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OCULAR DISEASE

Capillary Haemangioma

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Introduction

- Capillary haemangioma is the most common tumour of the orbit and periorbital area in childhood.
- Girls are affected more commonly than boys (3 : 1).
- It may present as a small isolated lesion of minimal clinical significance, or as a large disfiguring mass that can cause visual impairment and systemic complications.

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- **Hamartoma:** a disorganized overgrowth of mature tissues normally present in the involved area – and due to endothelial cell proliferation.

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- **Large or multiple** lesions may have associated visceral involvement, which can lead to serious complications such as thrombocytopenia (Kasabach–Merritt syndrome, with up to 50% mortality) and high-output cardiac failure, and systemic investigation should be considered.

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Incidence…

- Infantile haemangioma in the general population is around 5%, and a small proportion of these, especially if a large facial haemangioma is present, will have PHACE (PHACES) syndrome, which includes

✓ systemic features including eye involvement

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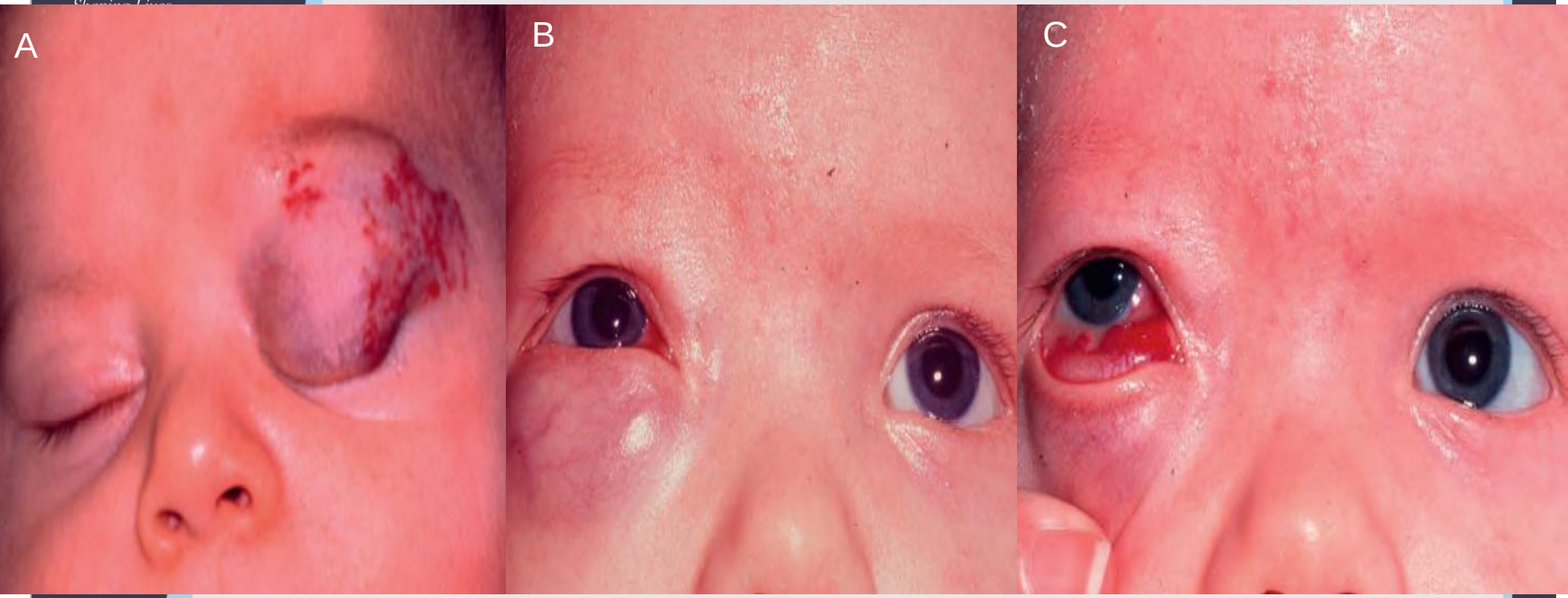
Capillary haemangioma. (A) Large preseptal tumour causing ptosis and purple cutaneous discoloration – there is a superficial component (strawberry naevus); (B) inferior preseptal tumour; (C) involvement of forniceal conjunctiva



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Diagnosis

- **Symptoms:** The lesion is usually noticed by the parent, usually in the first few months of life; approximately 30% are present at birth.

