



**ESPR**  
European Society of  
Paediatric Radiology

**56<sup>th</sup>** Annual Meeting &  
**42<sup>nd</sup>** Post Graduate Course  
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# Tumors of the Orbit and the Eyeball

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# Disclosures

I have no conflict of interest regarding this presentation.





# Objectives

- Present a **compartment-based approach** to tumors of the orbit and the eyeball
- Become familiar with **imaging findings** of (some) tumors of the orbit and the eyeball
- Highlight relevant imaging findings for **differential diagnosis** and **treatment planning** of tumors of the orbit and the eyeball

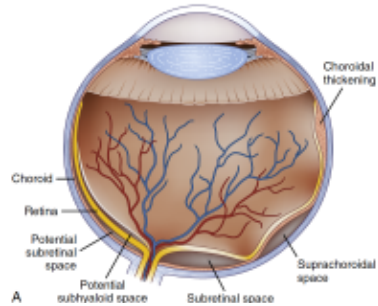




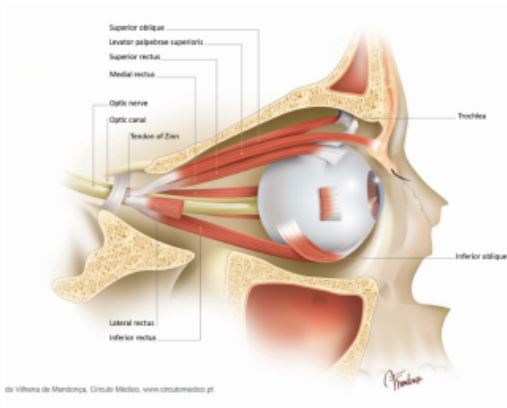
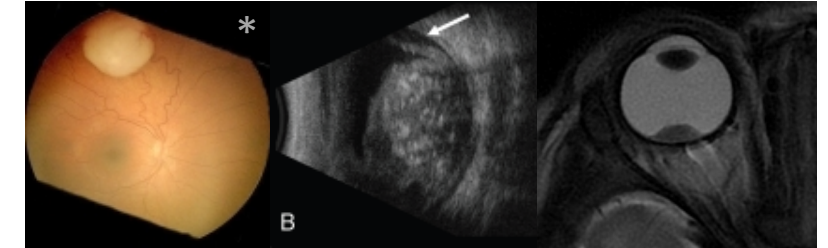
# Tumors of the Orbit and the Eyeball

- Children are affected by a **spectrum** of tumors of the orbit and eyeball that differs substantially from adults
- Most orbital tumors in children are **benign**
- Both benign and malignant lesions of the orbit may result in significant **morbidity**
- Imaging plays an essential role in early **diagnosis**, optimal **treatment** planning and **response** assessment

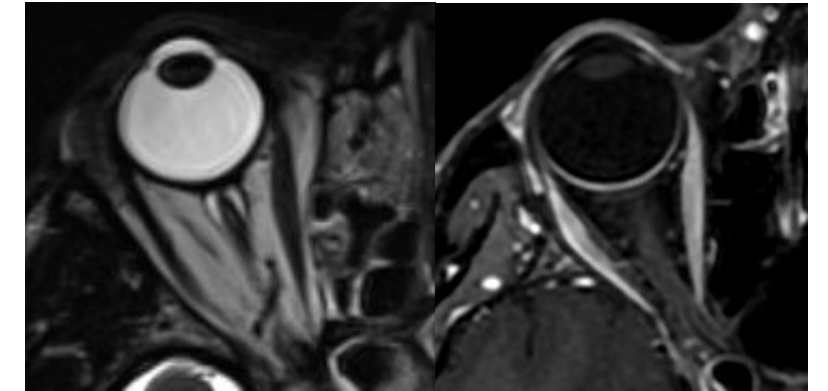




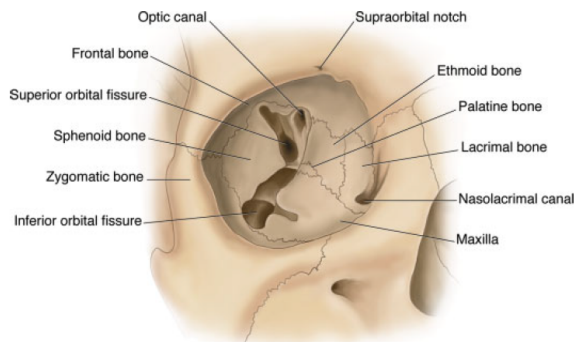
## Intraocular



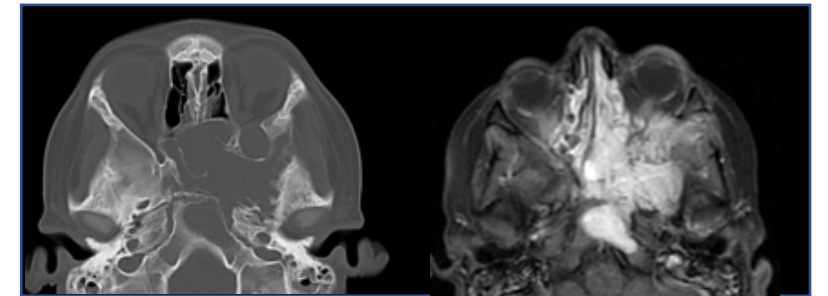
## Intraconal



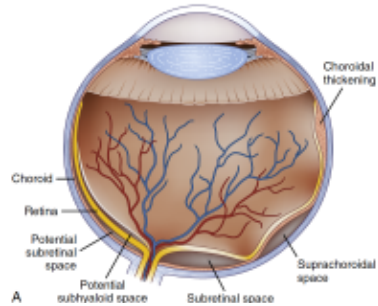
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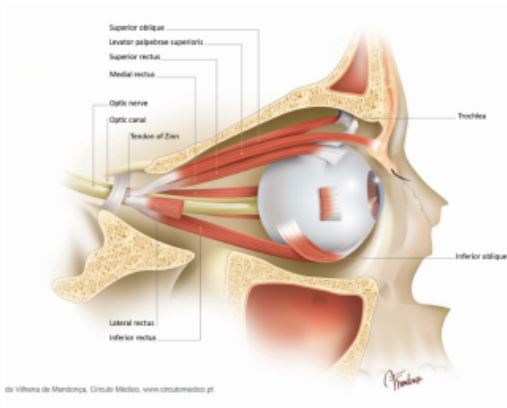
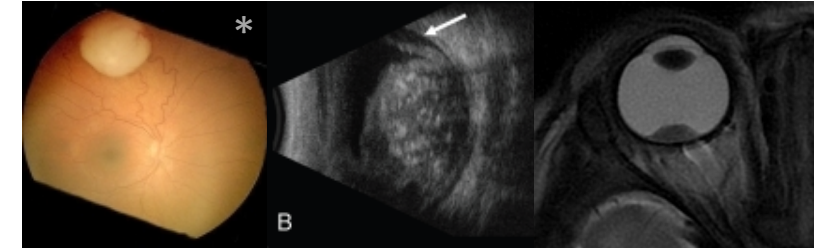
## Bony orbit



