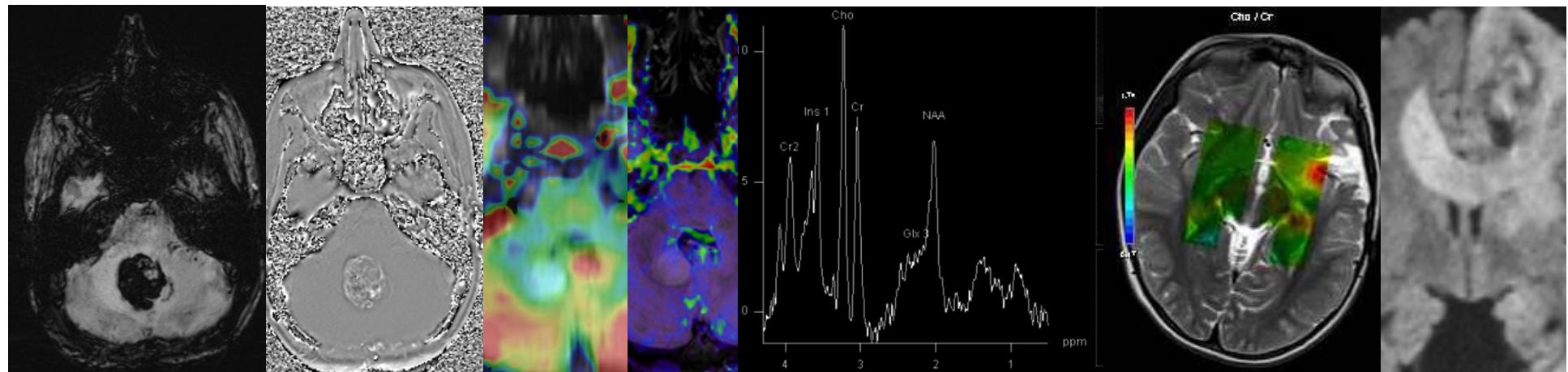




# Tumores Cerebrales Primarios Pediátricos



Sección de Neuroradiología Pediátrica  
Hospital Vall d'Hebron – Área Materno-Infantil  
Barcelona  
angel.sanchez.idi@gencat.cat





# Conflicto de intereses

- Ninguno

# Índice

- Objetivos
- Introducción
- Clasificación
- T. Fosa posterior

- T. Supraselar-Diencéfalo
- T. Supratentorial
- Blackjack
- Conclusiones





# Objetivos



Revisar los diferentes tipos de tumores cerebrales pediátricos



Caracterización tumoral según la semiología radiológica



Abordar el diagnóstico diferencial neurooncológico pediátrico



Aproximación oncogenética e introducción en la era molecular

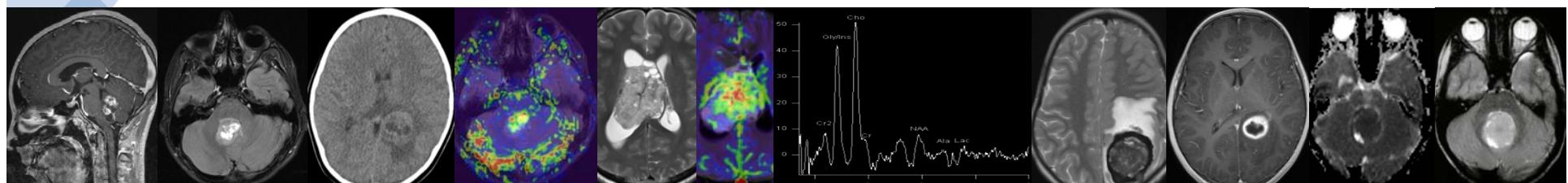


Dominar el blackjack: 21 tumores



# Introducción

- Tumores del SNC: 25% de todos los cánceres en la infancia
- Tumor sólido más frecuente y mortal. 54-70% **fosa posterior**
- Incidencia de 4/100.000 en menores de 15 años
- Pronóstico ha mejorado en los últimos años; **curación >80%**
- Mejor pronóstico astrocitoma pilocítico de cerebelo
- Alto porcentaje de **secuelas** y reducción de la calidad de vida
- Sintomatología relacionada con HTE, alteración marcha o ppcc
- Clínica insidiosa, aunque a veces aguda como crisis o hidrocefalia



# Etiología

Neurofibromatosis: Neurofibroma, Astrocytoma...

Esclerosis Tuberosa: Hamartoma, SEGA

Von Hippel-Lindau: Hemangioblastoma

Li-Fraumeni: Astrocytoma, MBL (mutación *TP53*)

Sd. Turcot: Astrocyt, MBL WNT (mutación *APC*)

Sd. Gorlin: Meduloblastoma SHH (*PTCH1/SUFU*)

Sd. Rubinstein-Taybi: Meduloblastoma (*CREBBP*)

Mayoría esporádicos sin causa conocida. Sd. genéticos → > incidencia

# Clasificación



Vall  
d'Hebron  
Barcelona Hospital Campus

IDI  
INSTITUT DE  
DIAGNÒSTIC PER  
LA IMATGE

SEHR  
SOCIETAT  
Catalana  
d'Radiologia  
XVI CURSO NACIONAL DE  
NEURORADIOLOGÍA



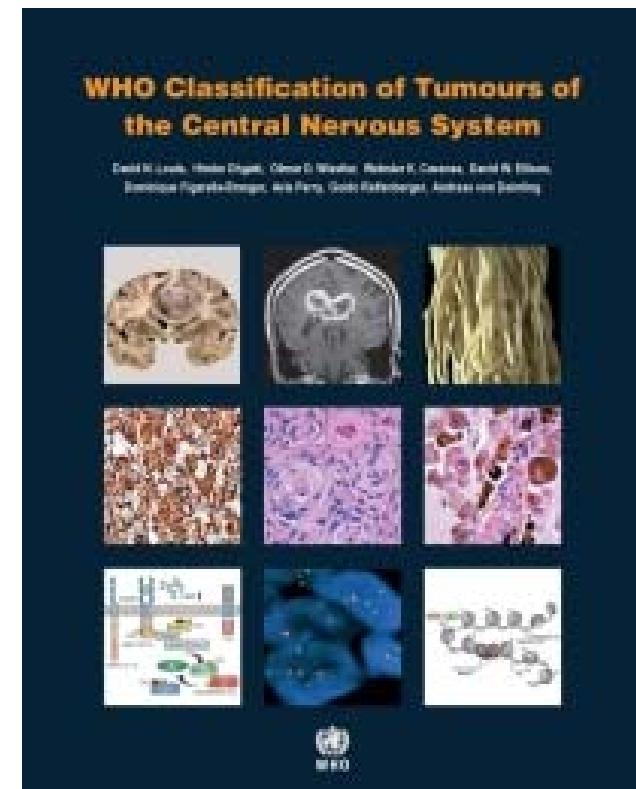
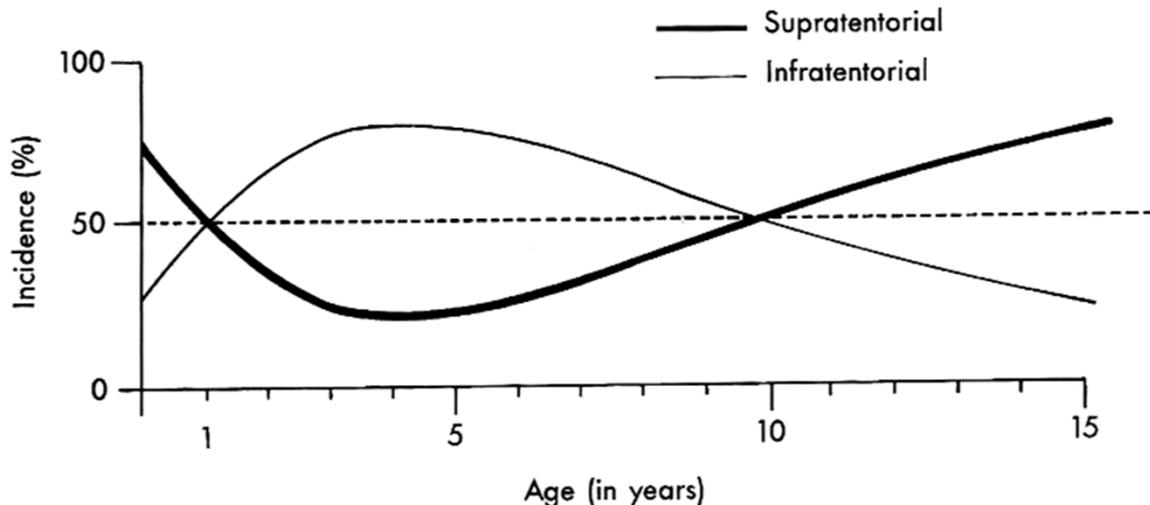
CrossMark

Acta Neuropathol (2016) 131:803–820  
DOI 10.1007/s00401-016-1545-1

REVIEW

## The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary

David N. Louis<sup>1</sup> · Arie Perry<sup>2</sup> · Guido Reifenberger<sup>3,4</sup> · Andreas von Deimling<sup>4,5</sup> ·  
Dominique Figarella-Branger<sup>6</sup> · Webster K. Cavenee<sup>7</sup> · Hiroko Ohgaki<sup>8</sup> ·  
Otmar D. Wiestler<sup>9</sup> · Paul Kleihues<sup>10</sup> · David W. Ellison<sup>11</sup>





# Clasificación

Diffuse astrocytic and oligodendroglial tumours		Neuronal and mixed neuronal-glial tumours					
Diffuse astrocytoma, IDH-mutant	9400/3	Dysembryoplastic neuroepithelial tumour	9413/0	Melanotic schwannoma	9560/1	Osteochondroma	9210/0
Gemistocytic astrocytoma, IDH-mutant	9411/3	Ganglioglioma	9492/0	Neurofibroma	9540/0	Osteosarcoma	9180/3
Diffuse astrocytoma, IDH-wildtype	9400/3	Ganglioglioma	9505/1	Atypical neurofibroma	9540/0		
Diffuse astrocytoma, NOS	9400/3	Anaplastic ganglioglioma	9505/3	Plexiform neurofibroma	9550/0		
Anaplastic astrocytoma, IDH-mutant	9401/3	Dysplastic cerebellar gangliocytoma (Lhermitte-Duclos disease)	9493/0	Perineurioma	9571/0	<b>Melanocytic tumours</b>	
Anaplastic astrocytoma, IDH-wildtype	9401/3	Desmoplastic infantile astrocytoma and ganglioglioma	9412/1	Hybrid nerve sheath tumours	9540/3	Meningeal melanocytosis	8728/0
Anaplastic astrocytoma, NOS	9401/3	Papillary glioneuronal tumour	9509/1	Malignant peripheral nerve sheath tumour	9540/3	Meningeal melanocytoma	8728/1
Glioblastoma, IDH-wildtype	9440/3	Rosette-forming glioneuronal tumour	9509/1	Epithelioid MPNST	9540/3	Meningeal melanoma	8720/3
Giant cell glioblastoma	9441/3	Diffuse leptomeningeal glioneuronal tumour		MPNST with perineural differentiation	9540/3	Meningeal melanomatosis	8728/3
Gliosarcoma	9442/3	Central neurocytoma	9506/1	<b>Meningiomas</b>			
Epithelioid glioblastoma	9440/3	Extraventricular neurocytoma	9506/1	Menignoma	9530/0	Lymphomas	
Glioblastoma, IDH-mutant	9445/3*	Cerebellar liponeurocytoma	9506/1	Meningothelial meningioma	9531/0	Diffuse large B-cell lymphome of the CNS	9680/3
Glioblastoma, NOS	9440/3	Paraganglioma	8693/1	Fibrous meningioma	9532/0	Immunodeficiency-associated CNS lymphomas	
Diffuse midline glioma, H3 K27M-mutant	9385/3*	<b>Tumours of the pineal region</b>		Transitional meningioma	9537/0	AIDS-related diffuse large B-cell lymphoma	
Oligodendroglioma, IDH-mutant and 1p/19q-codeleted	9450/3	Pineocytoma	9361/1	Psammomatous meningioma	9533/0	EBV-positive diffuse large B-cell lymphoma; NOS	
Oligodendroglioma, NOS	9450/3	Pineal parenchymal tumour of intermediate differentiation	9362/3	Angiomatous meningioma	9534/0	Lymphomatoid granulomatosis	9766/1
Anaplastic oligodendroglioma, IDH-mutant and 1p/19q-codeleted	9451/3	Pineoblastoma	9362/3	Microcystic meningioma	9530/0	Intravascular large B-cell lymphoma	9712/3
Anaplastic oligodendroglioma, NOS	9451/3	Papillary tumour of the pineal region	9395/3	Secretory meningioma	9530/0	Low-grade B-cell lymphomas of the CNS	
Oligoastrocytoma, NOS	9382/3	<b>Embryonal tumours</b>		Lymphoplasmacyte-rich meningioma	9530/0	T-cell and NK/T-cell lymphomas of the CNS	
Anaplastic oligoastrocytoma, NOS	9382/3	Medulloblastoma, genetically defined	9475/3*	Metaplastic meningioma	9530/0	Anaplastic large cell lymphoma, ALK-positive	9714/3
Other astrocytic tumours		Medulloblastoma, WNT-activated		Chordoid meningioma	9538/1	Anaplastic large cell lymphoma, ALK-negative	9702/3
Pilomyxoid astrocytoma	9421/1	Medulloblastoma, SHH-activated		Clear cell meningioma	9538/1	MALT lymphoma of the dura	9693/3
Pilomyxoid astrocytoma	9425/3	TP53-mutant		Atypical meningioma	9539/1	<b>Histiocytic tumours</b>	
Subependymal giant cell astrocytoma	9384/1	Medulloblastoma, SHH-activated and		Papillary meningioma	9538/3	Langerhans cell histiocytosis	9751/3
Pleomorphic xanthoastrocytoma	9424/3	TP53-wildtype		Rhabdoid meningioma	9538/3	Erdheim-Chester disease	9750/1
Anaplastic pleomorphic xanthoastrocytoma	9424/3	Medulloblastoma, non-WNT/non-SHH	9477/3*	Anaplastic (malignant) meningioma	9530/3	Rosai-Dorfman disease	
Ependymal tumours		Medulloblastoma, group 3		<b>Mesenchymal, non-meningotheelial tumours</b>		Juvenile xanthogranuloma	
Subependymoma	9383/1	Medulloblastoma, group 4		Solitary fibrous tumour / hemangiopericytoma**		Histiocytic sarcoma	9755/3
Myxopapillary ependymoma	9394/1	Medulloblastomas, histologically defined		Grade 1	8815/0	<b>Germ cell tumours</b>	
Ependymoma	9391/3	Medulloblastoma, classic	9470/3	Grade 2	8815/1	Germinoma	9064/3
Papillary ependymoma	9393/3	Medulloblastoma, desmoplastic/hodgular	9471/3	Grade 3	8815/3	Embryonal carcinoma	9070/3
Clear cell ependymoma	9391/3	Medulloblastoma with extensive nodularity	9471/3	Haemangioblastoma	9161/1	Yolk sac tumour	9071/3
Tanycytic ependymoma	9391/3	Medulloblastoma, large cell / anaplastic	9474/3	Haemangioma	9120/0	Choriocarcinoma	9100/3
Ependymoma, RELA fusion-positive	9396/3*	Medulloblastoma, NOS	9470/3	Epithelioid haemangiobendothelioma	9133/3	Teratoma	9080/1
Anaplastic ependymoma	9392/3	<b>Tumours of the sellar region</b>		Angiosarcoma	9120/3	Mature teratoma	9080/0
Other gliomas		Embryonal tumour with multilayered rosettes, C19MC-altered	9478/3*	Kaposi sarcoma	9140/3	Immature teratoma	9080/3
Chordoid glioma of the third ventricle	9444/1	Embryonal tumour with multilayered rosettes, NOS	9473/3	Ewing sarcoma / PNET	9384/3	Teratoma with malignant transformation	9084/3
Angiocentric glioma	9431/1	Medulloepithelioma	9501/3	Lipoma	8650/0	Mixed germ cell tumour	9085/3
Astroblastoma	9430/3	CNS neuroblastoma	9500/3	Angiolipoma	8881/0		
Choroid plexus tumours		CNS ganglioneuroblastoma	9490/3	Hibernoma	8880/0	<b>Tumours of the sellar region</b>	
Choroid plexus papilloma	9390/0	CNS embryonal tumour, NOS	9473/3	Liposarcoma	8650/3	Cranipharyngioma	9350/1
Atypical choroid plexus papilloma	9390/1	Atypical teratoid/rhabdoid tumour	9508/3	Desmoid-type fibromatosis	8821/1	Ademantinomatous craniopharyngioma	9351/1
Choroid plexus carcinoma	9390/3	CNS embryonal tumour with rhabdoid features	9508/3	Mycfibroblastoma	8625/0	Papillary craniopharyngioma	9352/1
<b>Tumours of the cranial and paraspinal nerves</b>		Schwannoma	9560/0	Inflammatory myofibroblastic tumour	8825/1	Granular cell tumour of the sellar region	9582/0
		Cellular schwannoma	9560/0	Benign fibrous histiocytoma	8830/0	Pituitary adenoma	9432/1
		Plexiform schwannoma	9560/0	Fibrosarcoma	8610/3	Spindle cell oncocyotoma	8290/0
				Undifferentiated pleomorphic sarcoma / malignant fibrous histiocytoma	8802/3	<b>Metastatic tumours</b>	
				Leiomyoma	8890/0	The morphology codes are from the International Classification of Diseases for Oncology (ICD-O) (7424). Behaviour is coded 0 for benign tumours; 1/1 for unspccified, borderline, or uncertain behaviour; 2 for carcinomas in situ and grade III; 3/3 for epithelial neoplasia; and 4/4 for malignant tumours. The classification is modified from the previous WHO classification, taking into account changes in our understanding of these lesions.	
				Leiomyosarcoma	8890/3	*These new codes were approved by the IAPC/WHO Committee for ICD-O. **Grade according to the 2013 WHO Classification of Tumours of Soft Tissue and Bone.	
				Rhabdomyoma	8900/0		
				Rhabdomyosarcoma	8900/3		
				Chondroma	9220/0		
				Chondrosarcoma	9220/3		
				Osteoma	9180/0		

