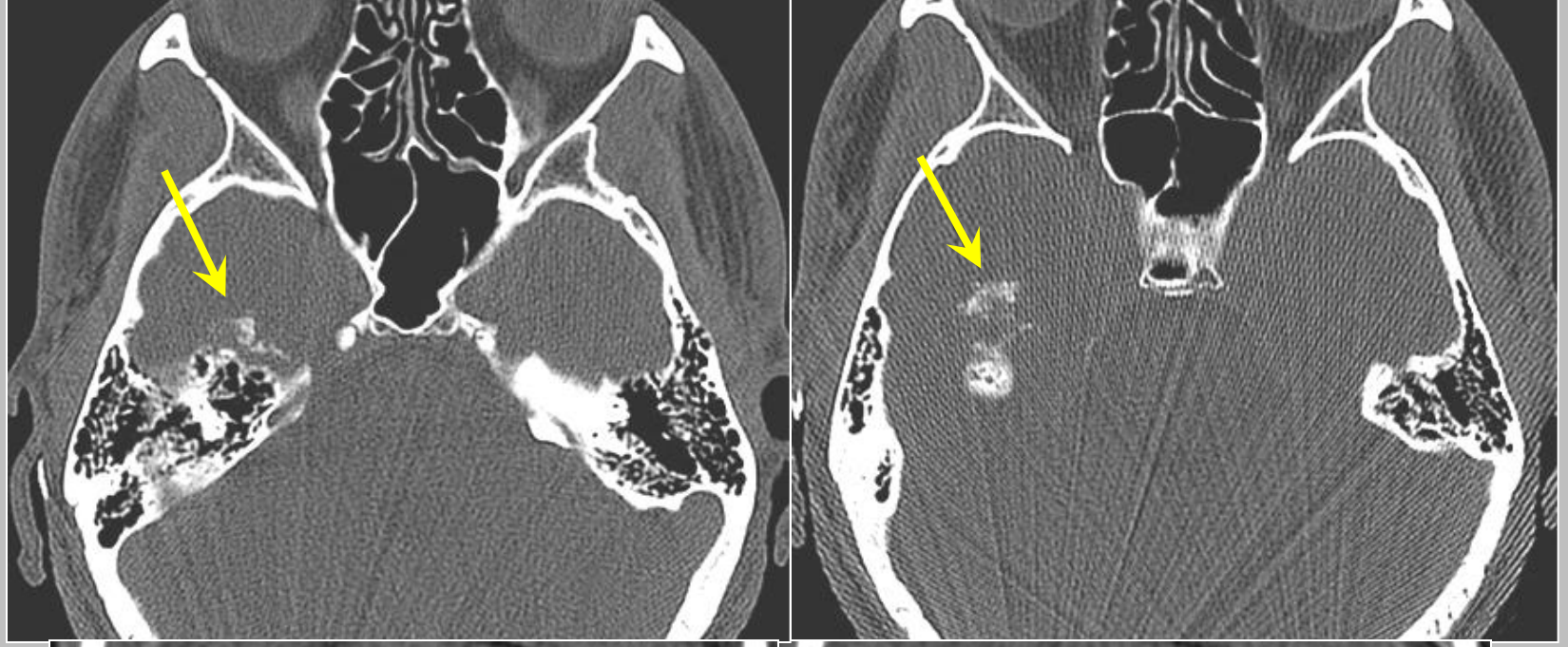
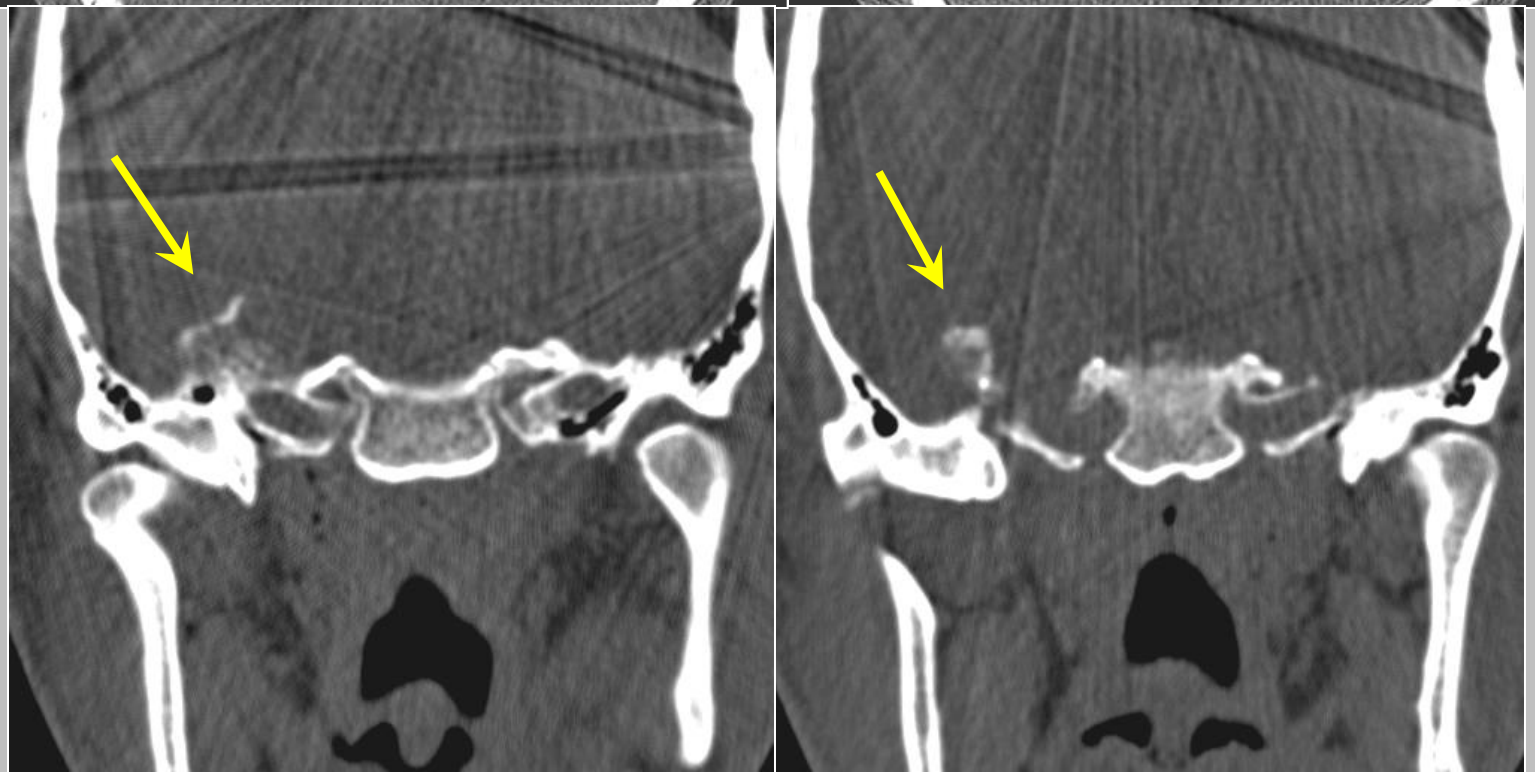