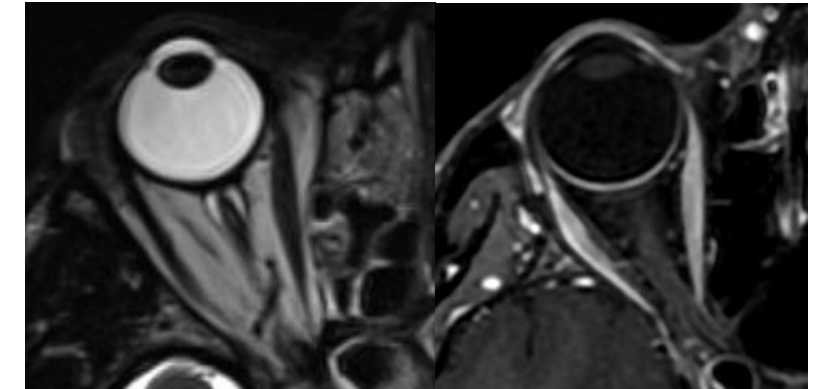




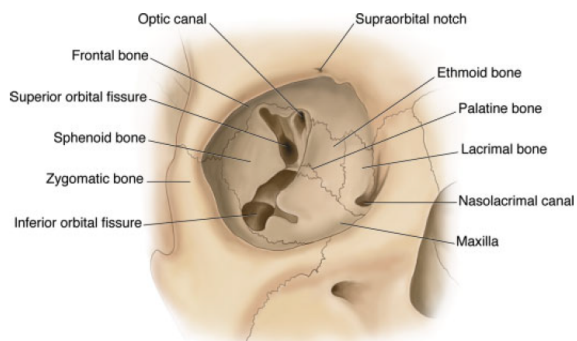
## Intraocular



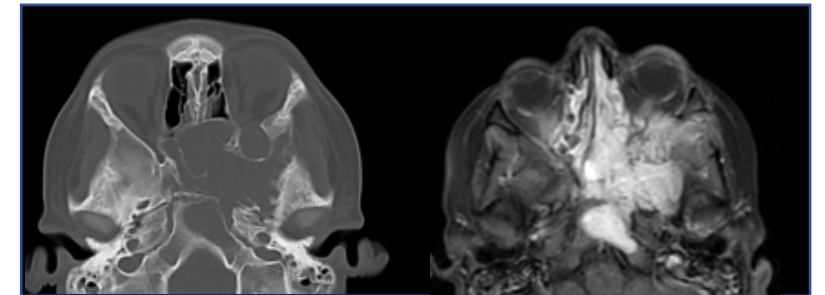
## Intraconal



## Extraconal



## Bony orbit





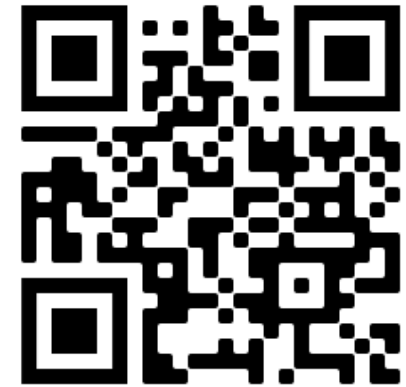


GUIDELINES



## Guidelines for magnetic resonance imaging in pediatric head and neck pathologies: a multicentre international consensus paper

Felice D'Arco<sup>1,2</sup> · Livja Mertiri<sup>1,3</sup>  · Pim de Graaf<sup>4</sup> · Bert De Foer<sup>5</sup> · Katarina S. Popović<sup>6</sup> · Maria I. Argyropoulou<sup>7</sup> · Kshitij Mankad<sup>1</sup> · Hervé J. Brisse<sup>8,9</sup> · Amy Juliano<sup>10</sup> · Mariasavina Severino<sup>11</sup> · Sophie Van Cauter<sup>12,13</sup> · Mai-Lan Ho<sup>14,15</sup> · Caroline D. Robson<sup>16</sup> · Ata Siddiqui<sup>2,17</sup> · Steve Connor<sup>2,17,18</sup> · Sotirios Bisdas<sup>19,20</sup> · on behalf of the Consensus for Magnetic Resonance Protocols Study (COMPS) Group



### Orbit

- Axial + coronal T2 WI TSE with FS
- Axial + coronal T1 WI TSE
- Axial DWI
- Axial + coronal post-Gad T1 TSE with FS

**High resolution**  
**Fat-suppressed**  
**Gad-enhanced**







# Orbital lesions: 3 step approach

## 1. Clinical features

- Age!
- Leucokoria, decreased vision, strabismus, skin changes...

## 2. Compartment + extent

- Intraocular, intraconal, extraconal, bony orbit, trans-spacial...
- Relevant neighbouring structures

## 3. Imaging characteristics: Signal intensity, vascular flow, ADC, perfusion





# Tumors of the Orbit and the Eyeball



Intraocular

Intraconal

Extraconal

Bony orbit

T2 and T1

Gad pattern

DWI

Perfusion

Likely malignant

Indetermined

Likely benign







# Tumors of the Orbit and the Eyeball



Intraocular

Intraconal

Extraconal

Bony orbit

T2 and T1

Gad pattern

DWI

Perfusion

Likely malignant

Indetermined

Likely benign







# Tumors of the Orbit and the Eyeball

Intraocular

**Retinoblastoma**

Medulloepithelioma

Intraconal

Extraconal

Non-neoplastic lesions

- Persistent fetal vasculature
- Coats disease
- Retinopathy of prematurity

Bony orbit





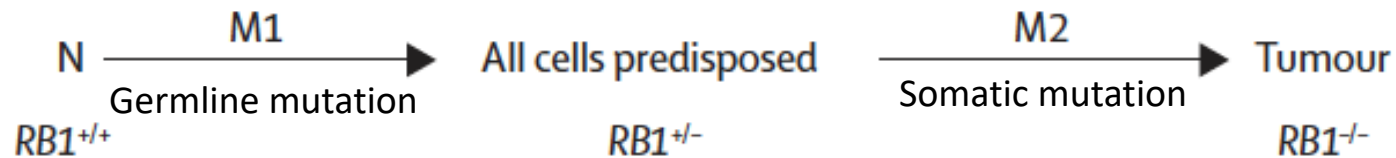
## Intraocular

## Retinoblastoma

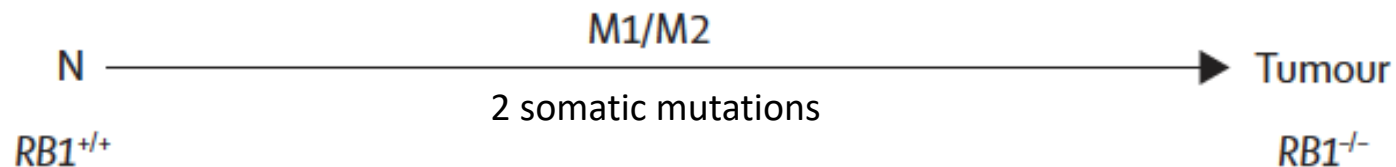
**90-95% < 5 years**

- Most common primary intraocular tumor
- Biallelic mutation in tumor suppressor **RB-1 gene** & **Two Hit Theory of Knudson**

