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FUNDACIÓN CARDIOINFANTIL



**FUNDACIÓN
CARDIOINFANTIL**
INSTITUTO DE CARDIOLOGÍA



**Hospital
Simón Bolívar**

CASO CLÍNICO

MOTIVO DE CONSULTA Y ENFERMEDAD ACTUAL

Paciente de 4 meses remitida de la consulta externa de neurología por cuadro clínico congénito consistente en proptosis izquierda de aumento progresivo asociado a opacidad corneal y quemosis. Madre niega otra sintomatología.

ANTECEDENTES

- ▶ Parto por cesárea
- ▶ Gestación a término 39 semanas
- ▶ No antecedentes prenatales de importancia
- ▶ Madre y hermanos con múltiples manchas café con leche más de 5 de diámetro mayor 2 cm en tórax, niega otros.
- ▶ Refiere estudios neuro imagenológicos familiares negativos.
- ▶ No estudios genéticos

REVISION POR SISTEMAS: Niega otros

EXAMEN FÍSICO

- ▶ TA 98/46
- ▶ FC: 144 x”
- ▶ FR: 30
- ▶ GCS: 15/15
- ▶ PC 37.5 CM

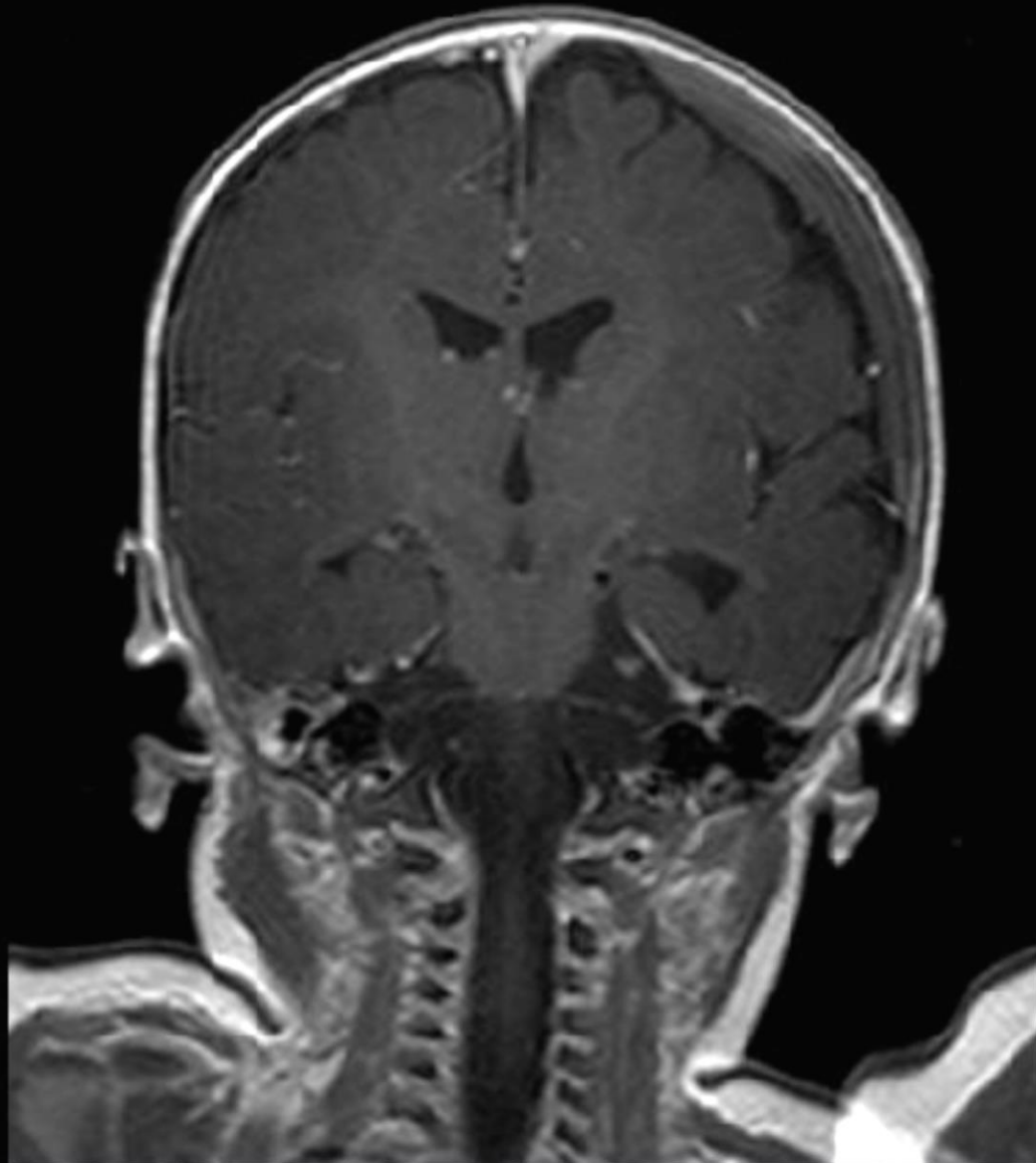
- ▶ CABEZA Y CUELLO: sin alteraciones, no signos meníngeos.
- ▶ ABDOMEN: Normal
- ▶ NEUROLOGICO: sin déficit aparente.
- ▶ PIEL Y FANERAS:

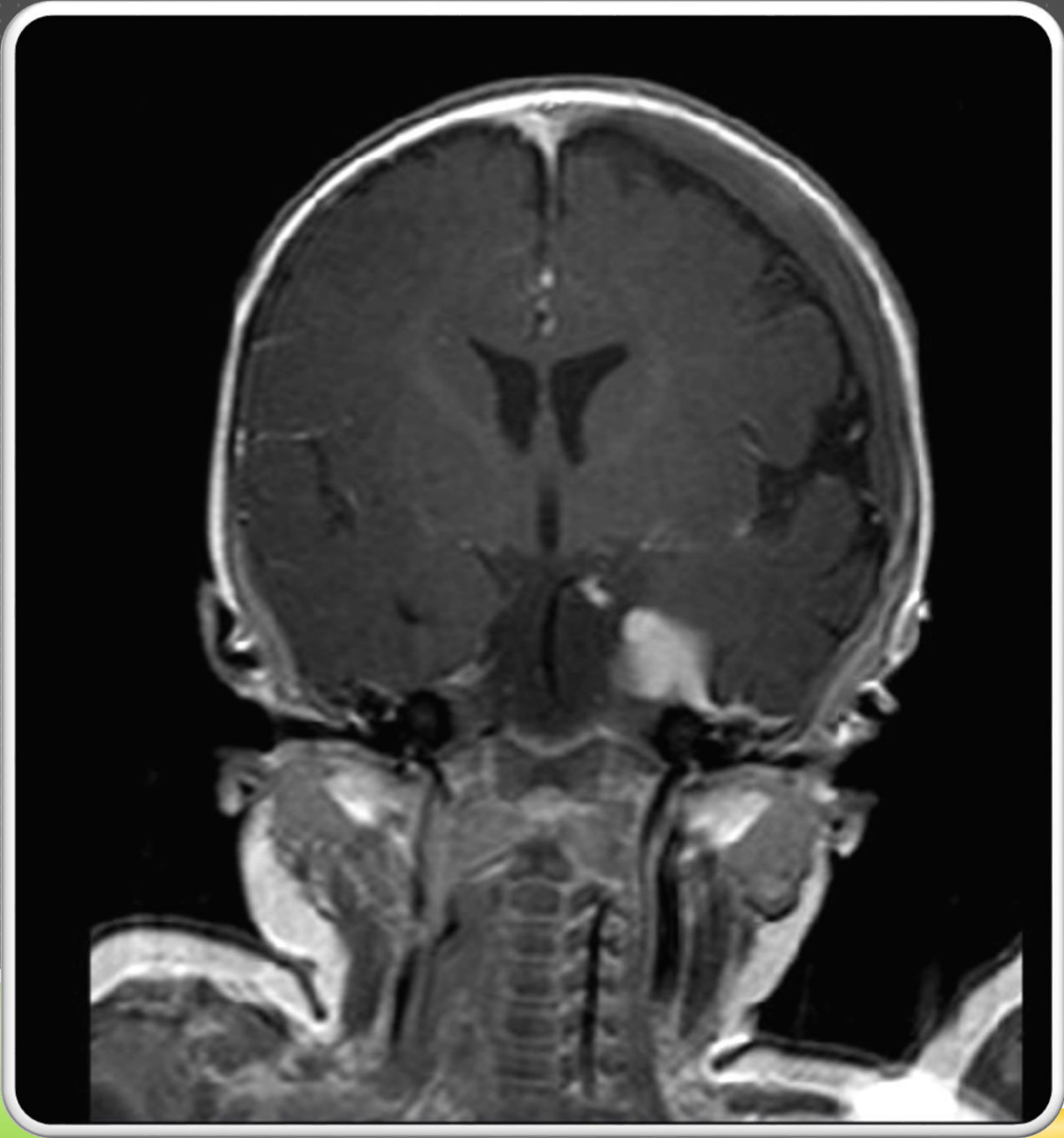
EXAMEN FÍSICO OFTALMOLÓGICO

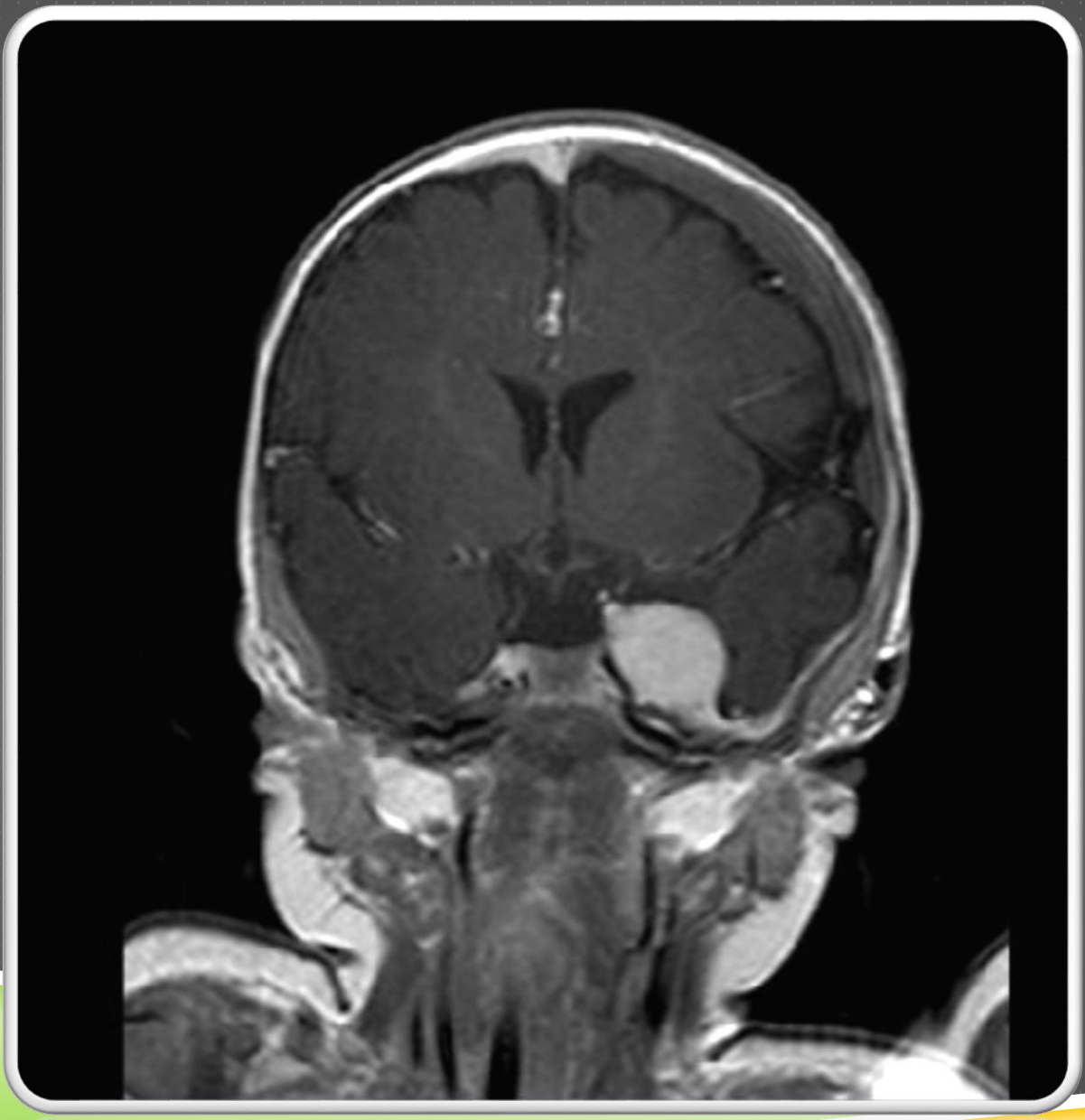
- ▶ OJO DERECHO: Conjuntiva sana, cámara anterior normal, pupila de 3mm reactiva, fondo de ojo normal
- ▶ OJO IZQUIERDO: Proptosis severa que condiciona a BUFTALMOS, aumento del diámetro corneal, edema corneal difuso con queratitis difusa, cámara anterior formada, pupila midriática 6 mm areactiva, PIO aumentada, fondo de ojo no visualizado.