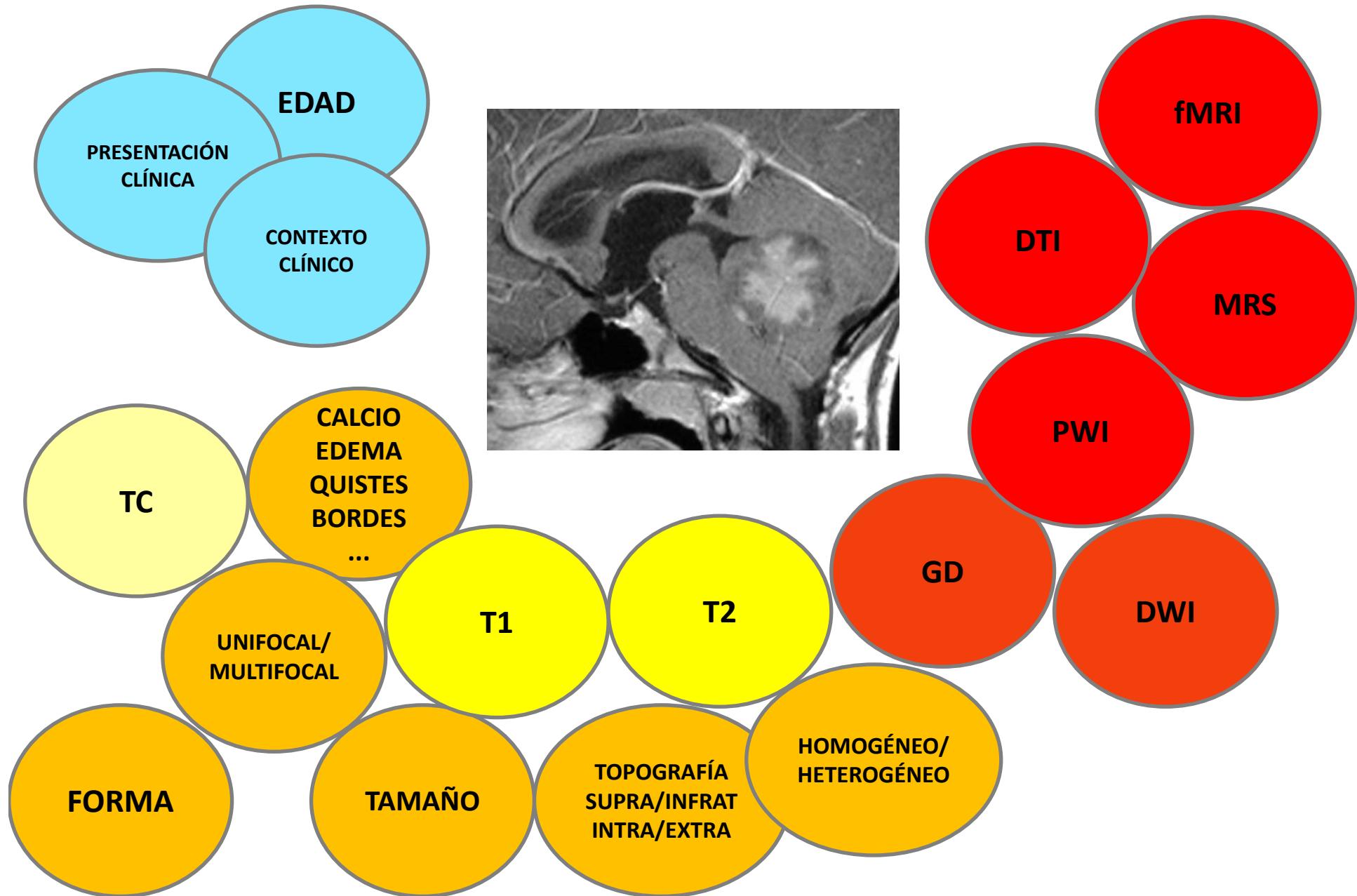
# Clasificación - Gradación

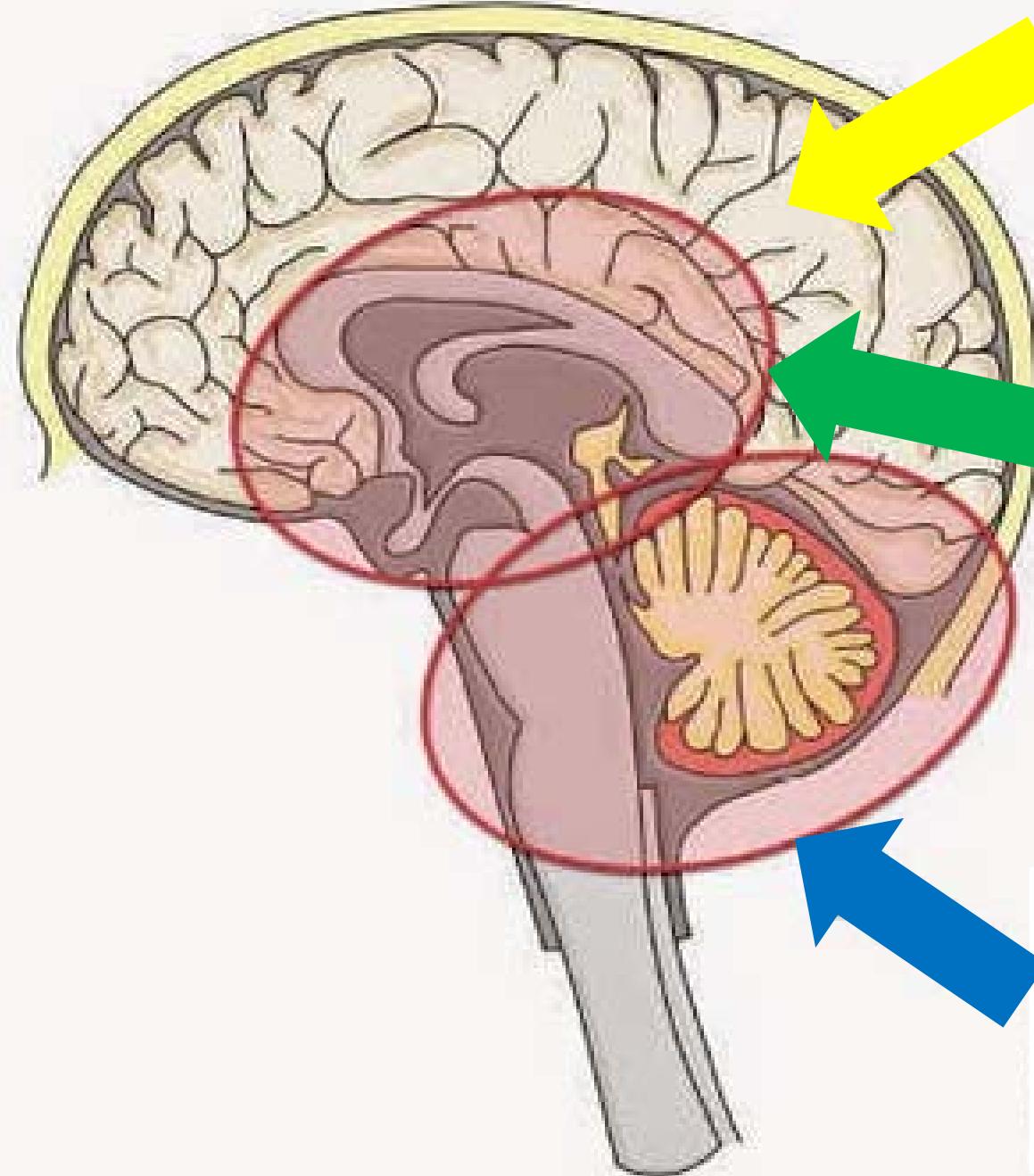


<b>WHO grades of select CNS tumours</b>			
<b>Diffuse astrocytic and oligodendroglial tumours</b>			
Diffuse astrocytoma, IDH-mutant	II	Desmoplastic infantile astrocytoma and ganglioglioma	I
Anaplastic astrocytoma, IDH-mutant	III	Papillary glioneuronal tumour	I
Glioblastoma, IDH-wildtype	IV	Rosette-forming glioneuronal tumour	I
Glioblastoma, IDH-mutant	IV	Central neurocytoma	II
Diffuse midline glioma, H3 K27M-mutant	IV	Extraventricular neurocytoma	II
Oligodendrogioma, IDH-mutant and 1p/19q-codeleted	II	Cerebellar liponeurocytoma	II
Anaplastic oligodendrogioma, IDH-mutant and 1p/19q-codeleted	III	<b>Tumours of the pineal region</b>	
		Pineocytoma	I
		Pineal parenchymal tumour of intermediate differentiation	II or III
		Pineoblastoma	IV
		Papillary tumour of the pineal region	II or III
<b>Other astrocytic tumours</b>		<b>Embryonal tumours</b>	
Pilocytic astrocytoma	I	Medulloblastoma (all subtypes)	IV
Subependymal giant cell astrocytoma	I	Embryonal tumour with multilayered rosettes, C19MC-altered	IV
Pleiomorphic xanthoastrocytoma	II	Medulloepithelioma	IV
Anaplastic pleiomorphic xanthoastrocytoma	III	CNS embryonal tumour, NOS	IV
<b>Ependymal tumours</b>		Atypical teratoid/rhabdoid tumour	IV
Subependymoma	I	CNS embryonal tumour with rhabdoid features	IV
Myxopapillary ependymoma	I	<b>Tumours of the cranial and paraspinal nerves</b>	
Ependymoma	II	Schwannoma	I
Ependymoma, RELA fusion-positive	II or III	Neurofibroma	I
Anaplastic ependymoma	III	Perineurioma	I
<b>Other gliomas</b>		Malignant peripheral nerve sheath tumour (MPNST)	II, III or IV
Angiocentric glioma	I	<b>Meningiomas</b>	
Chordoid glioma of third ventricle	II	Meningioma	I
<b>Choroid plexus tumours</b>		Atypical meningioma	II
Choroid plexus papilloma	I	Anaplastic (malignant) meningioma	III
Atypical choroid plexus papilloma	II	<b>Mesenchymal, non-meningotheelial tumours</b>	
Choroid plexus carcinoma	III	Solitary fibrous tumour / haemangiopericytoma	I, II or III
<b>Neuronal and mixed neuronal-glial tumours</b>		Haemangioblastoma	I
Dysembryoplastic neuroepithelial tumour	I	<b>Tumours of the sellar region</b>	
Gangliocytoma	I	Craniopharyngioma	I
Ganglioglioma	I	Granular cell tumour	I
Anaplastic ganglioglioma	III	Pituicytoma	I
Dysplastic gangliocytoma of cerebellum (Lhermitte-Duclos)	I	Spindle cell oncocytooma	I

PNET

# Abordaje





### T. Supratentoriales

Gliomas bajo y alto grado  
Ependimoma  
ETMR  
Tumores plexos coroideos  
Tumores glioneuronales

### T. Selar-Diencefálica

Craneofaringioma  
Tumores pineales  
Tumores germinales  
Glioma diencefálico

### T. Infratentoriales

Astrocitoma Cerebeloso  
Meduloblastoma  
Ependimoma  
Glioma troncoencefálico



# Fosa Posterior

- Representan el 54-70% de los tumores del SNC en pediatría

**Astrocitoma pilocítico juvenil**

**Meduloblastoma**

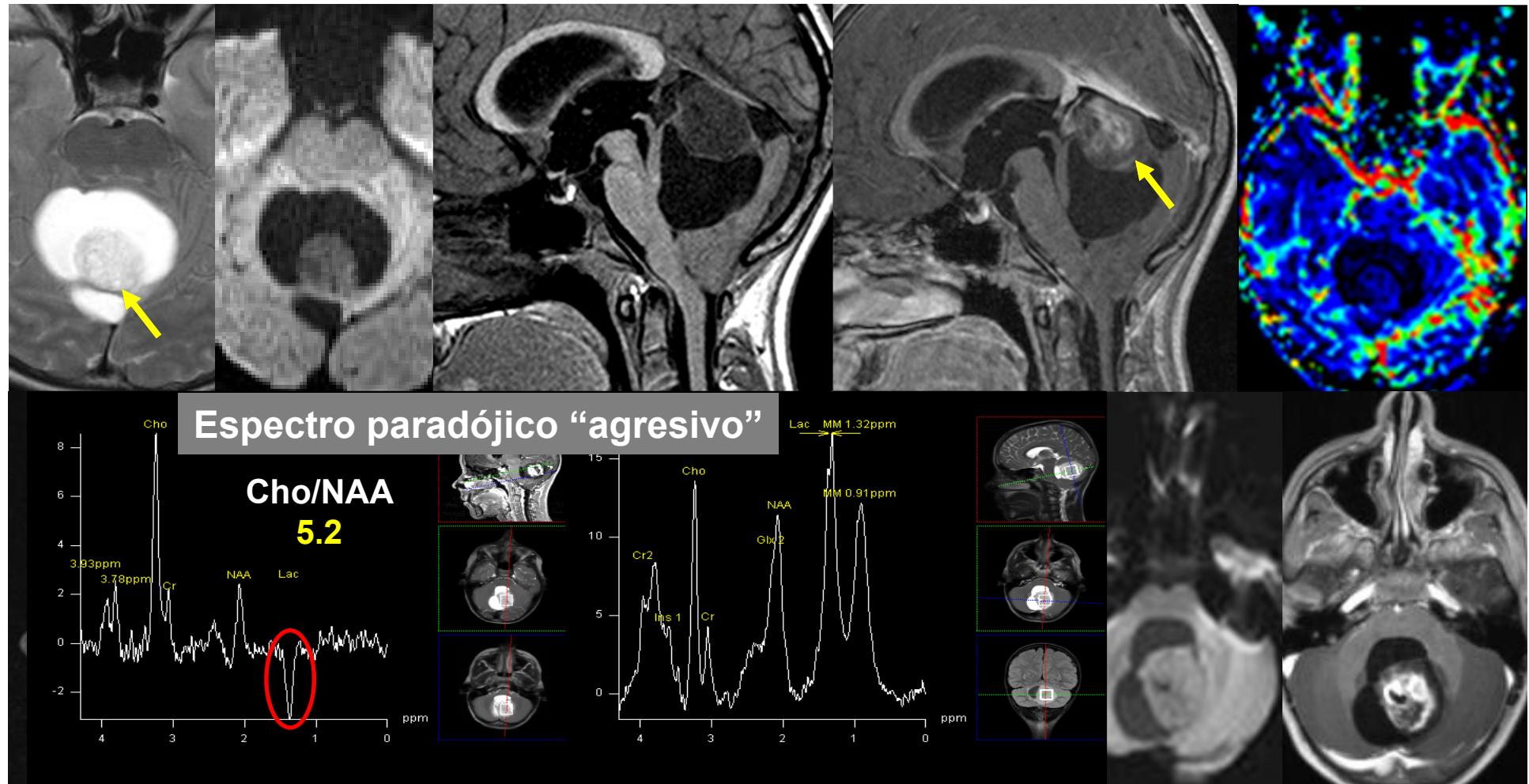
**Ependimoma**

**Glioma troncoencefálico**

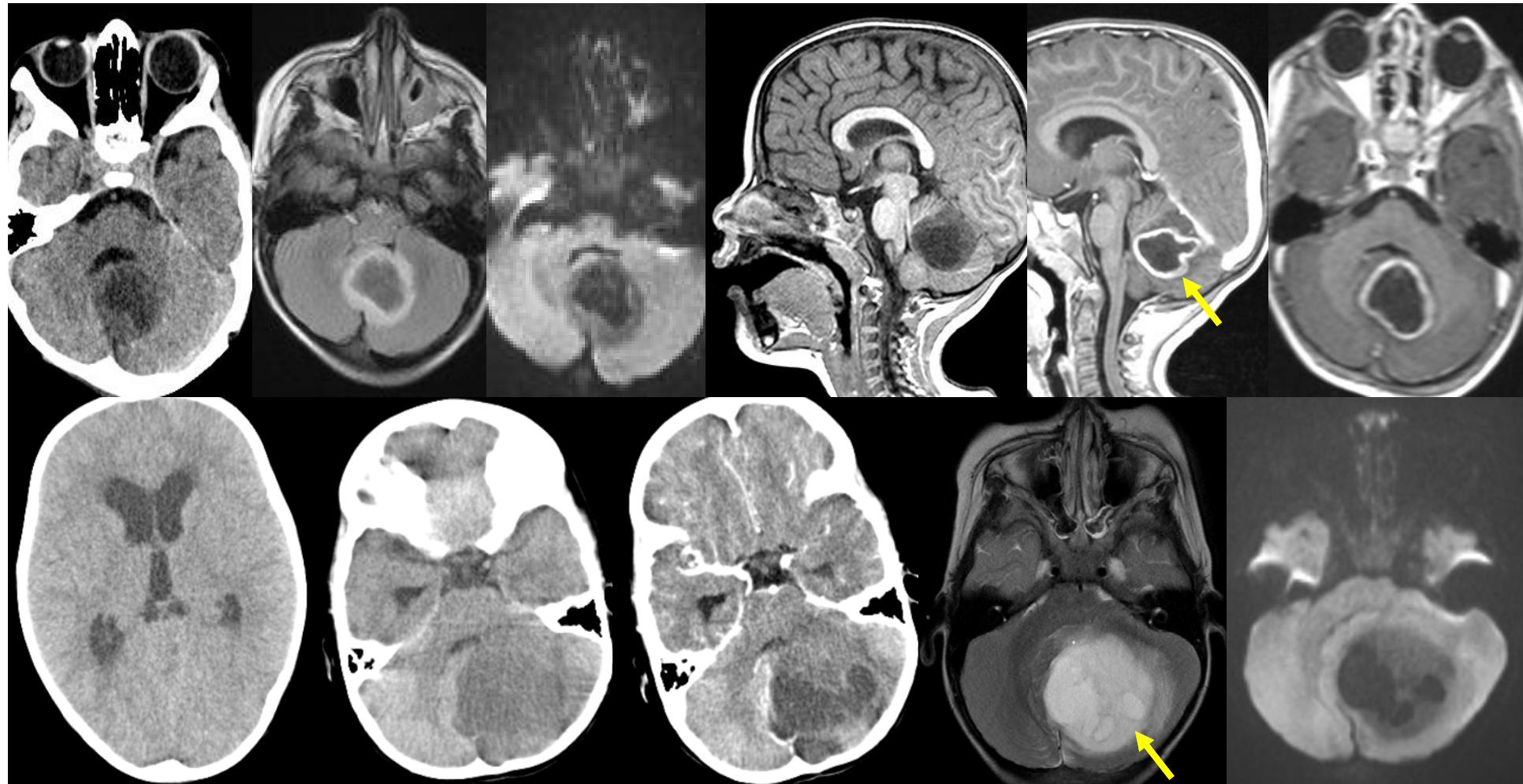


# Astrocitoma pilocítico juvenil

- WHO I. Tumor cerebral pediátrico + **común** (30-35%)
- Pico de incidencia 5-13 a - No diferencia de género
- Pronóstico excelente** / 90% ratio de supervivencia a los 25 a



# Astrocitoma pilocítico juvenil



- 60% **hemisférico cerebeloso + quiste con nódulo mural**
- 40% sólido, necrótico y realce en anillo o heterogéneo
- 95% realzan, 20% calcificación, no suelen sangrar

Q. Tumoral

Hiper FLAIR

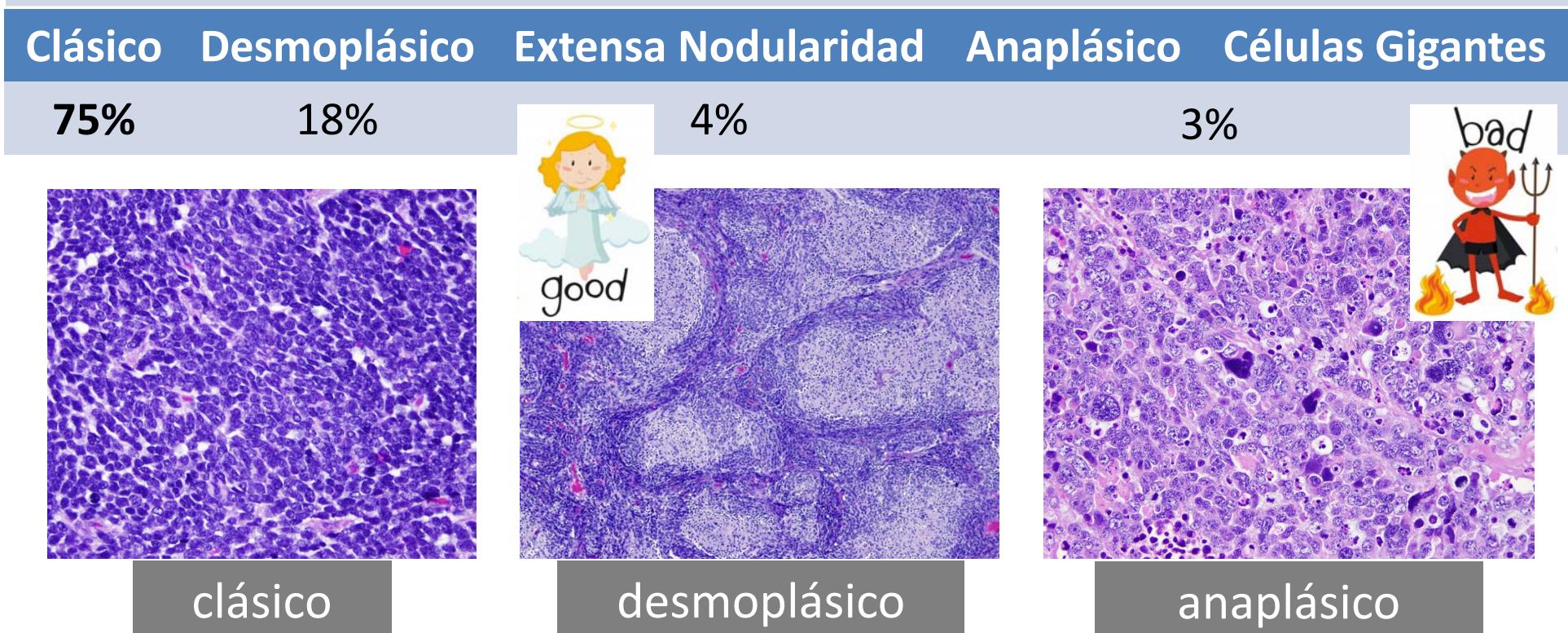
Realce Gd+



# Meduloblastoma

- Tumor embrionario WHO IV. **Tumor maligno pediátrico + común**
- Hipercelular, poco diferenciado con índice mitótico elevado
- Gran riesgo **diseminación SNC** vía LCR (30% al dx) y recurrencia