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- **Signs:** Extensive underlying orbital involvement should always be ruled out in a seemingly purely superficial lesion.

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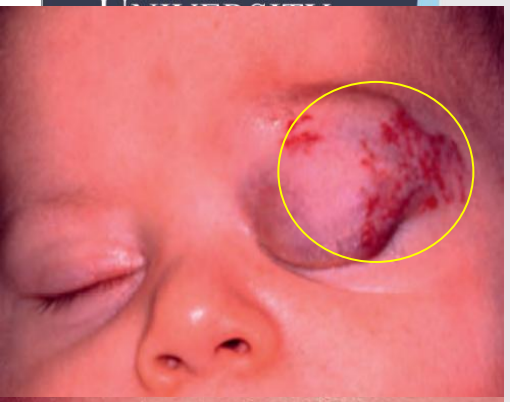
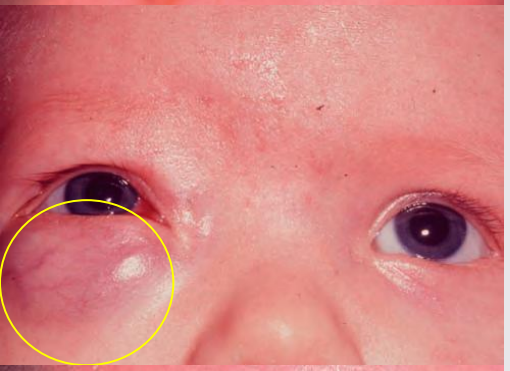
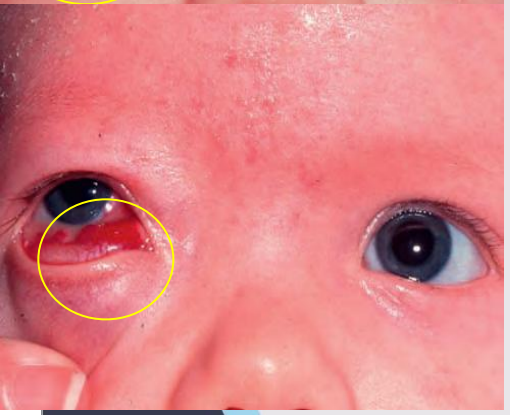


Superficial cutaneous lesions (‘strawberry naevi’) are bright red



Preseptal (deeper) tumours appear dark blue or purple through the overlying skin



Haemangiomatous involvement of the palpebral or forniceal conjunctiva is common.

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- A large tumour may enlarge and change in colour to a deep blue during crying, but both pulsation and a bruit are absent.
- Deep orbital tumours give rise to unilateral proptosis without skin discoloration.

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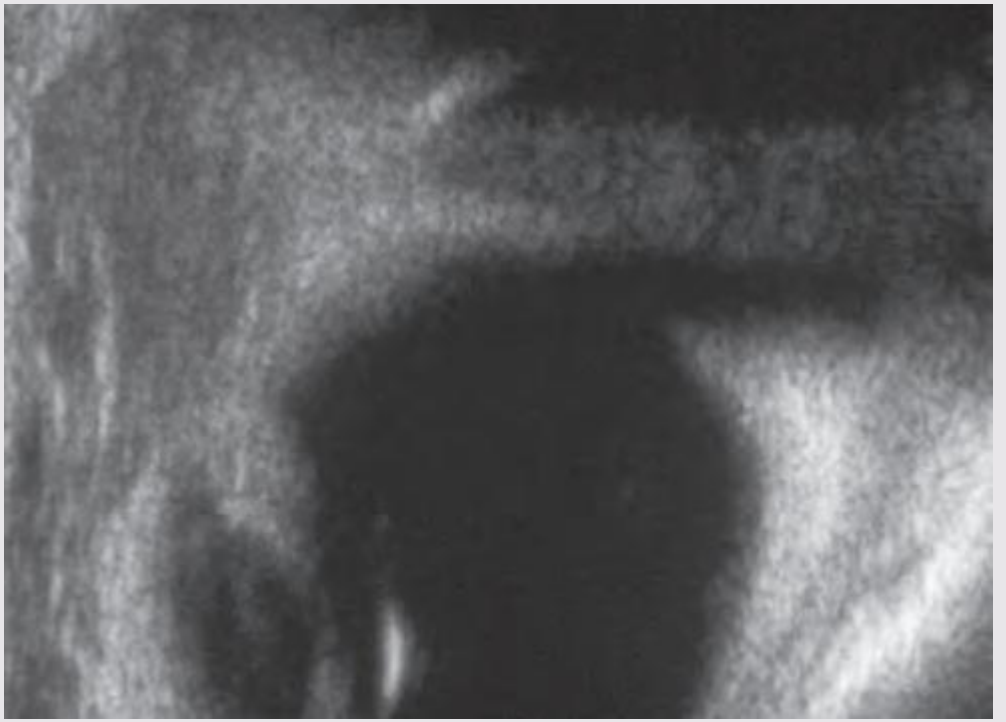
Investigation