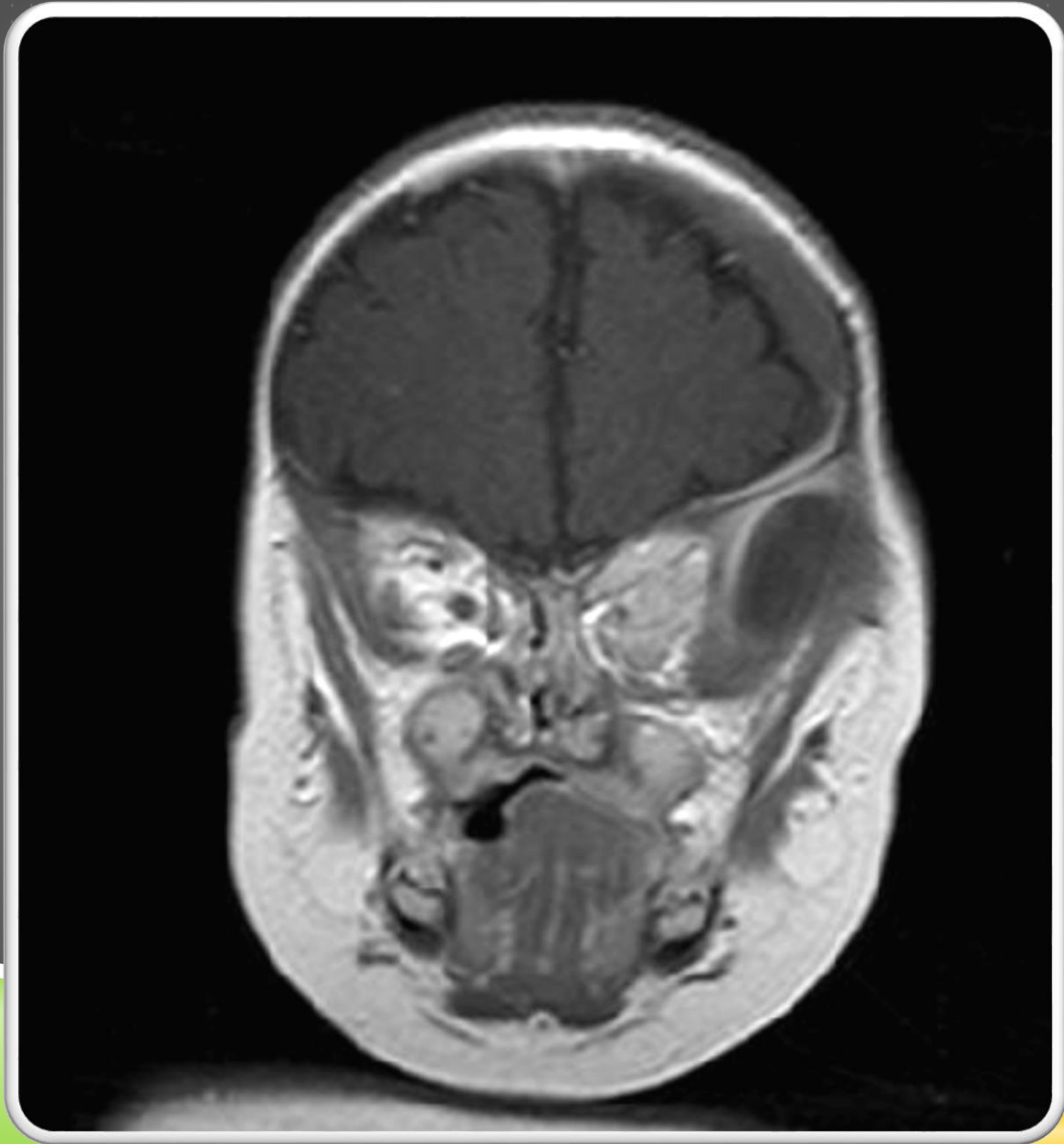


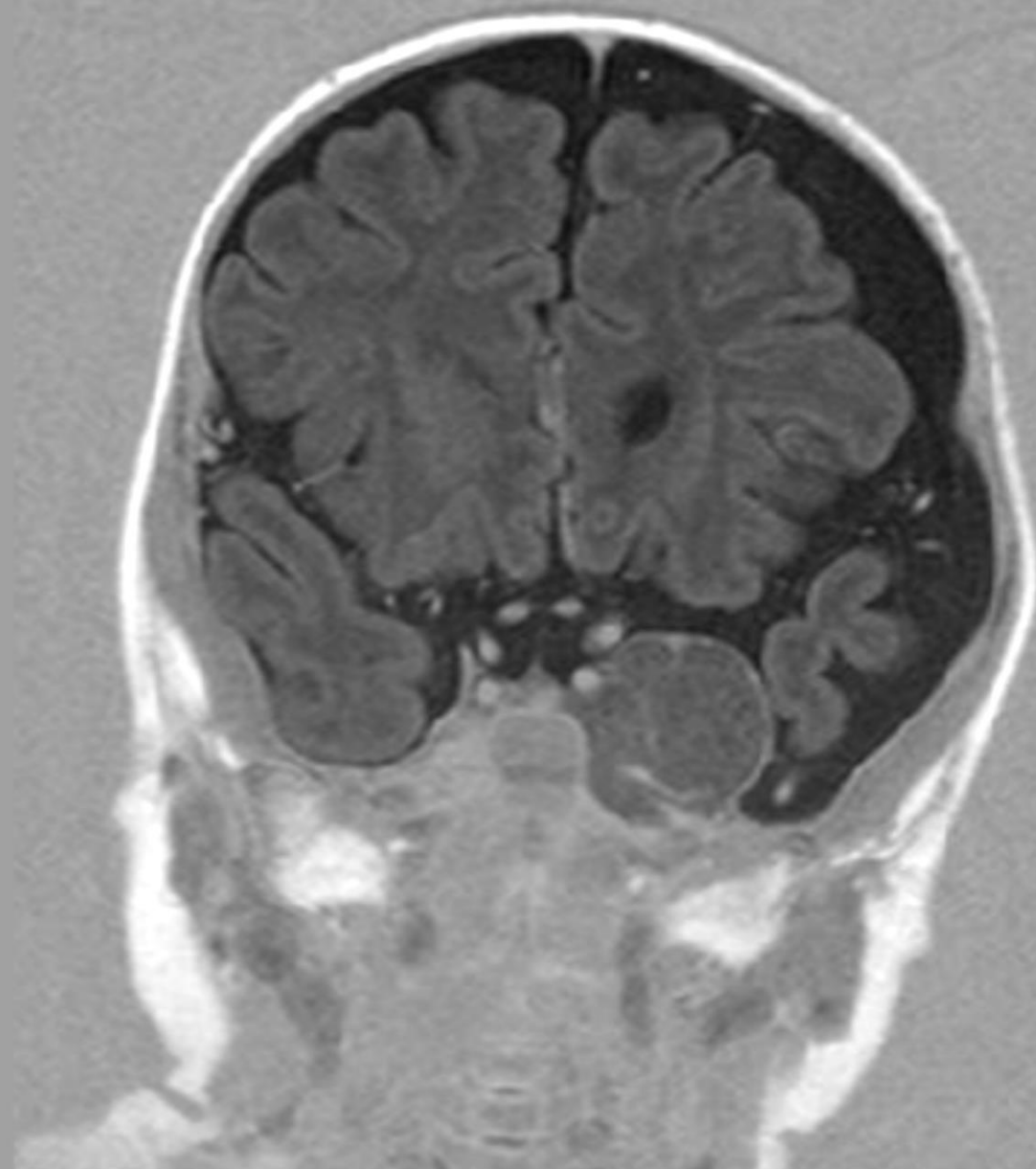


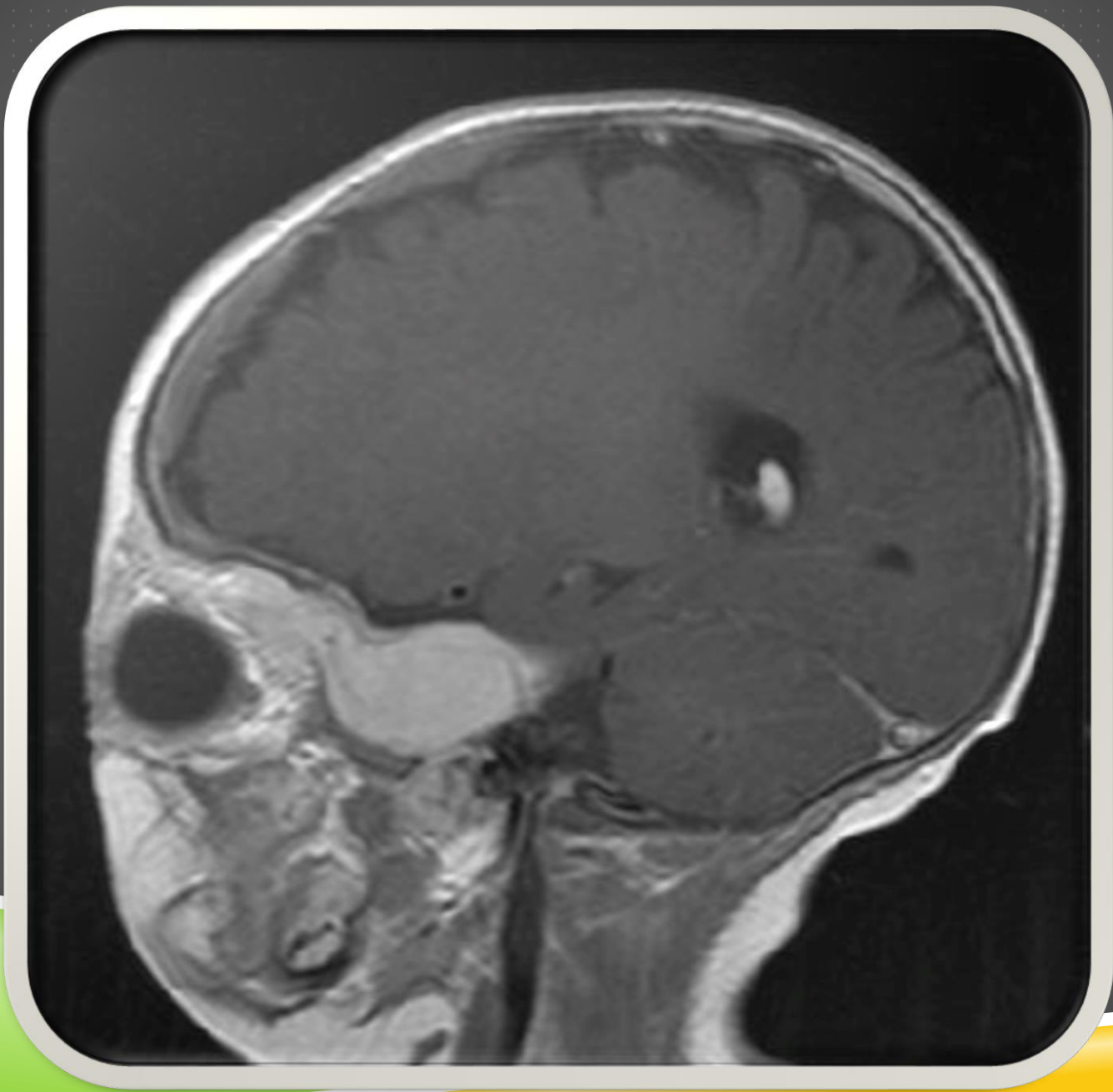


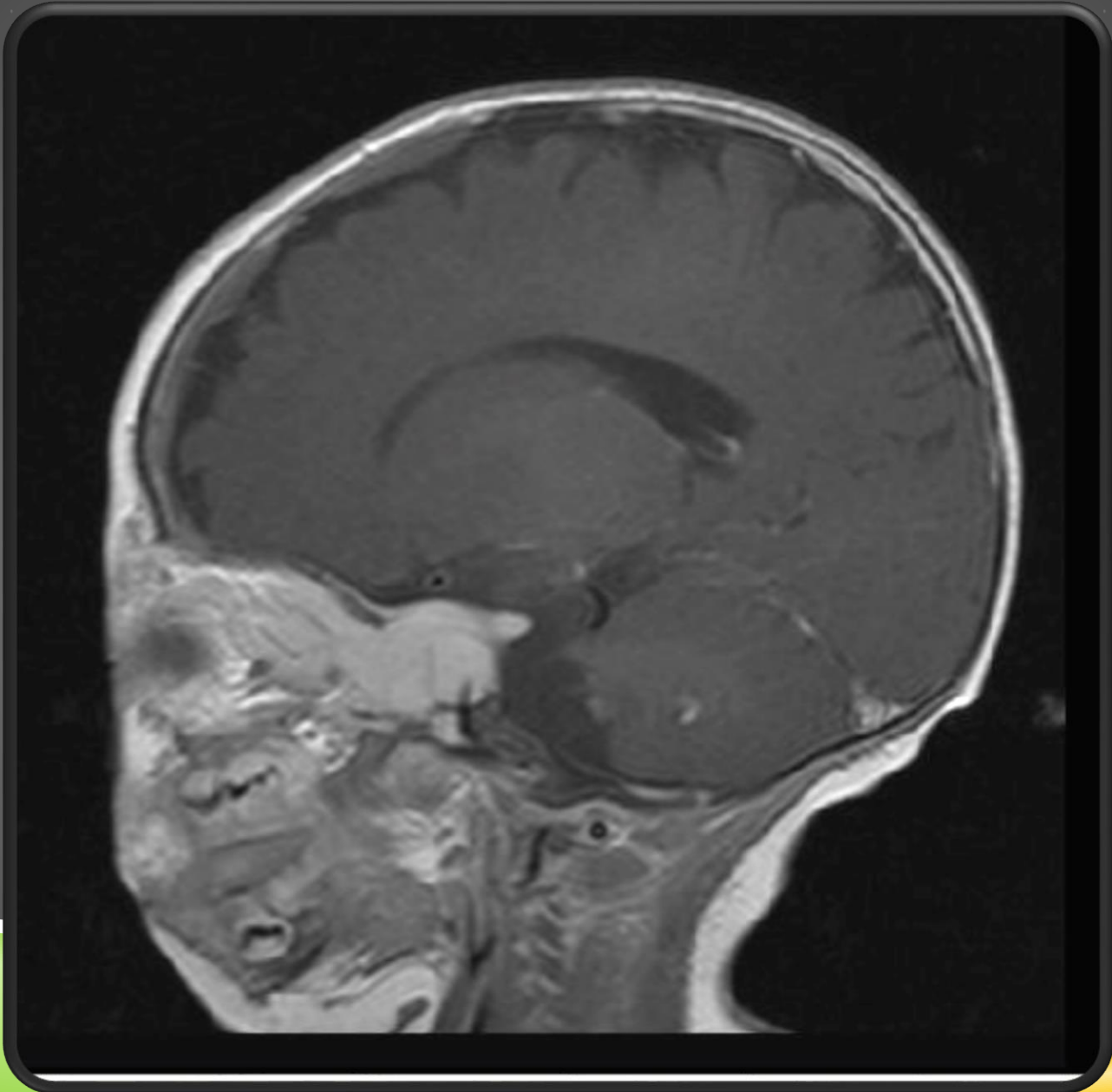


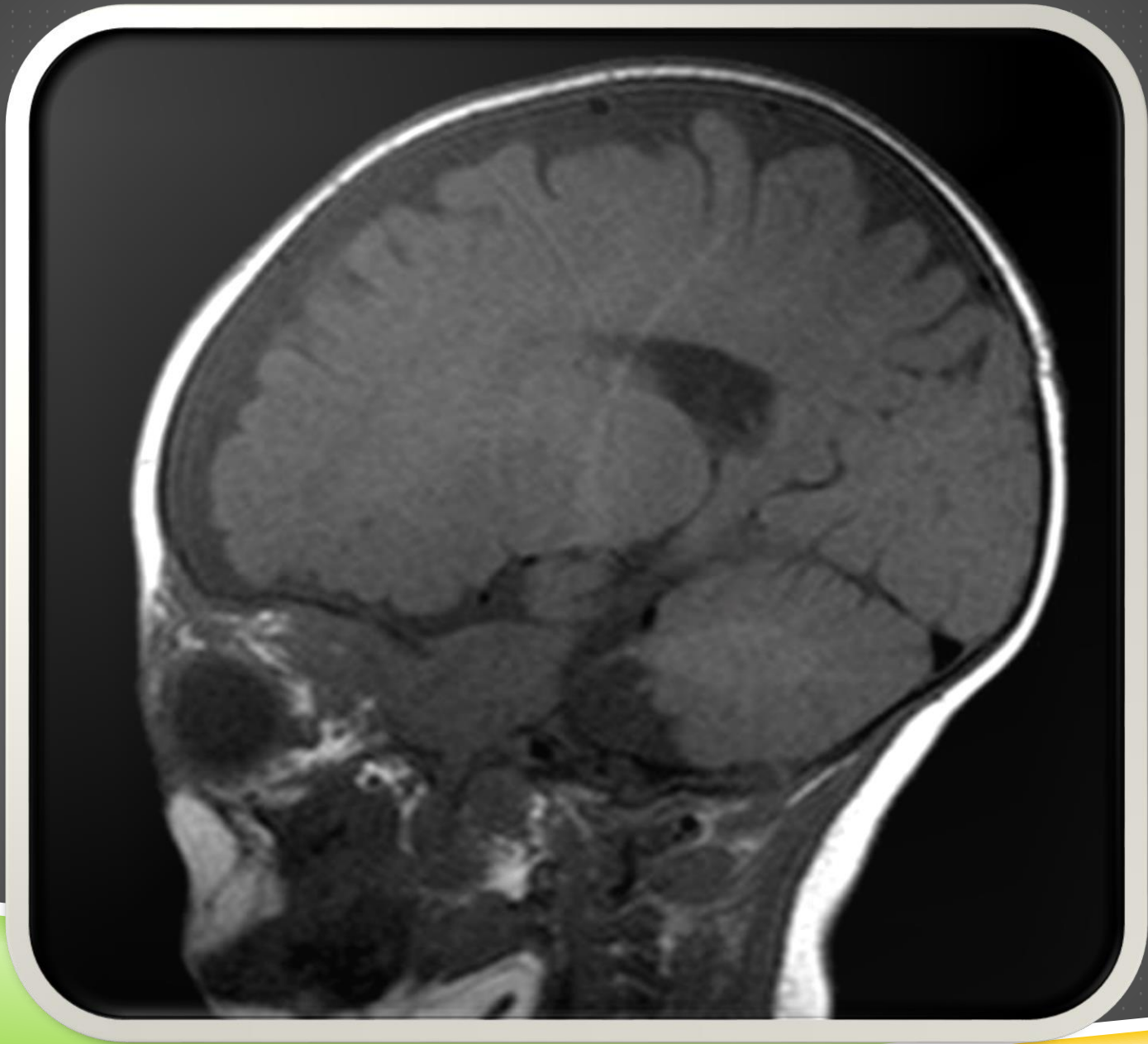




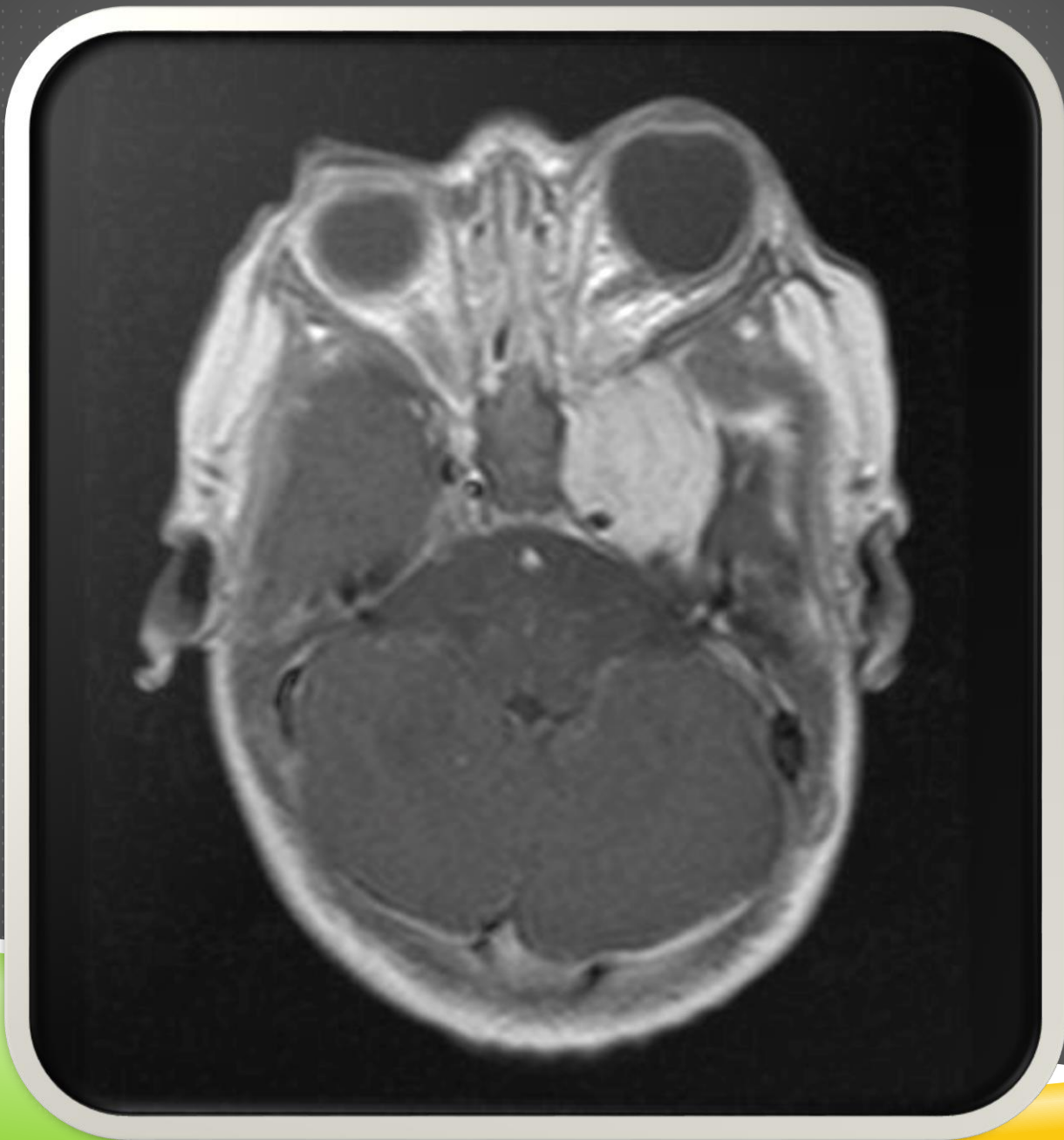


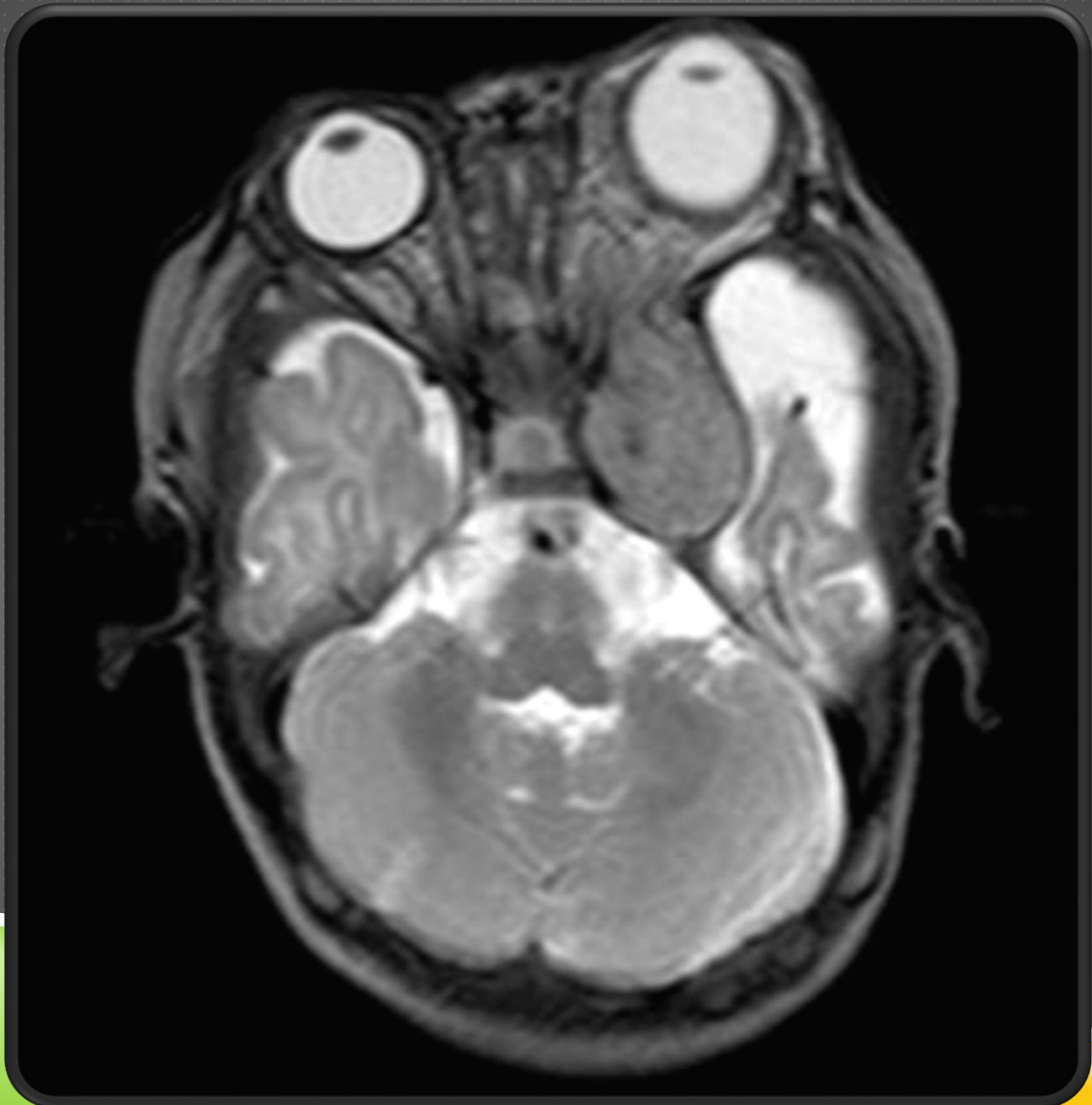