### Heritable retinoblastoma **Avoid radiation!**



### Non-heritable retinoblastoma



### Heritable retinoblastoma (50%)

- Presentation 12 months
- Often bilateral

### Non heritable retinoblastoma

- Presentation 24 months

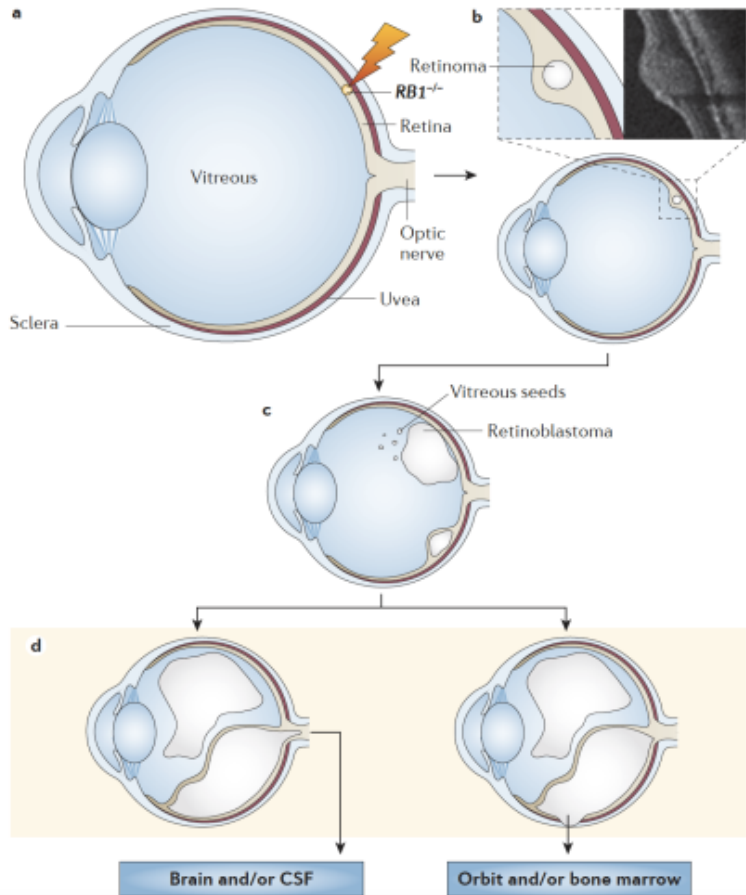




## Intraocular

## Retinoblastoma

**Dedicated MR protocol**  
**Brain MR** !

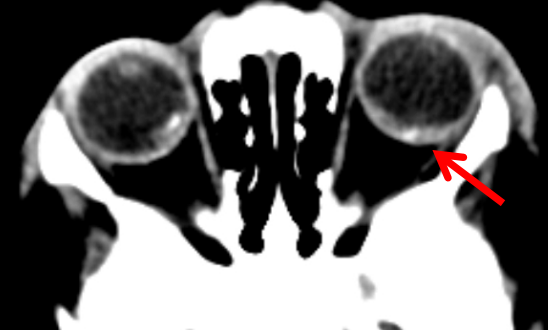
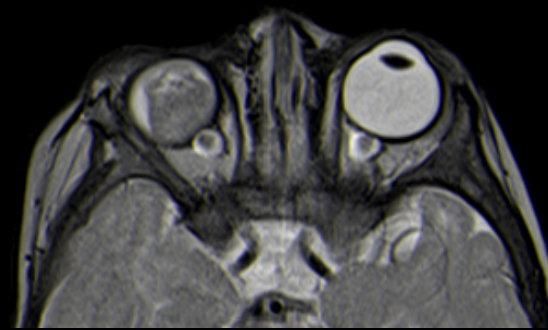
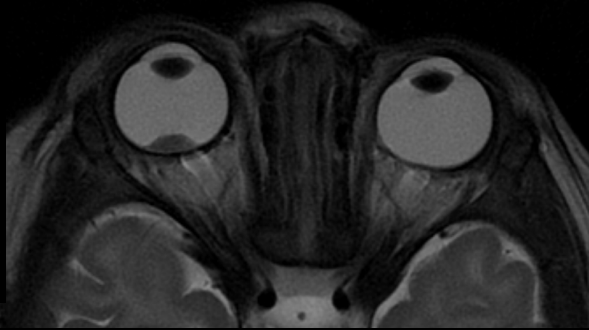
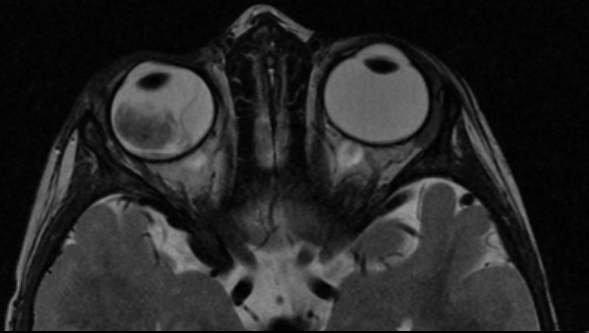


- **Calcification** in 95%, **bilateral** in 40%
- Growth pattern (endophytic, exophytic, diffuse)
- **Tumor extension**
  - **Anterior eye** segment
  - **Optic nerve** and meningeal sheath
  - **Ocular wall**: choroid + sclera
  - **Extraocular**: retrobulbar fat
- **Brain**: pineal gland, supra- or parassellar region, malformations

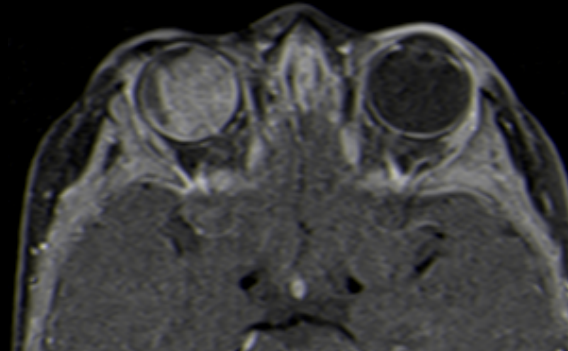
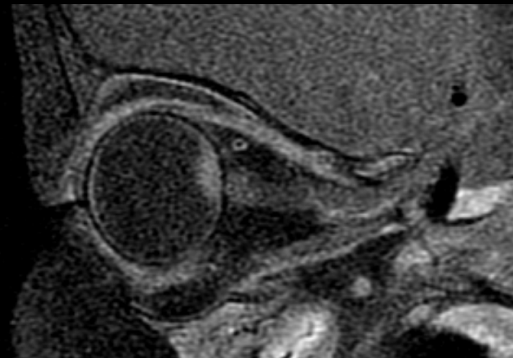
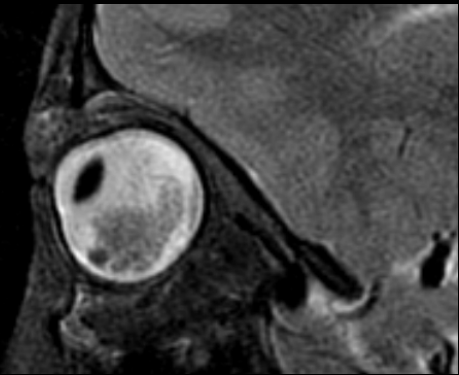


# Retinoblastoma

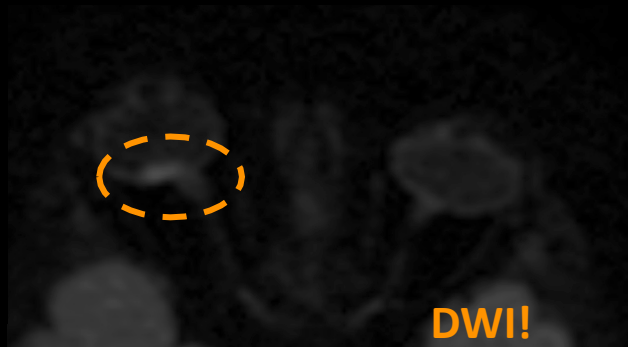
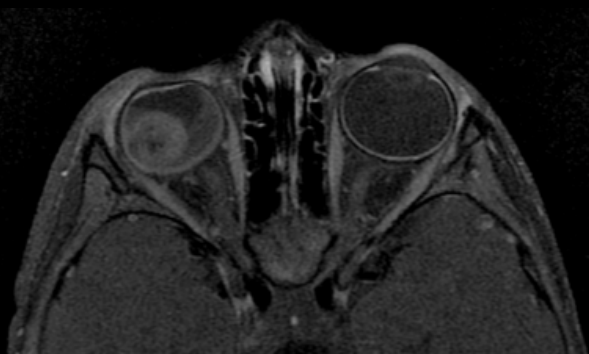
**Calcifications!**