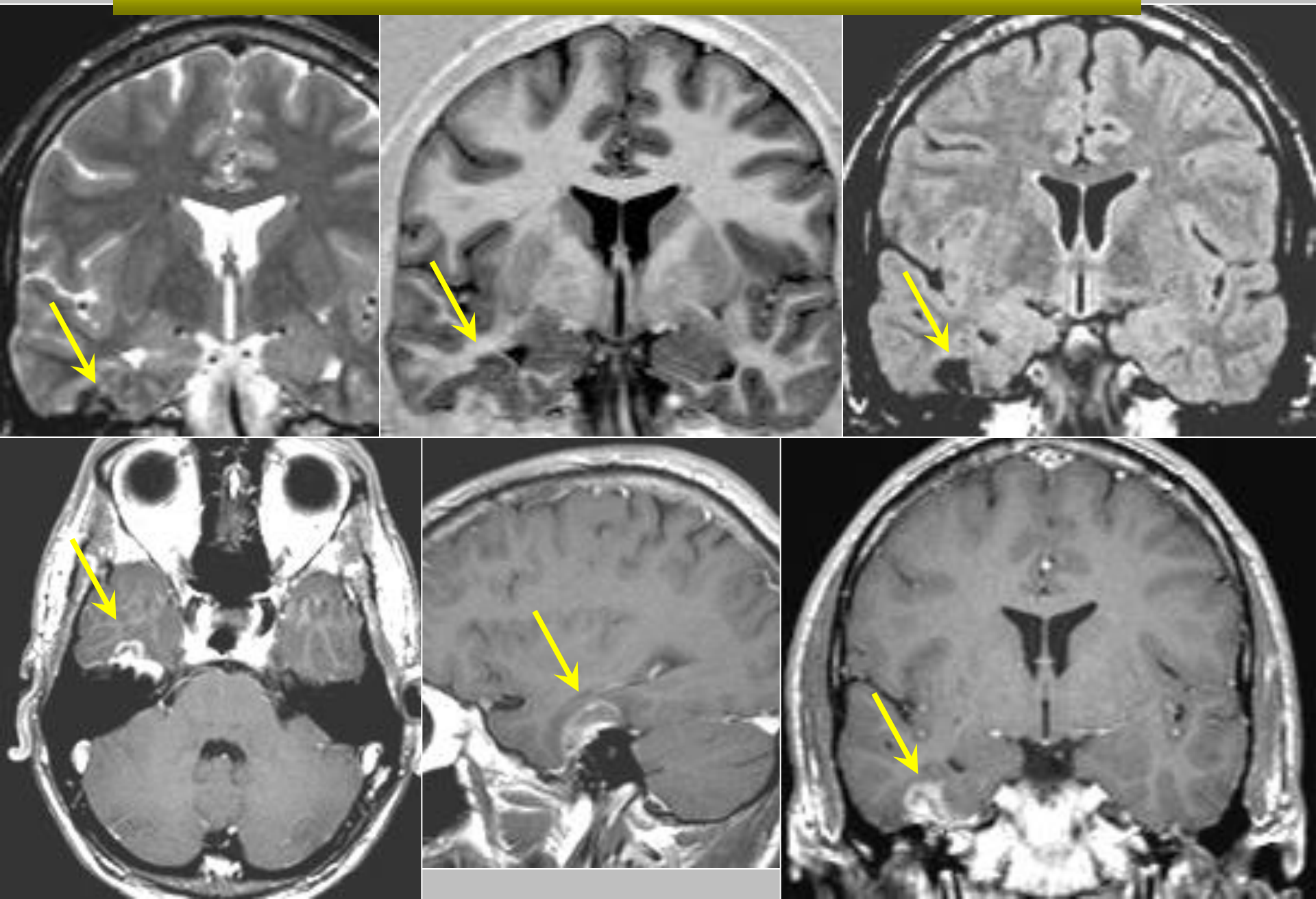


Meningioangiomatosi

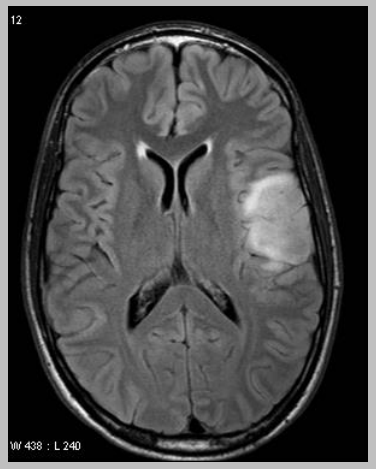
- condizione amartomatosa
- Classicamente associata a NF2
- Sede: leptomeningi, corteccia e sostanza bianca sottocorticale
- Istologia: proliferazione di vasi sanguigni circondati da orletti di cellule meningoteliali ⇨ proliferazione corticale meningovascolare e calcificazioni leptomeningee