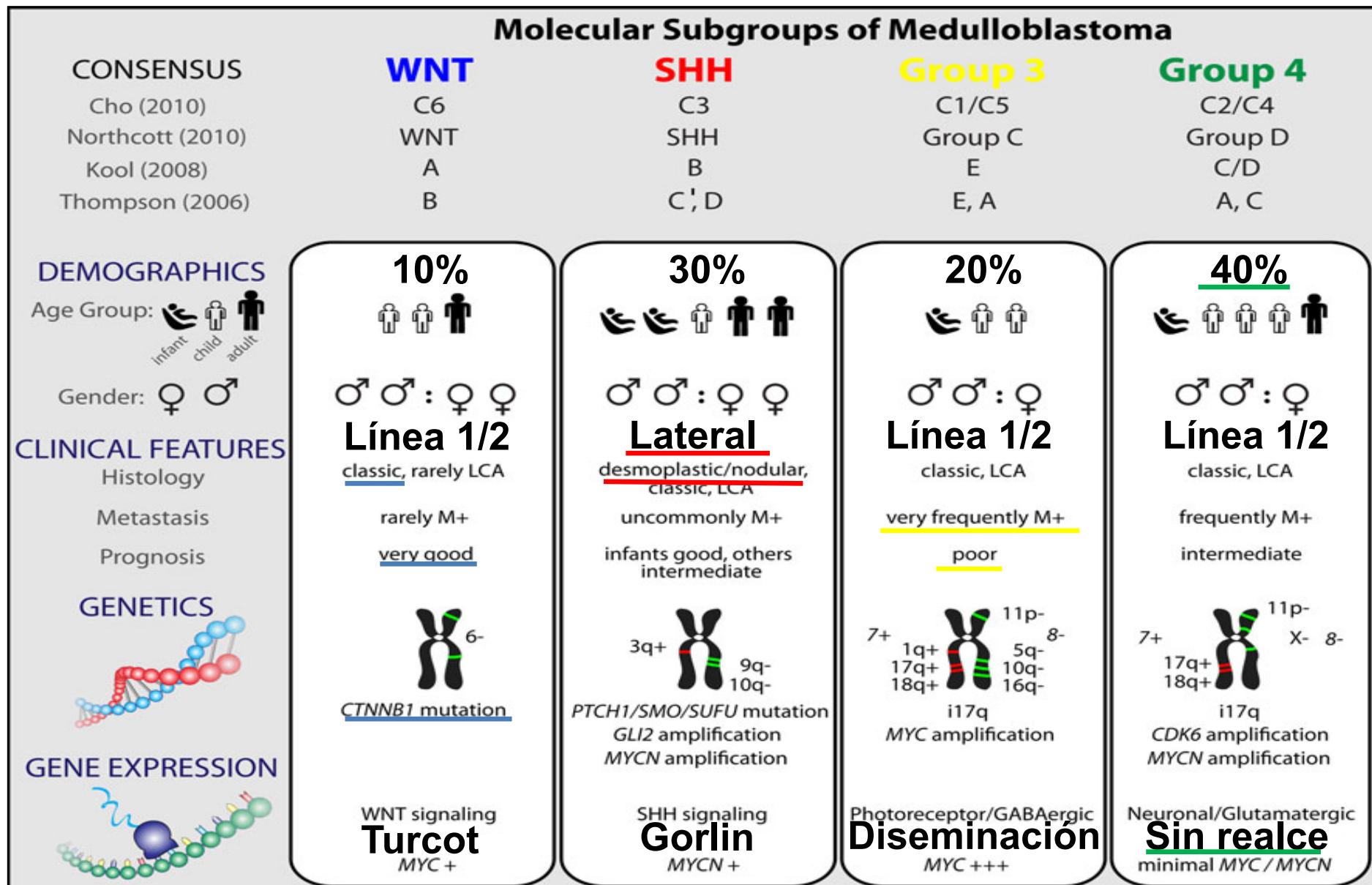
## Clasificación Histopatológica



Louis DN, et al. The 2007 WHO classification of tumours of the CNS. Acta Neuropathol 2007;114(2): 97-109



# Medulloblastoma



Taylor MD, et al. Molecular subgroups of medulloblastoma: the current consensus. *Acta Neuropathol* 2012;123:465–472



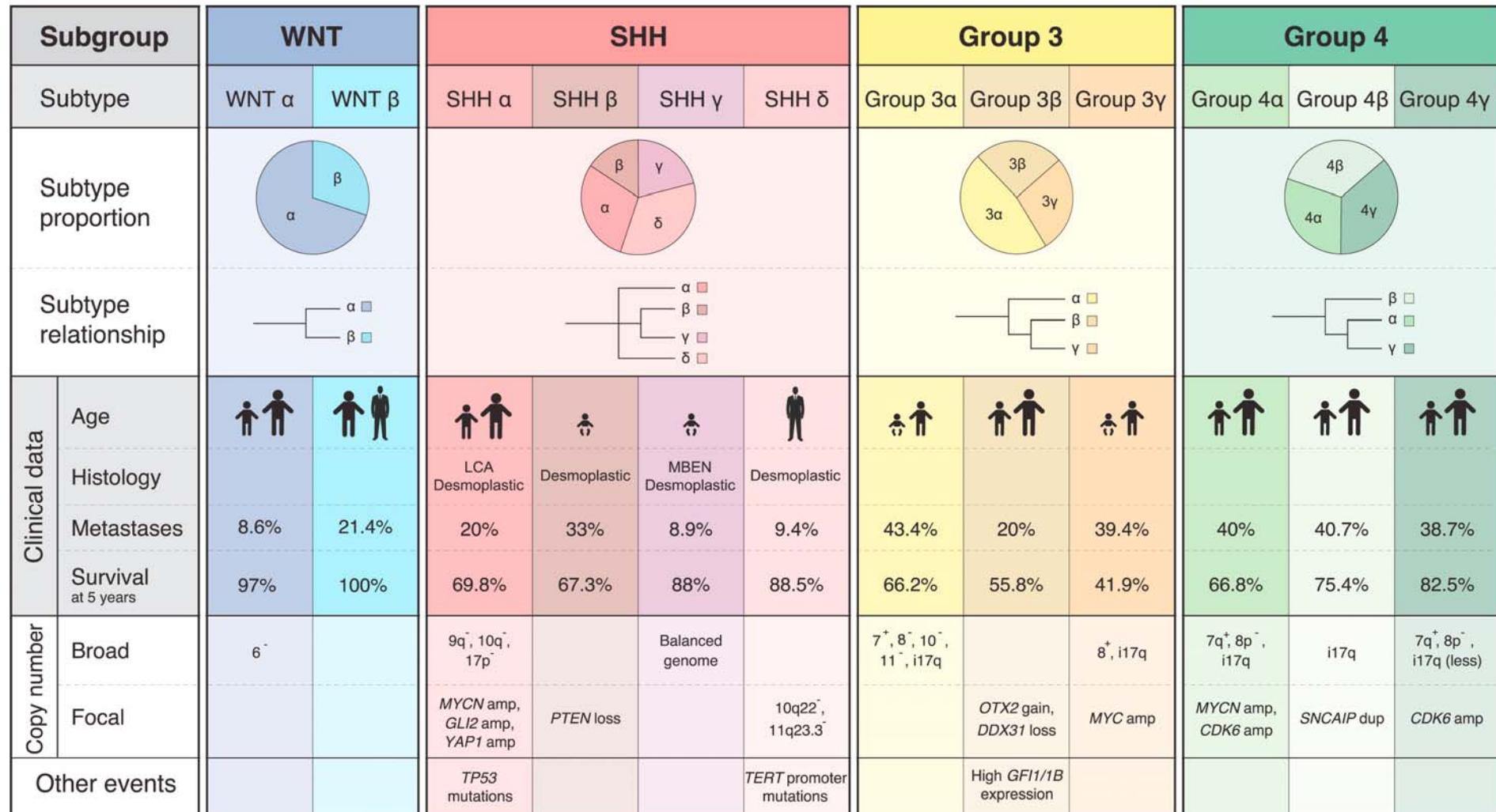
# MB – Diagnóstico integrado

Genetic profile	Histology	Prognosis
Medulloblastoma, WNT-activated	Classic	Low-risk tumour; classic morphology found in almost all WNT-activated tumours
	Large cell / anaplastic (very rare)	Tumour of uncertain clinicopathological significance
Medulloblastoma, SHH-activated, TP53-mutant	Classic	Uncommon high-risk tumour
	Large cell / anaplastic Desmoplastic / nodular (very rare)	High-risk tumour; prevalent in children aged 7–17 years Tumour of uncertain clinicopathological significance
Medulloblastoma, SHH-activated, TP53-wildtype	Classic	Standard-risk tumour
	Large cell / anaplastic Desmoplastic / nodular	Tumour of uncertain clinicopathological significance Low-risk tumour in infants; prevalent in infants and adults
	Extensive nodularity	Low-risk tumour of infancy
Medulloblastoma, non-WNT/non-SHH, group 3	Classic	Standard-risk tumour
	Large cell / anaplastic	High-risk tumour
Medulloblastoma, non-WNT/non-SHH, group 4	Classic	Standard-risk tumour; classic morphology found in almost all group 4 tumours
	Large cell / anaplastic (rare)	Tumour of uncertain clinicopathological significance

Louis DN, et al. The 2016 WHO Classification of Tumors of the CNS: a summary. *Acta Neuropathol.* 2016;131(6):803-820

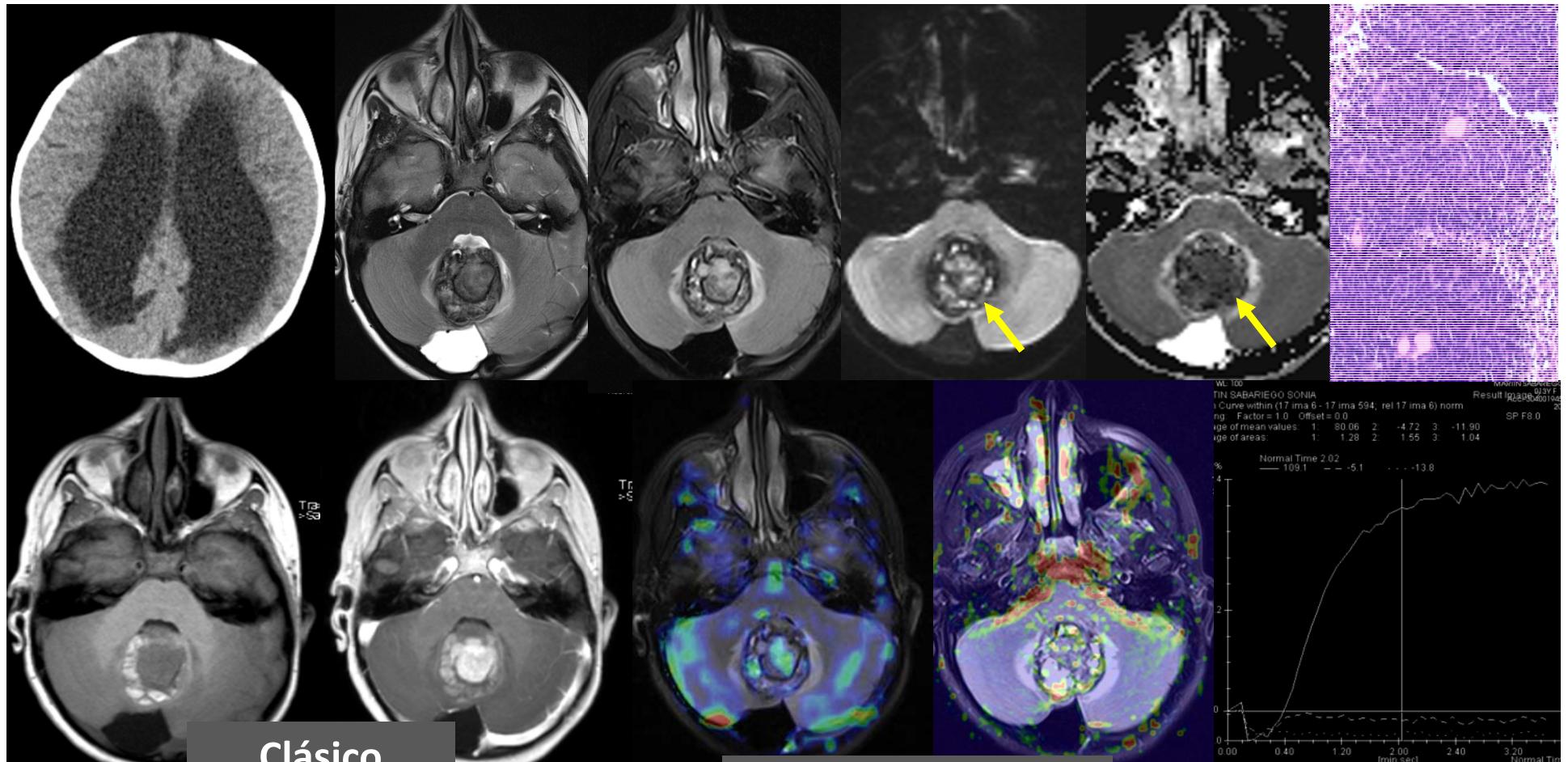


# Medulloblastoma



Age (years): 0-3 >3-10 >10-17 >17

# MB - WNT



Clásico  
Línea ½

10%

Muy buen px  
Sd. Turcot

Hiperdensidad 90%

Calcificación 20%

Hemorragia rara

Hidrocefalia 95%

MB Clásico  
Beta-catenina +

MB - SHH

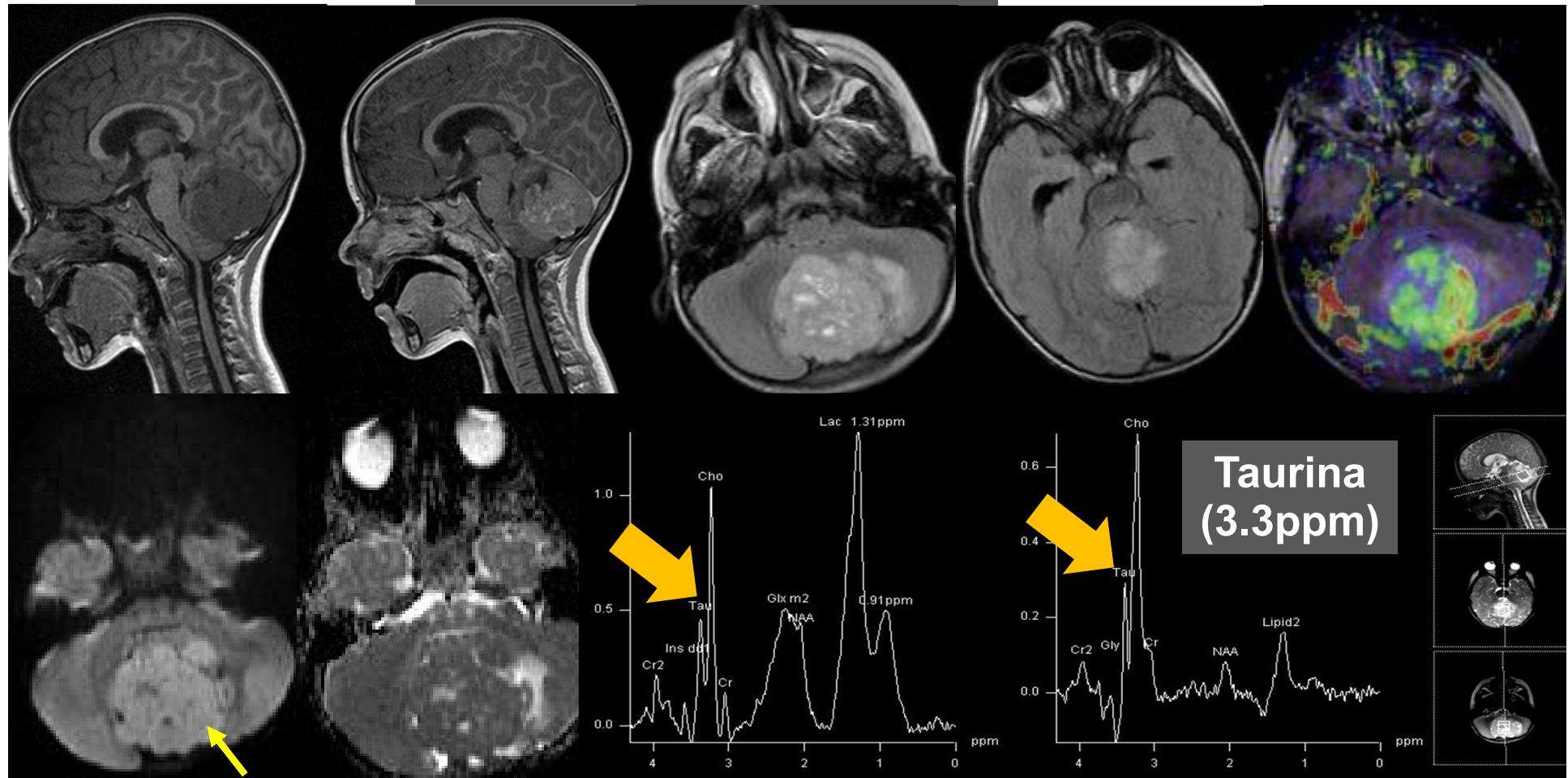
## Medulloblastoma EN SHH activa, p53 salvaje