**Bilateral retinoblastoma**



**Trilateral retinoblastoma**



**DWI!**



**SWI!**





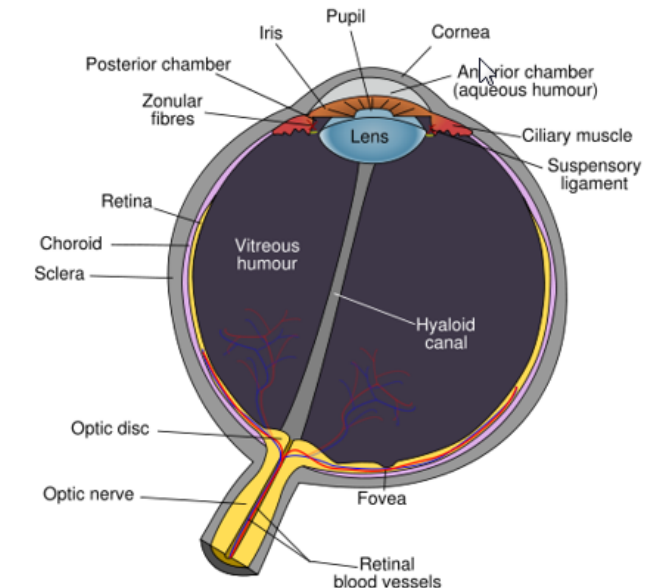
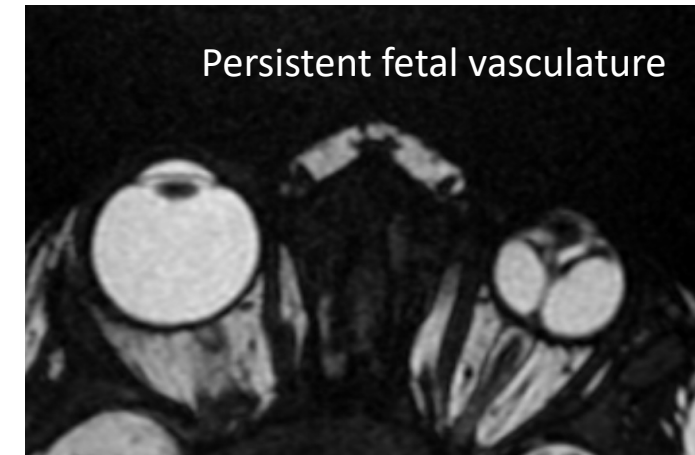
## Intraocular

## Leucokoria



**? Globe size**  
**Calcifications**  
**Uni or bilateral**

- Retinoblastoma
- Persistent fetal vasculature
- Coats disease
- Retinopathy of prematurity







# Tumors of the Orbit and the Eyeball

Intraocular

**Intraconal**

Extraconal

Bony orbit

**Optic pathway glioma**

**Optic nerve sheath meningioma**

Orbital schwannoma

Non-neoplastic lesions

- Lymphatic malformations
- Veno-lymphatic malformations





## Intraconal

# Optic pathway glioma

- Low-grade glial neoplasms, most commonly pilocytic astrocytoma **WHO grade 1**
- Most occur in children with NF1
- **NF1** vs **sporadic** optic pathway gliomas
  - NF1: more commonly in **anterior optic pathway**, frequently **bilateral**
  - Sporadic: often centered in **chiasm** and **optic tracts**
- **Gad** enhancement may **wax and wane** (sometimes resolves without treatment!)

**Peak age 2 – 8 years**

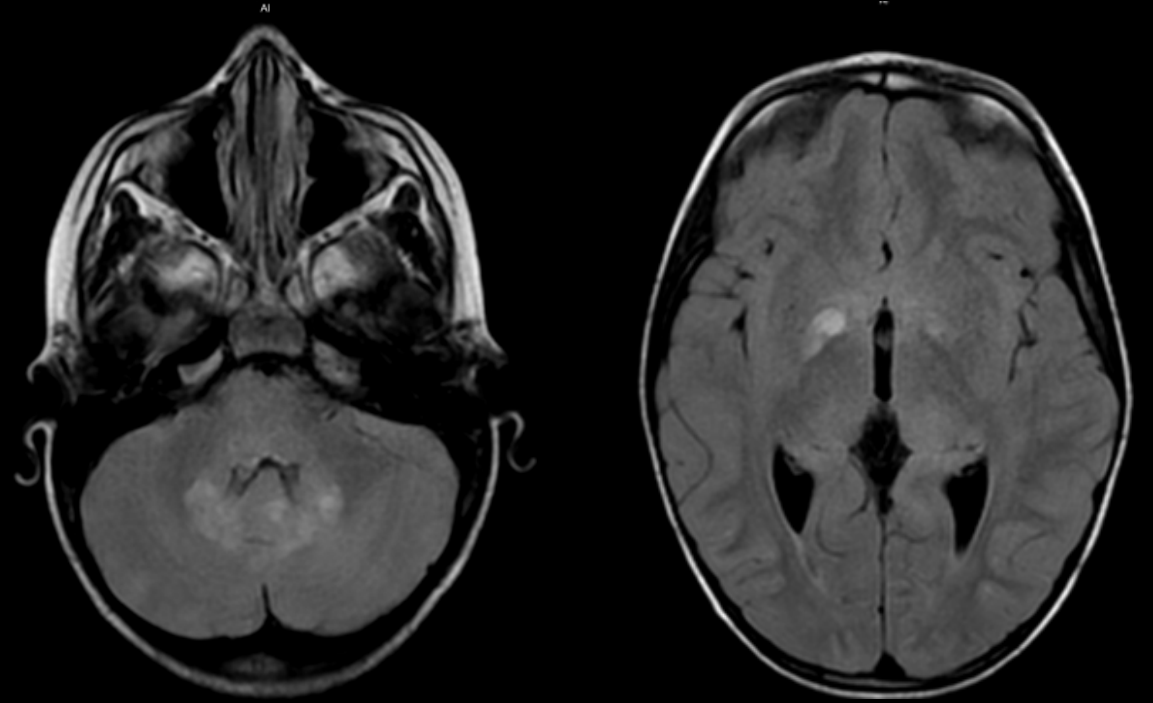


# Optic nerve glioma



Courtesy Dr Júlia Duarte

## Neurofibromatosis type 1



- FASIs – focal areas of signal intensity
- Spheno-orbital dysplasia
- Buphthalmos
- Nerve sheath tumors