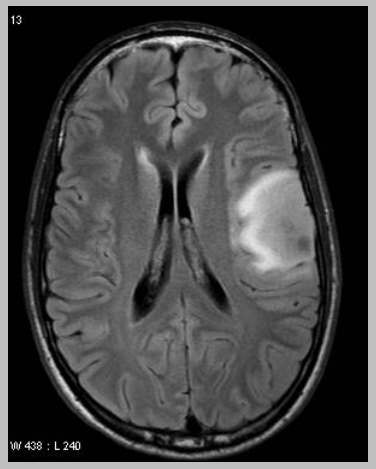



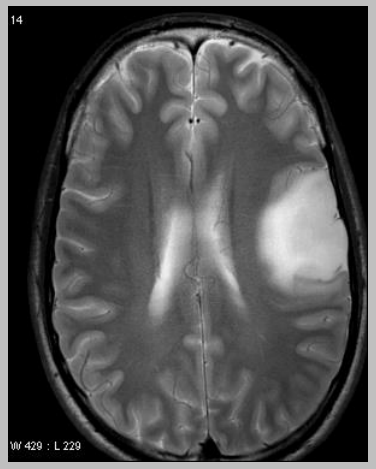
Meningioangiomatosi

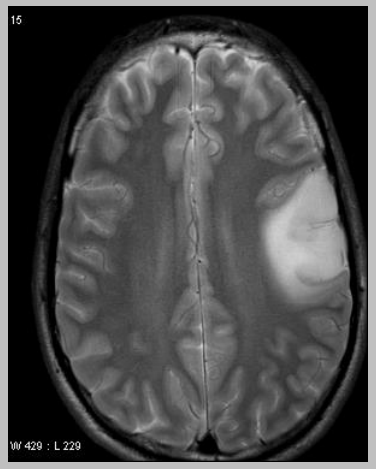


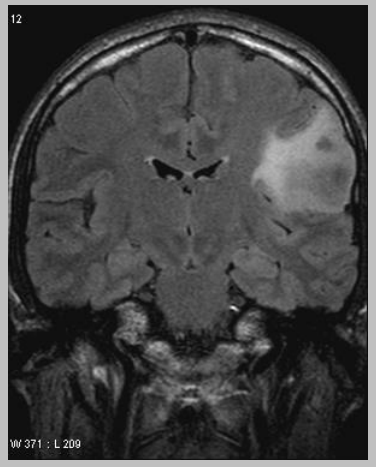
GLIOMA BASSO GRADO

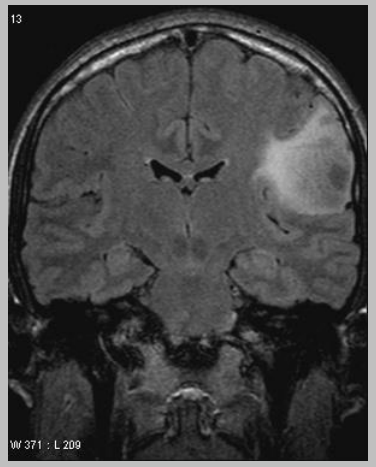


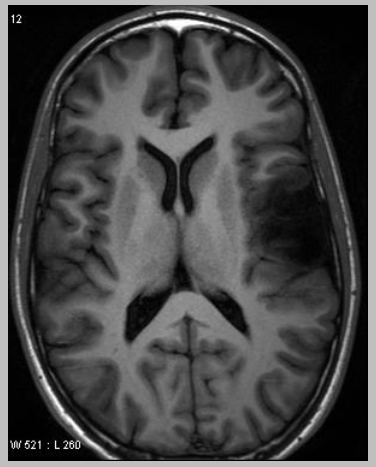


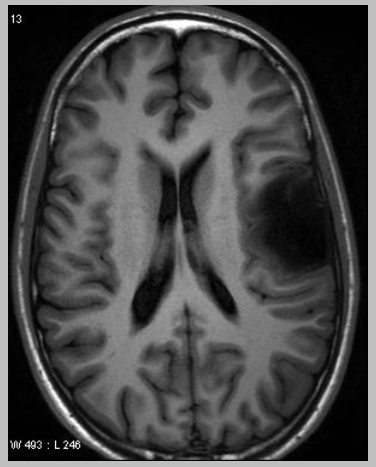


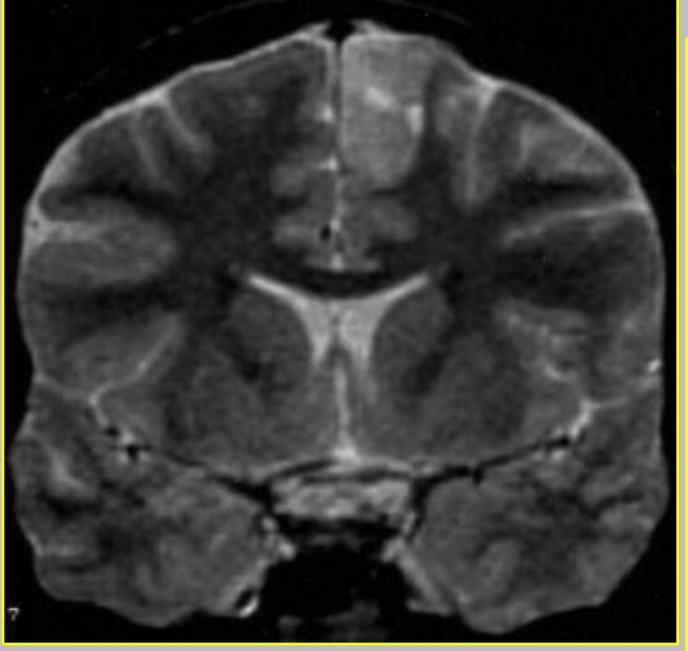


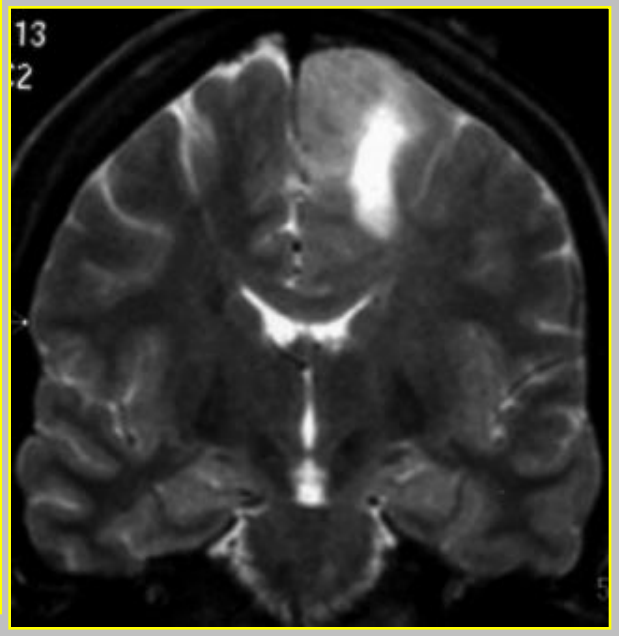


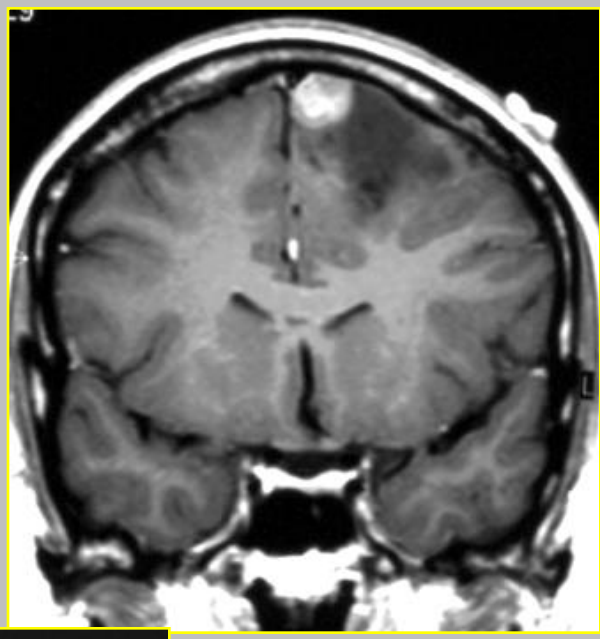


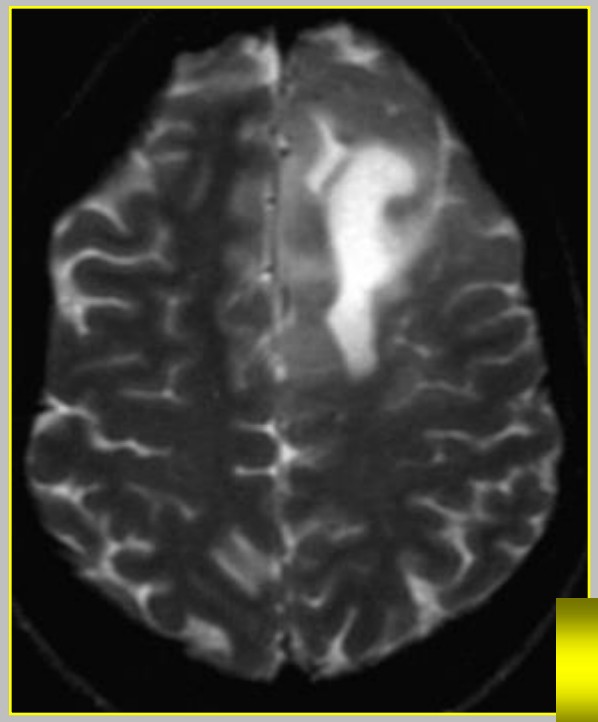


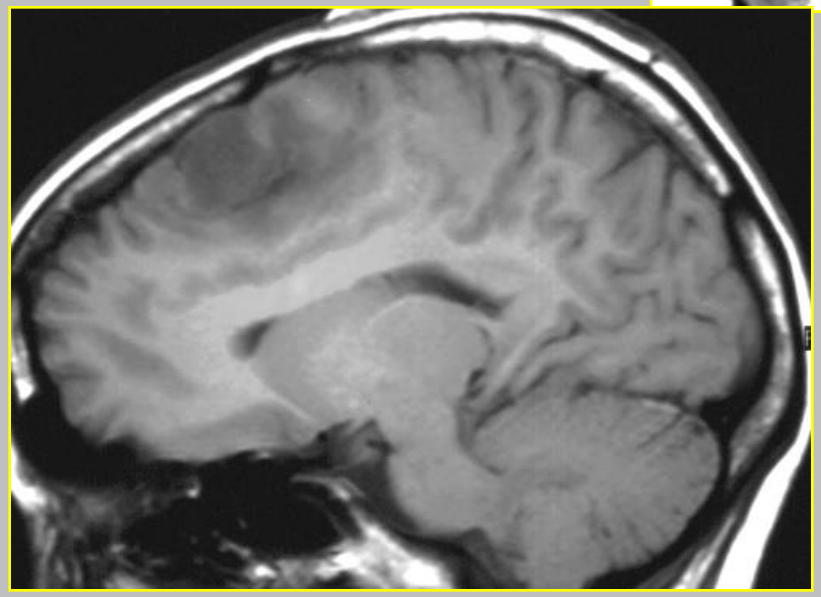












Oligodendroglioma grado III°

Conclusioni

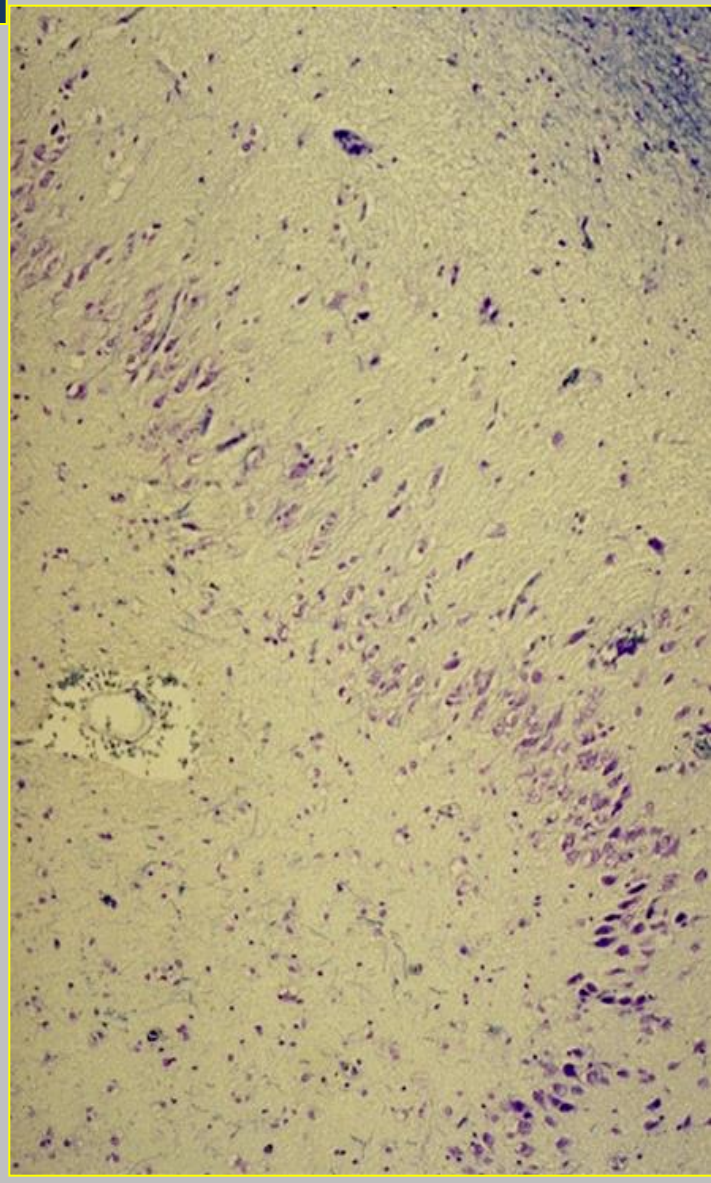
- I tumori costituiscono il 30% delle lesioni nei pazienti operati per epilessia focale farmaco-resistente
- La maggioranza dei tumori epilettogeni sono di basso grado ed a lenta crescita
- Queste lesioni possono essere riconosciute e caratterizzate tramite uno studio RM mirato
- Frequente sovrapposizione delle caratteristiche RM dei vari istotipi
- Il riconoscimento di caratteristiche aggressive alla RM è fondamentale per la tempistica dell'intervento

Sclerosi ippocampale (HS)

- Costituisce il più frequente substrato istologico dell'epilessia del lobo temporale (50-70% di tutte le lobectomie temporali)

Istologia

- Perdita neuronale
- Proliferazione astrocitaria (gliosi)



Sclerosi Ippocampale – Aspetti RM - Analisi visiva

Criteri primari (rilevabili a carico dell'ippocampo)

atrofia

aumentato segnale in T2 e FLAIR

ridotto segnale in T1 IR

perdita architettura interna

Criteri secondari ipsilaterali (extra-ippocampali)