¿POSIBILIDADES
DIAGNÓSTICAS?

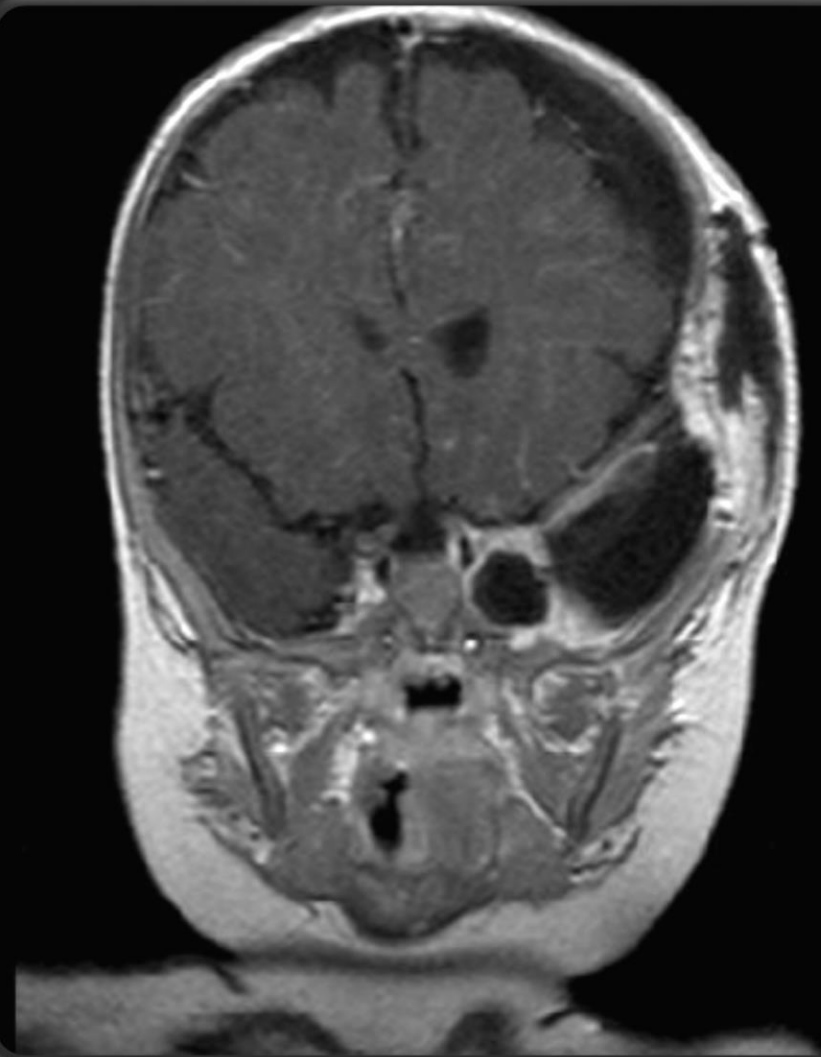
¿OPCIONES
TERAPEUTICAS?

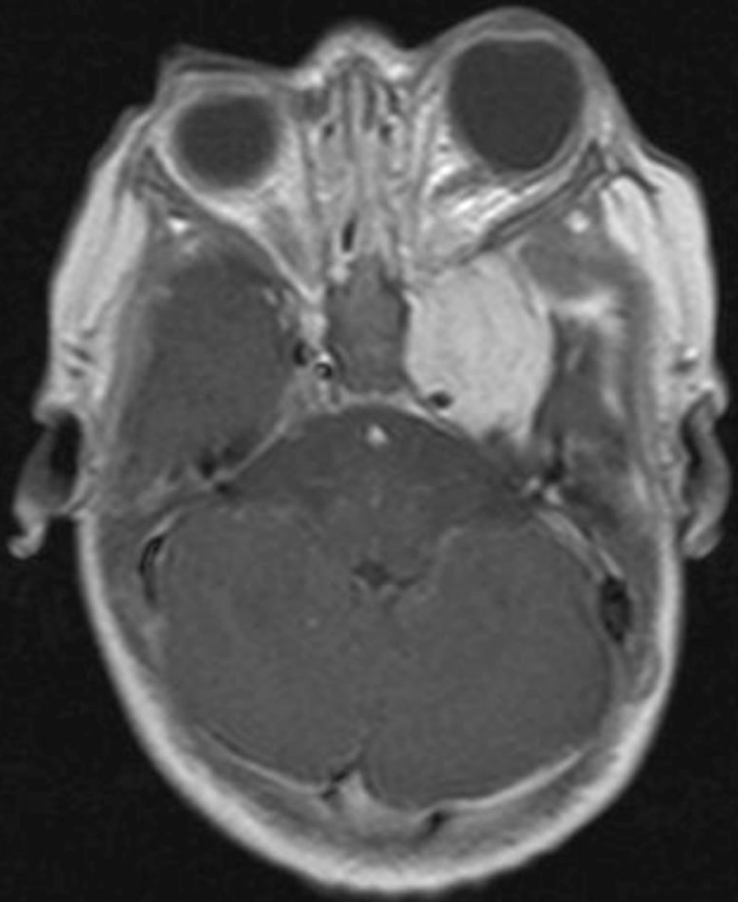
¿CIRUGÍA?

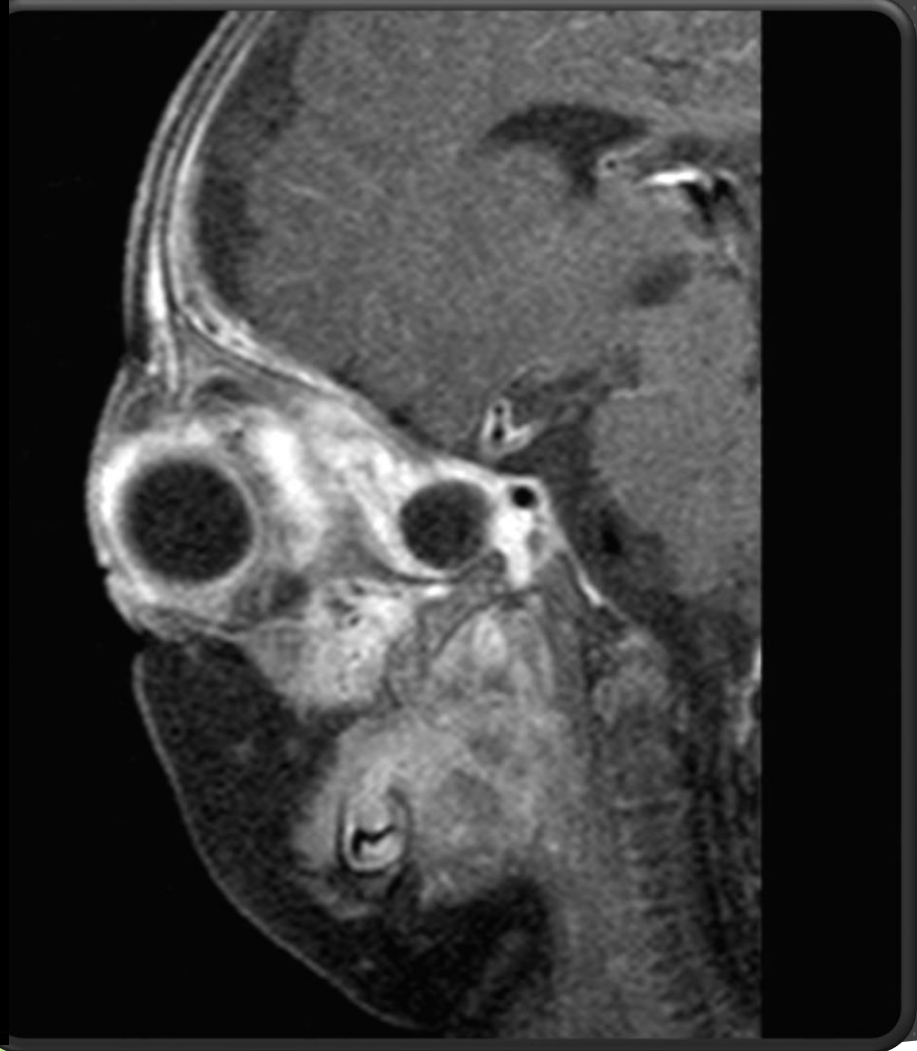
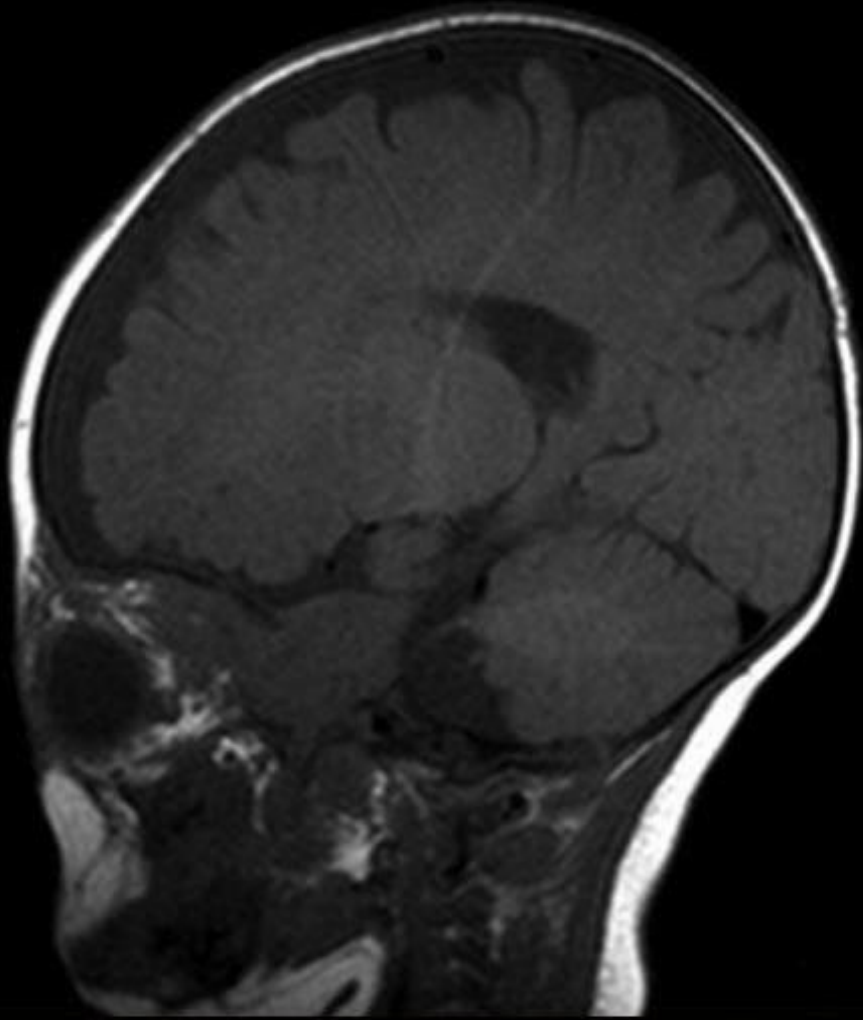
¿RADIOTERAPIA?

¿RADIOCIRUGÍA?