Vall  
d'Hebron  
Barcelona Hospital Campus

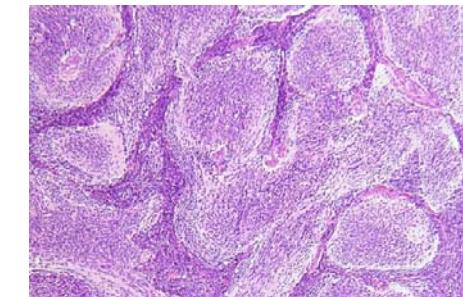
IDI  
INSTITUT DE  
DIAGNÒSTIC PER  
LA IMATGE

XVI CURSO NACIONAL DE  
SENR  
CURSO NACIONAL DE  
NEURORADIOLOGÍA

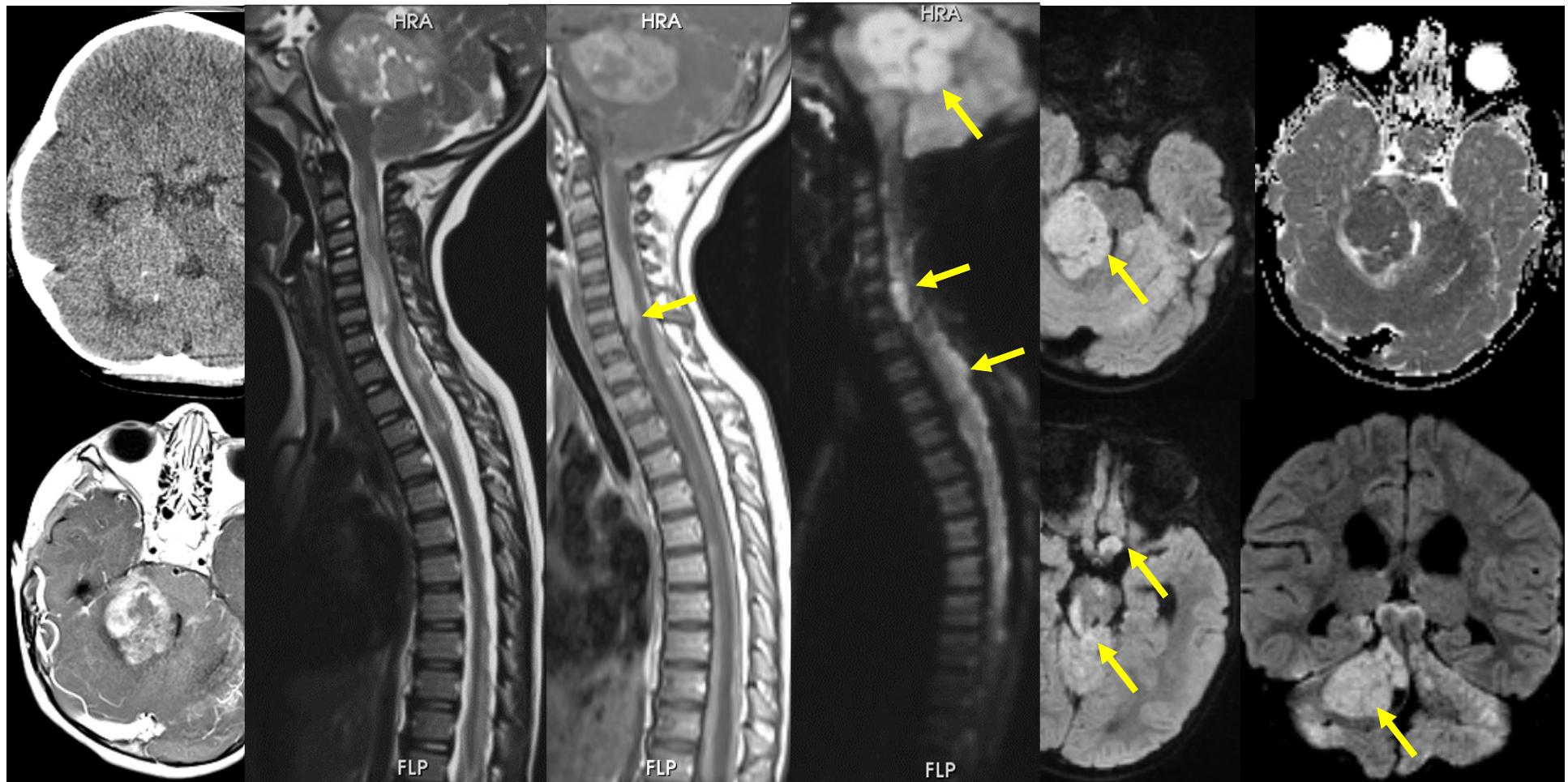


Lateral  
30%  
Buen px  
Sd. Gorlin

MB EN en < 2 a:  
criterio Dx mayor sd. Gorlin

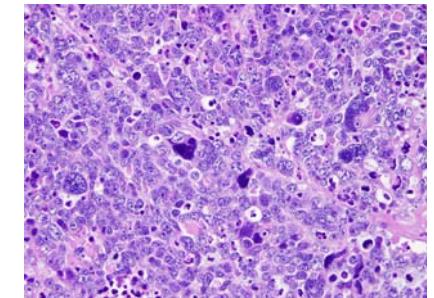


# MB - SHH



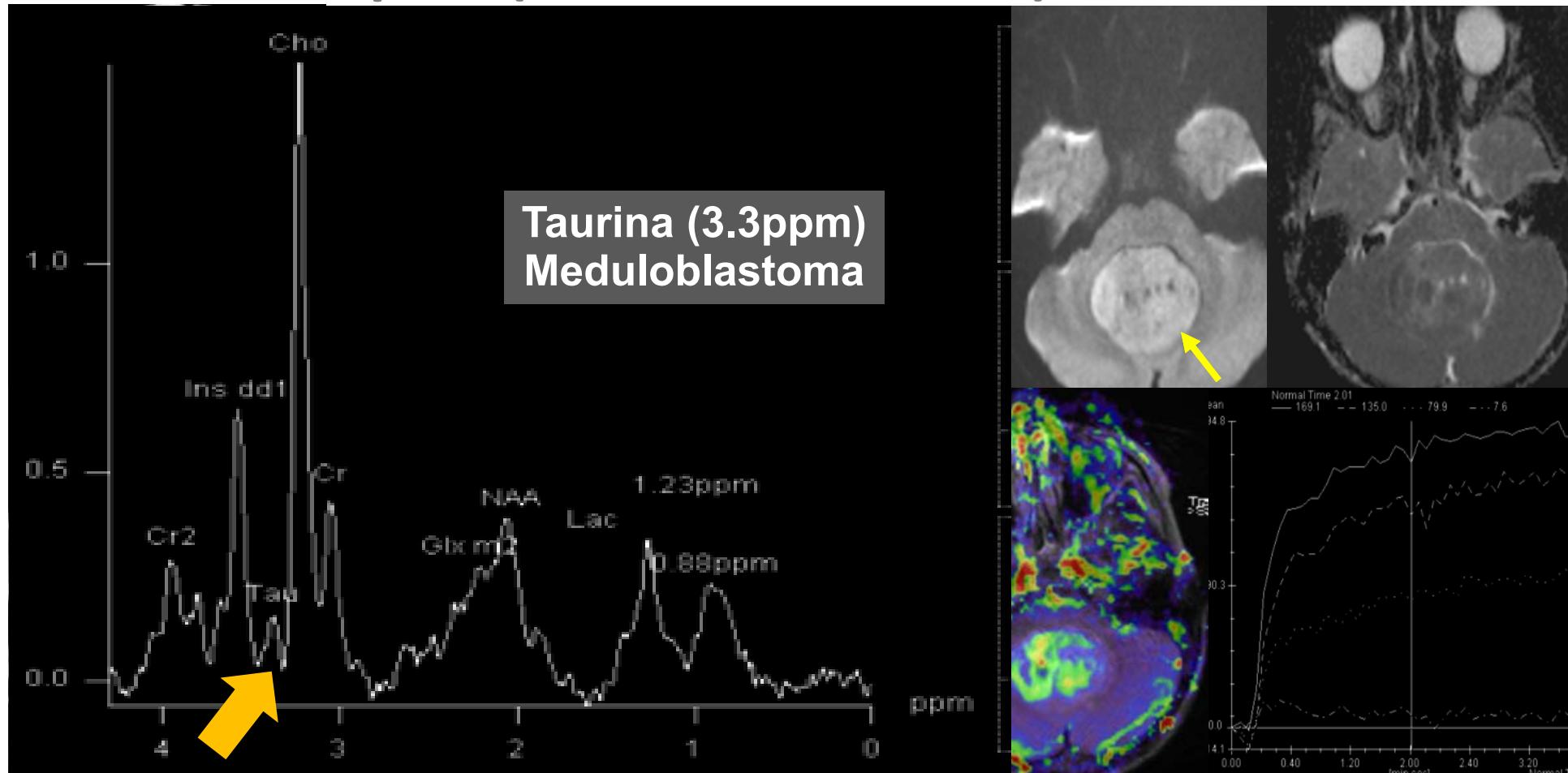
Lateral  
30%  
Buen px  
Sd. Gorlin

Medulloblastoma Anaplásico  
SHH activa, p53 mutado





# MB – Grupo 3 (No WNT / No SHH)

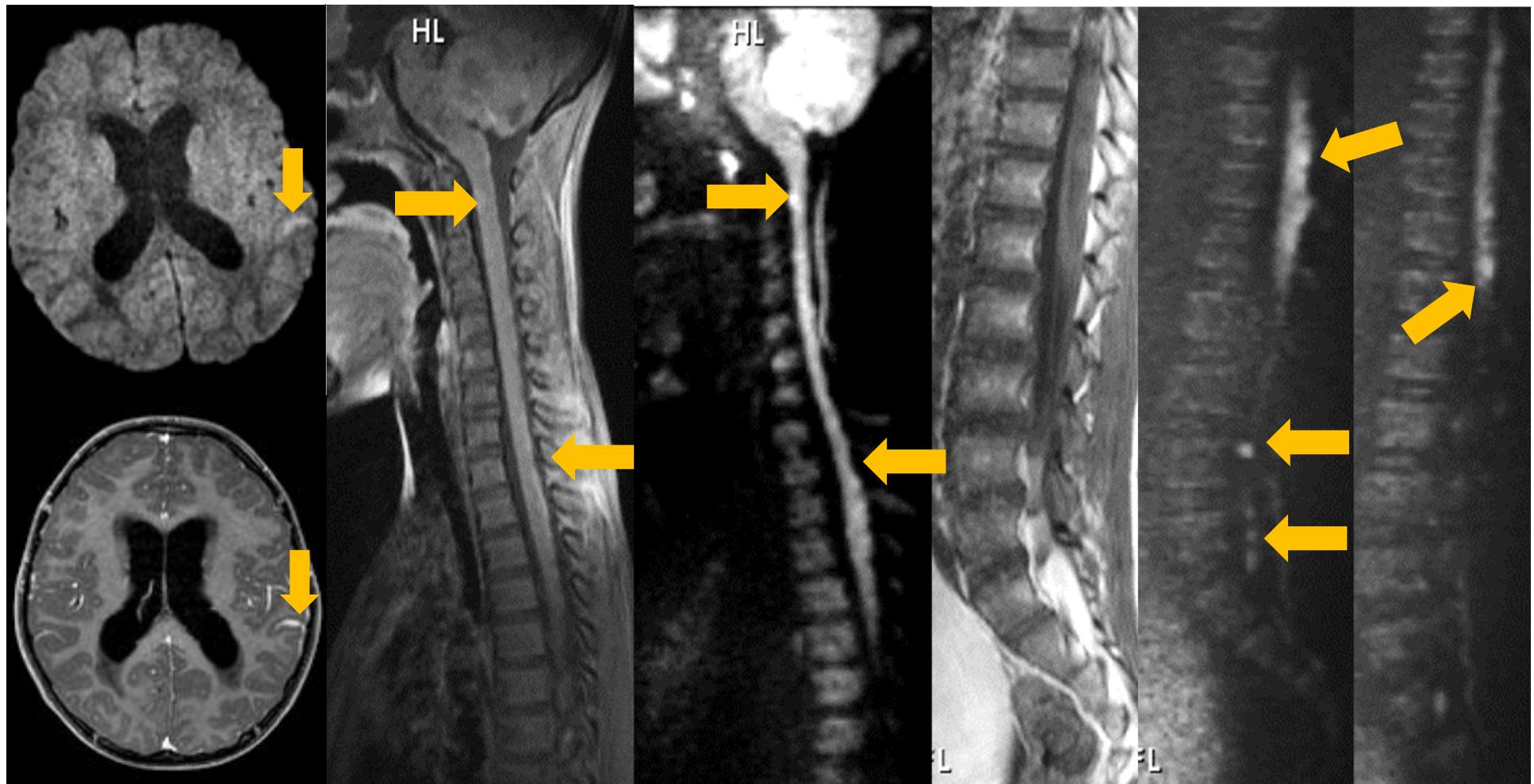


Clásico/ Anaplásico  
Línea 1/2  
20%  
Mal px

Meduloblastoma G3 Anaplásico  
Beta-catenina y P-53 negativos

Kovanlikaya A, et al. Quantitation of taurine with MR spectroscopy. Radiology. 2005;236(3):1020-5

# MB – Grupo 3 (No WNT / No SHH)

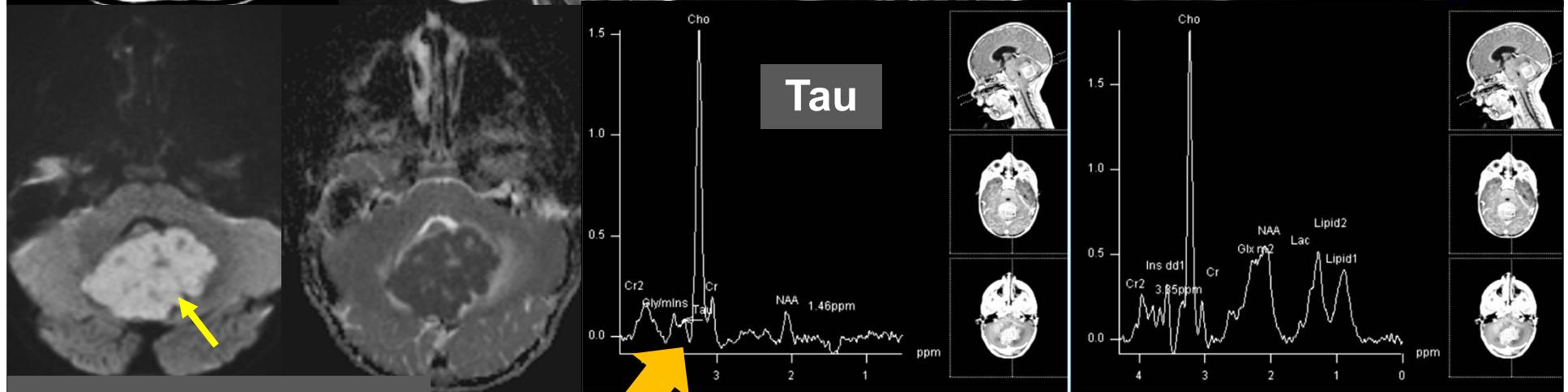
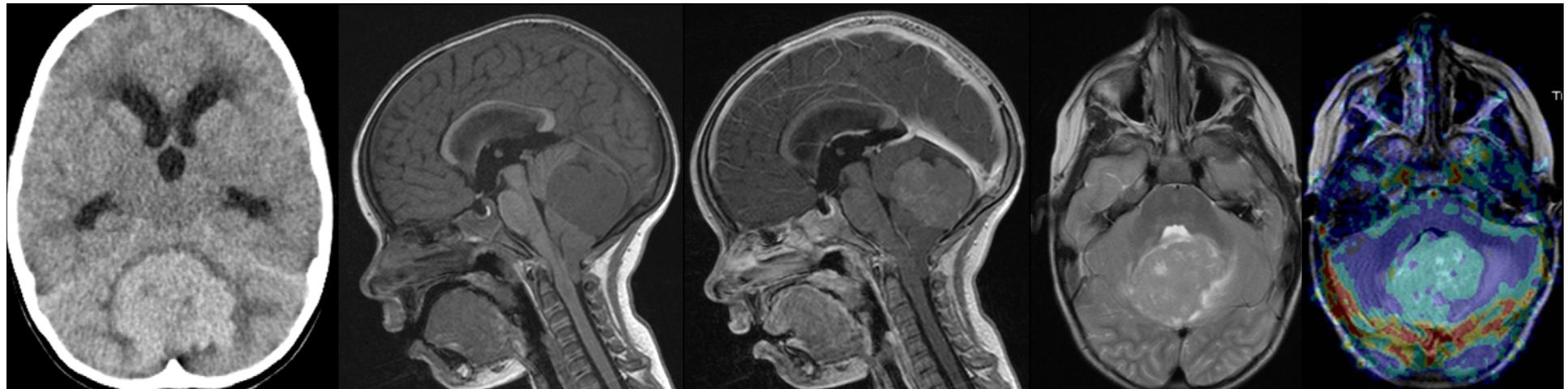


Alta incidencia de diseminación leptomeníngea craneal y espinal

Misma neuroimagen, en <2 años → ATRT

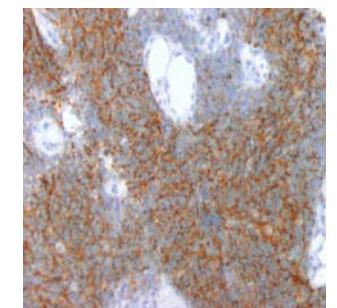


# MB – Grupo 4 (No WNT / No SHH)



Clásico/ Anaplásico  
Línea 1/2  
40%  
Intermedio px  
No realce

Meduloblastoma G4 Clásico  
Beta-catenina y P-53 negativos  
N-myc amplificado



# Ependimoma



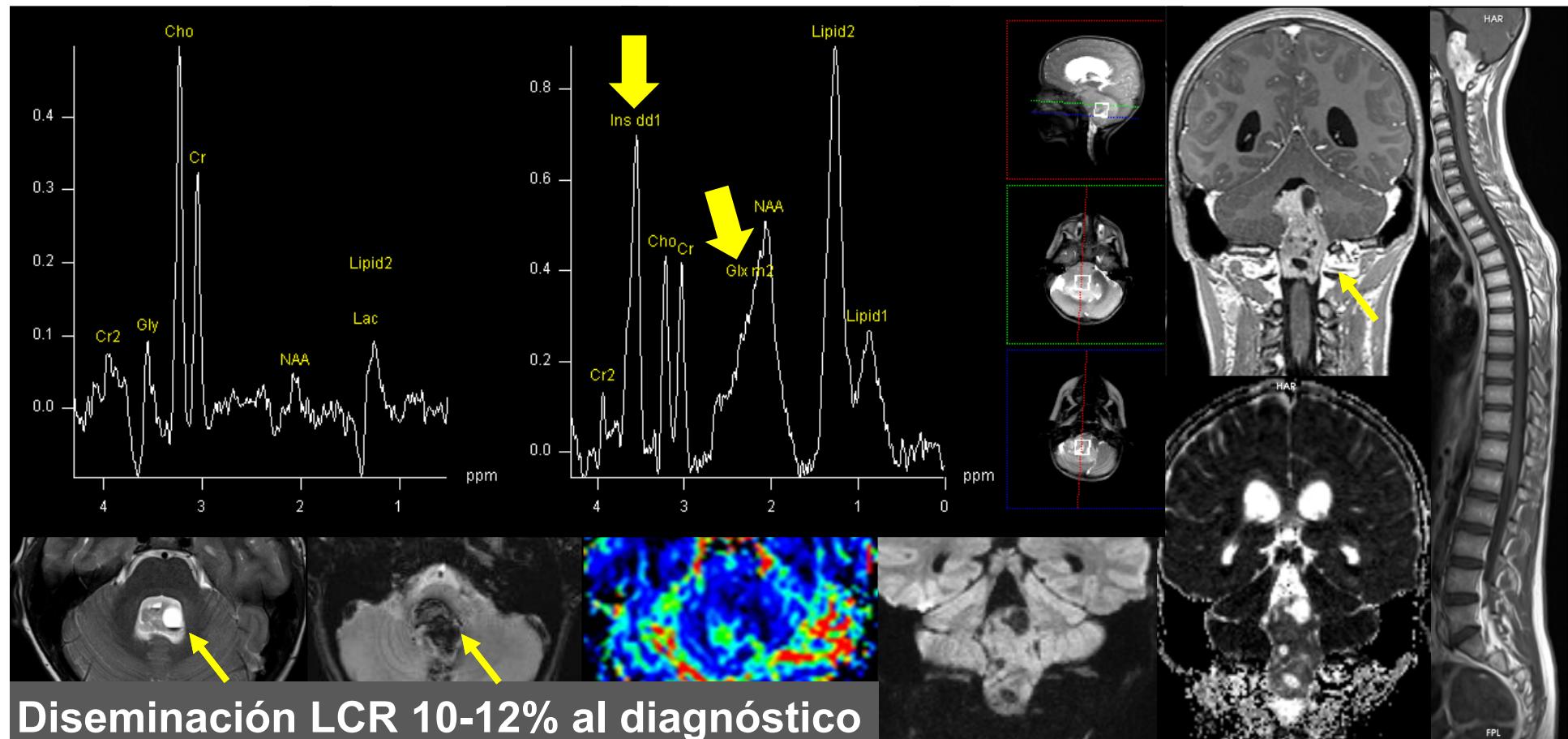
Vall  
d'Hebron  
Barcelona Hospital Campus

IDI  
INSTITUT DE  
DIÀGNOSTIC PER  
LA IMATGE

SENR  
SOCIetat  
Catalana  
de Radiologia  
XVI CURSO NACIONAL DE  
NEURORADIOLOGÍA

>% anaplásico a < edad

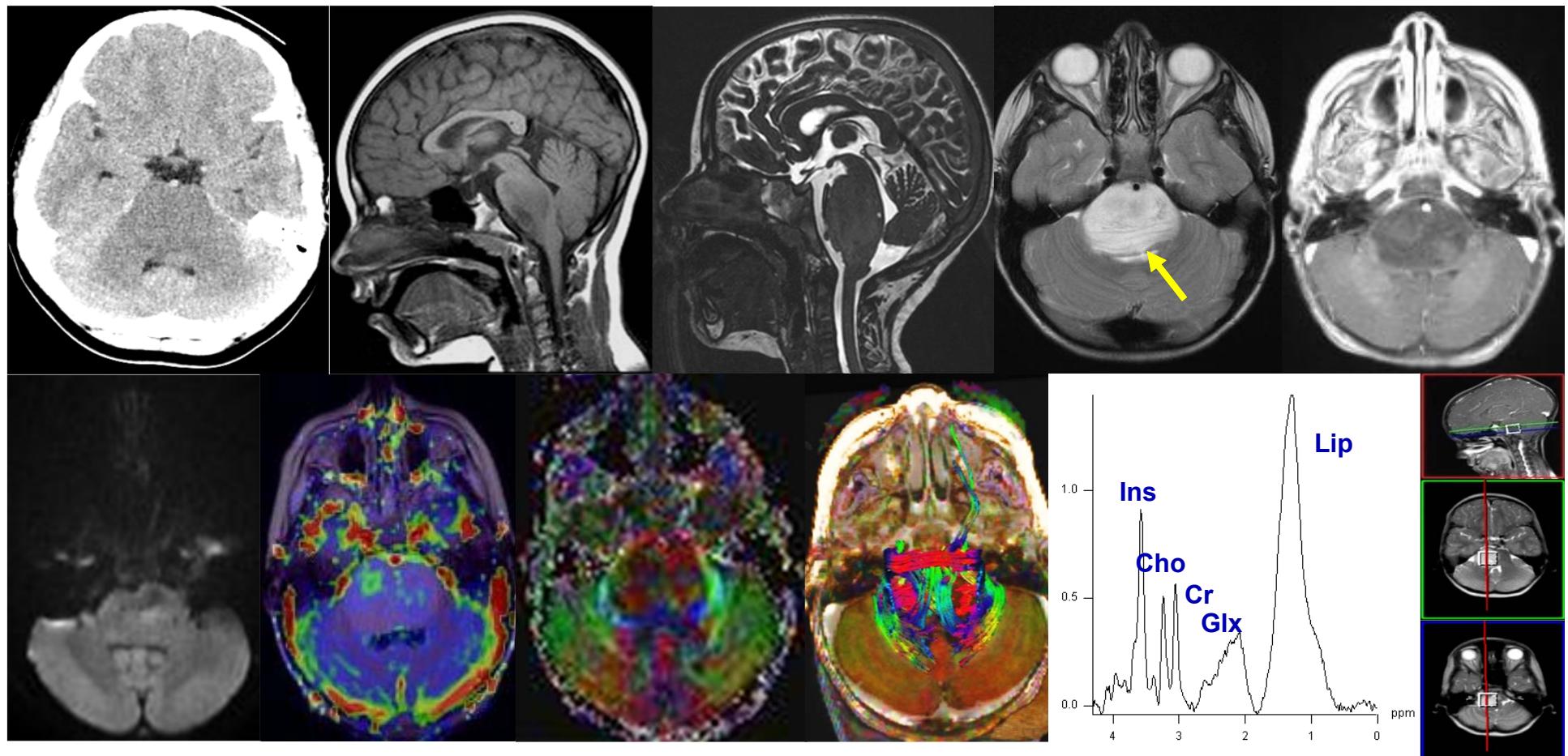
- WHO II. Variantes: E. anaplásico WHO III, E. mixopapilar WHO I, subependimoma WHO I, E. RELA fusión-positiva WHO II-III
- InfraT (70%) Masa línea  $\frac{1}{2}$  4V. **Crecimiento plástico**. Rodea vasos
- Heterogéneo**: 50% calcio, sangre, quistes, Gd+, DWI +/-, PWI +/-