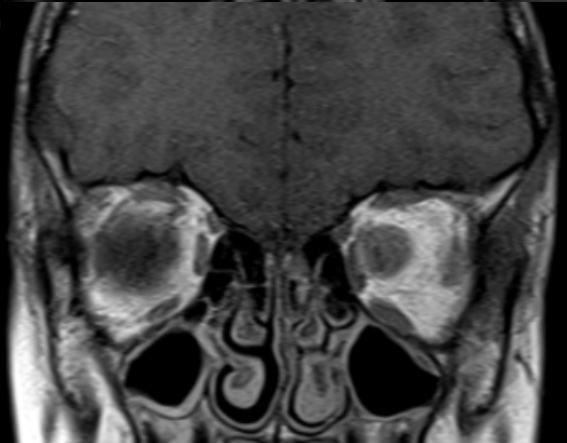
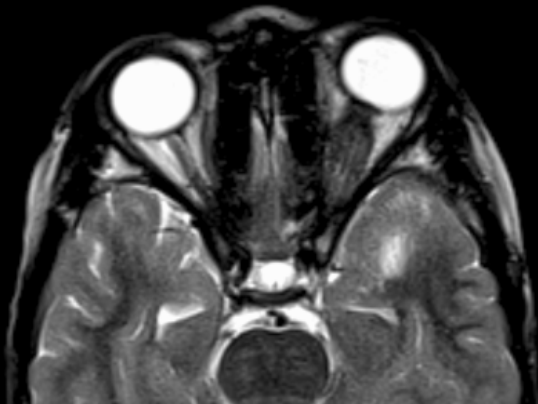
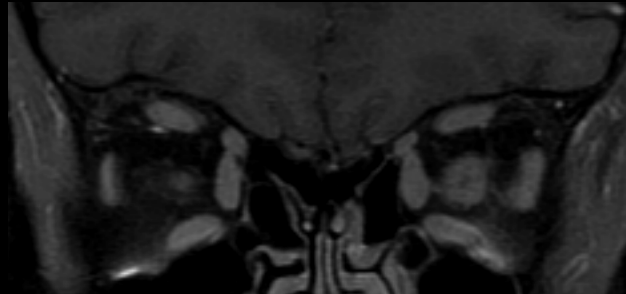
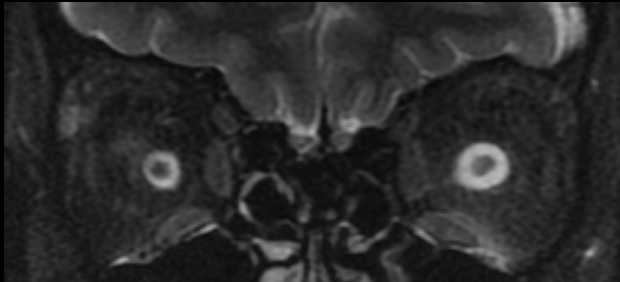
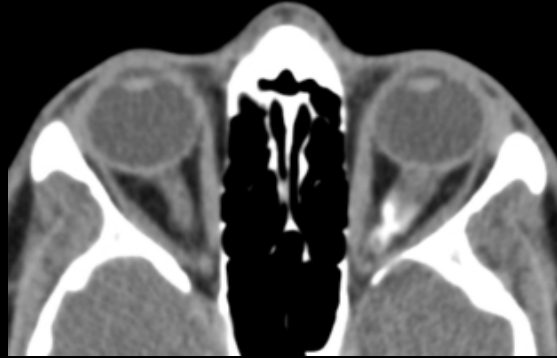
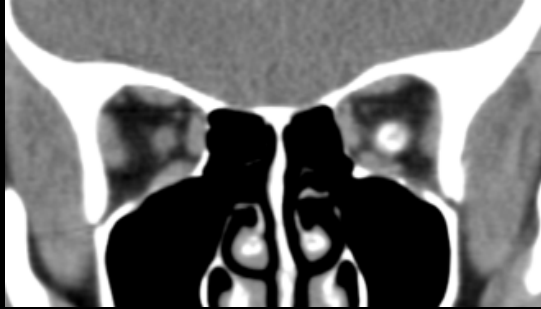
## Intraconal

# Optic nerve sheath meningioma

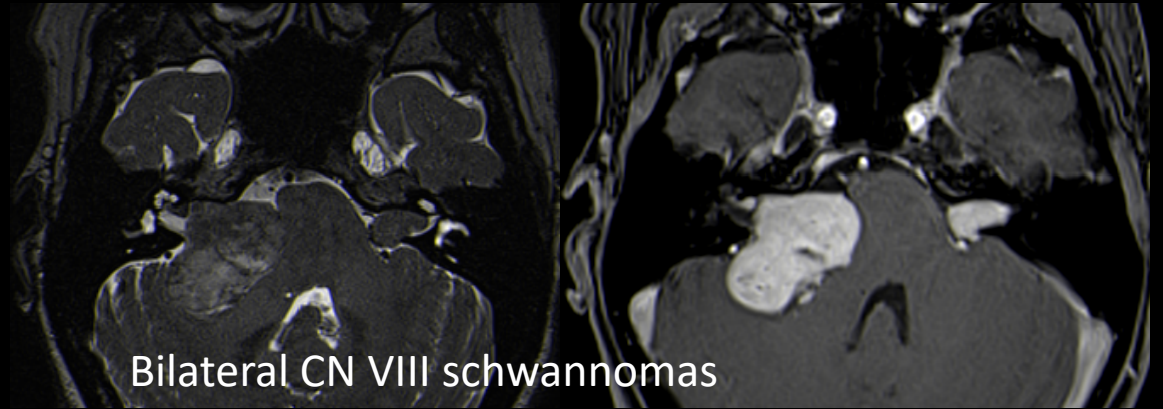
- Arise from the meninges surrounding the optic nerve
- One-third of these tumors occur in children with **NF2**
- 97% have **vision loss** at presentation
- Tubular, global, fusiform or focal enlargement of the **optic nerve sheath**
- **“Tram-track calcifications”** in one-fourth of cases