IMÁGENES POSOPERATORIO







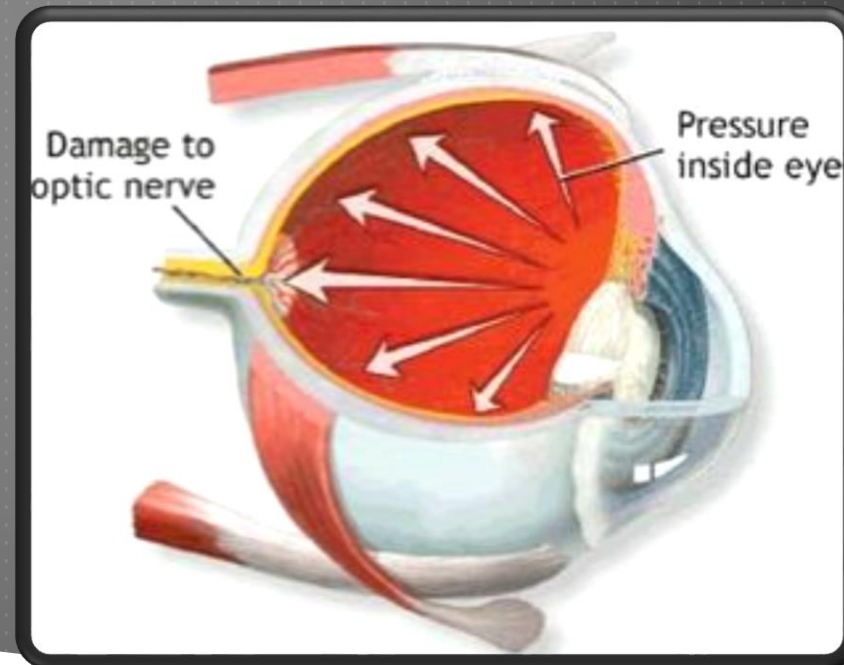
BUFTALMOS

▶ DEFINICIÓN

Aumento anormal en el diámetro AP del globo ocular secundario a un elevación de la presión intraocular cuya causa puede ser multifactorial

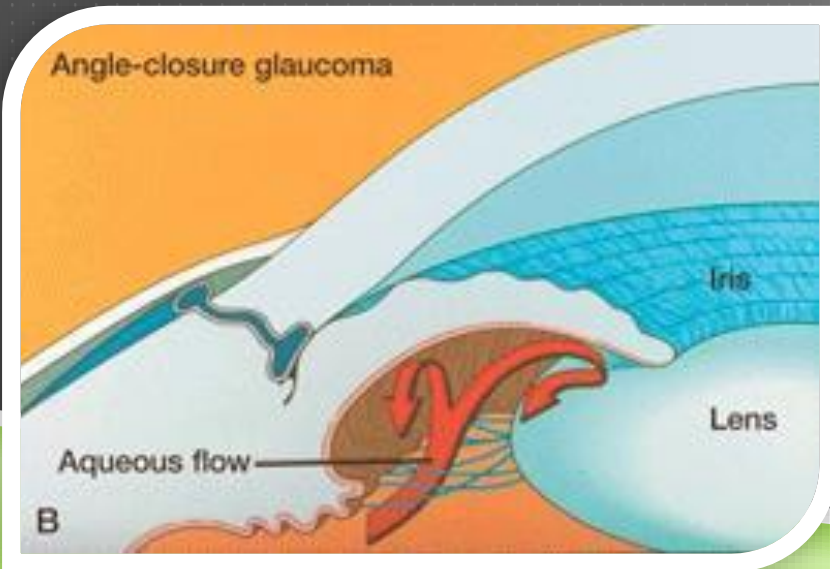
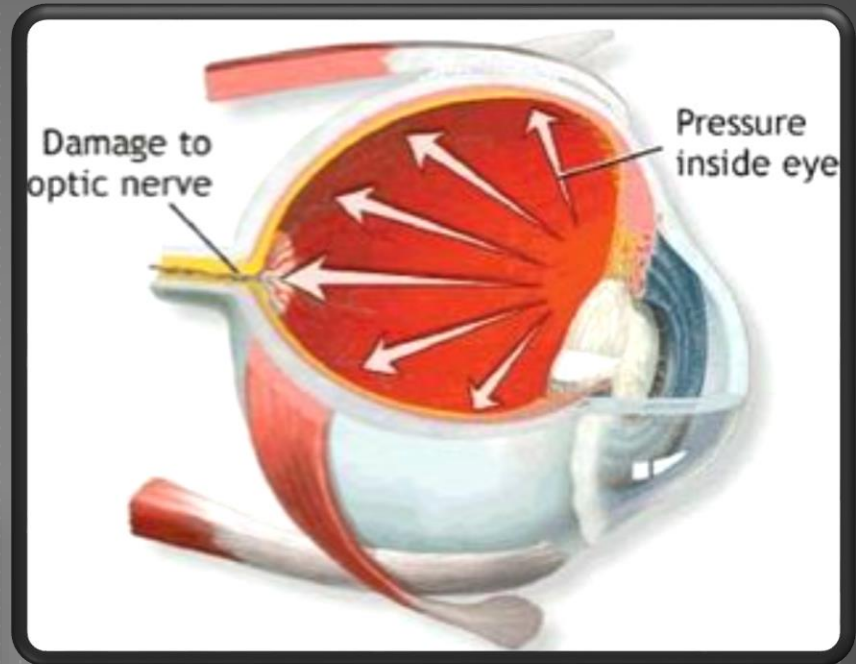
FISIOPATOLOGÍA

- ▶ Hipertensión intraocular
- ▶ Aumento del diámetro anteroposterior y corneano
- ▶ Lesión del nervio óptico
- ▶ Ruptura de capas corneales
- ▶ Opacidad corneal
- ▶ pseudoproptosis



Rehany U, Zegal Z. Congenital unilateral buphthalmos. A clinicopathological study. *Eye* (1999) **13**, 778–780

FISIOPATOLOGÍA



EPIDEMIOLOGÍA

- ▶ 1 de C/ 100.000 nacidos vivos
- ▶ Más frecuente en mujeres 2/1
- ▶ 70 % bilateral
- ▶ 90 % congénito
- ▶ 10 % generan ceguera

Rehany U, Zegal Z. Congenital unilateral buphthalmos. A clinicopathological study.
Eye (1999) **13**, 778–780

► Manifestaciones clínicas

Triada clásica



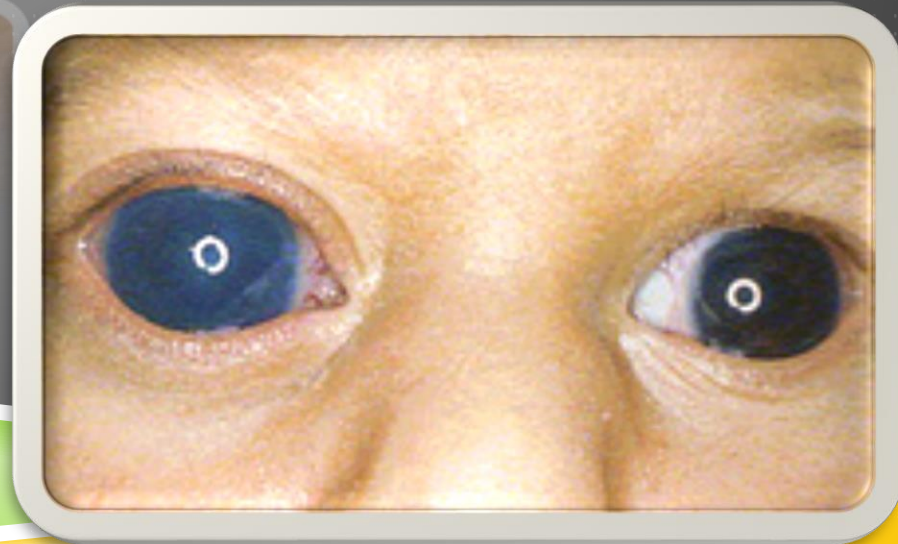
Fotofobia

Blefaroespasma

Epifora

- Proptosis
- Opacidades corneales (estriás de habb)
- Edema conjuntival
- Pérdida progresiva de la agudeza visual
- Astigmatismo

MANIFESTACIONES CLÍNICAS



BUFTALMOS



BUFTALMOS

Table 15.2: Differentiating features of **buphthalmos**, keratoglobus and megalocornea

Features	Buphthalmos	<i>Keratoglobus</i>	<i>Megalocornea</i>
Laterality	Usually bilateral	Bilateral	Almost always bilateral
Sex predilection	M:F:5:3	Uncertain	Male (90%)
Hereditary pattern	Autosomal recessive	Uncertain	X-linked recessive
Vision	Markedly impaired	Impaired	Not impaired
Corneal transparency	Opacities due to rupture in Descemet's membrane	Stromal haze due to fragmentation of Bowman's membrane	Not affected
Intraocular pressure	Raised	Normal	Normal
Angle of anterior chamber	Developmental anomalies seen	Normal	Normal
Optic disk	May be cupped	No cupping	No cupping

Rehany U, Zegal Z. Congenital unilateral *buphthalmos*. A clinicopathological study. *Eye* (1999) **13**, 778–780

ETIOLOGÍA

Anormalidades oculares

- ▶ Aniridia
- ▶ Hipoplasia del iris
- ▶ Síndrome Axenfeld-Rieger
- ▶ Anomalía de Peter
- ▶ Ectropión de úvea congénito
- ▶ Estafiloma congénito de córnea
- ▶ Córnea plana
- Microcórnea
- Vítreo primario hiperplásico persistente
- Nanophthalmos
- Distrofia polimorfa posterior
- Megalocórnea

PATOLOGÍAS EXTRAOCULARES

- ▶ Glaucoma por facomatosis.
- ▶ Trisomía.
- ▶ Síndrome de Pierre Robin.
- ▶ Mucopolisacáridos
- ▶ Síndrome de Prader Willis.
- ▶ Tumores de LA FOSA MEDIA

SCHWANNOMAS TRIGEMINALES

GENERALIDADES

- ▶ Segunda localización más común para schwannomas intracraneales.
- ▶ 0.8 - 8 % de los schwannomas.
- ▶ 0.07-0.36% de tumores del sistema nervioso.
- ▶ Lesiones de bajo grado WHO I.
- ▶ Mayor incidencia en la cuarta o quinta década de la vida.

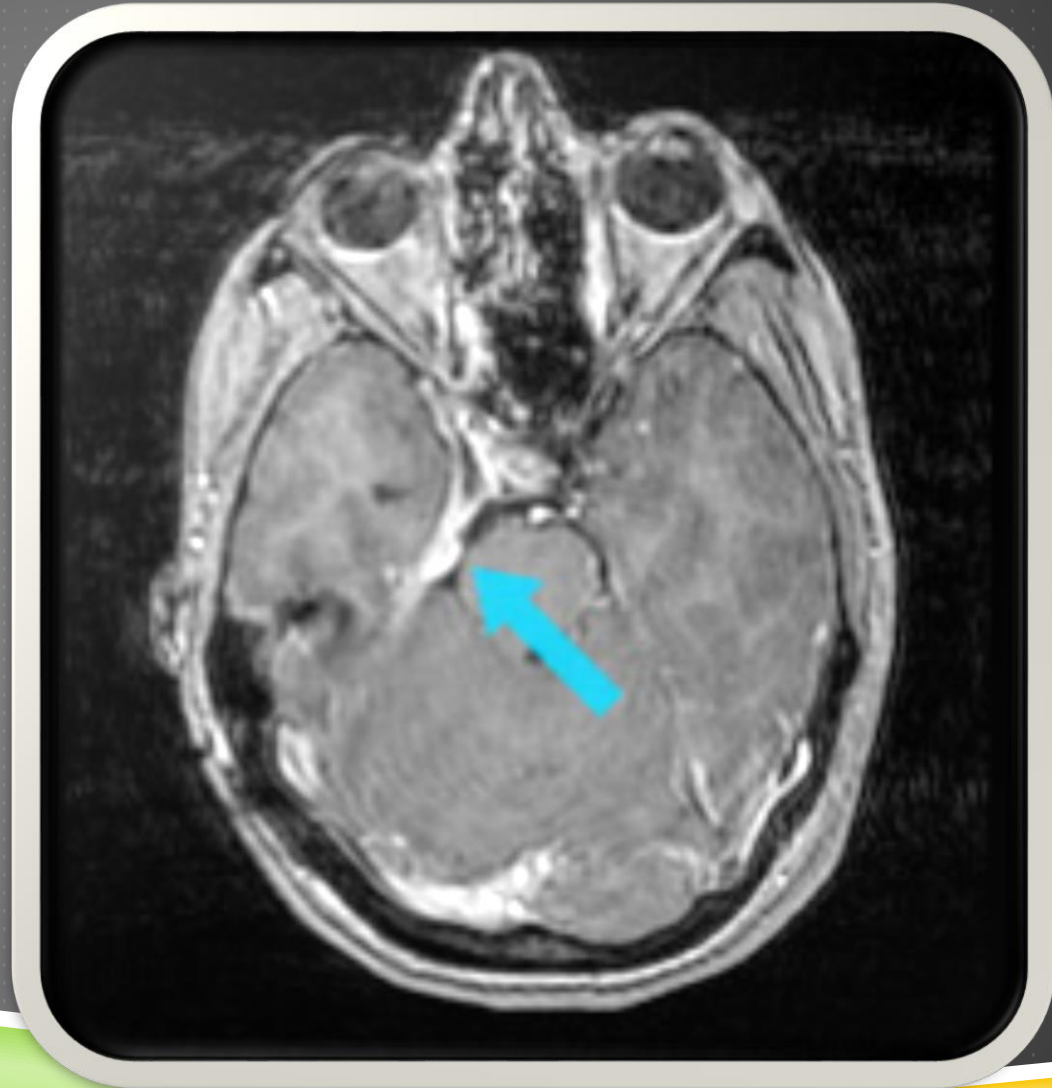
CLASIFICACIÓN

- ▶ TIPO A: FOSA MEDIA
- ▶ TIPO B: FOSA POSTERIOR
- ▶ TIPO C: FOSA MEDIA Y POSTERIOR
- ▶ TIPO D: EXTRACRANEAL

VandeVyver V, Lemmerling M, Van Hecke W, et al. MRI finding of the normal and diseased trigeminal nerve ganglion and branches: a pictorial review. JBR-BTR. 2007; 90: 272-277

TIPO A

- Fosa media (ganglionar).
- 50 % de los schwannoma trigeminales
- 20 % infiltran el seno cavernoso.
- 15 % compromiso oculomotor al momento del diagnóstico.
- 5% compromiso del nervio facial y vestibulococlear



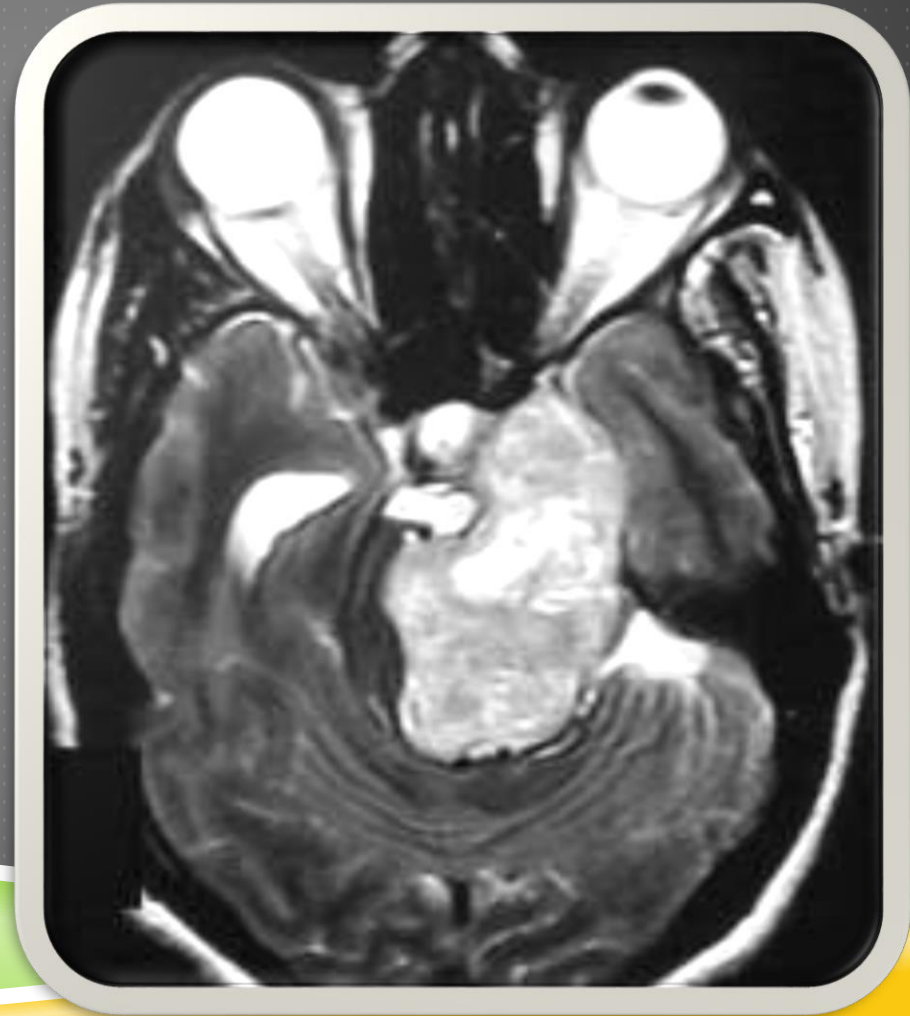
TIPO B “Root Tumor”

- ▶ 30 % de los TS.
- ▶ Infratentoriales.
- ▶ 20 % compromiso de pares bajos.
- ▶ Compromiso piramidal 30%
- ▶ Hipertensión intracraneal 30 %



TIPO C “Dumbell”

- ▶ 34.5 %
- ▶ Sintomatología combinada
- ▶ Origen a nivel de fosa posterior con extensión a fosa media.