# Glioma troncoencefálico

- 5-11% de los tumores intracraneales. 15-30% de los infraT
- Tipos: difuso intrínseco pontino (80%), exofítico bulbar y tectal M
- **DIPG:** infiltra >75% área TE, mal definido, Gd +/- . DWI -. Mal px





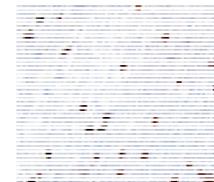
Vall  
d'Hebron  
Barcelona Hospital Campus

IDI  
INSTITUT DE  
DIAGNÒSTIC PER  
LA IMATGE

SENRA  
XVI CURSO NACIONAL DE  
NEURORRADIOLOGÍA

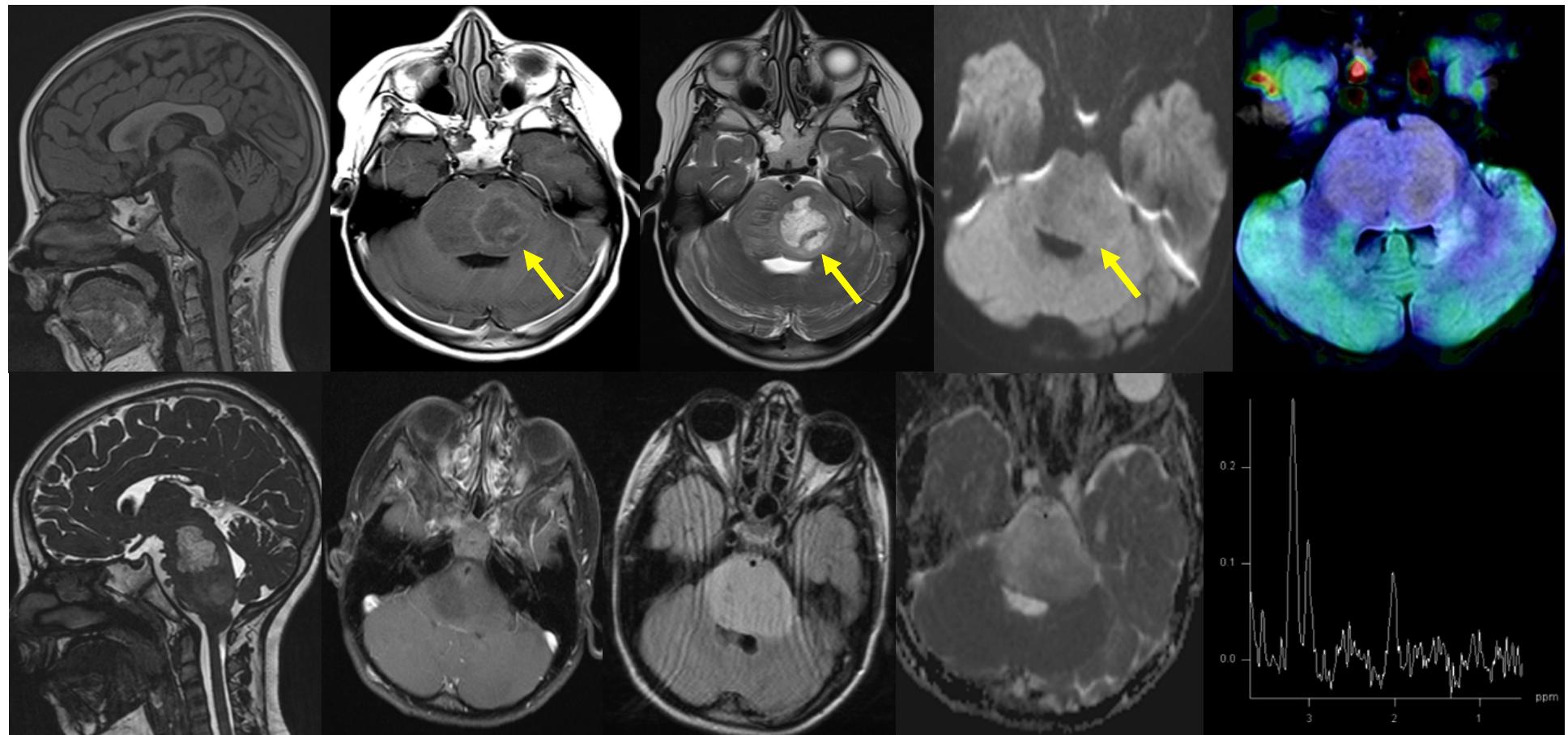


# Glioma difuso línea media

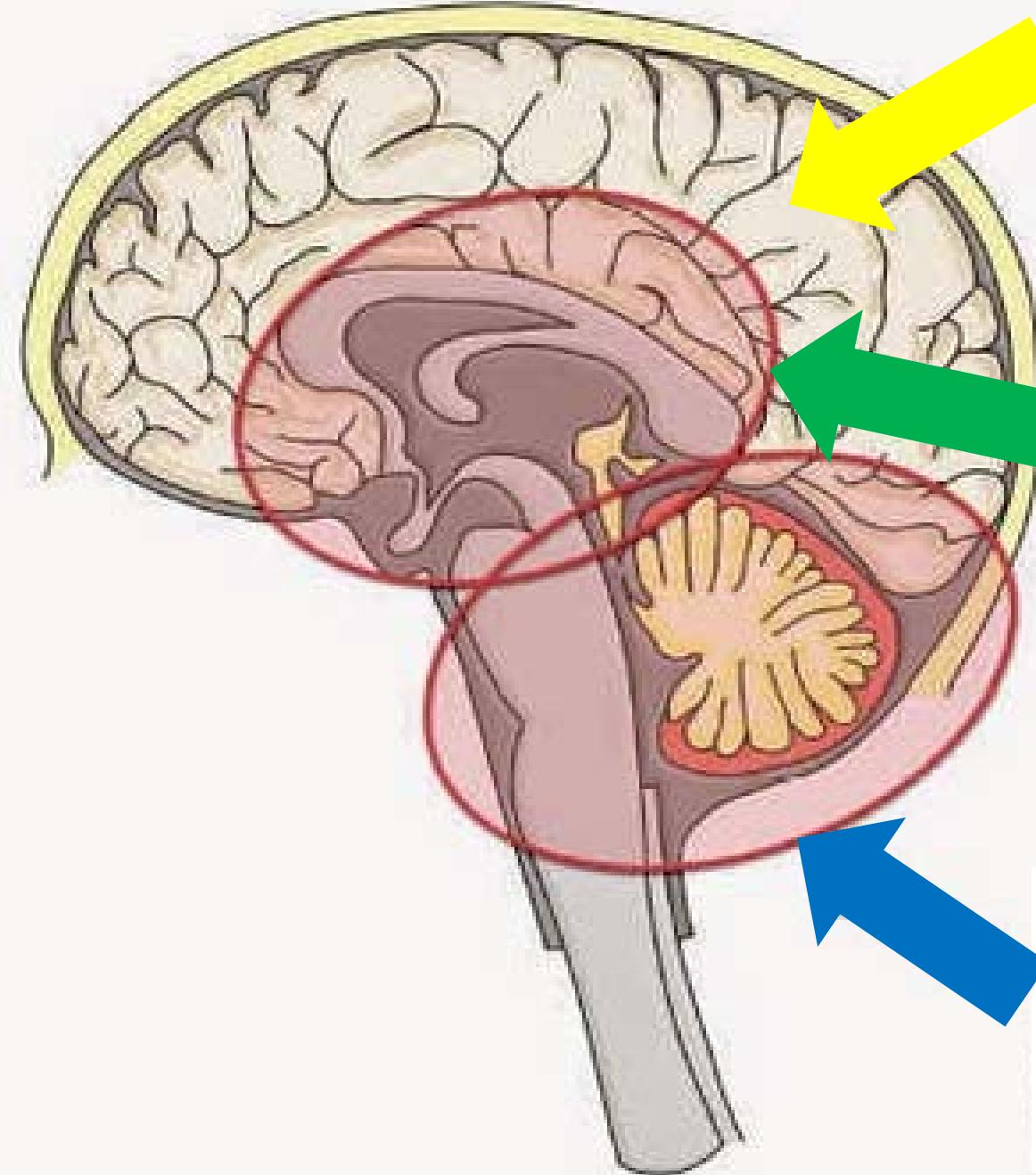


- WHO IV. Mutación **H3 K27M**: f. posterior (Pons), tálamo y médula
- Sólidos, sin necrosis, con infiltración local, Gd -, DWI -, metástasis
- Clínica: HTE, ppcc, ataxia. AP: baja celularidad y atipia

DIPG



Aboian MS, et al. Pediatric Diffuse Midline Gliomas with Histone H3 K27M Mutation. AJNR. 2017;38(4):795-800



### T. Supratentoriales

Gliomas bajo y alto grado  
Ependimoma  
ETMR  
Tumores plexos coroideos  
Tumores glioneuronales

### T. Selar-Diencefálica

Craneofaringioma  
Tumores pineales  
Tumores germinales  
Glioma diencefálico

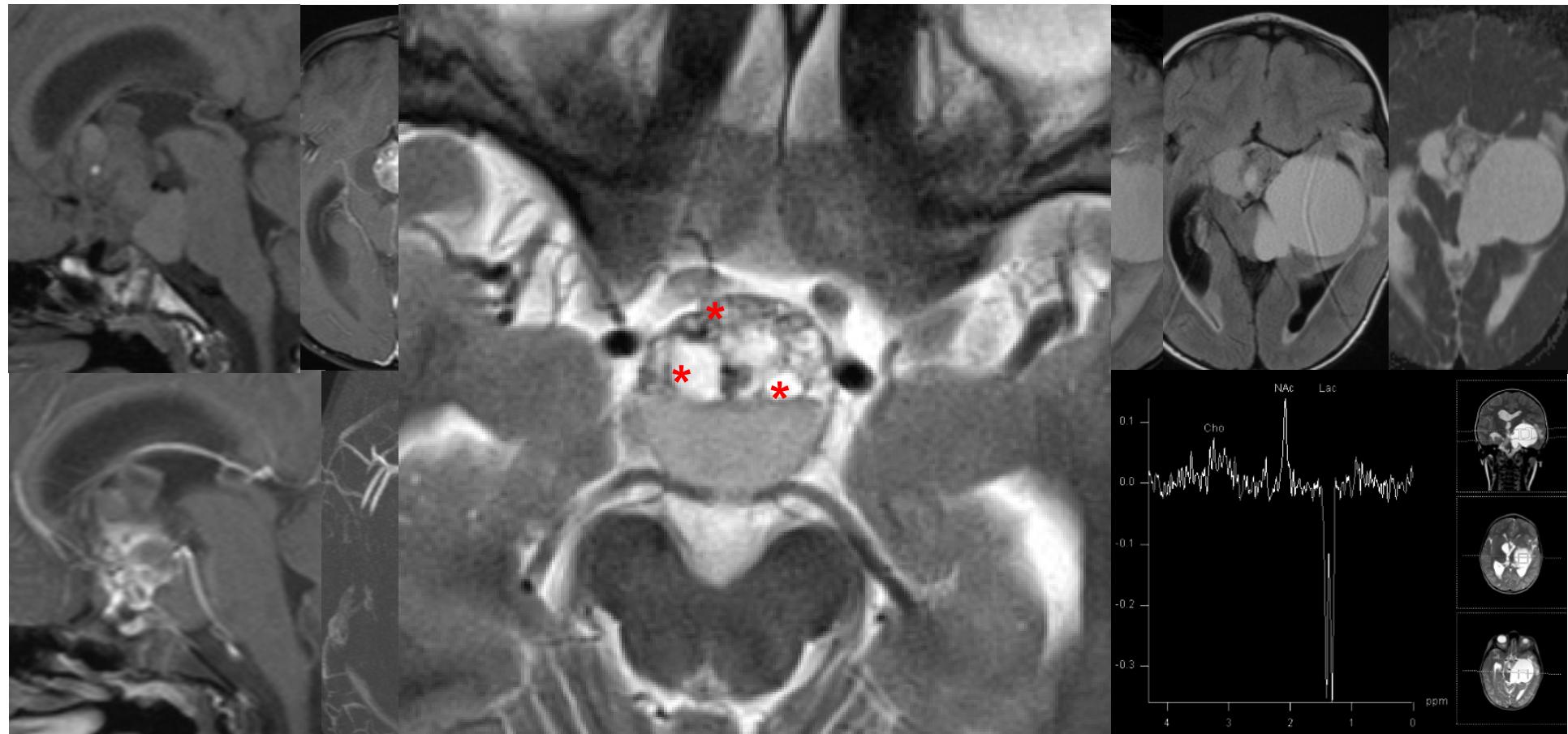
### T. Infratentoriales

Astrocitoma Cerebeloso  
Meduloblastoma  
Ependimoma  
Glioma troncoencefálico



# Craneofaringioma

- WHO I. T. pediátrico intracranial **no glial + común** (1-5%)
- Adolescentes (**adamantinomatoso**) / adultos (papilar)
- **90's rule:** supraselar, quistes, nivel, nódulos sólidos + calcio y Gd+

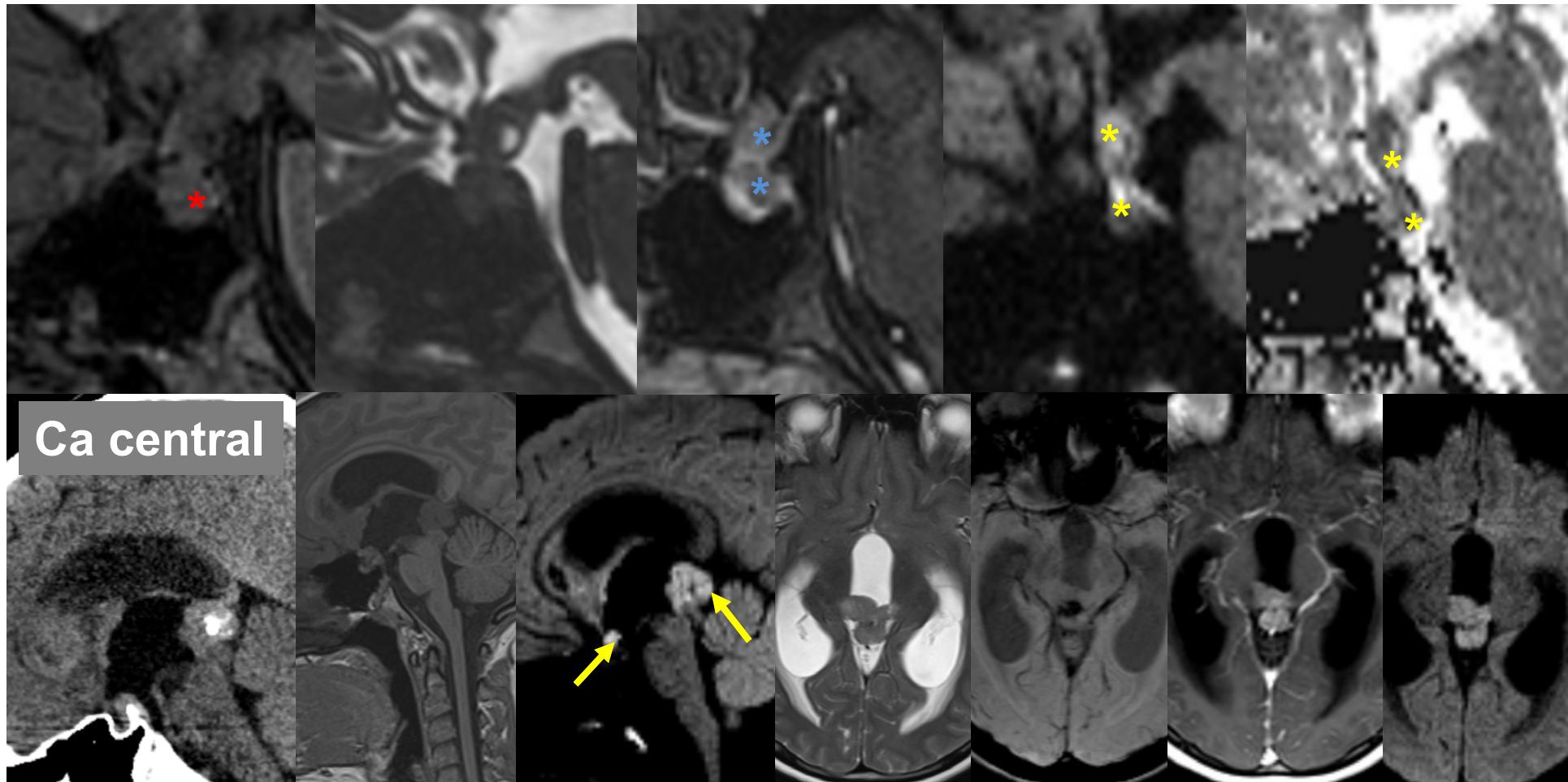


Castillo M, Mukherji SK. Imaging of the Pediatric Head, Neck and Spine. Philadelphia. Lippincott-Raven, 1996



# Tumor germinal

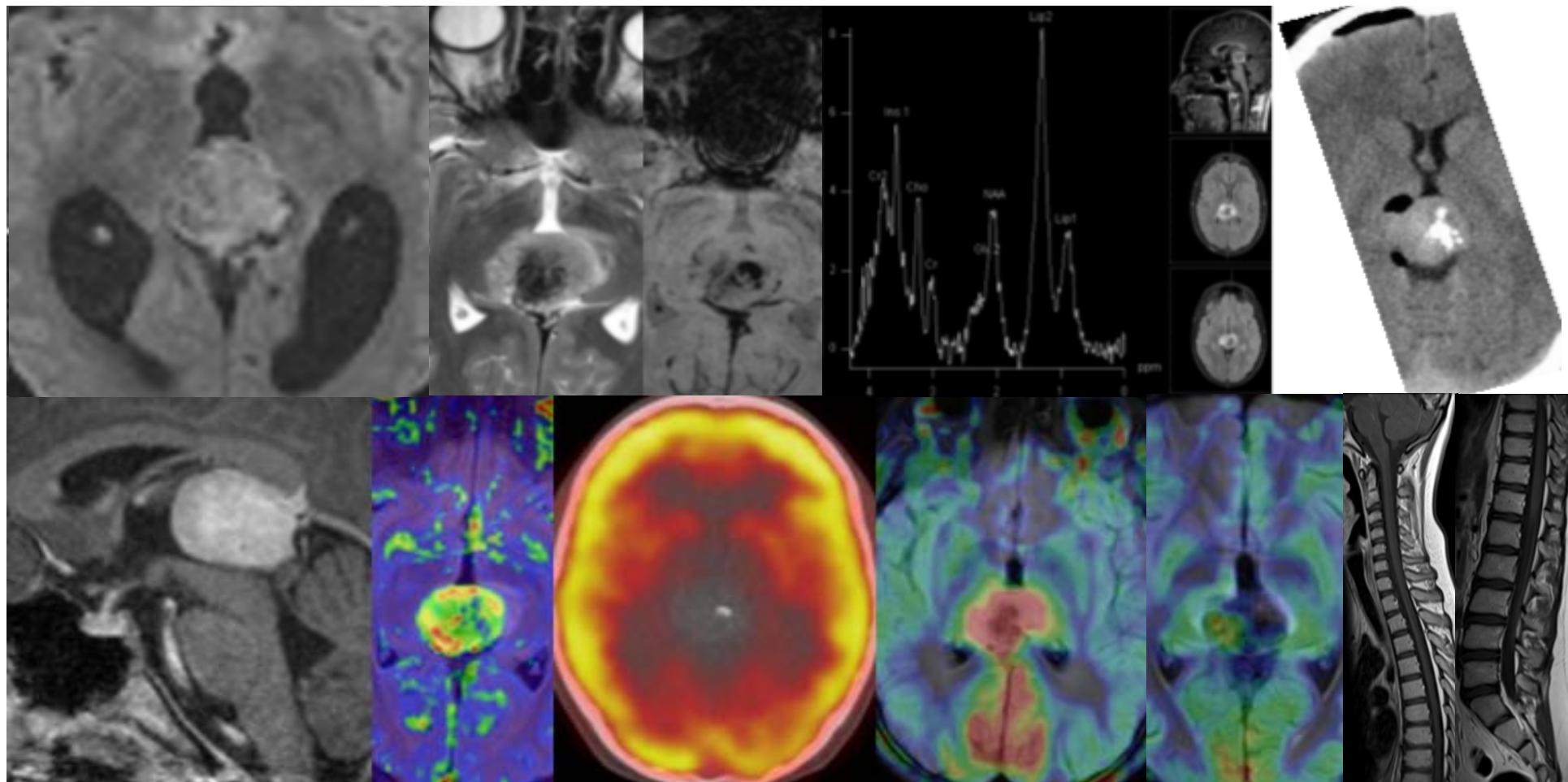
- WHO I. Loc.: pineal (50-65%) / suelo 3V (6-13%). B-HCG / A-FP
- **Celular:** hiperD, DWI + y Gd +. Diseminación LCR. Hidrocefalia