- Imaging is generally performed for other than very small lesions, mainly to rule out more extensive orbital disease.
- Ultrasound shows medium internal reflectivity

Ultrasound of a preseptal lesion with an intraorbital component



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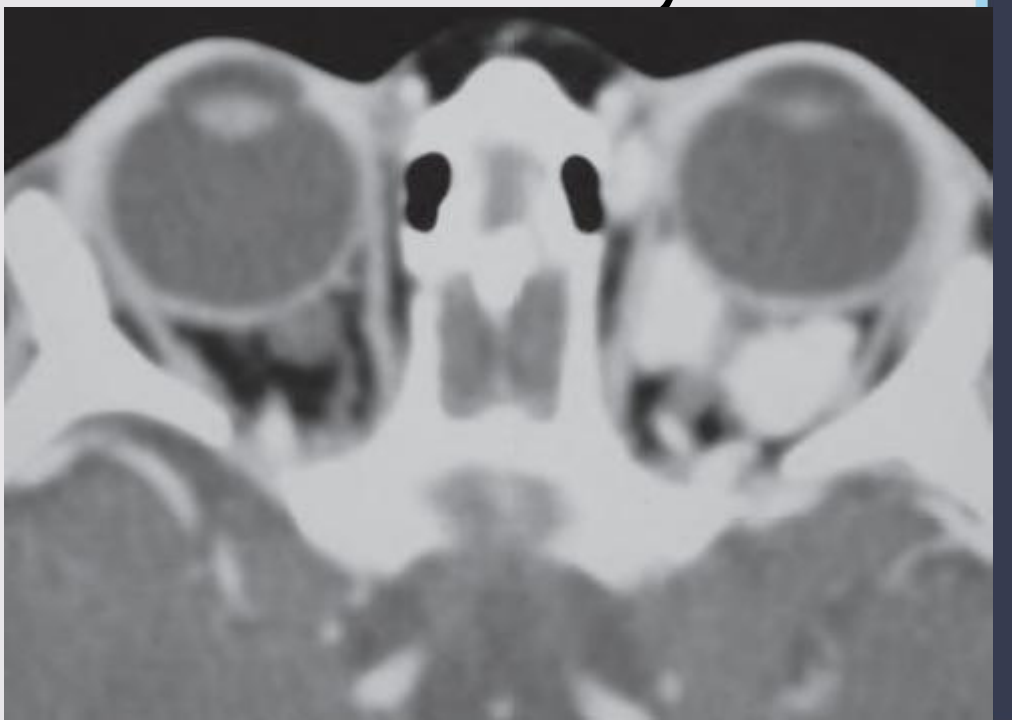
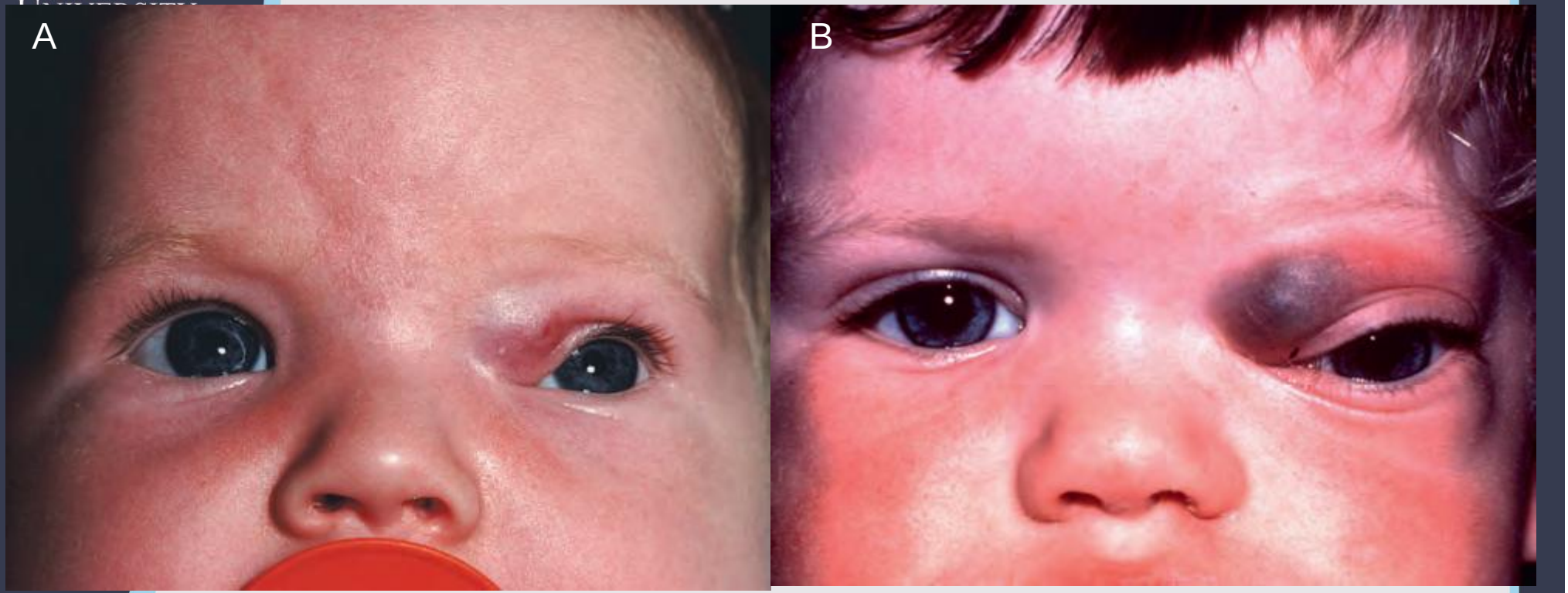
- MRI or CT the lesion appears as a soft tissue mass in the anterior orbit or as an extraconal mass with finger-like posterior expansions).
- The orbital cavity may show enlargement but there is no bony erosion