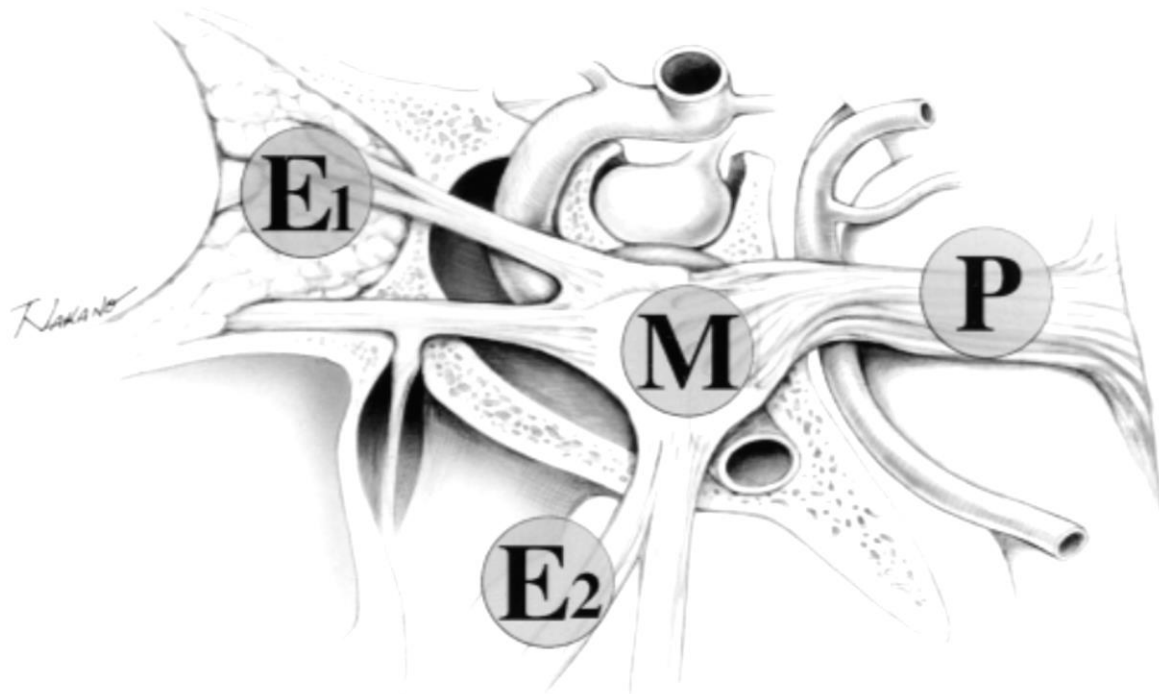


TIPO D

- ▶ 10.7 % de los casos
- ▶ Orbita, fosa infratemporal y pterigopalatina.
- ▶ Origen a nivel VI, V2 o V3
- ▶ Mayor porcentaje de malignidad



CLASIFICACIÓN



Kasunari Y, Takeshi K. Trigeminal neurinomas extending into multiple fossae: surgical methods and review of the literature. *J Neurosurg* 91:202-211, 1999

CLASIFICACIÓN

TABLE 4
*Classification and incidence of TNs**

Authors & Year	Tumor Type & No. (%)						Total
	M	P	E	MP	ME	MPE	
McCormick, et al., 1983	61 (50.8)	38 (31.7)	—	21 (17.5)	—	—	120
assorted studies, 1984–1997	100 (35.5)	58 (20.6)	22 (7.8)	91 (32.3)	10 (3.5)	1 (0.4)	282
present study	4 (14.8)	5 (18.5)	1 (3.7)	10 (37.0)	5 (18.5)	2 (7.4)	27
total	165 (38.5)	101 (23.5)	23 (5.4)	122 (28.3)	15 (3.5)	3 (0.7)	429

* — = not specified.

Kasunari Y, Takeshi K. Trigeminal neurinomas extending into multiple fossae: surgical methods and review of the literature. *J Neurosurg* 91:202–211, 1999

MANIFESTACIONES CLÍNICAS

- ▶ Hipoestesia facial (70 %)
- ▶ Neuralgia del trigémino (13 a 38.5 %)
- ▶ Parálisis del sexto par (26 %)
- ▶ Hipoacusia
- ▶ Tinnitus
- ▶ Alteraciones de la marcha
- ▶ Síntomas visuales
 - ▶ Proptosis
 - ▶ Diplopía
 - ▶ Disminución de la agudeza visual
 - ▶ Buftalmos

MANIFESTACIONES CLÍNICAS

Case	Age	Sex	Chief Complaint	Duration	+	-	Cranial Nerve	+	-	Other Findings	Location
10	33	F	rt lat diplopia	2 wks	-	-	VI	-	-	hand numbness	6, D
11	47	F	ataxia	2 yrs	-	-	VIII, IX	-	cerebellar	dementia	7, D
12	66	F	rt hearing loss	3 mos	+	V ₁₋₃	VII, VIII	-	-	-	6, D
13‡	1	M	proptosis	11 mos	-	V ₁	II-IV, VI	+	-	papilledema	7, V ₁
14	41	F	nasal obstruction	3 mos	-	V ₂	-	-	-	-	8, V ₂
15	58	F	rt hearing loss	1 yr	-	V ₃	-	-	-	serous otitis media	3, V ₃
16	57	M	proptosis	2 yrs	-	V ₁	II	+	-	-	2, O

* Cranial nerves are given in roman numerals. + = feature present; - = feature absent.

† Maximum diameter is given in cm. Abbreviations: P = posterior fossa primarily; M = middle fossa primarily; D = dumbbell growth pattern above and below the tentorium; V₁ = V₁ tumor involving the cavernous sinus, middle fossa, and orbit; V₂ = V₂ neurilemoma extending from the cavernous sinus through the foramen rotundum into the pterygopalatine and infratemporal fossae, sphenoid sinus, and maxillary sinus; V₃ = V₃ neurilemoma extending from the middle fossa through the foramen ovale to the infratemporal fossa; O = extracranial V₁ tumor extending through the orbital apex.

‡ Patients with associated neurofibromatosis.

pairment of either sensory or motor function of the trigeminal nerve. Other cranial nerves were affected in 11 patients; in four (Cases 4, 7, 10, and 11) these were the sole findings on examination. Impairment of the fifth cranial nerve was most common (seven patients).

arising from the intracranial trigeminal branches and extending extracranially. Another patient (Case 16) had a neurilemoma of the first division of the fifth nerve (V₁) originating in the orbital apex and growing intracranially. Although all four patients had decreased s

MANIFESTACIONES CLÍNICAS

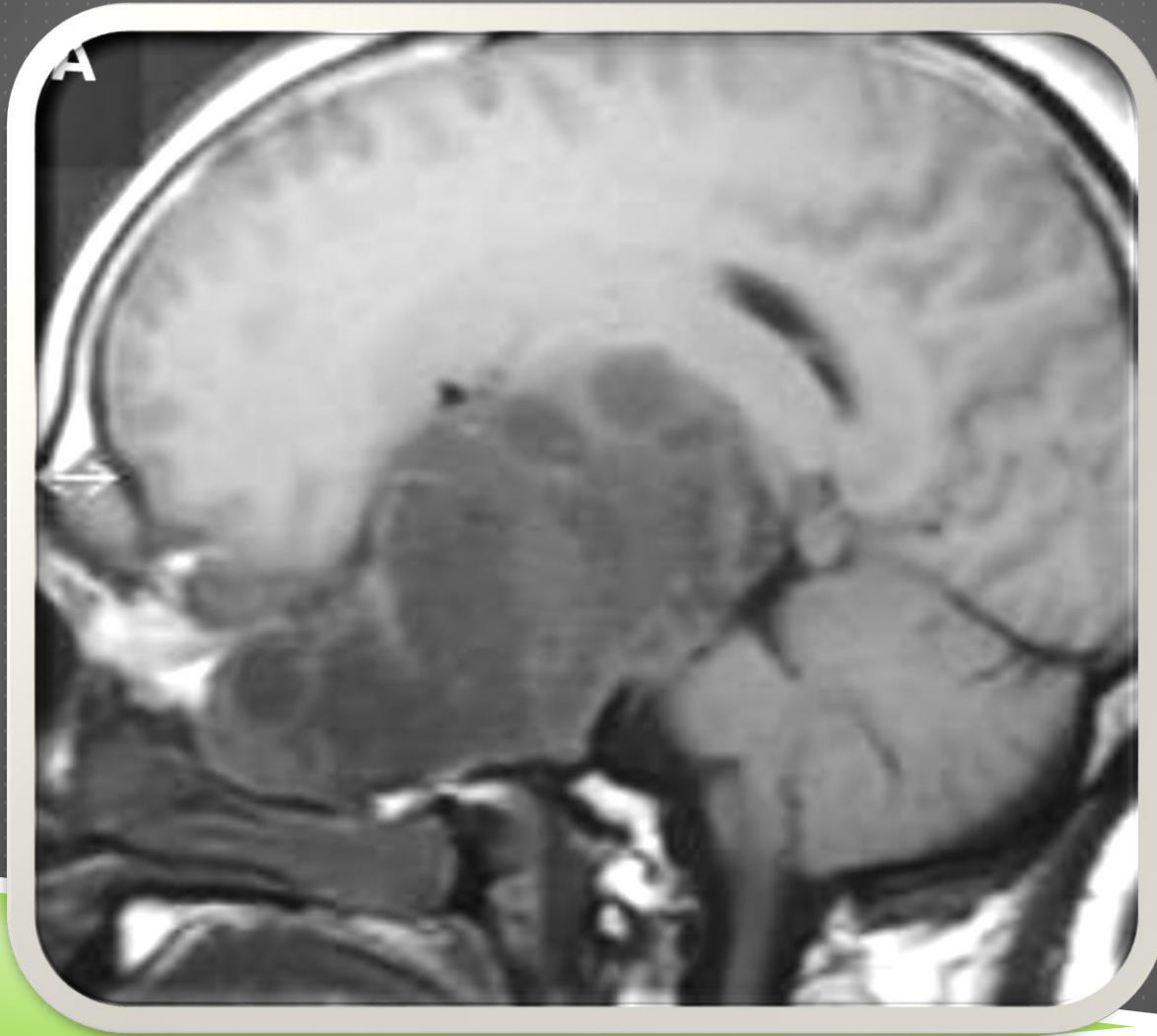
TABLE 2

Symptoms and findings at admission in 27 patients with TNs

Presentation	No. of Cases (%)
initial symptoms	
trigeminal	15 (56)
pain	3 (11)
paresthesia	2 (7)
hypesthesia	11 (41)
double vision	2 (7)
hearing loss	2 (7)
visual disturbance	3 (11)
exophthalmos	2 (7)
ataxia	2 (7)
headache	1 (4)
incidental	5 (19)
abnormal findings	
facial hypesthesia	14 (52)
facial pain	3 (11)
masticatory weakness	3 (11)
CN sign	
II	4 (15)
III	3 (11)
IV	2 (7)
VI	4 (15)
VII	2 (7)
VIII	3 (11)
IX-X	1 (4)
cerebellar/long tract signs	5 (19)

Kasunari Y, Takeshi K. Trigeminal neurinomas extending into multiple fossae: surgical methods and review of the literature. *J Neurosurg* 91:202-211, 2007

DX IMAGENOLÓGICO









TRATAMIENTO

- ▶ QUIRÚRGICO
- ▶ RADIOCIRUGÍA
- ▶ RADIOTERAPIA

RADIOCIRUGÍA

- ▶ Lesiones menores de 3 cm recurrente o residuales.
- ▶ Rangos de control 84-100 %
- ▶ 160 pacientes reportados
- ▶ Dosis promedio 13.1-18 Gy.

RADIOCIRUGÍA

TABLE 144-2 Studies on Radiosurgery for Trigeminal Schwannomas

SERIES	N	MODALITY	MEAN RADIATION DOSE (Gy)	TUMOR GROWTH CONTROL RATE (%)	MEAN FOLLOW-UP (mo)	TOTAL MORBIDITY (%)	NONTRIGEMINAL MORBIDITY (%)
Huang et al., ⁵⁶ 1999	16	GK	15.3	100	44	N/A	N/A
Mabanta et al., ⁵⁸ 1999	7	LINAC	13.1	100	32	N/A	N/A
Kida et al., ⁵⁷ 2000	19	GK	14.7	100	20.7	10.4	0
Pollock et al., ⁵⁹ 2002	10	GK	18	90	43	30	0
Nettel et al., ⁶⁰ 2004	23	GK	15	91	40	8.7	4.3 (facial weakness)
→ Wang et al., ⁶¹ 2005	35	GK	13.6	96.2	76	5.7	0
Hasegawa et al., ⁵⁴ 2007	37	GK	14.2	84	54	2.7	0
Peker et al., ⁵⁵ 2007	15	GK	16	100	61	6.7	0

GK, Gamma Knife; LINAC, linear accelerator; N/A, not available.

RADIOTERAPIA

- ▶ Tumores inoperables y recurrentes
- ▶ Dos estudios
- ▶ Wallner and col.
 - ▶ 8 pacientes
 - ▶ Seguimiento 2-15 años
 - ▶ Control del tumor en el 50 % 1.4 a 7 años
- ▶ Zabel and col.
 - ▶ 13 pacientes
 - ▶ 33 meses seguimiento
 - ▶ Control del crecimiento 100 %

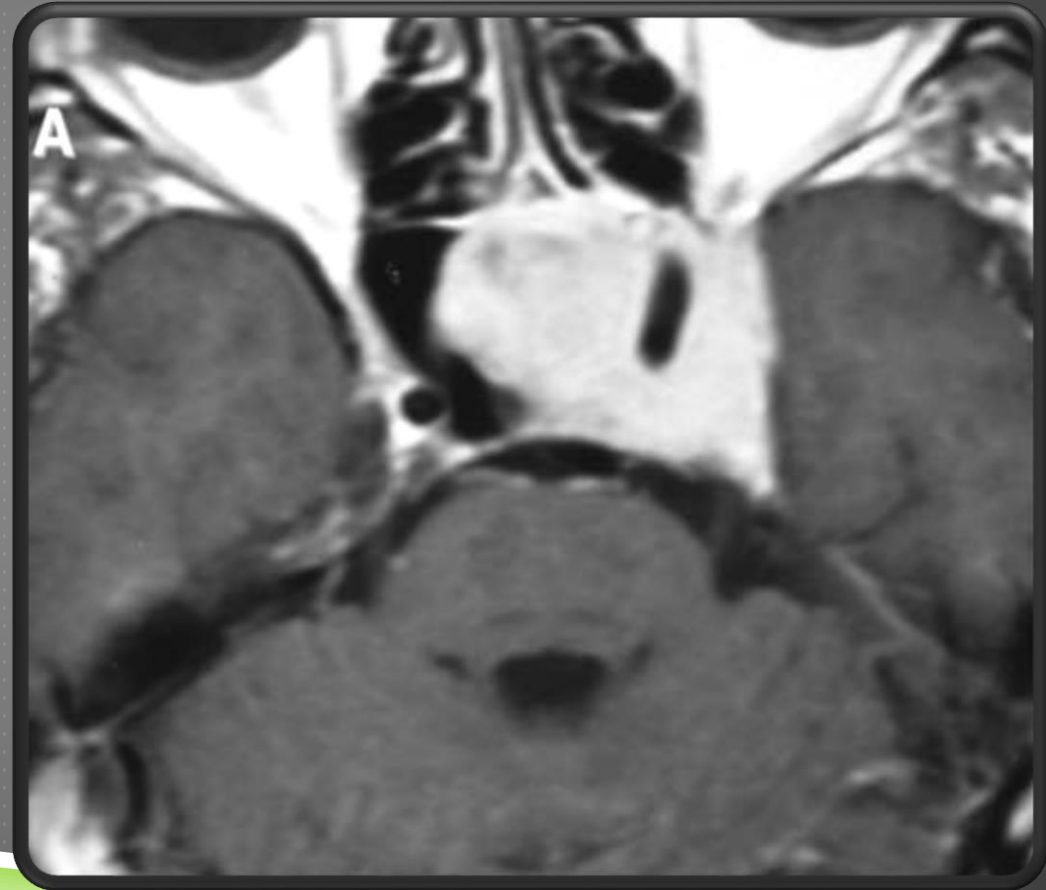
DIAGNÓSTICOS DIFERENCIALES

MENINGIOMA

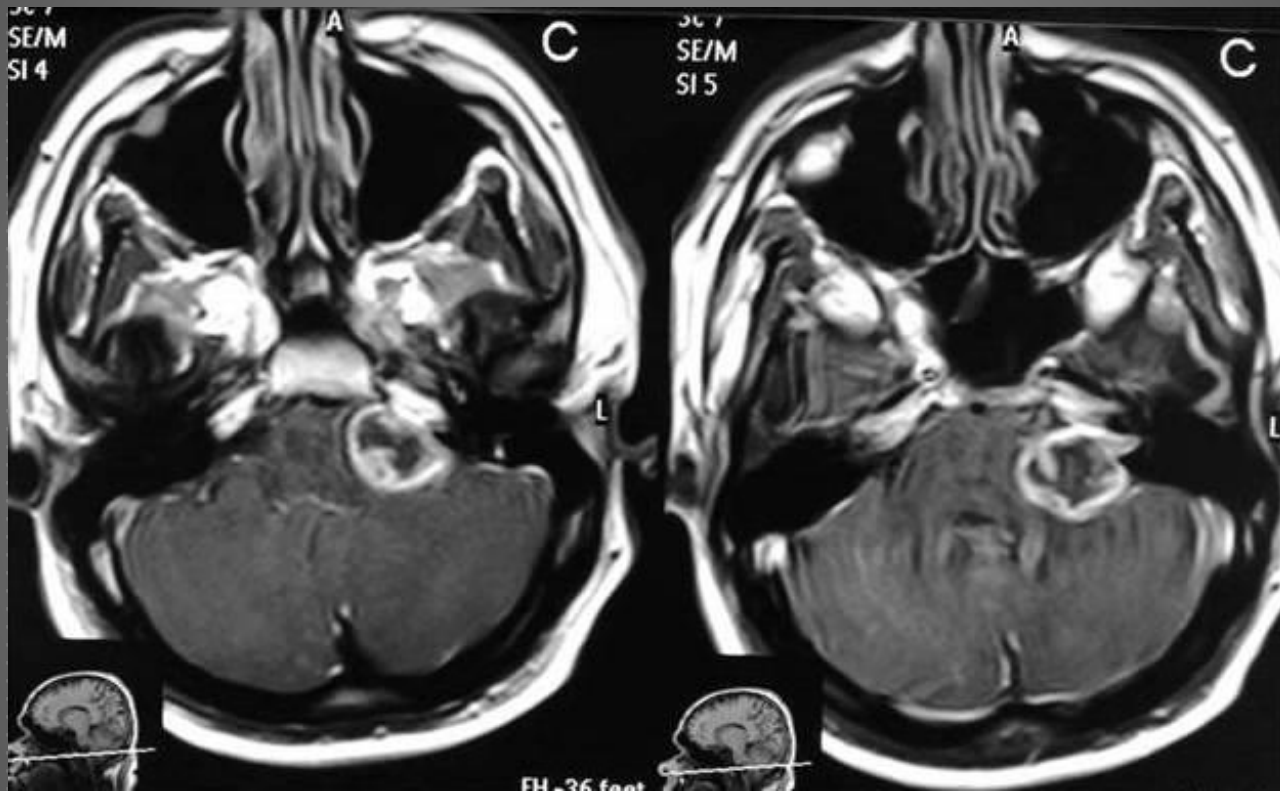


ADENOMA PITUITARIO

- ▶ 6-10 % invasión al seno cavernoso.
- ▶ Captación del contraste de manera homogénea