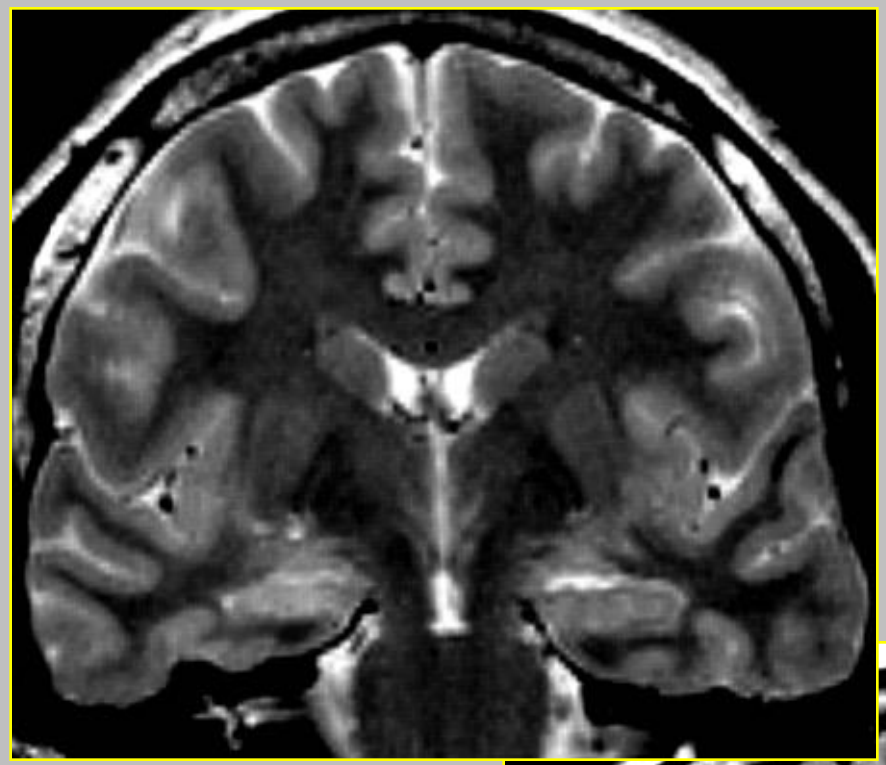
assottigliamento del fornice

atrofia corpi mamillari

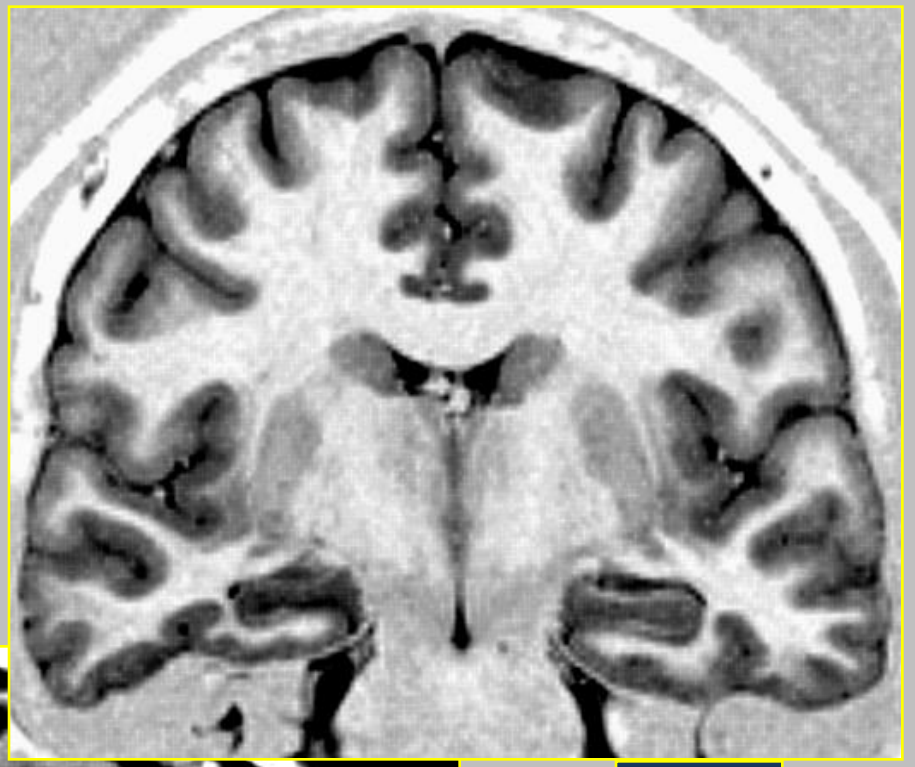
ridotto volume del lobo temporale & sostanza bianca locale