Axial enhanced CT shows a homogeneous intraconal orbital soft tissue mass



Growth of capillary haemangioma. (A) At presentation; (B) several months later



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Treatment

- The natural course is characterized by rapid growth 3–6 months after diagnosis, followed by a slower phase of natural resolution in which 30% of lesions resolve by the age of 3 years and about 75% by the age of 7.

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- Treatment is indicated principally for

✓Amblyopia

✓Secondary to induced astigmatism,

✓Anisometropia,

✓Strabismus,

✓Less commonly for optic nerve compression

✓Exposure keratopathy.

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- **Beta-blockers:**

✓ Oral propranolol is now widely used, and seems most effective in the proliferative stage; prescription and monitoring should generally be carried out by a paediatrician.

✓ Topical preparations including timolol

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- **Steroids:**

✓ Injection of triamcinolone acetonide (1–2 ml total of 40 mg/ml over several injection sites) or

✓ Betamethasone (4 mg/ml) into a cutaneous or preseptal tumour is usually effective in early lesions.

✓ Regression usually begins within 2 weeks but, if necessary, second and third injections can be given after about 2 months.

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- ✓ It is advisable not to inject deeply into the orbit for fear of causing occlusion of the central retinal artery.
- **Complications** include
- ✓ Skin depigmentation
- ✓ Necrosis,
- ✓ fat atrophy and systemic effects such as adrenal suppression.

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- ✓ Topical high-potency steroids (e.g. clobetasol propionate cream) are sometimes appropriate but are slow to exert their effect.
- ✓ Systemic steroids administered daily over several weeks may be used, particularly if there is a large orbital component.

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- ✓ **Laser** may be used to close blood vessels in superficial skin lesions less than 2 mm in thickness.
- ✓ **Interferon alfa-2a and vincristine** may be used for some steroid-resistant sight-threatening lesions.