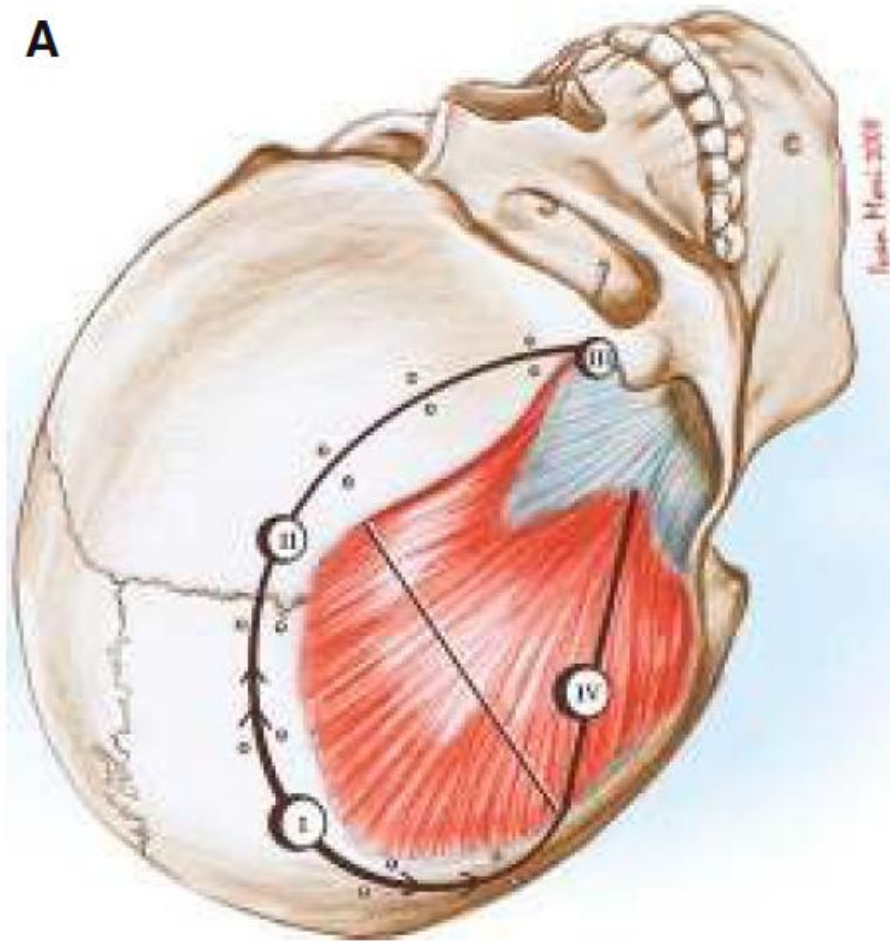


SCHWANNOMA VESTIBULAR

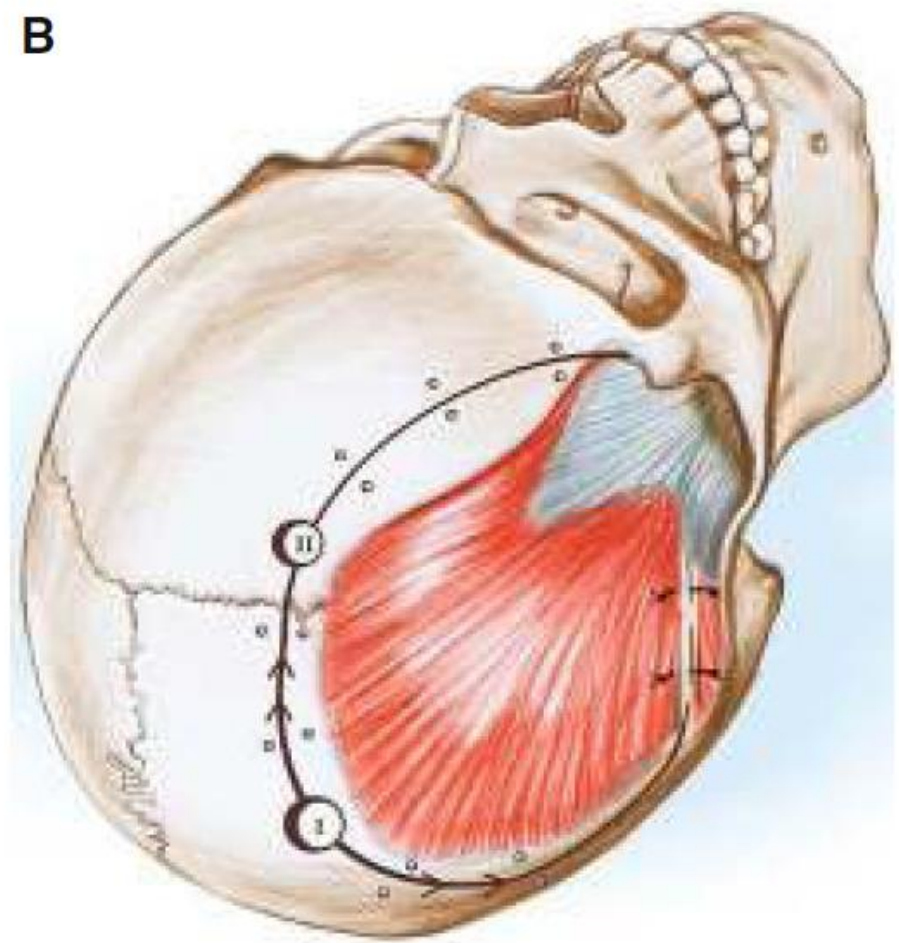


ABORDAJE QUIRÚRGICO AL SENO CAVERNOSO

A

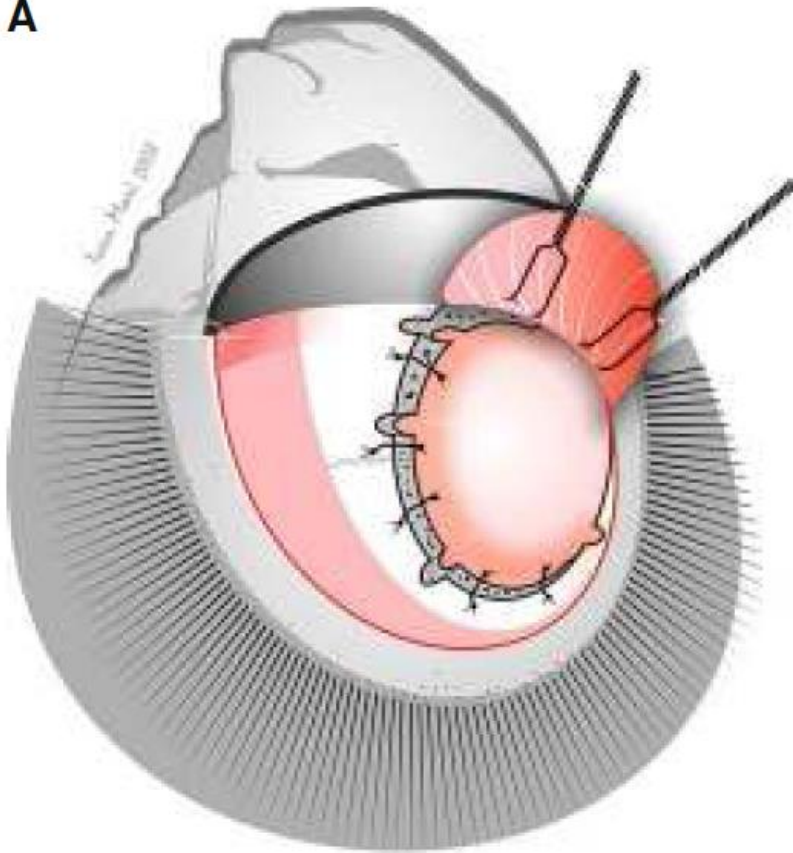


B

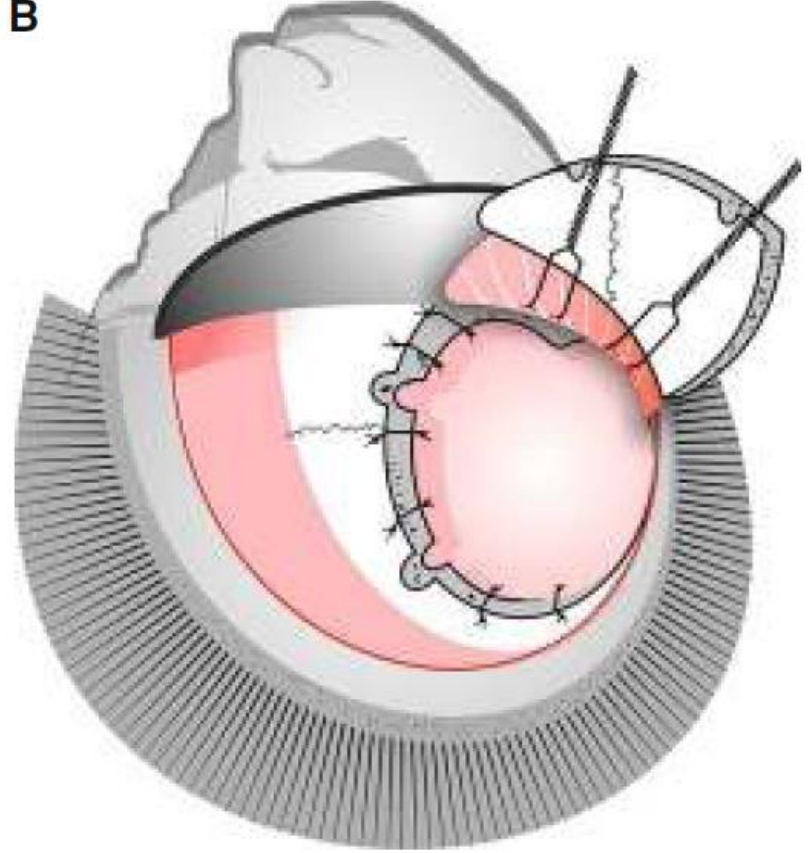


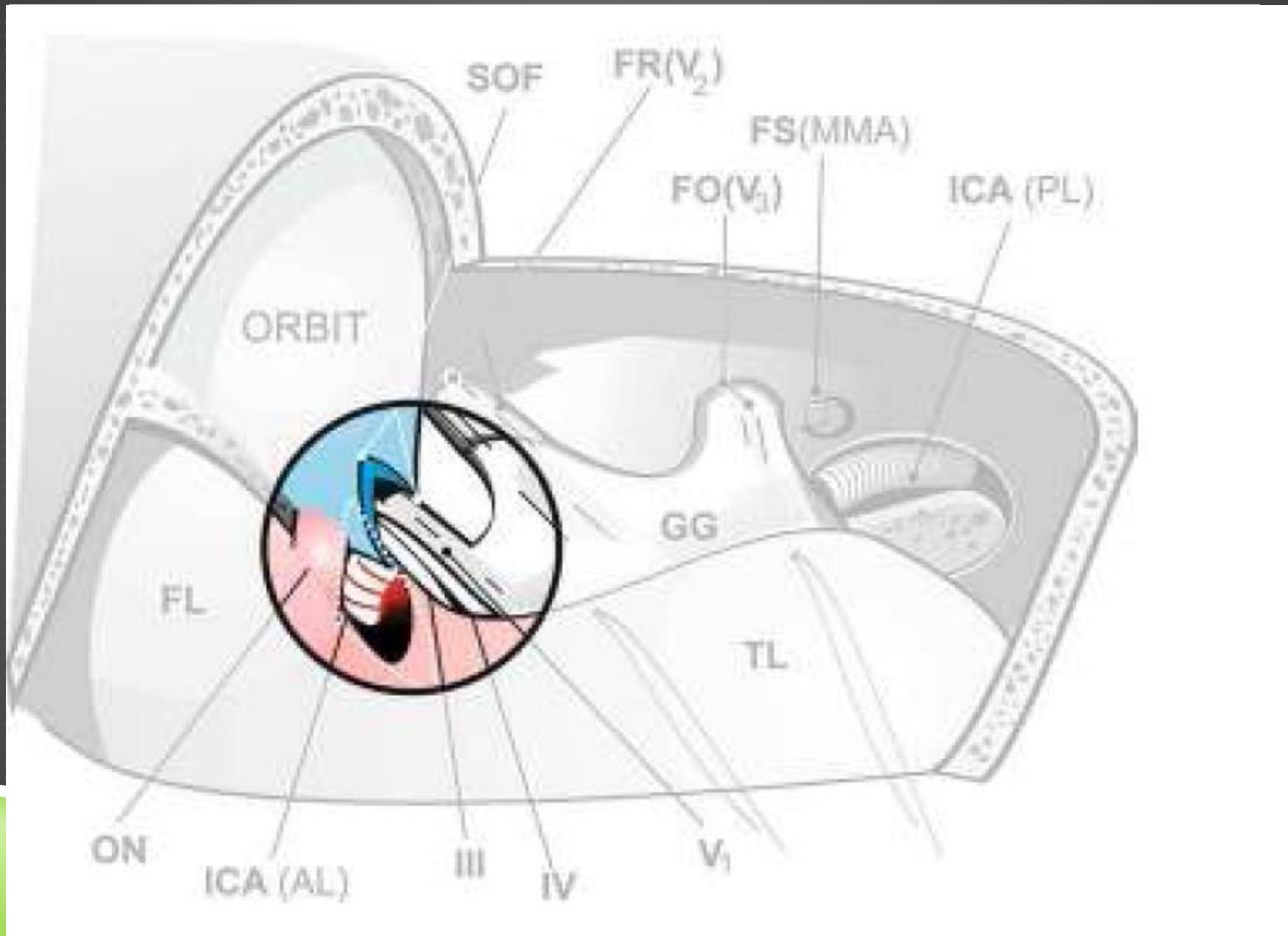
PRESERVACIÓN MUSCULAR

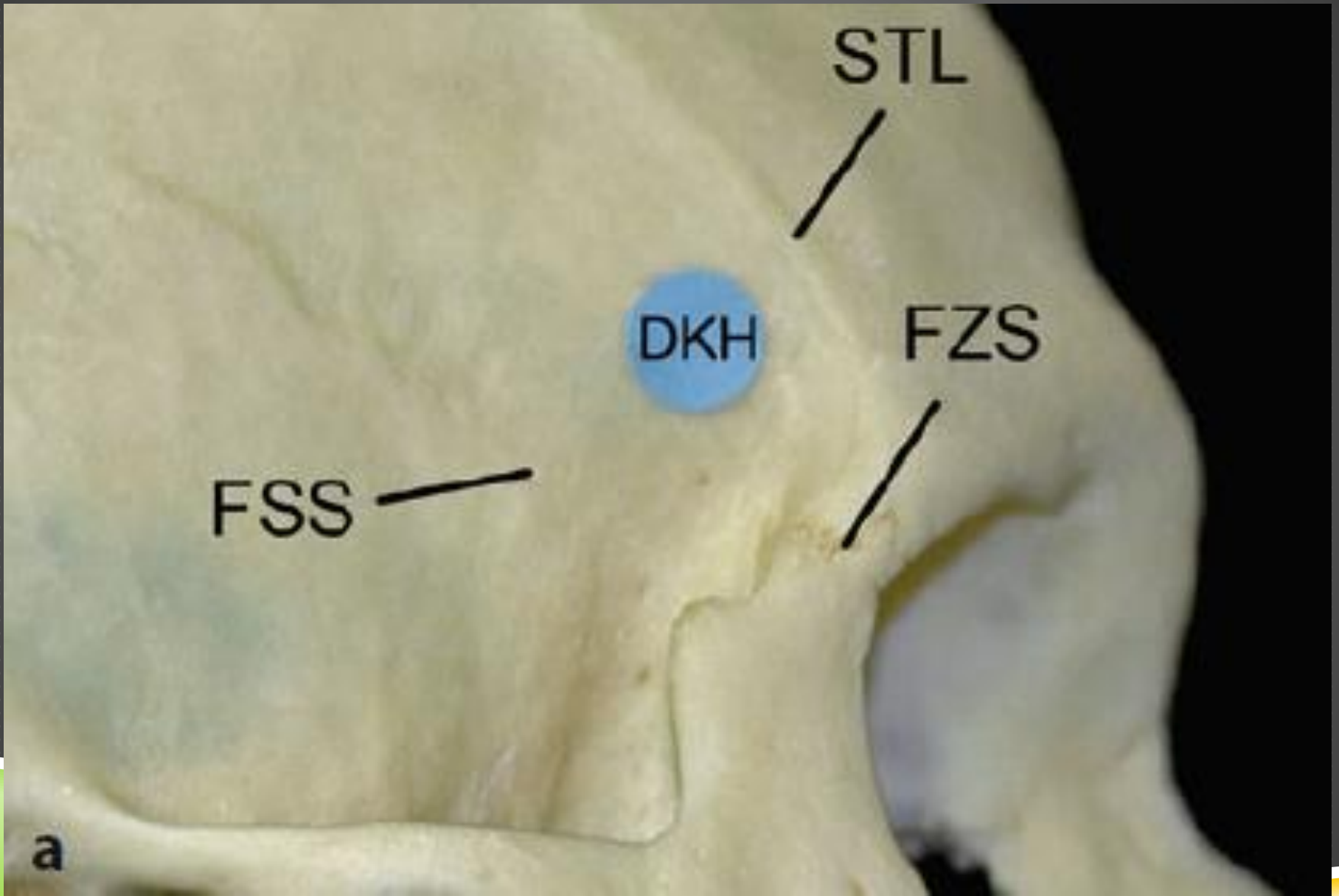
A



B







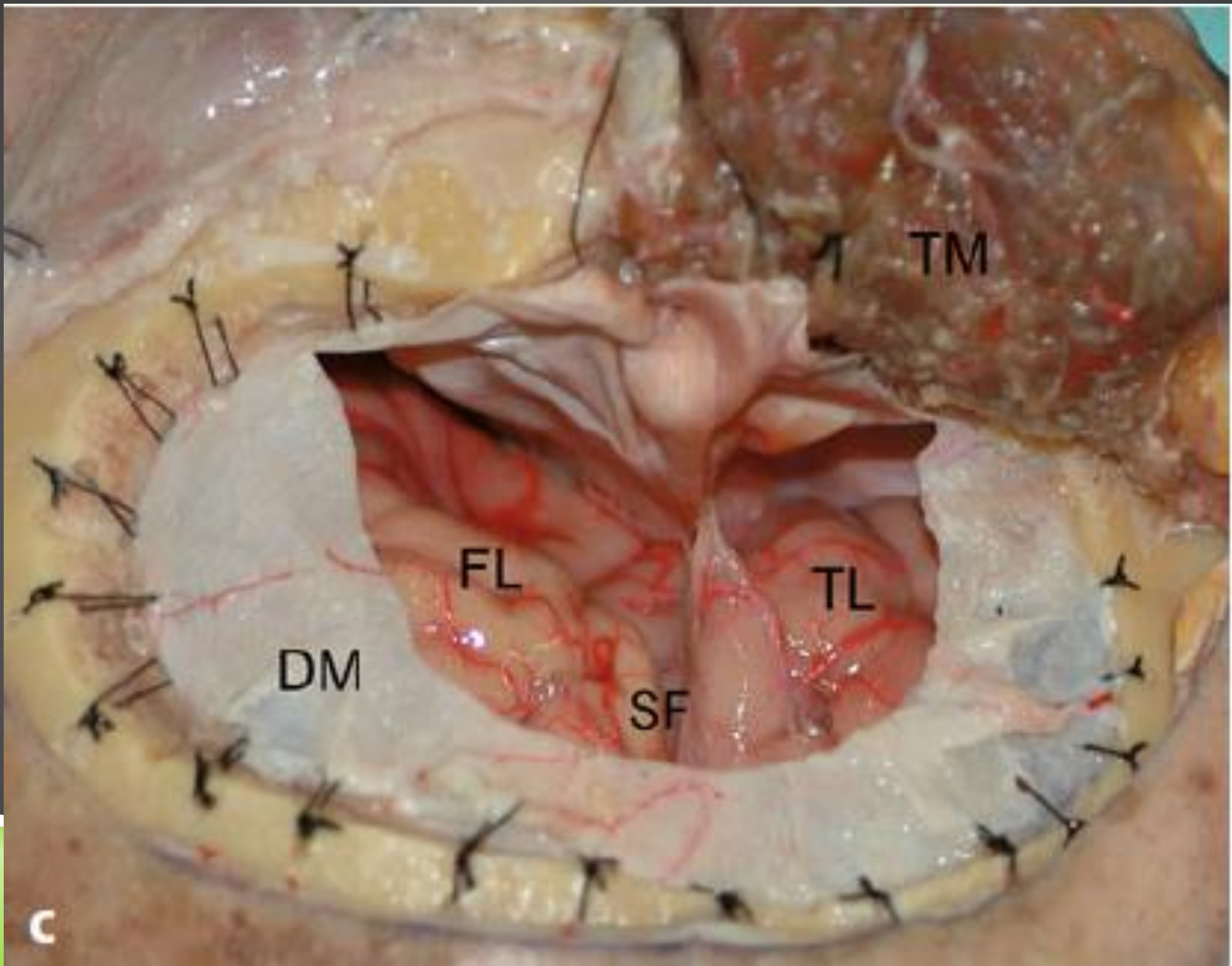
STL

DKH

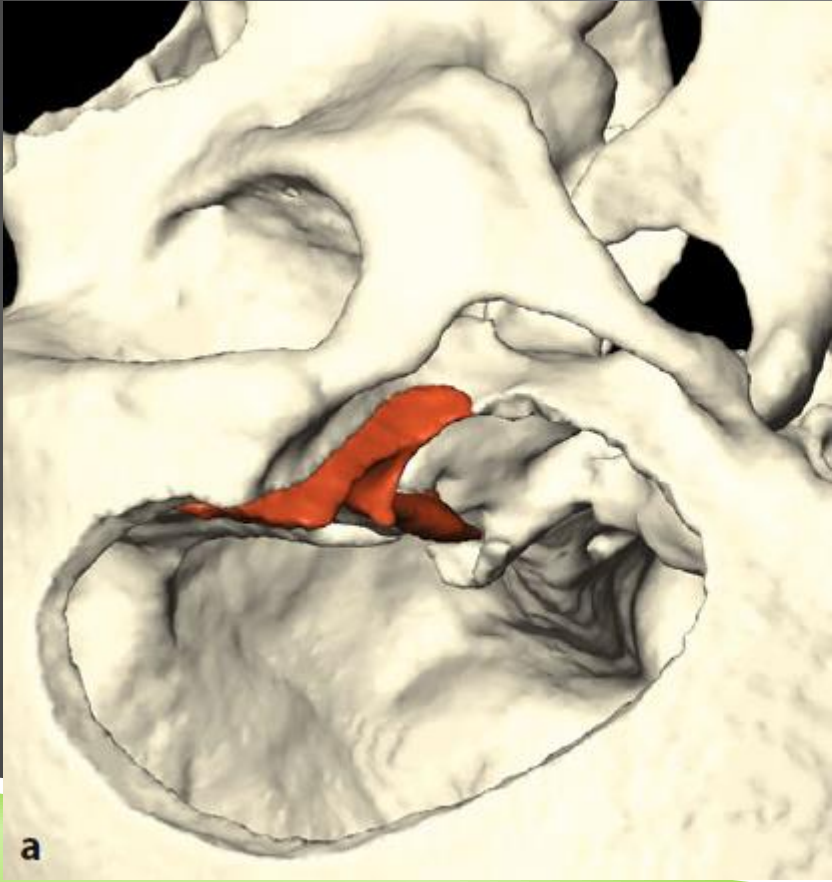
FZS

FSS

a



C



TRIÁNGULOS

- ▶ 4 SENO CAVERNOSO

- ▶ ÓPTICO
- ▶ OCULOMOTOR
- ▶ TROCLEAR
- ▶ OFTÁLMICA

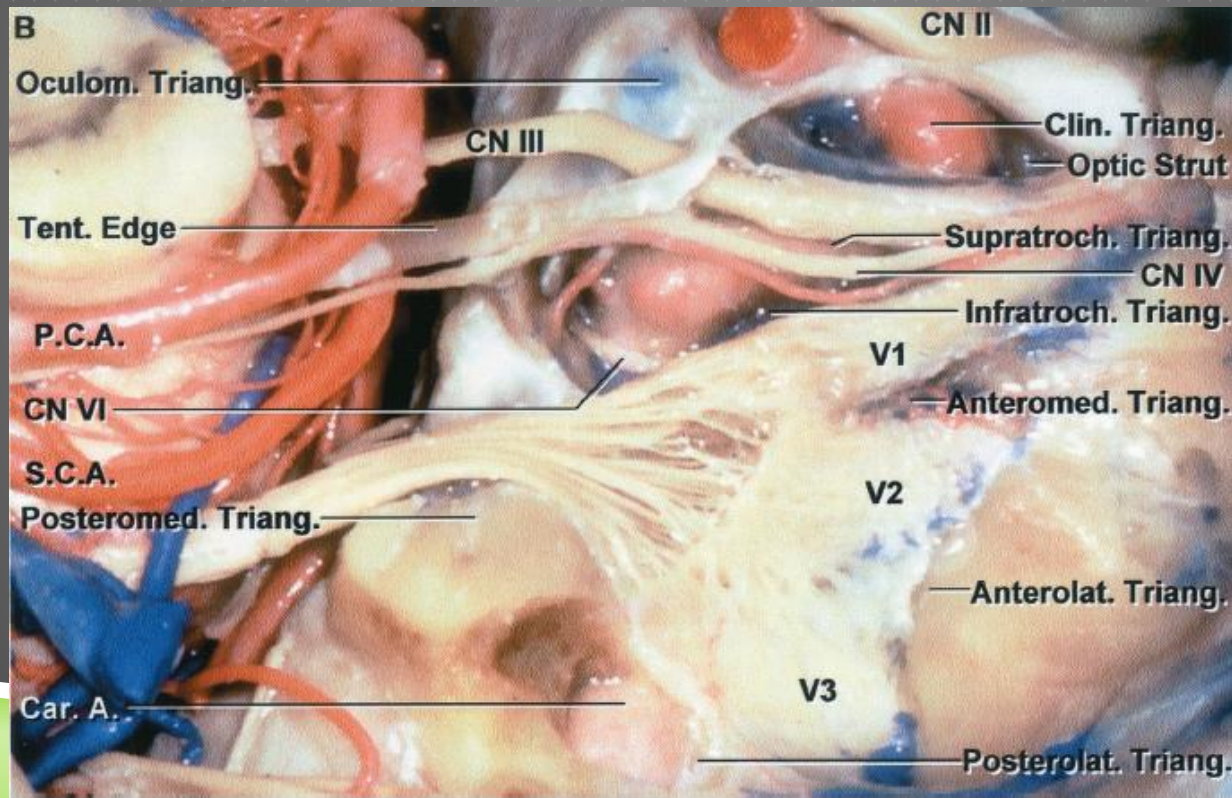