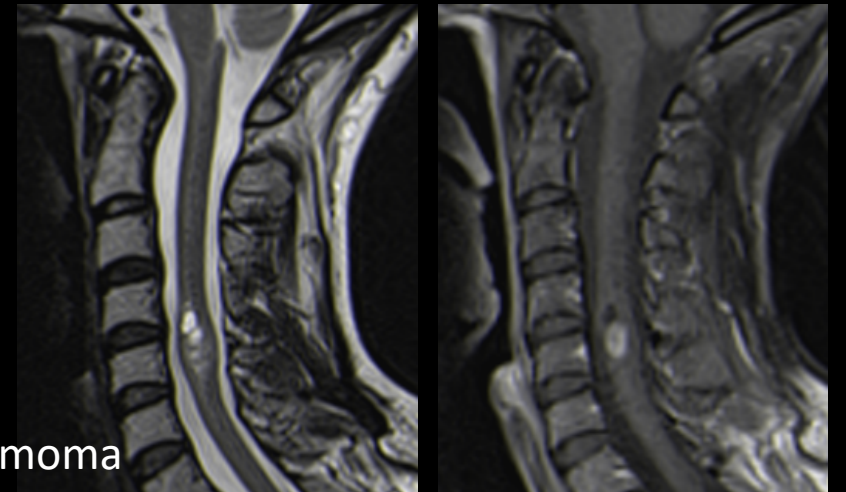
# Optic nerve sheath meningioma



## Neurofibromatosis type 2



## Ependymoma







# Tumors of the Orbit and the Eyeball

Intraocular

**Rhabdomyosarcoma**

Intraconal

**Infantile hemangioma**

**Extraconal**

Plexiform neurofibroma

Leukemia, Lymphoma

Bony orbit

Non-neoplastic lesions

- Venous varix
- Idiopathic orbital inflammation





## Extraconal

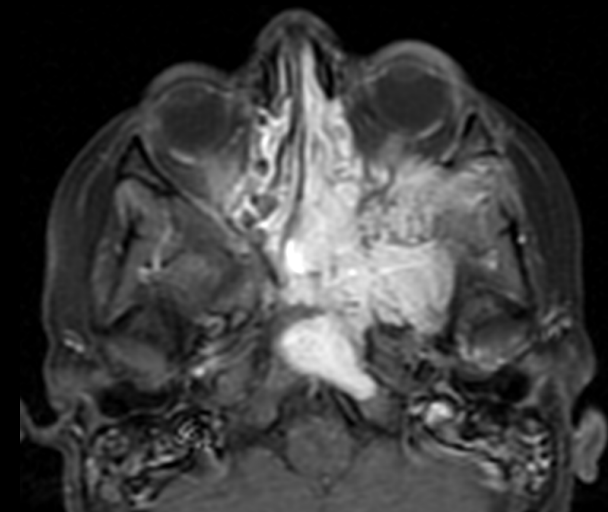
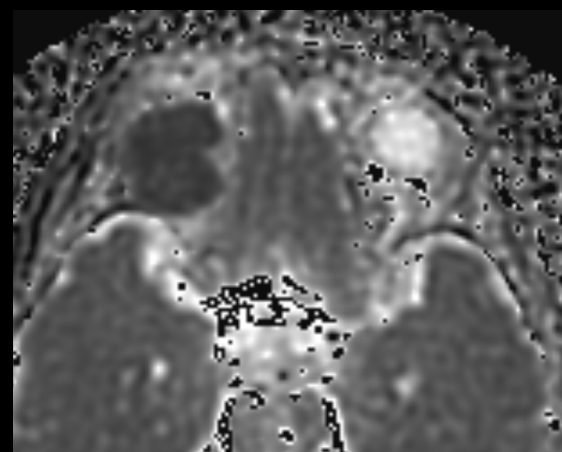
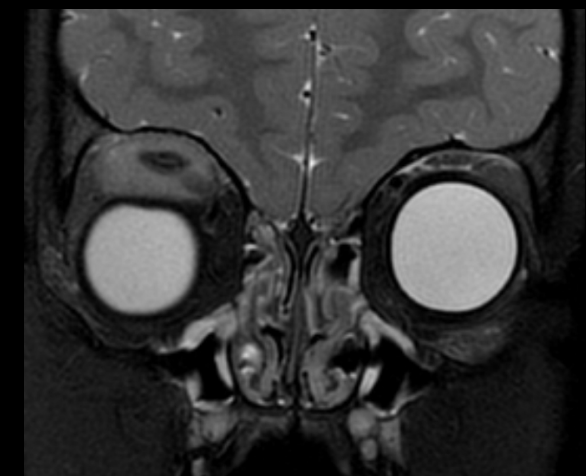
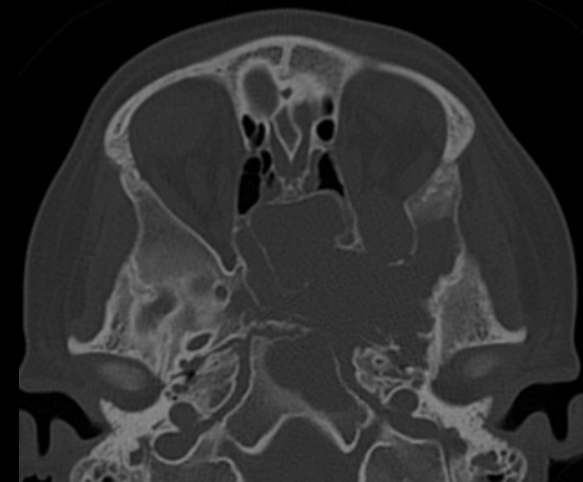
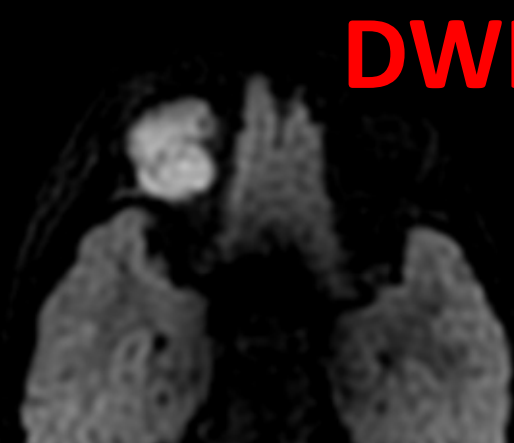
# Rhabdomyosarcoma

**6 – 8 years**

- Most common mesenchymal tumor of childhood (45% in head and neck)
- Classification
  - Orbital** (25% - 35% of head and neck rhabdomyosarcoma)
  - Parameningeal** (nasopharynx, pterygopalatine fossa, infratemporal fossa)
  - Nonparameningeal and nonorbital**
- **Unilateral** (but may be multicentric), **superonasal** quadrant
- Variable enhancement, reduced diffusivity (**ADC**  $< 1.159 \times 10^{-3} \text{ mm}^2/\text{sec}$ )



# Rhabdomyosarcoma



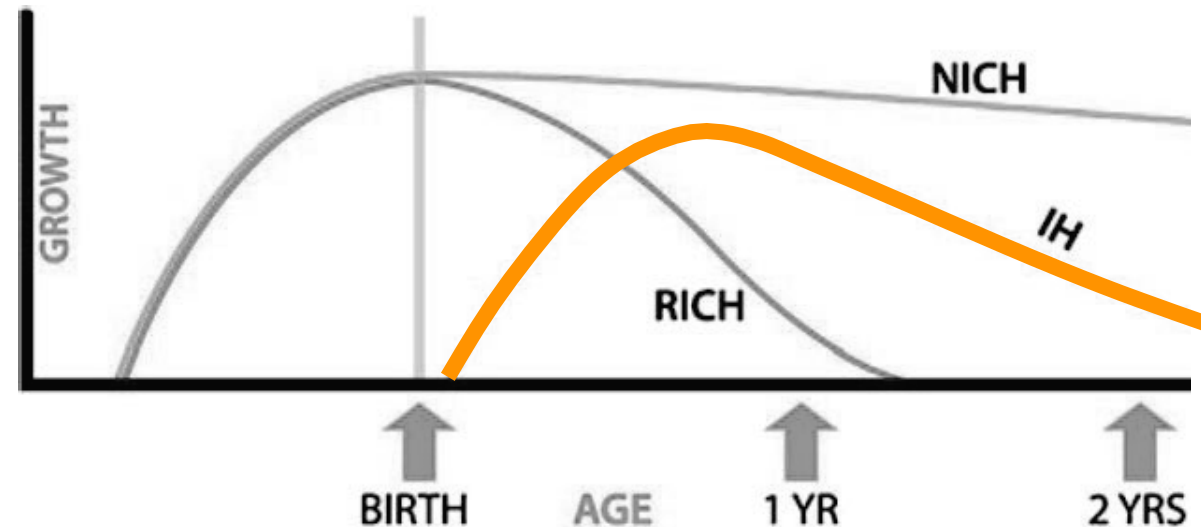




## Extraconal

# Infantile hemangioma

- Most common vascular **tumor** of infancy
- Present **shortly after birth**
- Triphasic growth
  - Proliferative phase: **flow voids, Gad**
  - Involution phase: fatty replacement
- Association with **PHACE(S) Syndrome\***