# Pineoblastoma

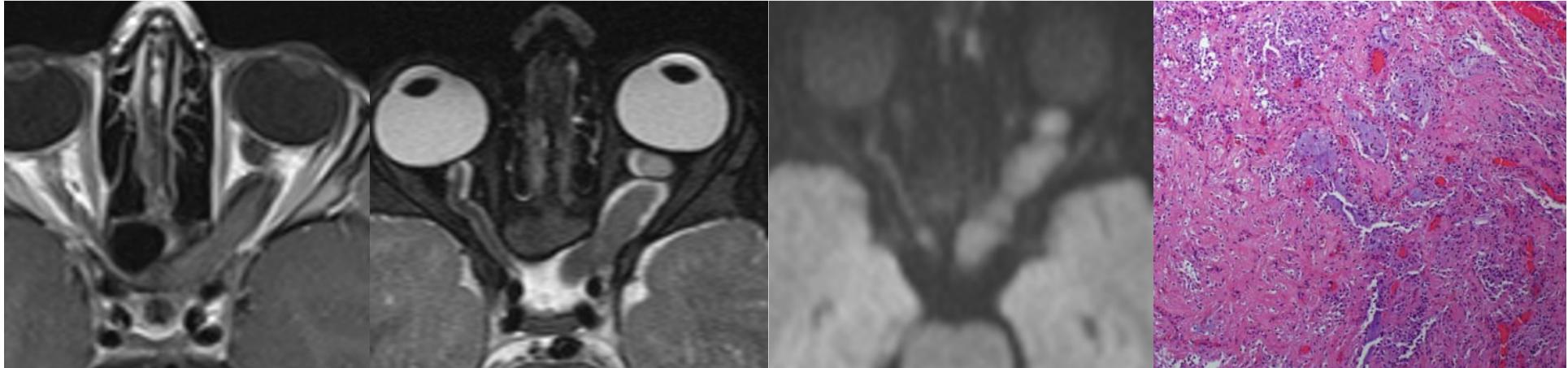
- WHO IV. Pinealocitos. Hidrocefalia y sd. Parinaud. **Blasted** Calcio
- >4cm hipercelular: hiperD, DWI+, Gd+, +/- necrosis y sangre





# Glioma optoquiasmático

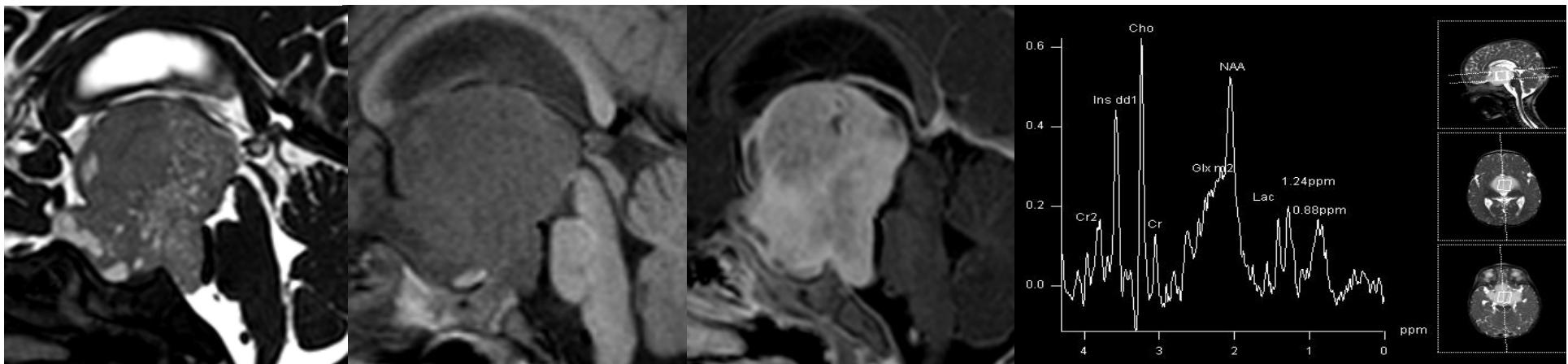
- WHO I. Engrosamiento fusiforme y tortuoso. **NF1** → Pilocítico Gd-

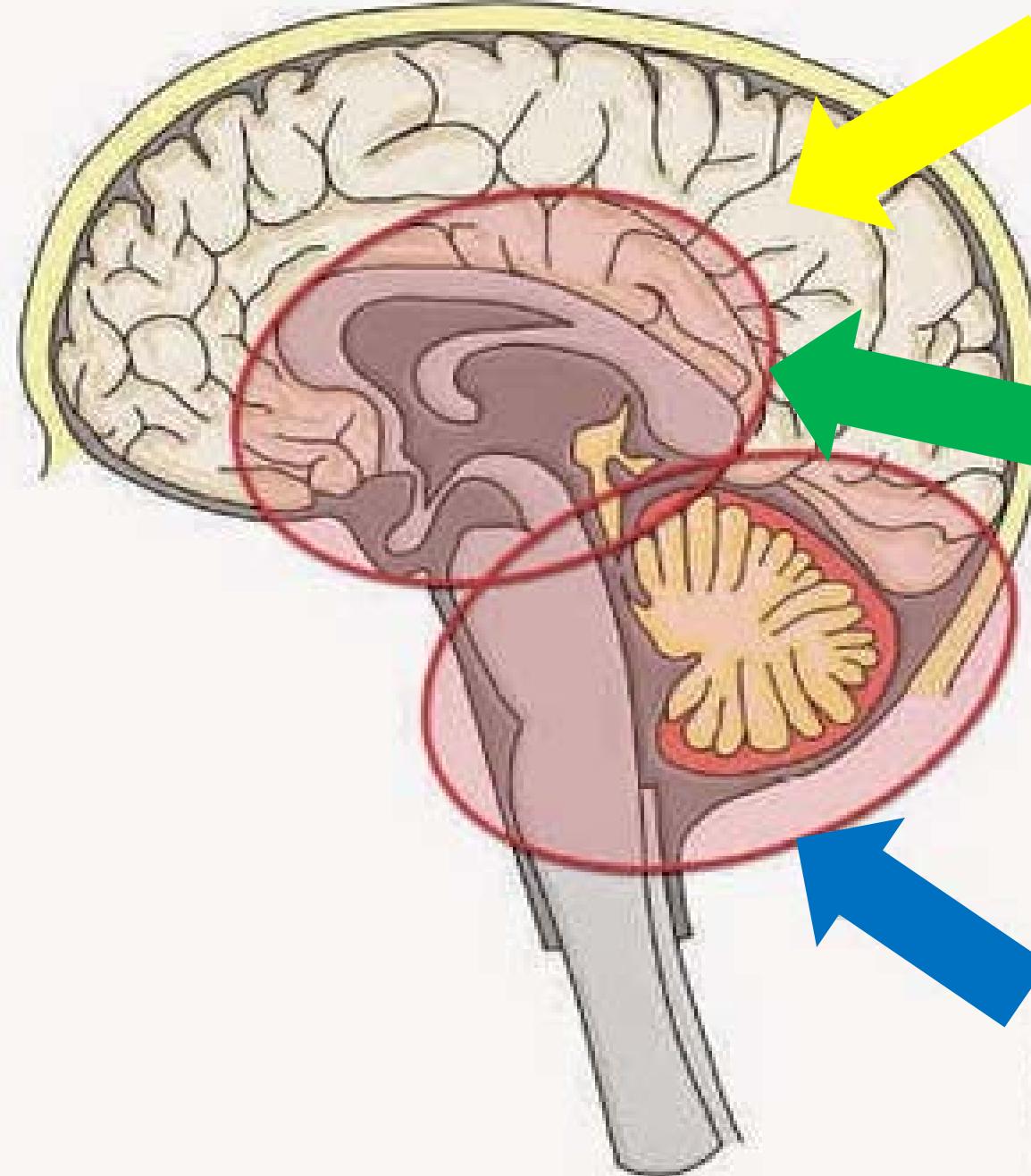


# Glioma hipotalámico

## Pilocítico vs Pilomixoide

- WHO I. Gran lesión supraselar homogénea, Gd+++ y quistes





### T. Supratentoriales

Gliomas bajo y alto grado  
Ependimoma  
ETMR  
Tumores plexos coroideos  
Tumores glioneuronales

### T. Selar-Diencefálica

Craneofaringioma  
Tumores pineales  
Tumores germinales  
Glioma diencefálico

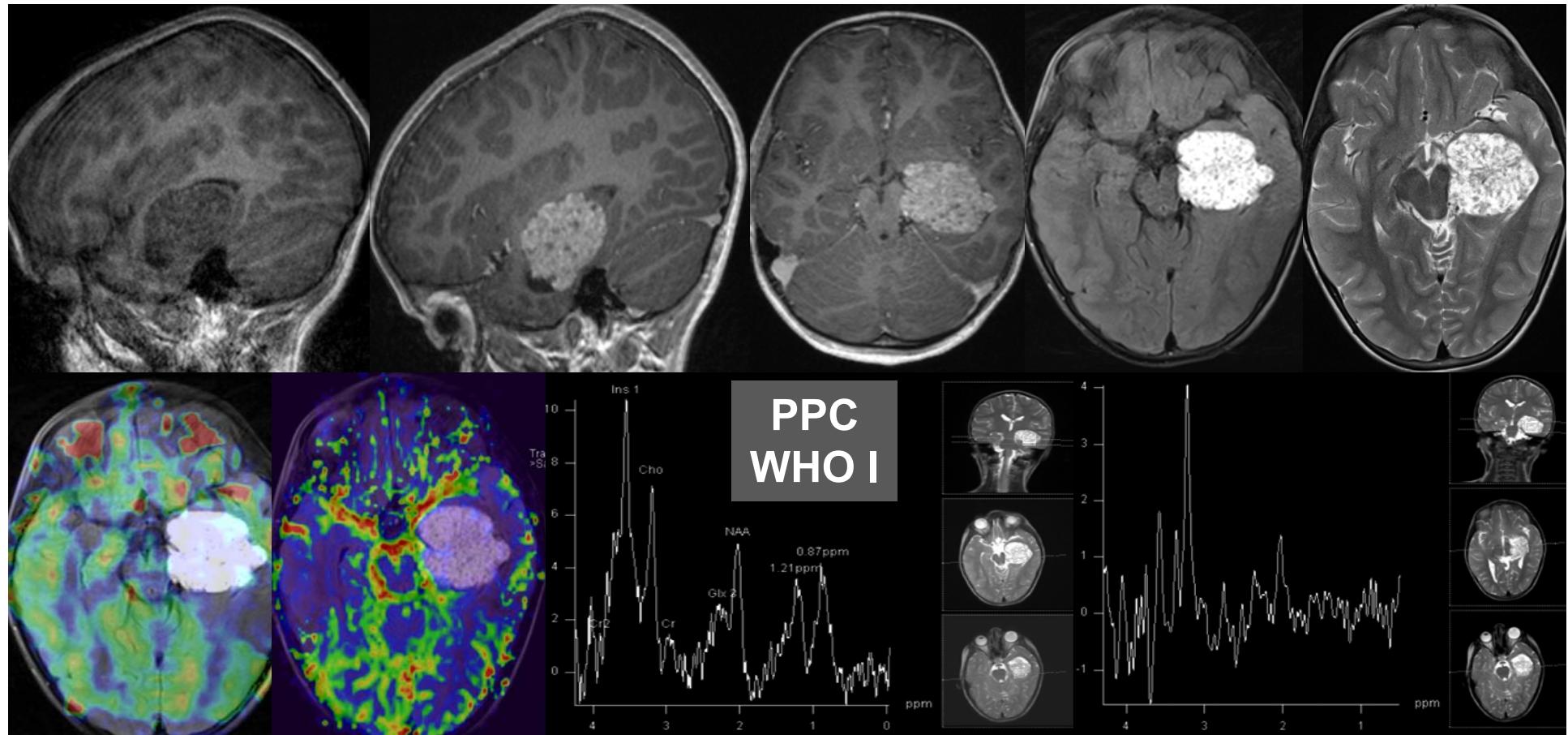
### T. Infratentoriales

Astrocitoma Cerebeloso  
Meduloblastoma  
Ependimoma  
Glioma troncoencefálico



# Tumores plexos coroideos

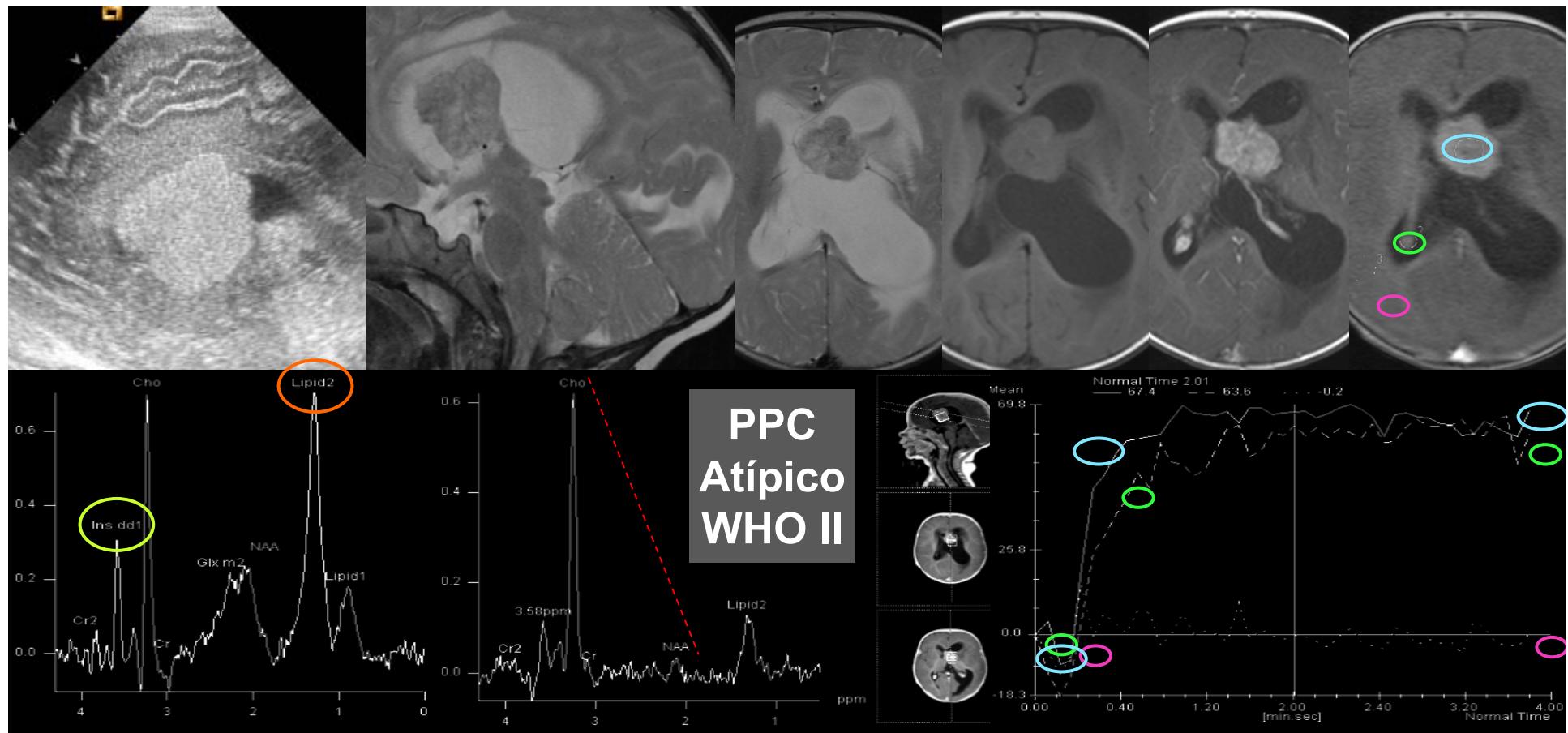
- Papiloma WHO I, Papiloma Atípico WHO II y Carcinoma WHO III
- 3-5% T. intracraneales pediatría. PCC → T. intraV (**atrio**) +frec <5a
- PPC: Masa polilobulada “**coliflor**” delimitada Gd+++ e hidrocefalia





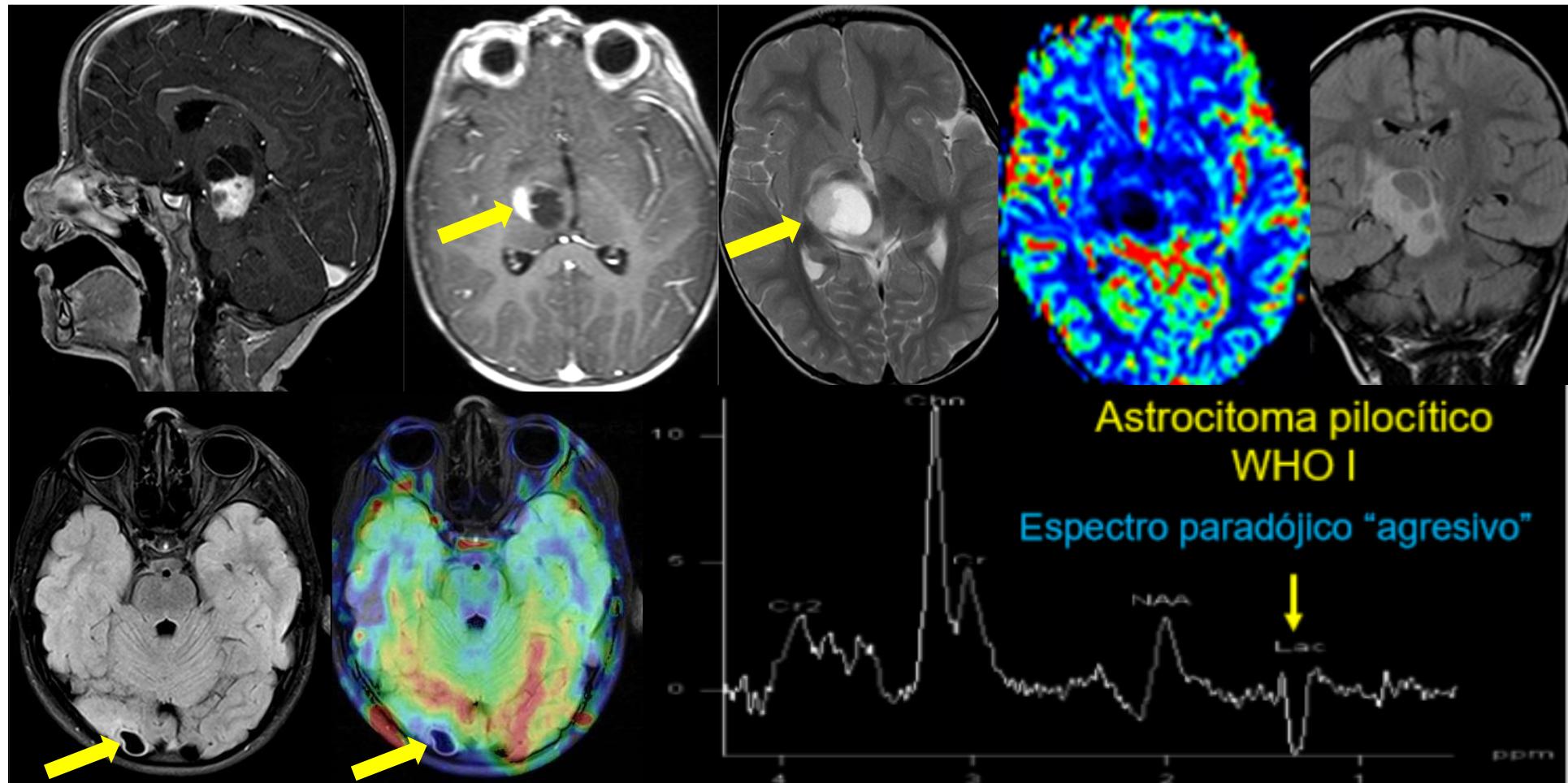
# Tumores plexos coroideos

- Papiloma WHO I, Papiloma Atípico WHO II y Carcinoma WHO III
- 3-5% T. intracraneales pediatría. PCC → T. intraV (**atrio**) +frec <5a
- PPC: Masa polilobulada “**coliflor**” delimitada Gd+++ e hidrocefalia