- ▶ **AGUJERO ÓPTICO Y FISURA ORBITARIA SUPERIOR**

- ▶ 2 Techo del seno cavernoso (repliegues dúrales)
 - ▶ Clinoideo
 - ▶ Oculomotor

- ▶ 2 pared lateral (estructuras nerviosas)
 - ▶ Supratroclear
 - ▶ Infratroclear (parkinson)

CLINOIDEO (DOLENC)

- ▶ Remover clinoides anterior

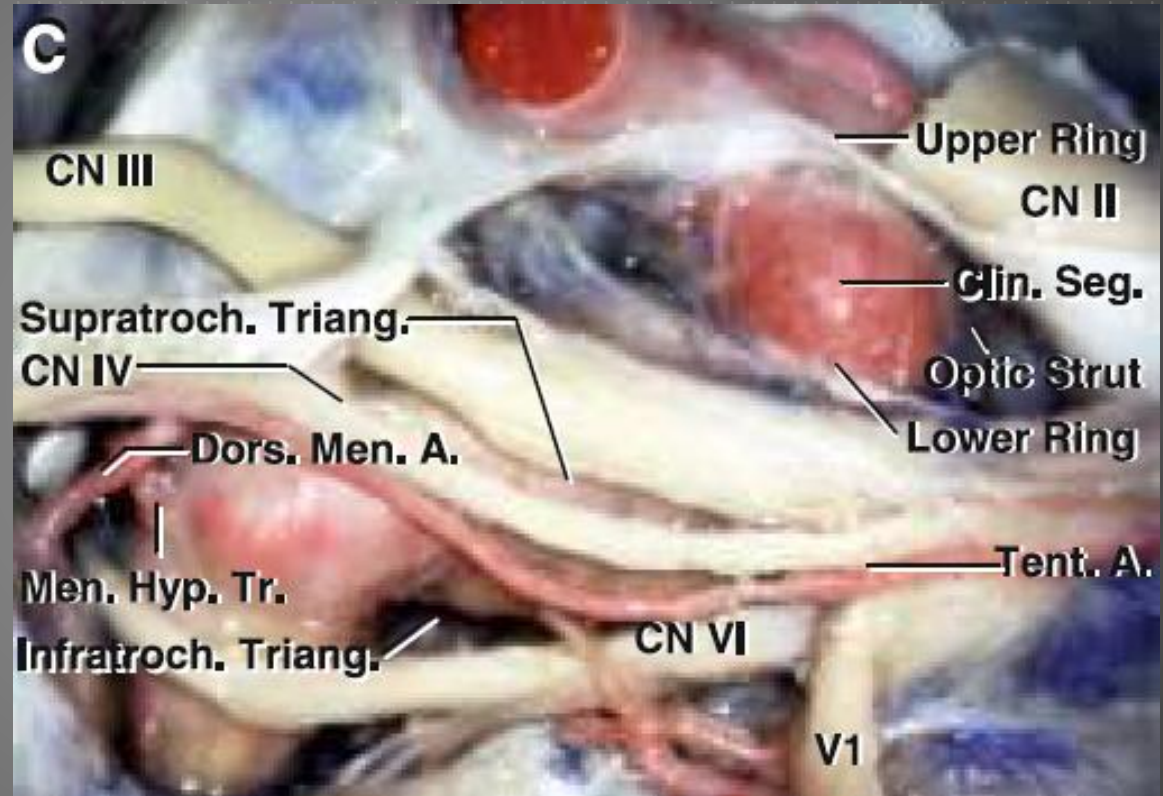


SUPRATROCLEAR

Borde inferior del III par

Borde superior de IV PAR

Unión entre los dos en la entrada hendidura



INFRATROCLEAR (PARKINSON)

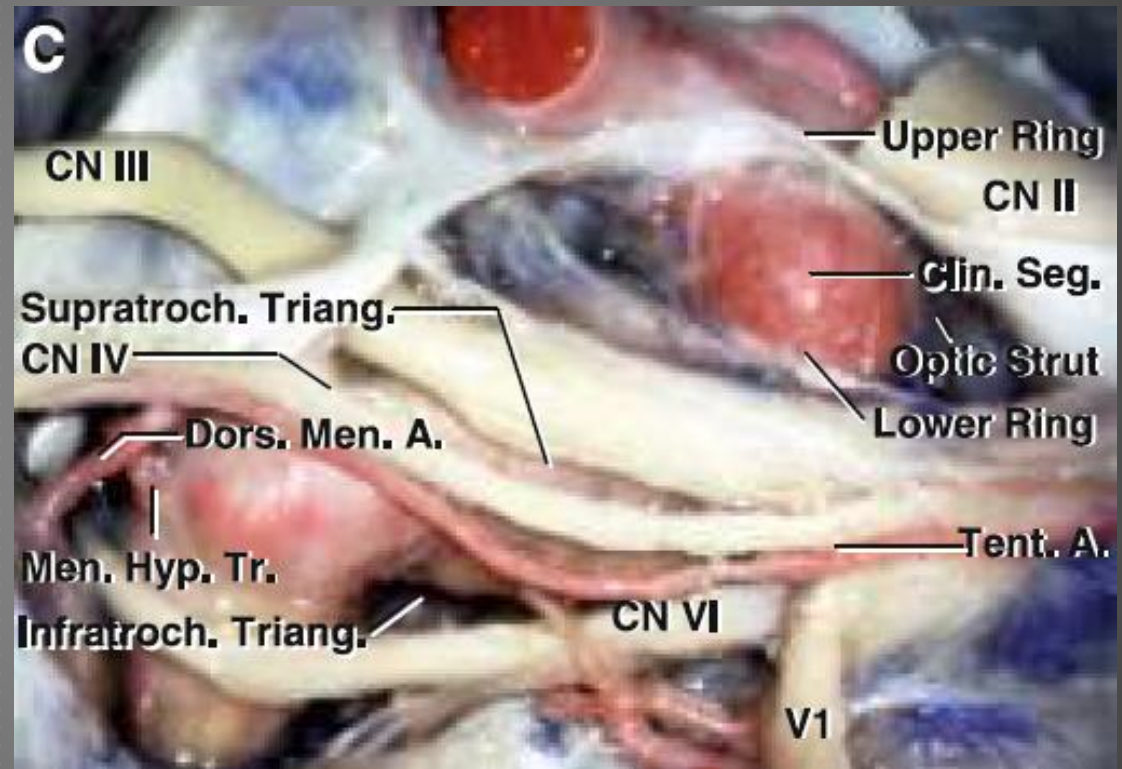
Borde inferior IV par

Borde superior de V I

Línea que une a estos
en:

Hendidura esfenoideal

Cavum de Meckel

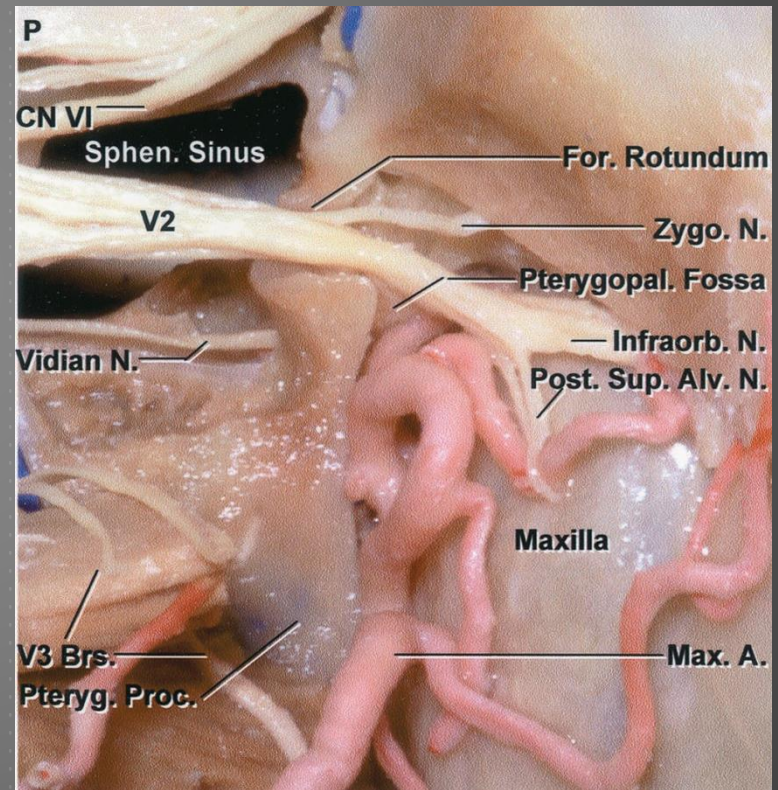


TRIÁNGULOS DE LA FOSA MEDIA

- ▶ 4 TRIÁNGULOS
 - ▶ Divisiones de el trigémino
 - ▶ Agujeros
 - ▶ Ganglio de Gasser

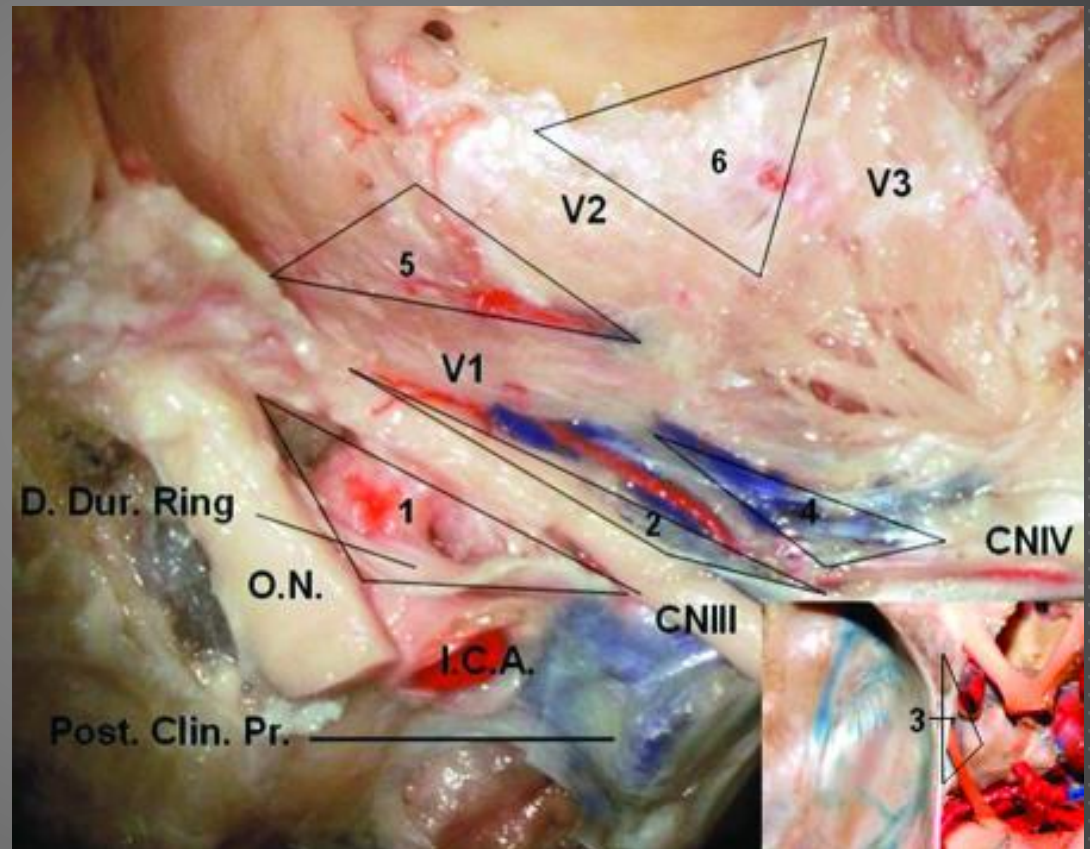
TRIÁNGULO ANTERIOMEDIAL

- ▶ Entre VI Y V2
- ▶ Línea pasa entre los dos
- ▶ Seno esfenoidal



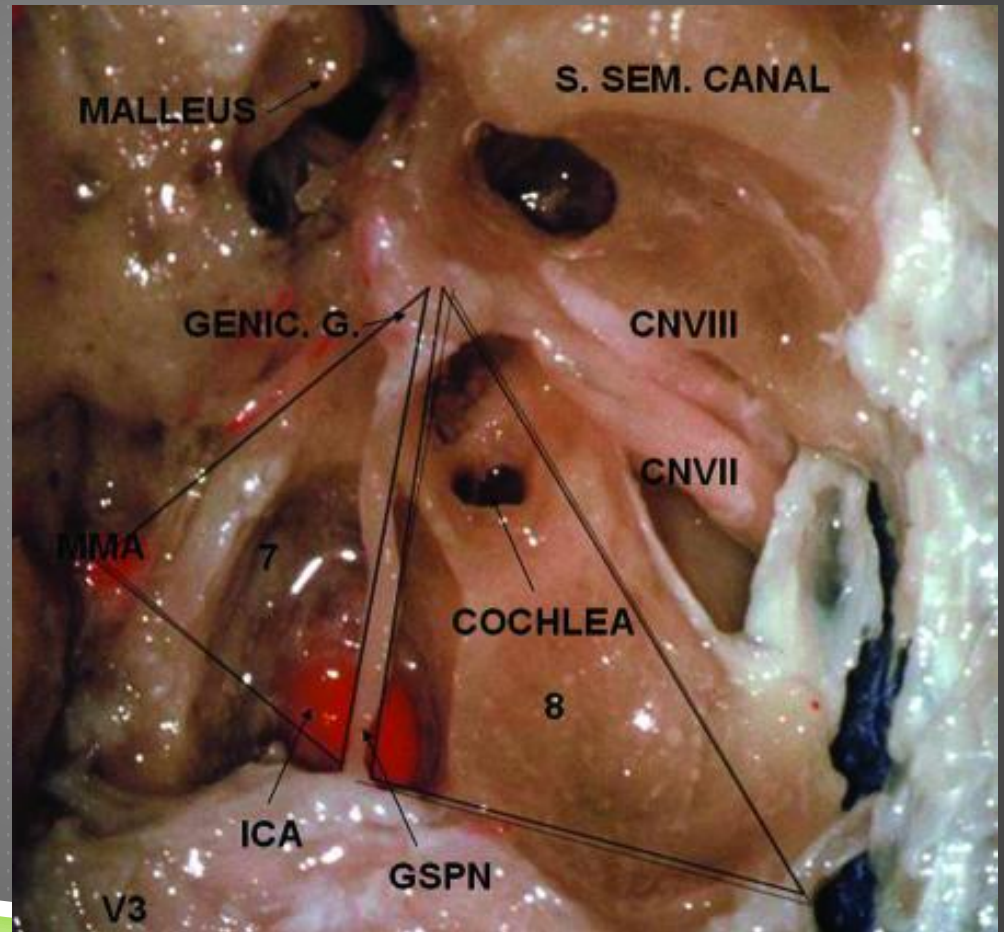
TRIÁNGULO ANTEROLATERAL

- ▶ ENTRE:
- ▶ V2 Y V3
- ▶ Seno esfenoidal

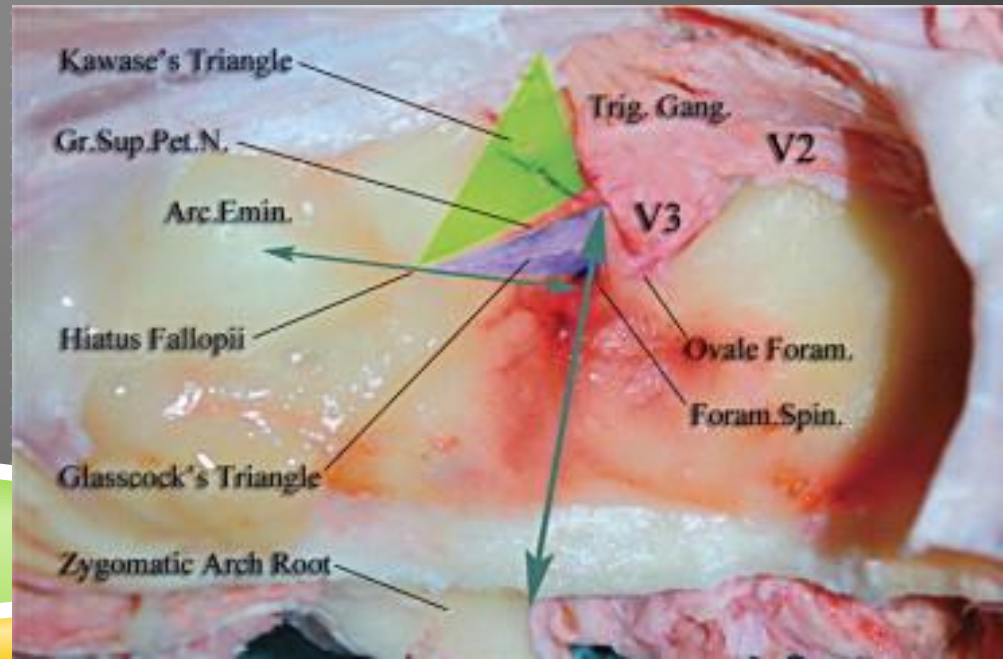
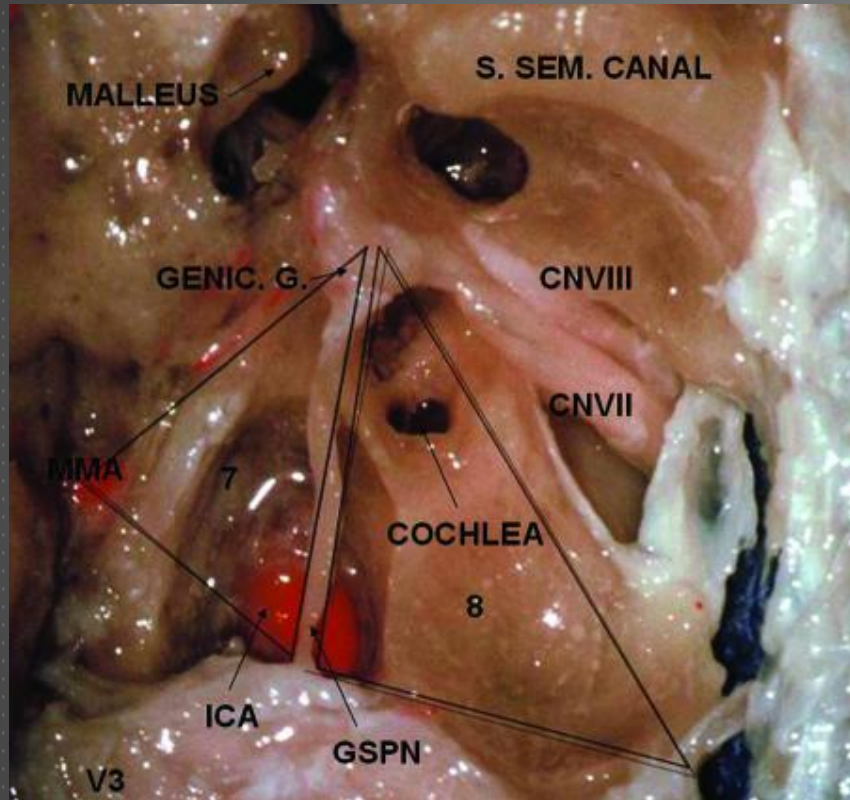


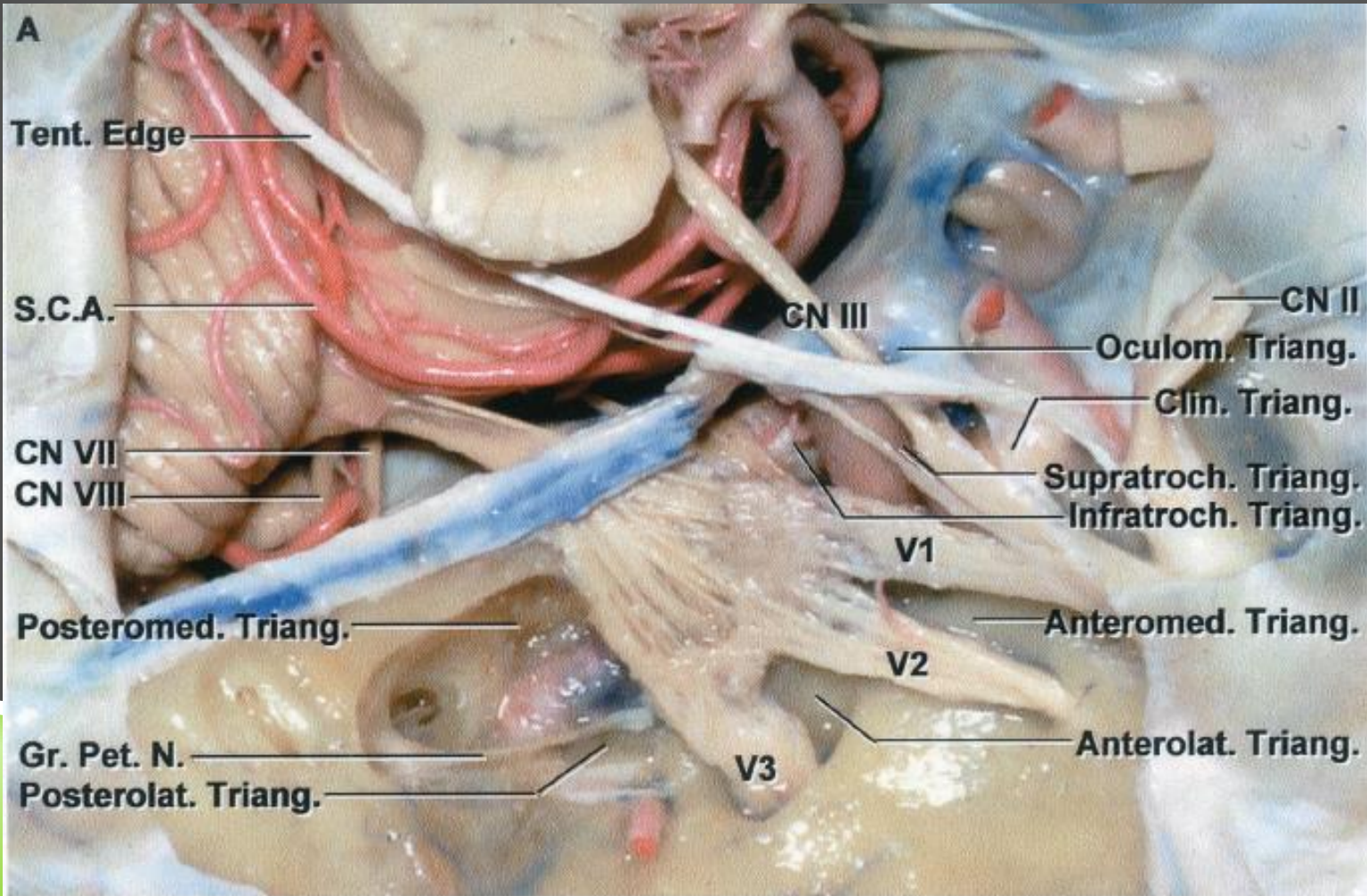
POSTEROLATERAL GLASSCOCK

- ▶ V3
- ▶ Petroso mayor
- ▶ Menínea media
- ▶ Fosa infratemporal



POSTEROMEDIAL (KAWASE)





CASO NÚMERO 2

Paciente de 1 año buftalmos congénito de crecimiento progresivo, asociado aparición de manchas café con leche en superficie corporal; no antecedentes personales o familiares de importancia. Niegan otra sintomatología.

Examen físico: parálisis del 6 par derecho, agudeza visual conservada pupilas isocóricas normorreactivas, moviliza 4 extremidades. Manchas café con leche en número 4 la de mayor tamaño 3 cm