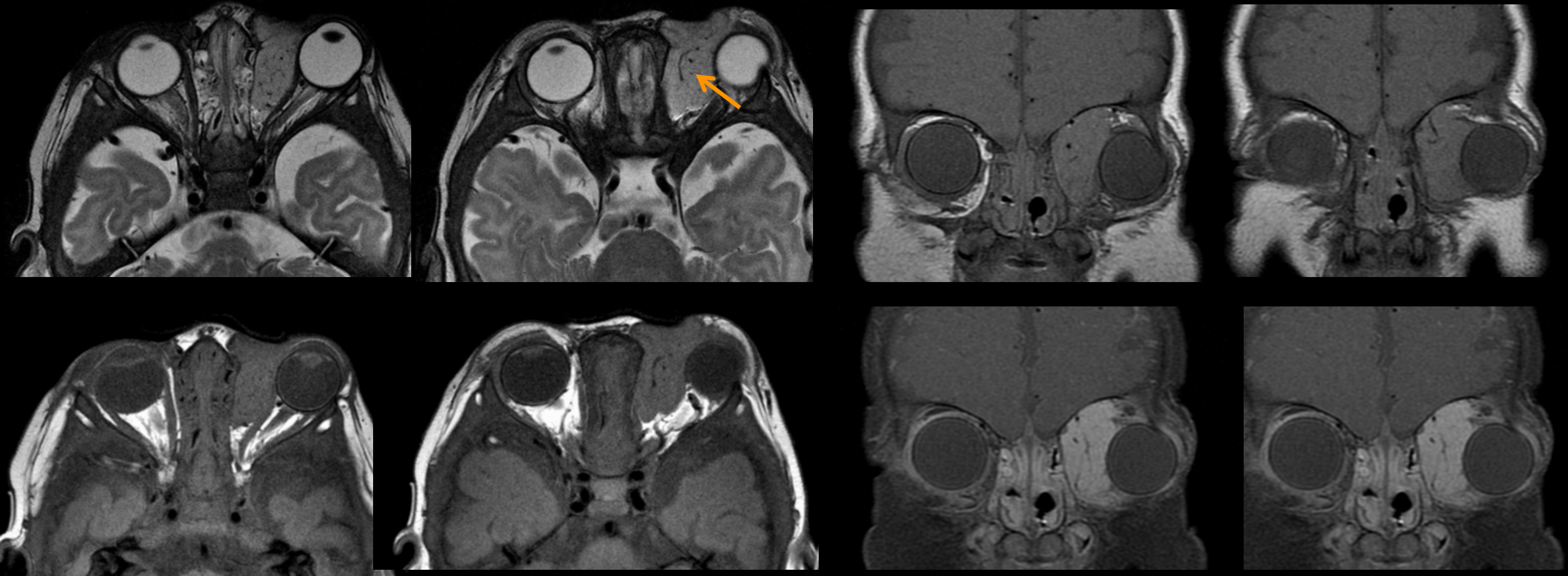


\* Posterior fossa anomalies, Hemangioma, Arterial anomalies, Coartation of the aorta, Cardiac defects, Eye abnormalities, Sternal clefting / Supraumbilical raphe syndrome



# Infantile hemangioma

Flow voids! Intense Gad enhancement!







## ISSVA Classification - Vascular Anomalies (2018)

<b>Tumors</b>	Benign: <b>Infantile hemangioma</b> , congenital hemangioma (...)	
	Locally aggressive or borderline	
	Malignant	
<b>Malformations</b>	Simple	Capillary, lymphatic, venous malformations
		Arteriovenous malformation and fistula
	Combined	CVM, CLM, LVM, CLVM, CAVM, CLAVM
	Of major named vessels	
	Associated with other anomalies	