# Astrocitoma pilocítico

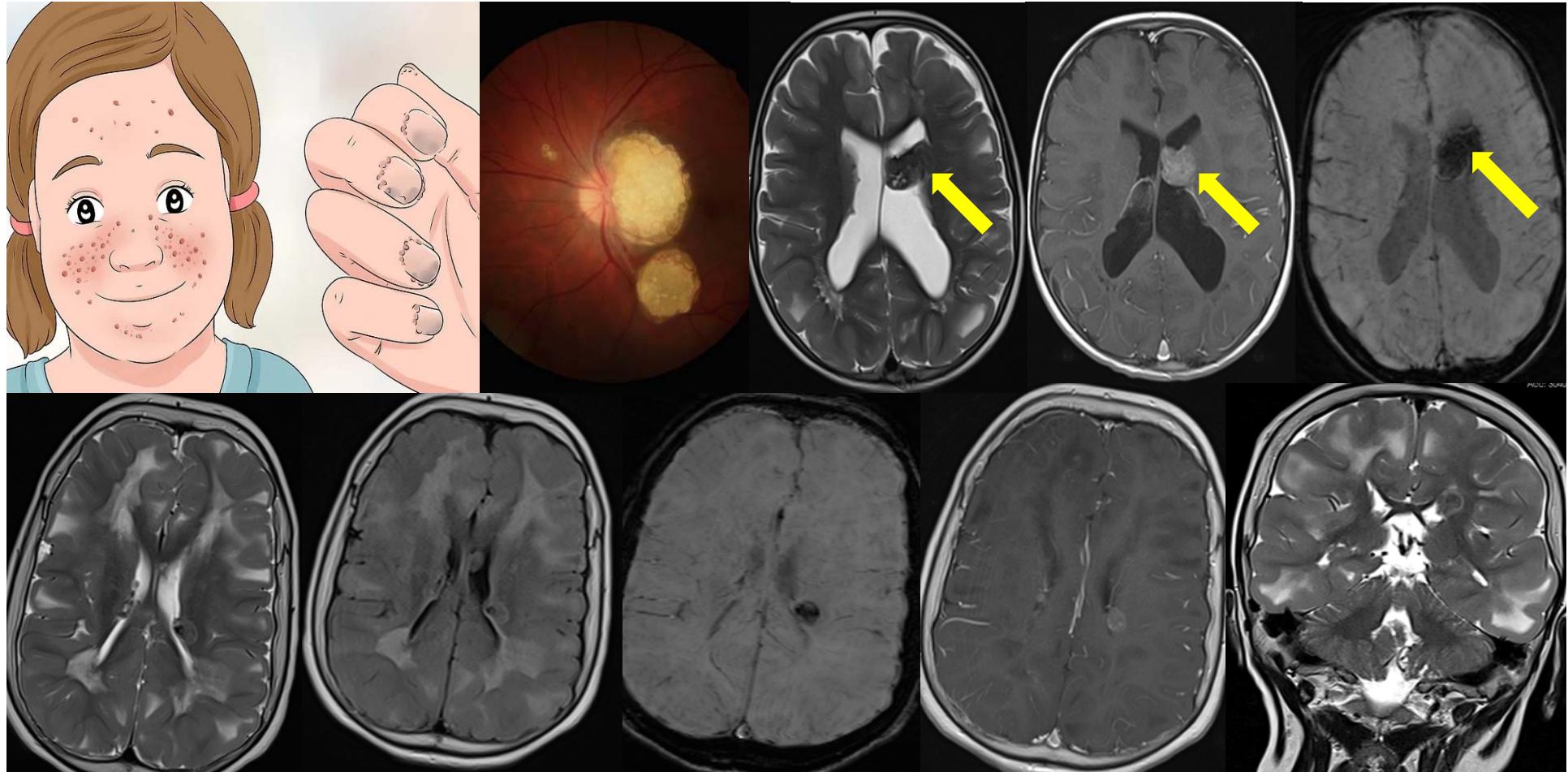
- WHO I. Tumor cerebral 1º + freq en la infancia. **BRAF**
- 75% en las 2 primeras décadas de la vida. **Excelente pronóstico**





# SEGA

- Astrocytoma subependimario de células gigantes. WHO I. <20a
- Sólo en ET (5-15%). <10mm Monro → hidrocefalia. Calcio y Gd+

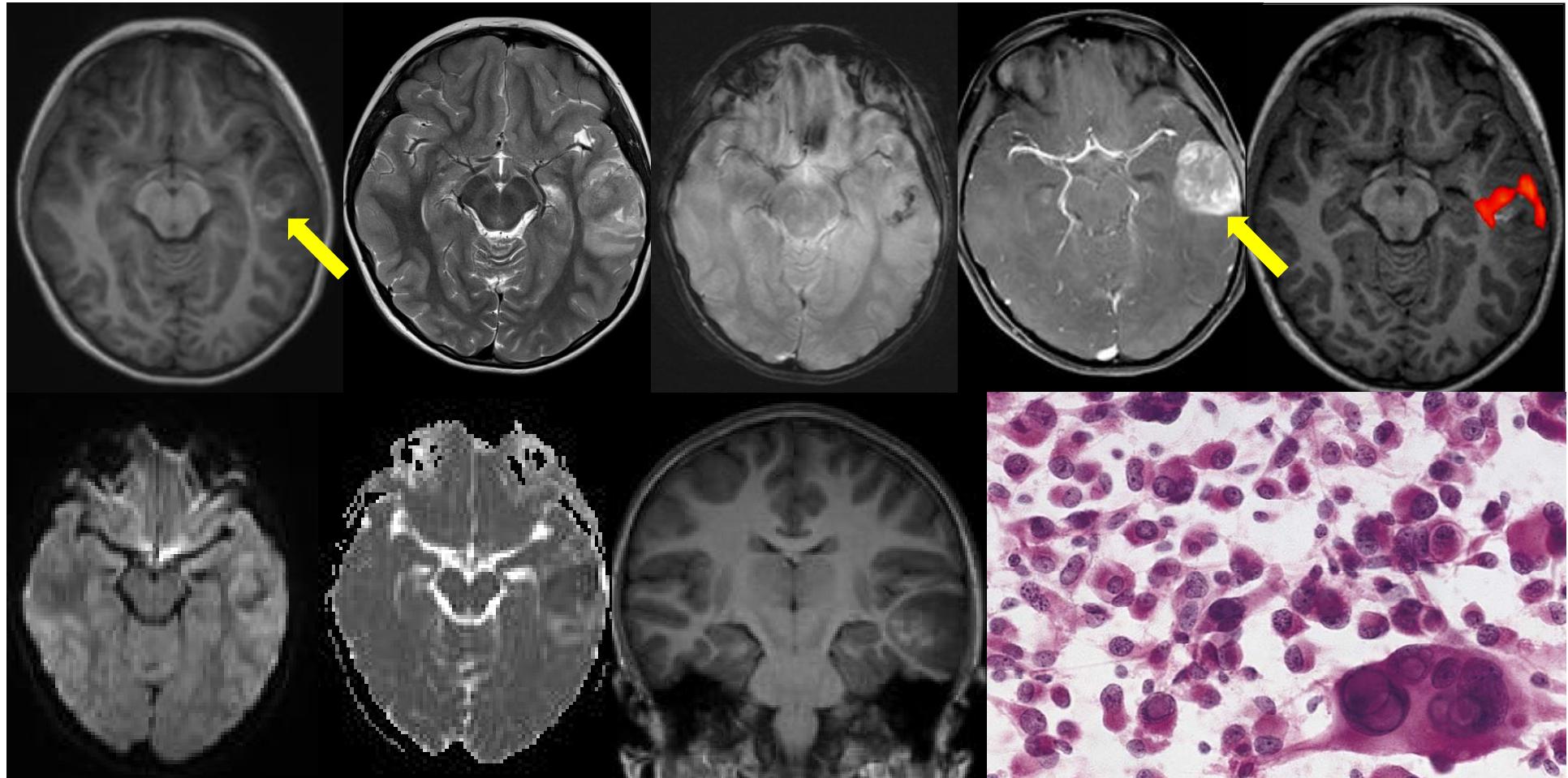


DDX Heterotopia subependimaria



# Xantoastrocitoma pleomórfico

- WHO II. Raro (1%). >10a. Epilepsia **Temporal** (75%) >Front >Pariet
- Cortical **Hiper T1** Sólido Q+/- Gd ++ **Cola dural** (70%) Erosión ósea

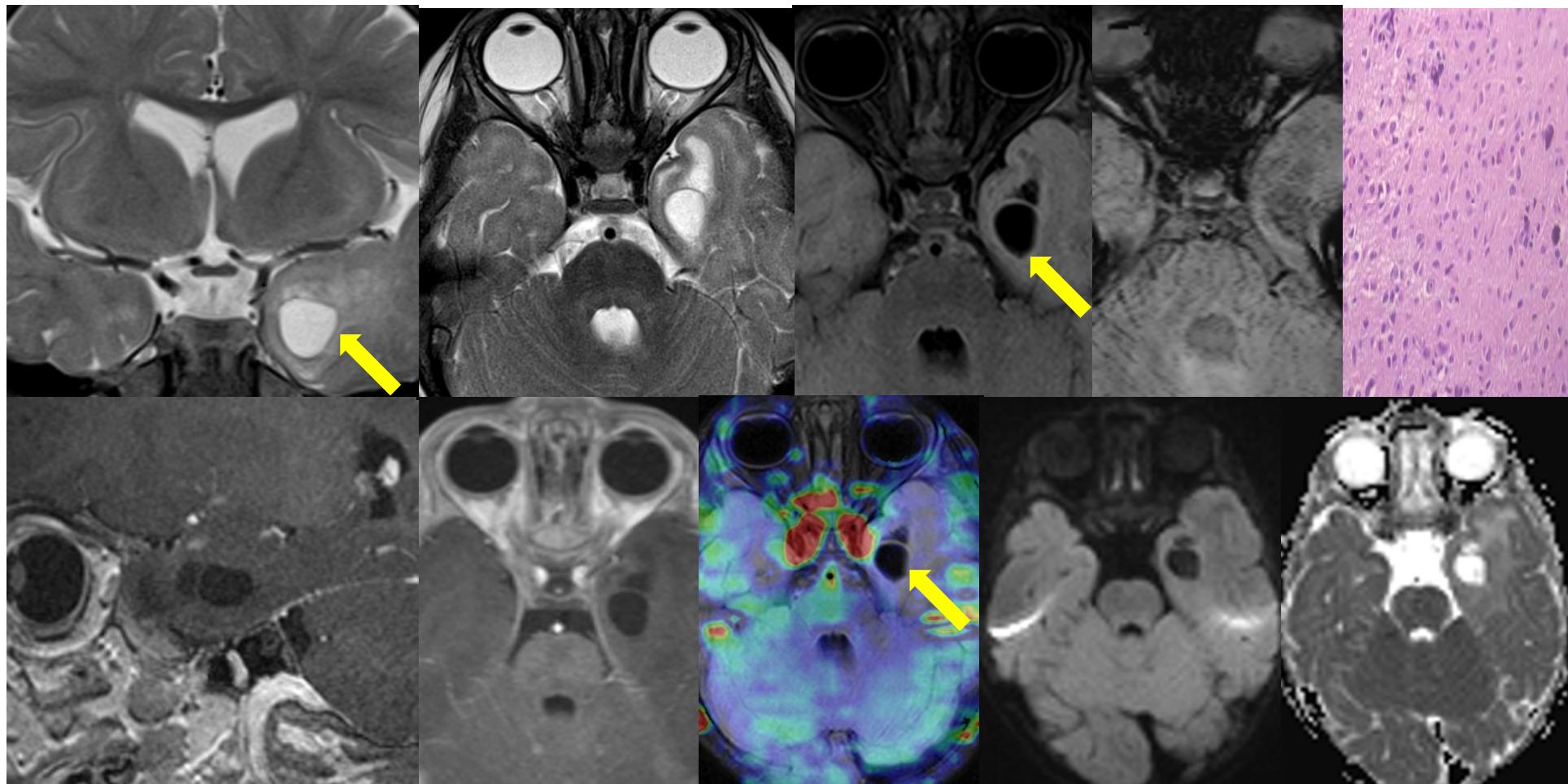




# Ganglioglioma

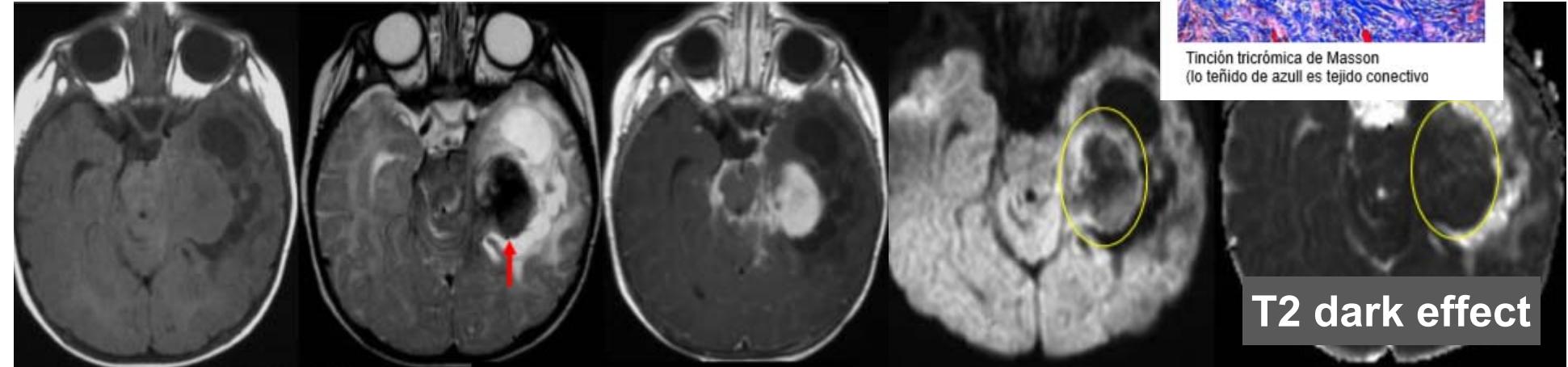
# Gangliocitoma

- WHO II. 10% T. 1os pediatría. BRAF Epilepsia **Temporal**. Hipotál.
- Lesión heterogénea, **infiltrante**, sangre, Ca+, Q+, Gd+/-



# DIG

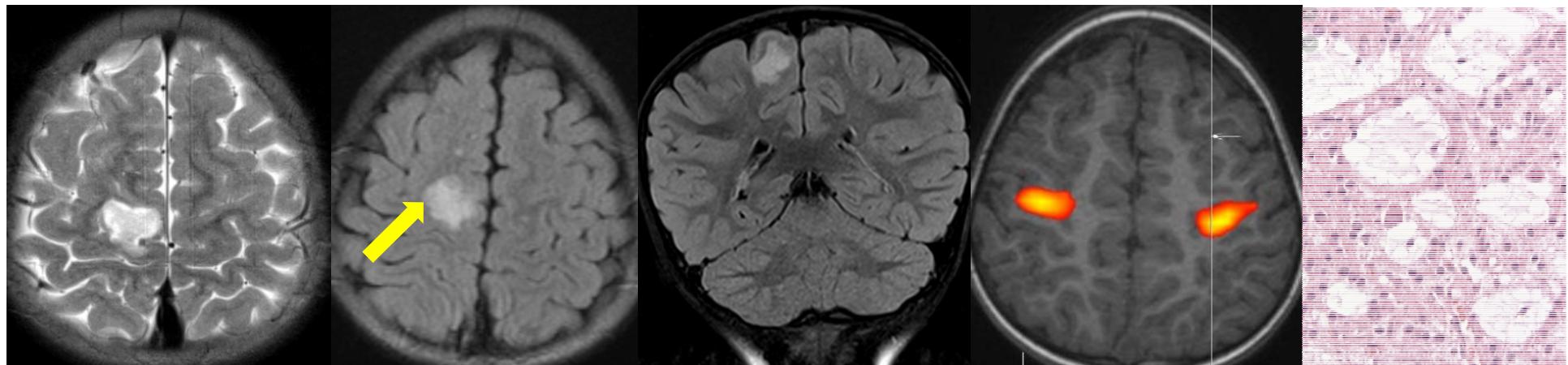
- Desmoplásico Infantil Ganglioglioma. WHO I-II



# DNET

Multinodular and Vacuolating tumor

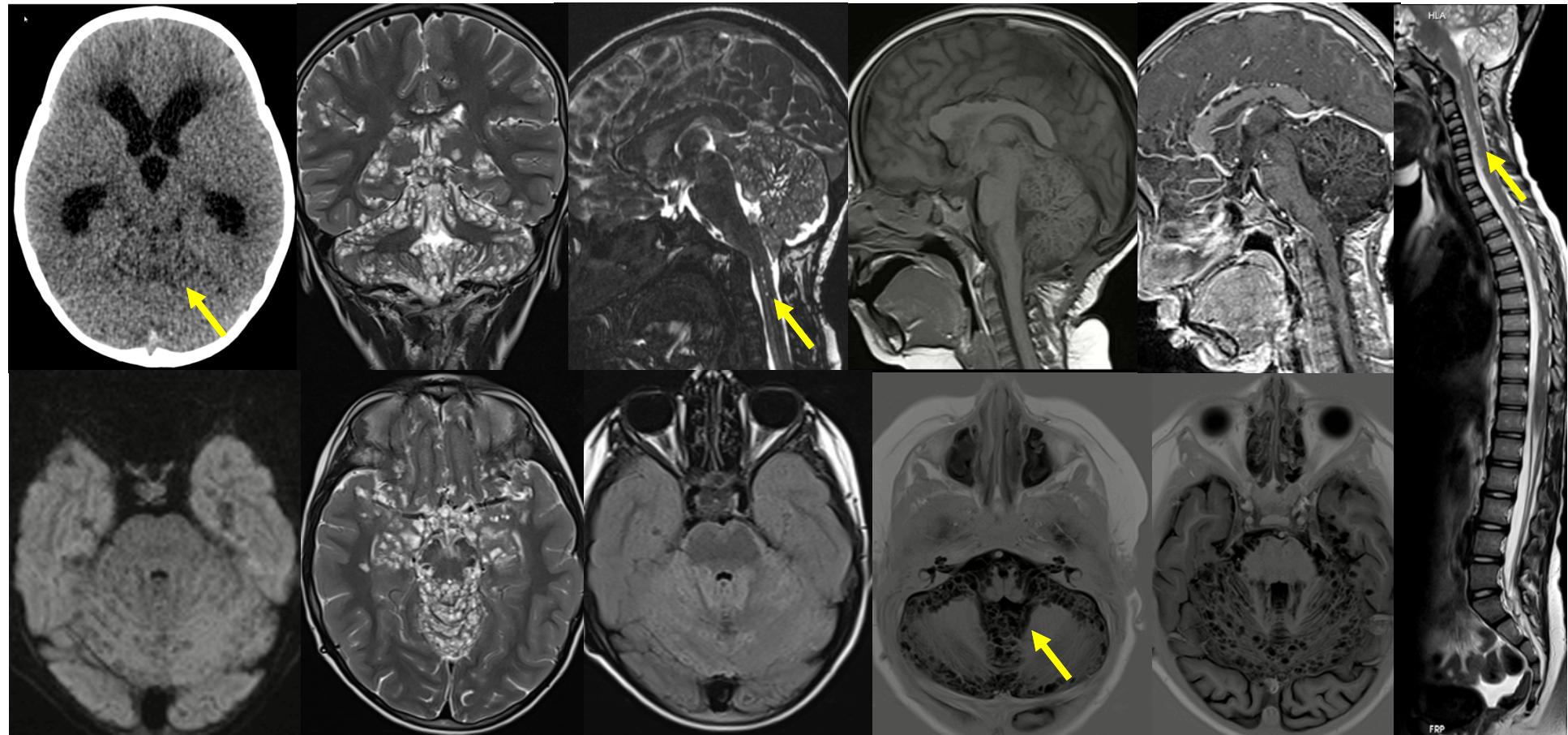
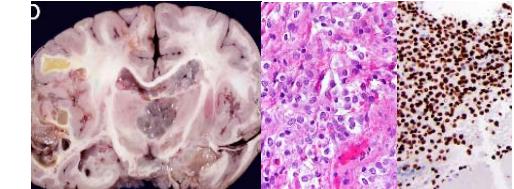
- T. Disembrioplásico NeuroEctodérmico WHO I **Bubbly** Halo FLAIR