dilatazione corno temporale

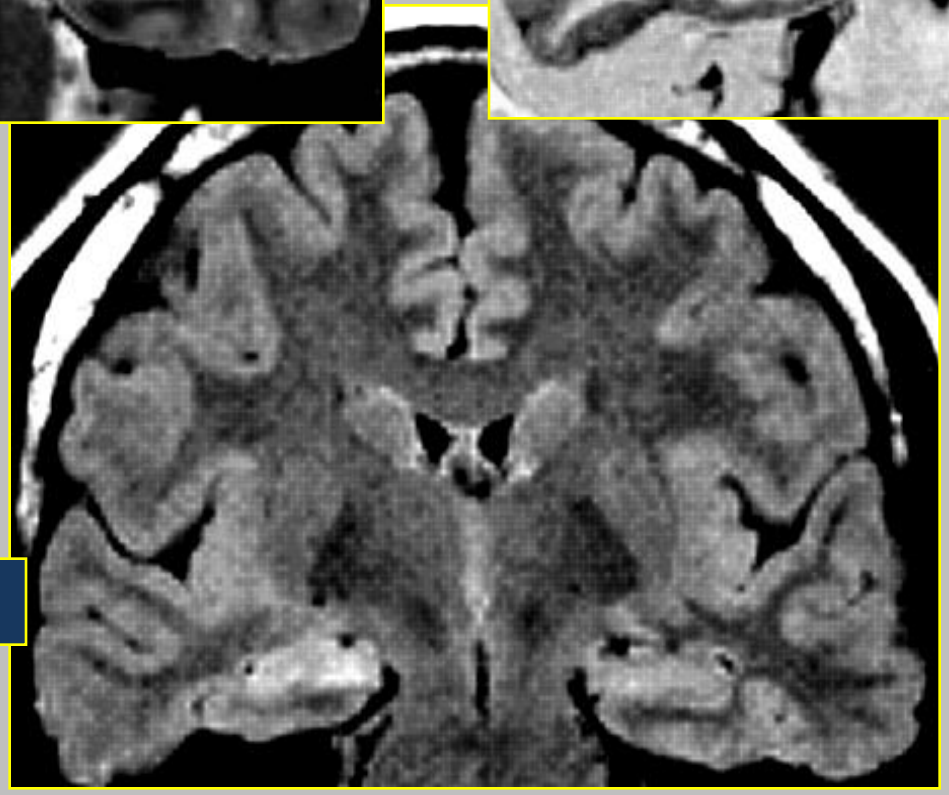
aumentato segnale dell'amigdala in T2



T2 TSE



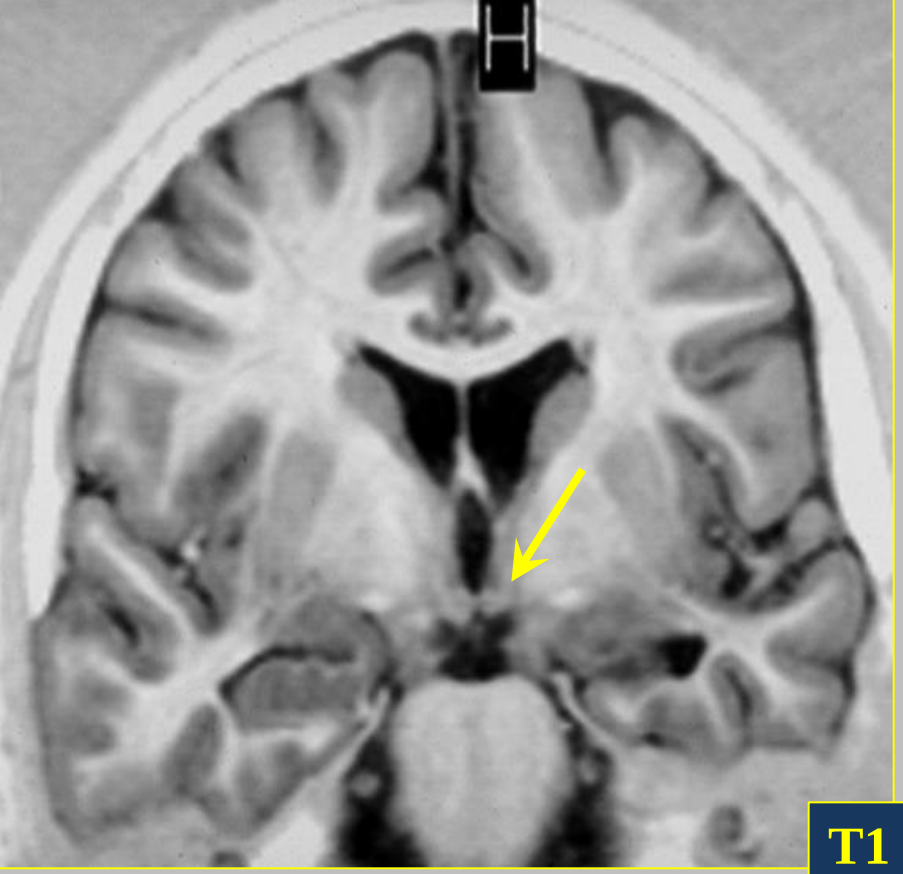
T1 IR

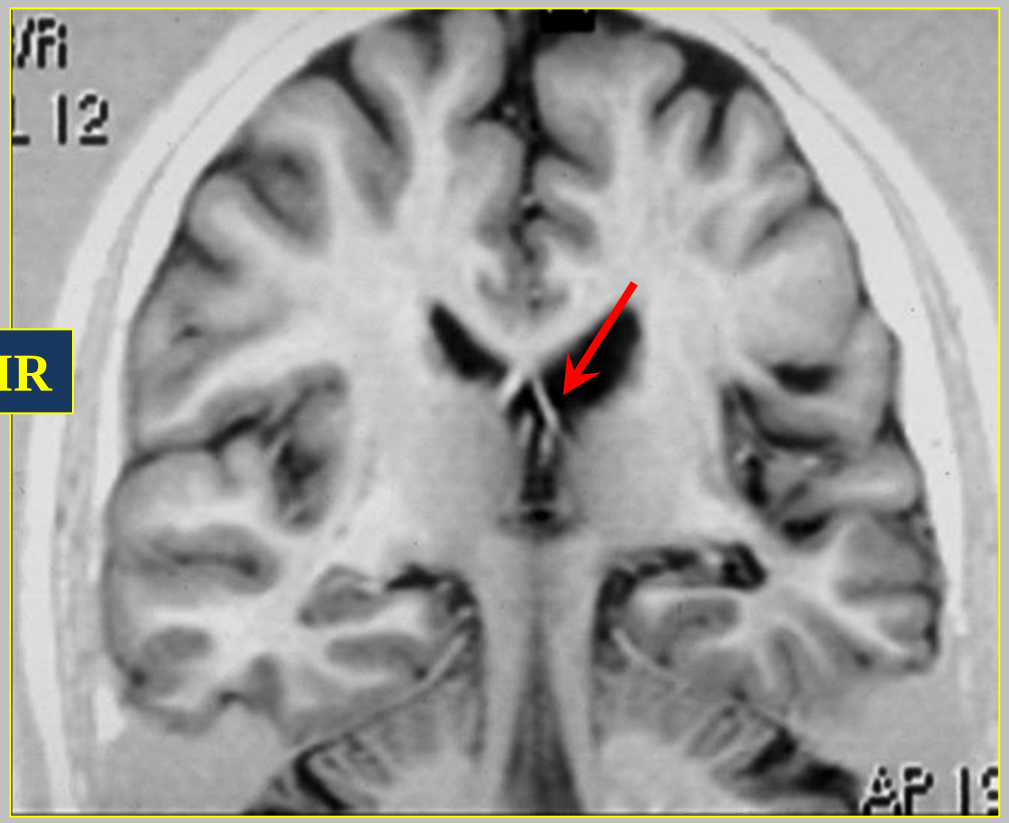


T2 FLAIR

Sclerosi Ippocampale

Sclerosi ippocampale



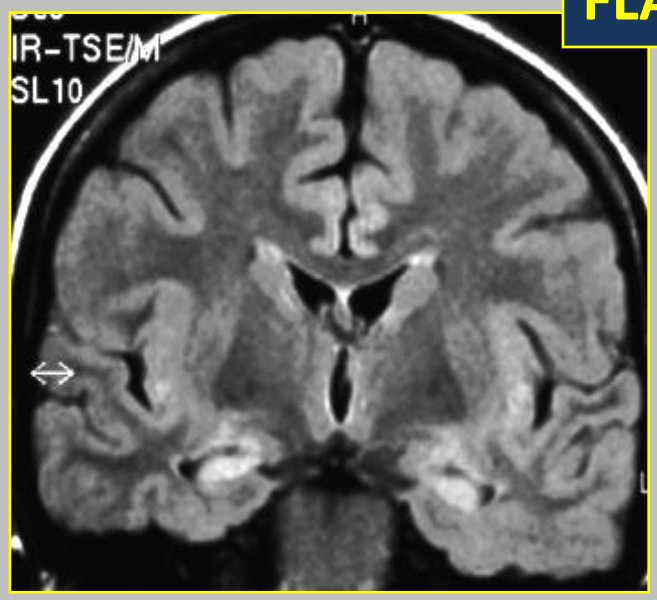


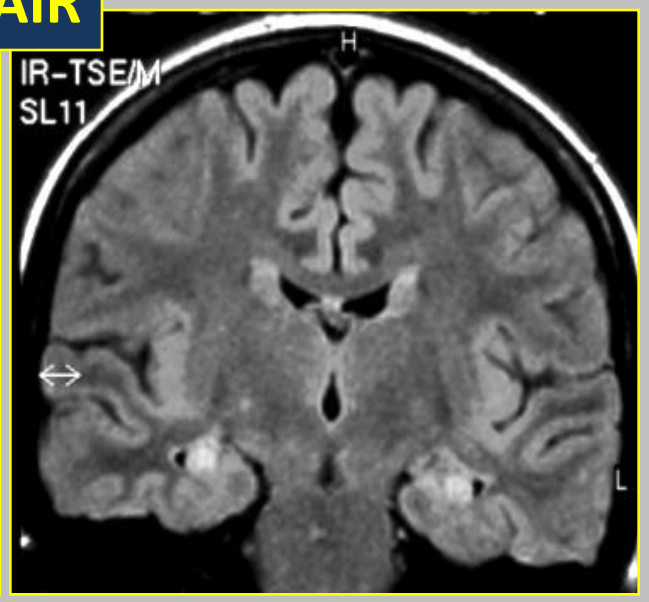
T1 IR

Criteri secondari extra-ippocampali

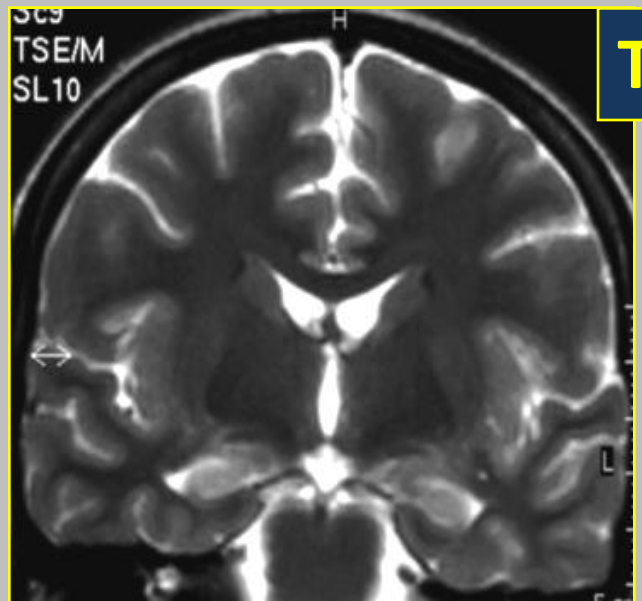