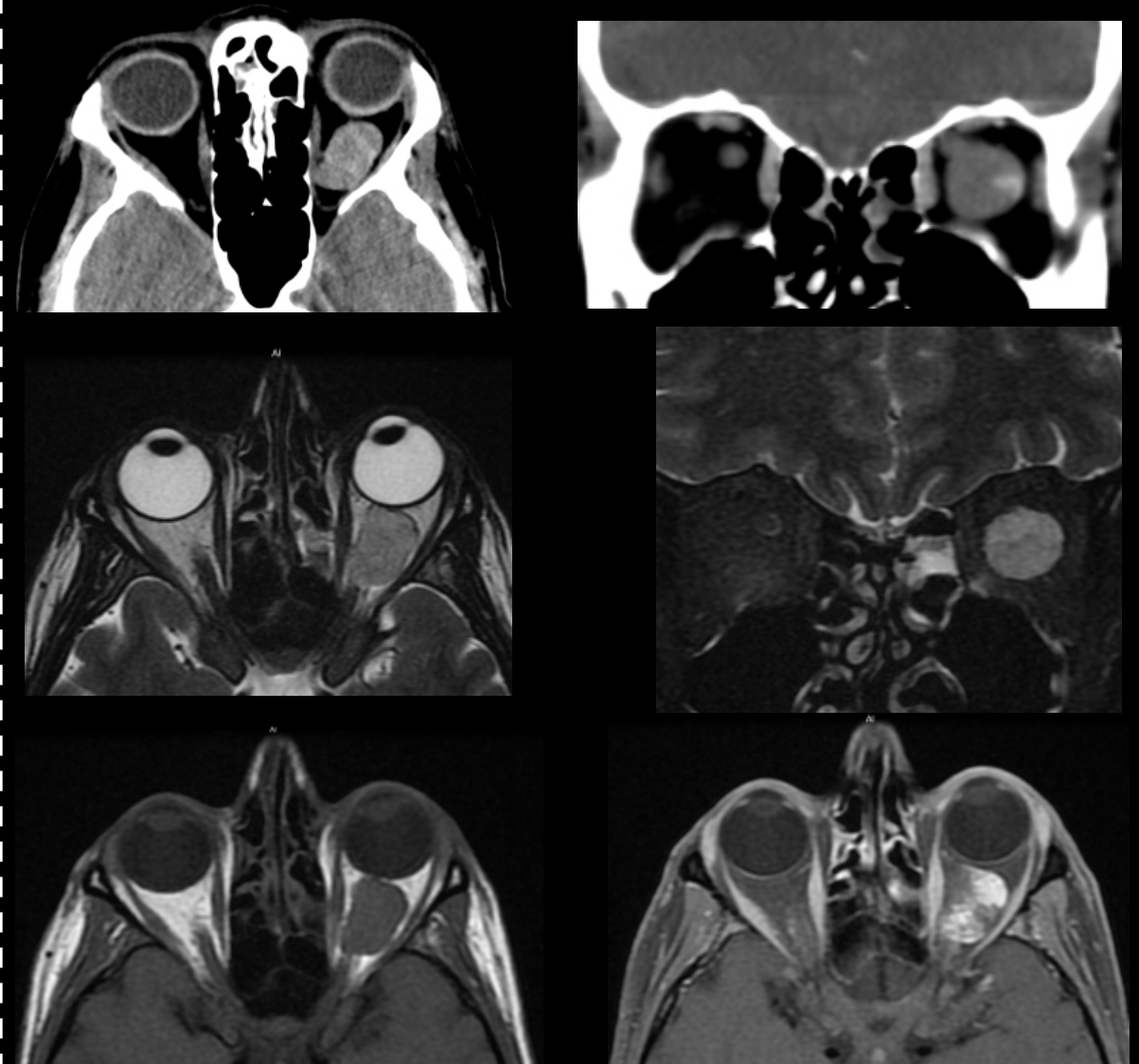
# Lymphatic malformations

Fluid-fluid levels! Macro or micro-cystic



# Venous malformations

Fleboliths! Patchy Gad enhancement







# Tumors of the Orbit and the Eyeball

Intraocular

## **Metastatic neuroblastoma**

Langerhans cell histiocytosis

Intraconal

Osteosarcoma

Extraconal

Juvenile ossifying fibroma

**Bony orbit**

Non-neoplastic lesions

- **Dermoid and epidermoid cysts**
- Fibrous dysplasia





## Bony orbit

## Metastatic neuroblastoma

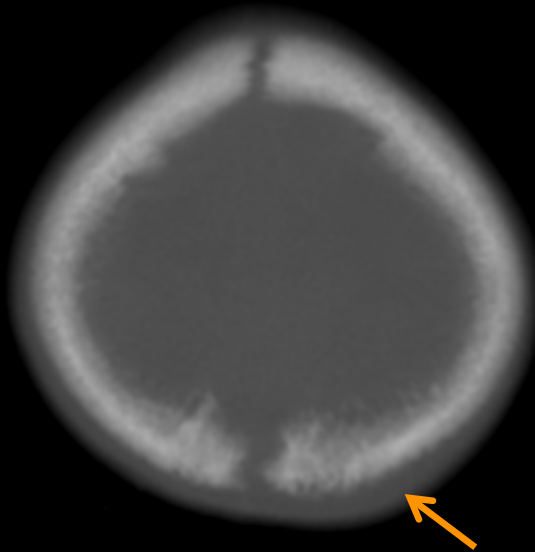
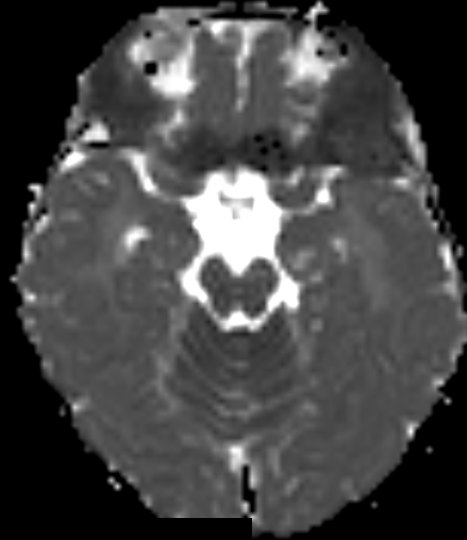
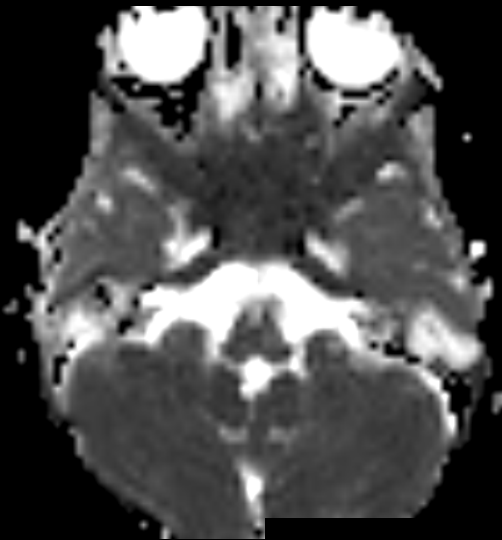
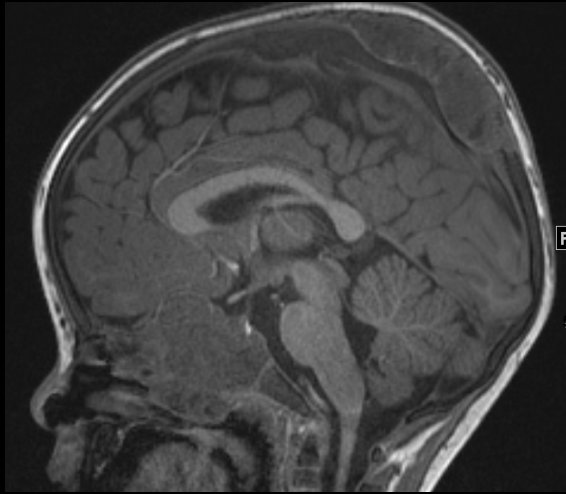
<2 years

- Most common childhood malignancy to **metastize** to the orbit (25-30% of cases)
- Proptosis + periorbital ecchymosis (**“raccoon eyes”**)
- **Lateral** or **posterior** orbital wall
- Complementary role of CT and MR
  - CT: spiculated “hair-on-end” **periosteal reaction**
  - MRI: avid **Gad** enhancement



# Metastatic neuroblastoma

Periosteal reaction!

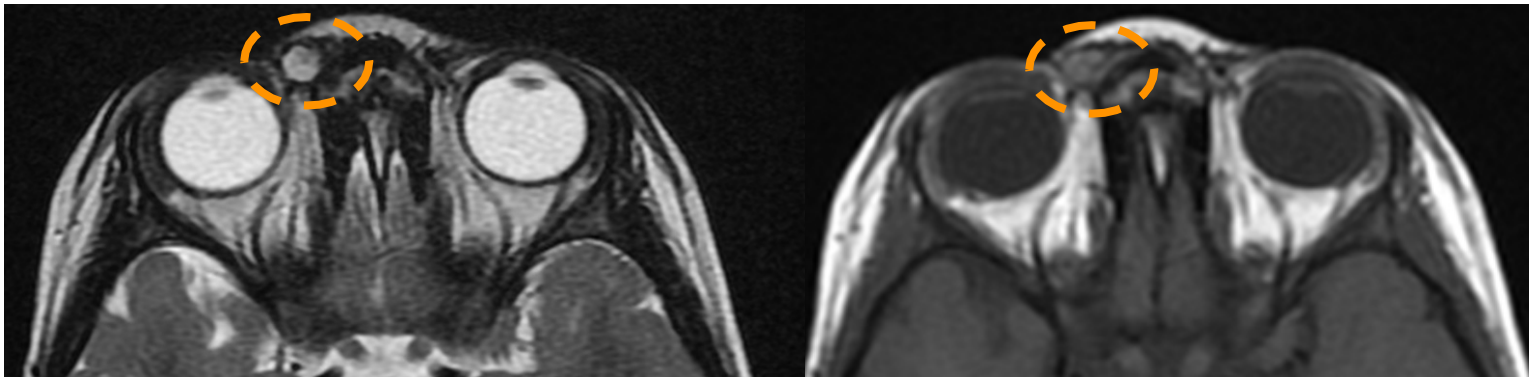


Courtesy Dr Felice D'Arco



## Bony orbit

## Dermoid and epidermoid cysts



Subcutaneous or intraorbital  
Bone remodeling  
Decreased diffusibility

- Most common **congenital** orbital lesion
- Sequestered epithelial rest in bony **sutures** (+ mesodermal derivatives in dermoids)  
(Frontozygomatic suture, fronto-ethmoidal suture)
- Inflammation secondary to rupture!





## Conclusion

- Children may be affected by a wide spectrum of orbital lesions, including both **benign** and **malignant tumors**
- **Clinical features**, a **compartment-based approach** and specific **imaging characteristics** help to narrow the differential diagnosis
- Accurate evaluation of **extension** of tumors of the orbit and the eyeball is crucial for **treatment planning** and **response assessment**







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