# T.difuso glioneuronal leptomeníngeo

- Tumor quístico-mucoide extensión LCR a través de **Virchow-Robin** al espacio SA, intraV e intraxial → hidrocefalia
- Citología similar al oligodendroglioma. **OLIG2 +**

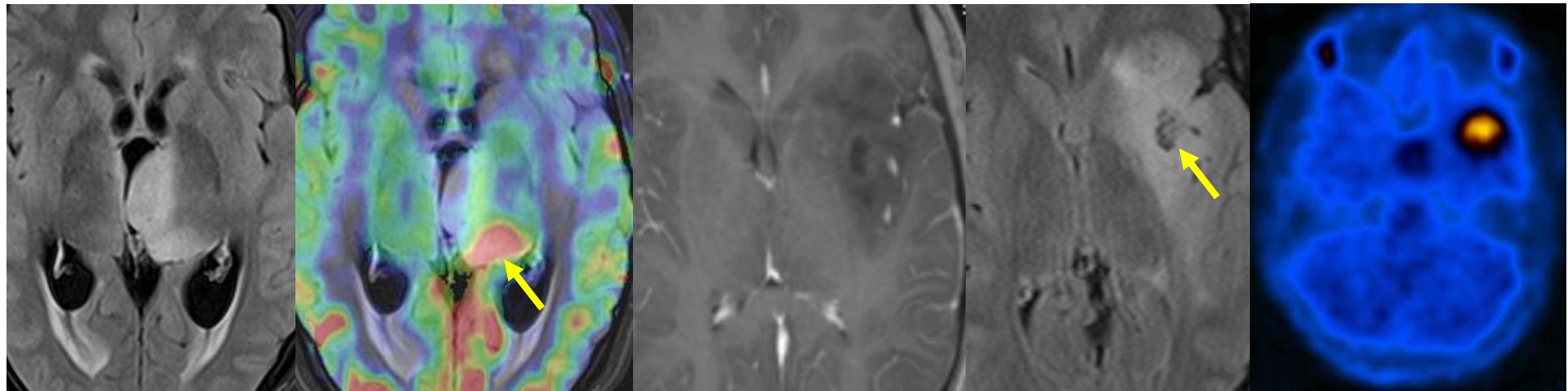


Gardiman MP, et-al. Diffuse leptomeningeal glioneuronal tumors: a new entity?. Brain Pathol. 2010;20 (2): 361-6



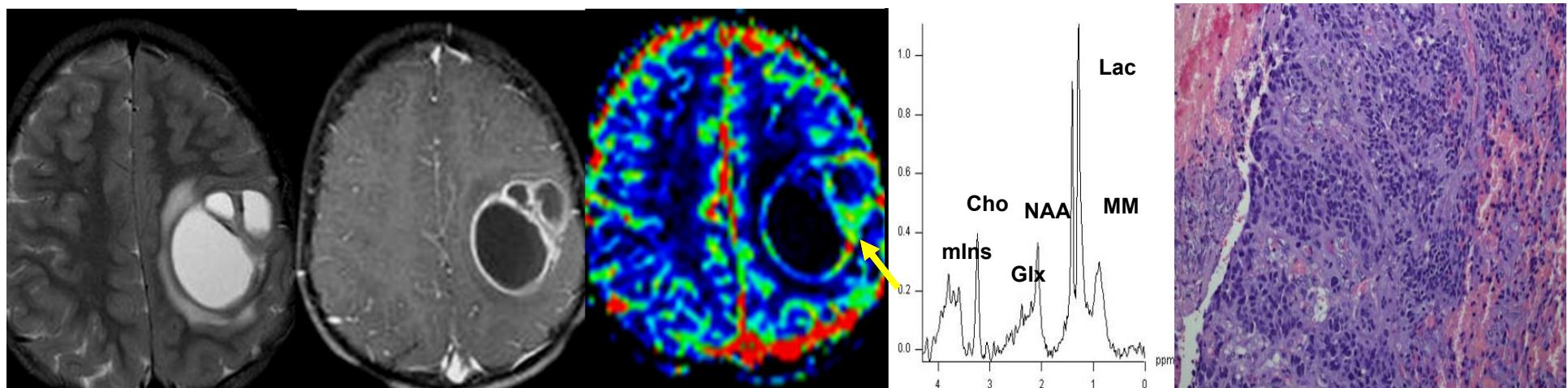
## Astrocitoma anaplásico

- WHO III. Lesión **infiltrativa** mal definida. Gd+/-, DWI +/-, **PWI+**



## Ependimoma anaplásico

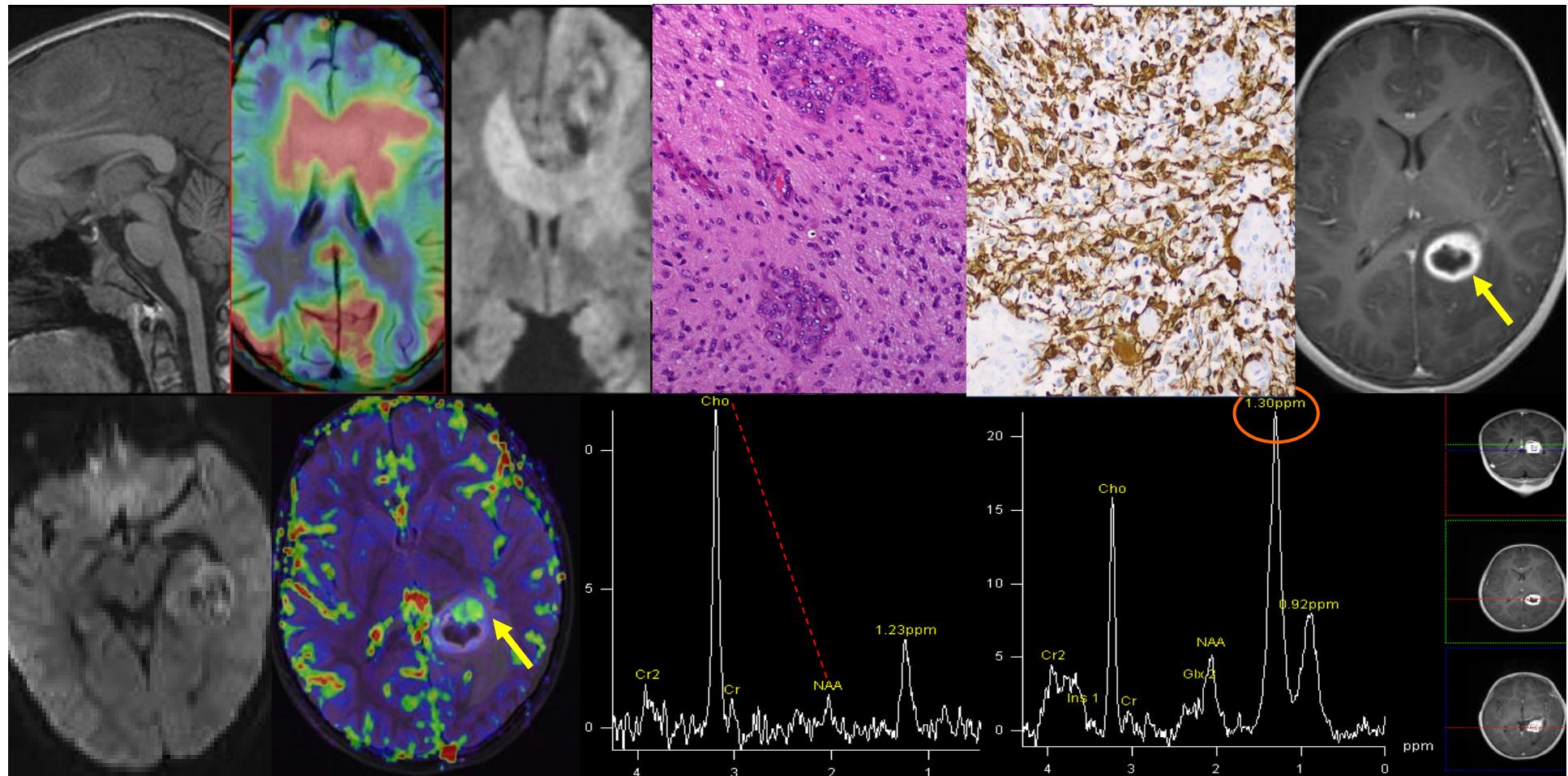
- WHO III. Gran lesión **heterog** sólido-quística **Gd+** Cerca ventrículo





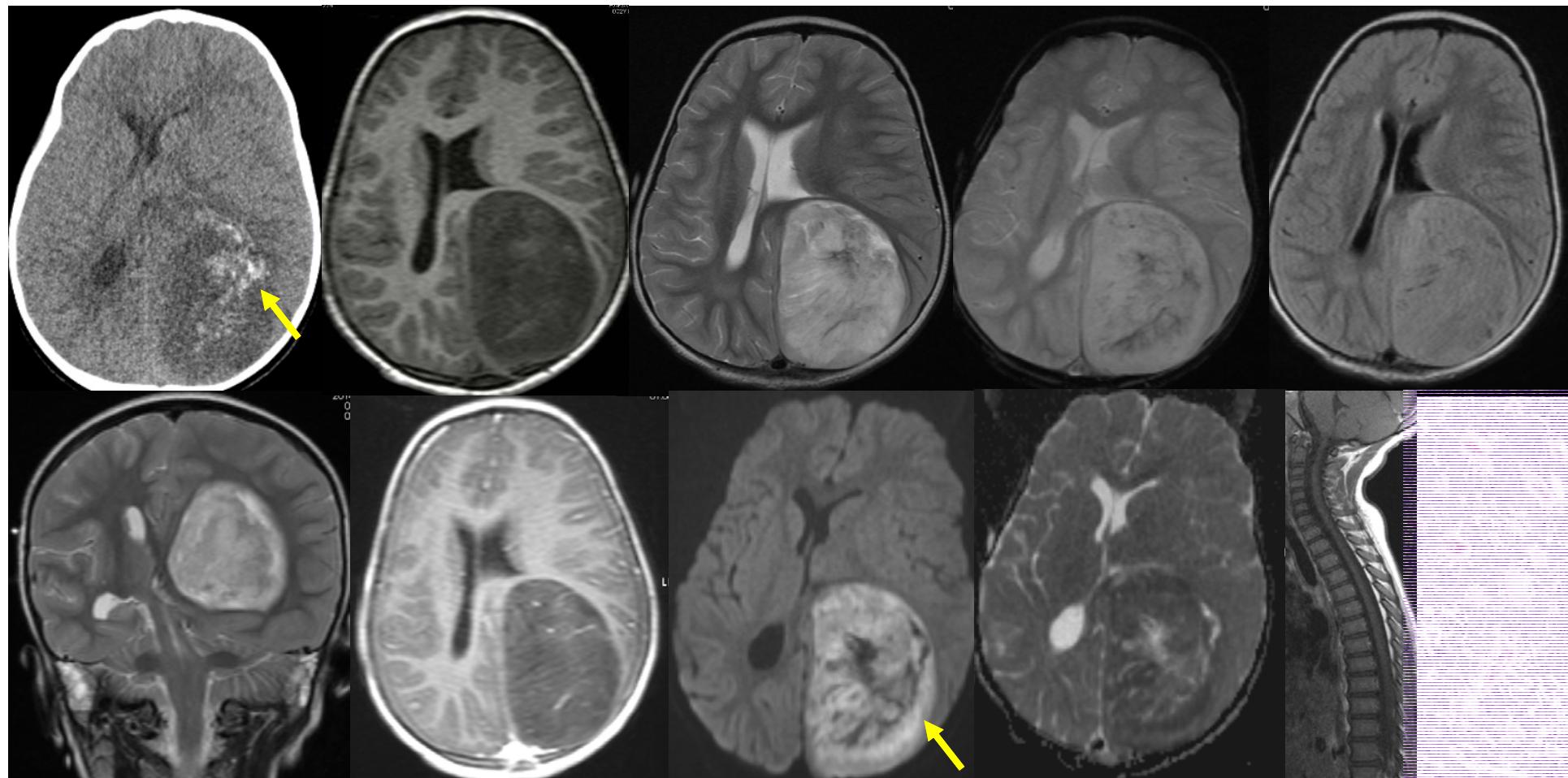
# Glioblastoma

- WHO IV. Poco frecuente. **Células gigantes**. Invasión CC y GGBB
- Masa heterogénea, agresiva, mal definida, **necrosis**, sangre, Gd+



# ETMR

- T. Embrionario Multicapas Rosetas: PNET + ETANTR + Ependimobl
- WHO IV. <2-4a. SupraT. **C19MC** amplificación en el cr. 19q. Mal px
- No formaciones epiteliales y sí rosetas ependimoblásticas





Vall  
d'Hebron  
Barcelona Hospital Campus

IDI  
INSTITUT DE  
DIAGNÒSTIC PER  
LA IMATGE

XVI CURSO NACIONAL DE  
NEURORRADIOLOGÍA



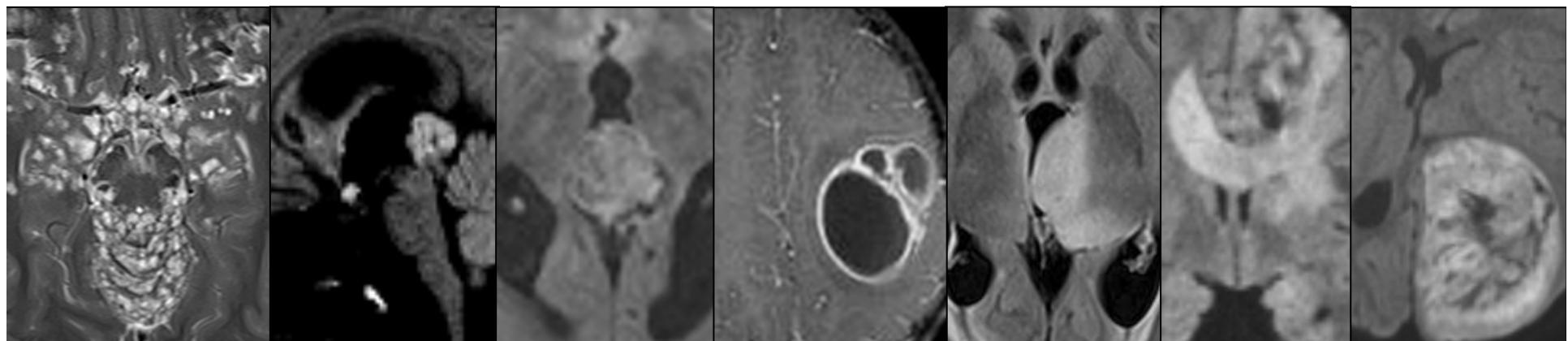
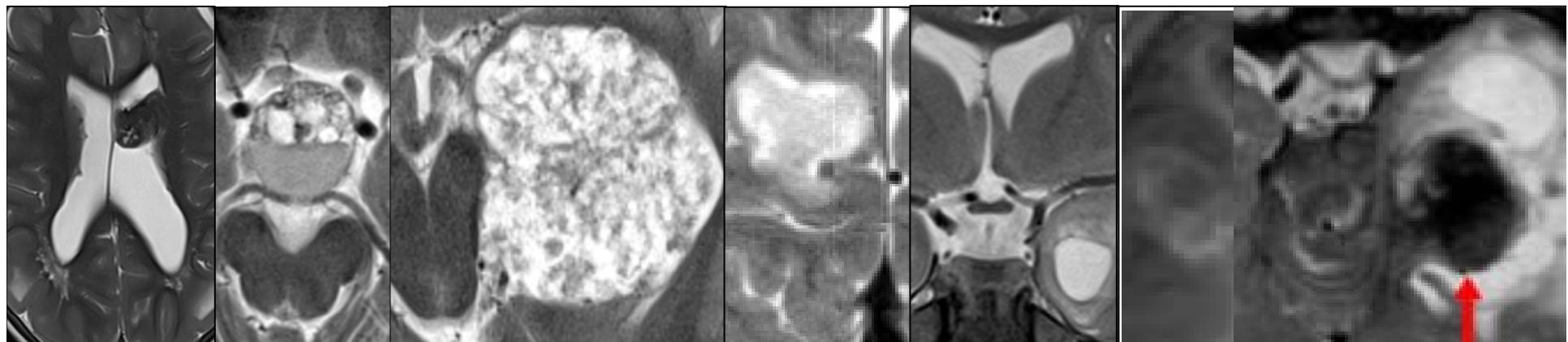
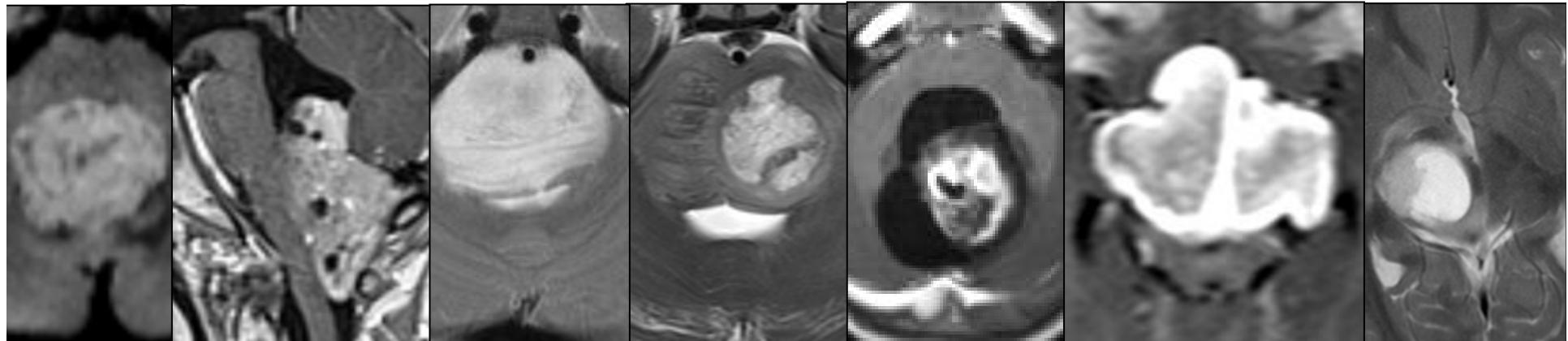
# Blackjack



Vall  
d'Hebron  
Barcelona Hospital Campus

IDI  
INSTITUT DE  
DIAGNOSTIC PER  
LA IMATGE

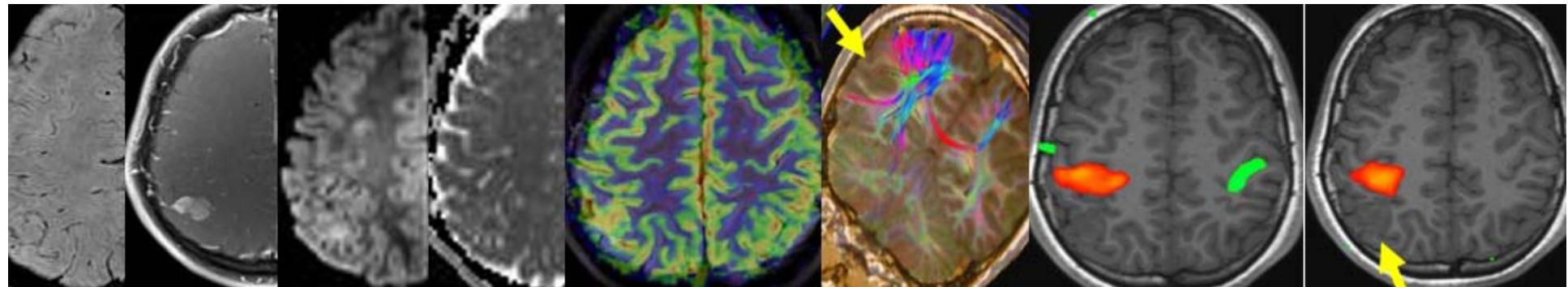
SENR  
XVI CURSO NACIONAL DE  
NEURORRADIOLOGIA





# Conclusiones

- Trabajo en equipo: onco, neurocir, neurología, UCI, RT y AP
- Síndromes prooncogénicos y facomatosis. TC sin contraste
- RM: técnica prínceps en el manejo onconeuroradiológico
- Si sospecha de tumor de alto grado → T1Gd y DWI espinal
- Imagen multimodal → info estructural, bio, fisio y metabólica
- Radiólogo clínico, patólogo y ahora también...  
... genetista y molecular





XVI CURSO NACIONAL DE  
NEURORRADIOLOGÍA  
*Neurorradiología en la Patología Tumoral Cerebral*



S.E.N.R.  
Sociedad Española  
de Neurorradiología

20-21  
febrero  
2020  
Madrid



# ¡Muchas gracias por vuestra atención!

Sección de Neurorradiología Pediátrica  
Hospital Vall d'Hebron – Área Materno-Infantil  
Barcelona  
angel.sanchez.idi@gencat.cat