Sclerosi ippocampale bilaterale

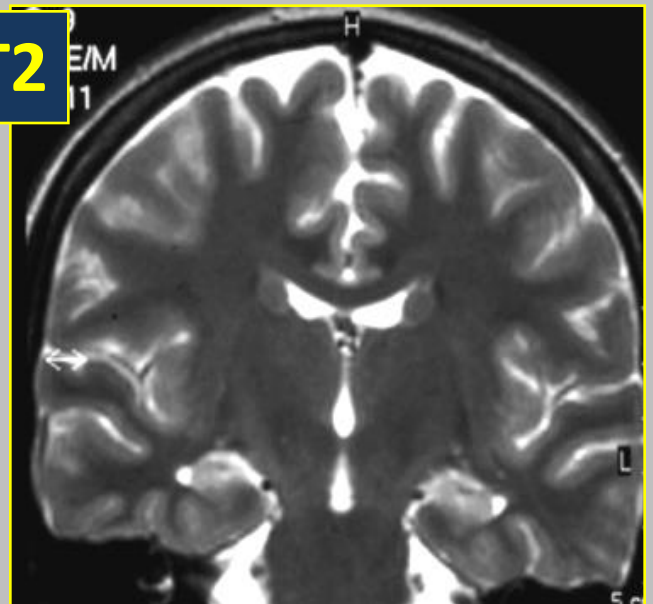
FLAIR

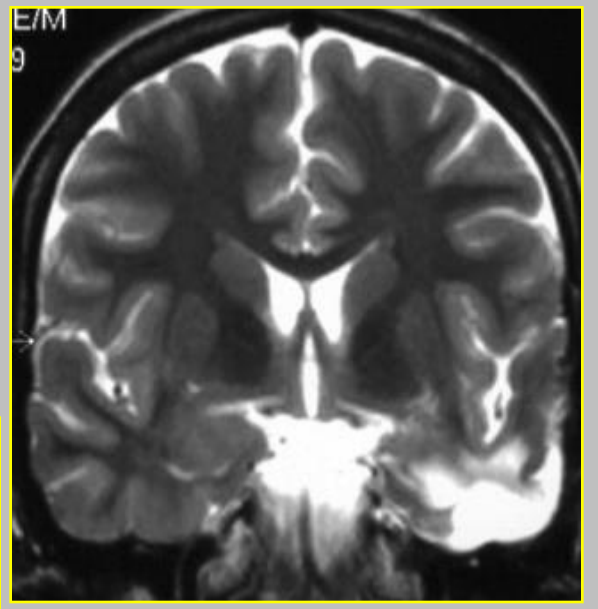




T2







Controllo Post-operatorio

Conclusioni

- La RM è una metodica dalle grandi potenzialità diagnostiche ma deve essere usata con criterio

- I migliori risultati si ottengono solo grazie ad una stretta collaborazione tra Neuroradiologi, Neurologi, Neurofisiologi e Neurochirurghi

Conclusioni

- Il trattamento chirurgico mira a controllare le crisi oltre che a rimuovere il tumore
- L'intervento chirurgico nei pazienti con EPFR deve essere pianificato per ogni soggetto basandosi sulla storia clinica, sulle caratterisitche EEG e sugli aspetti RM
- I migliori risultati si ottengono solo tramite una stretta collaborazione tra Neurologi, Neurochirurghi, Neuroradiologi e Neuropatologi

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PER
L’ATTENZIONE