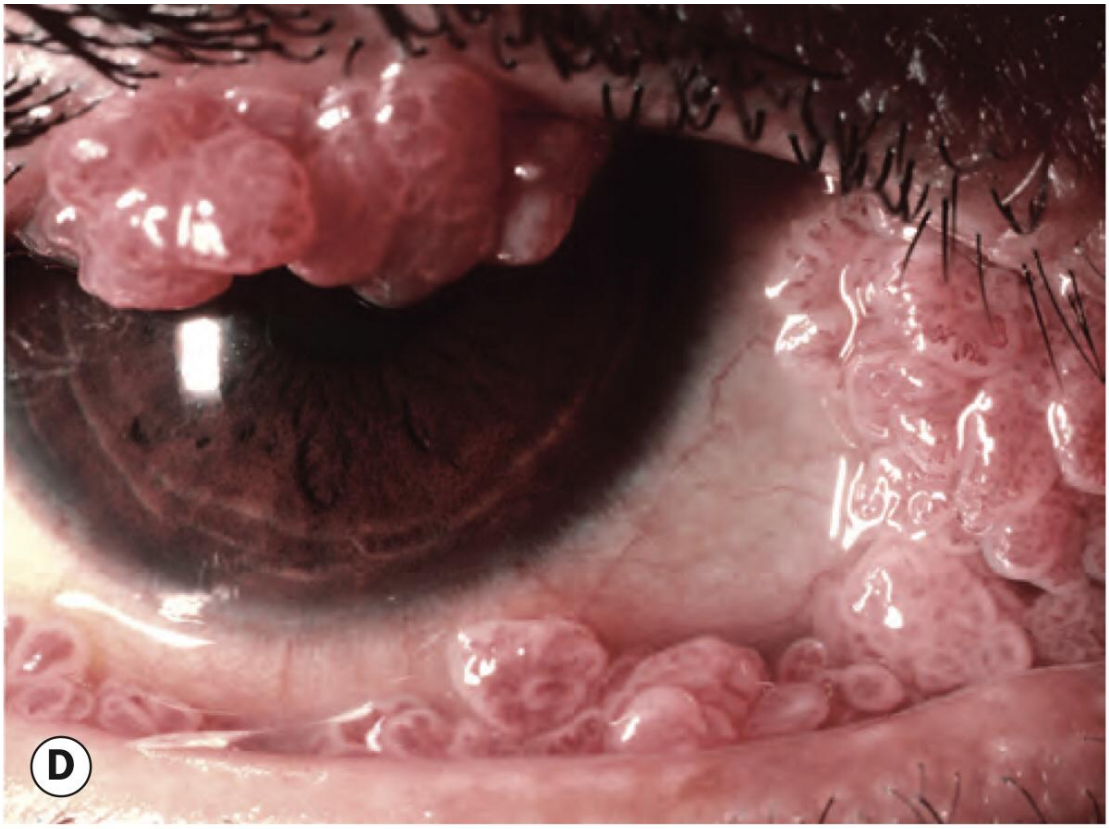
OCULAR TUMOURS



Oncology

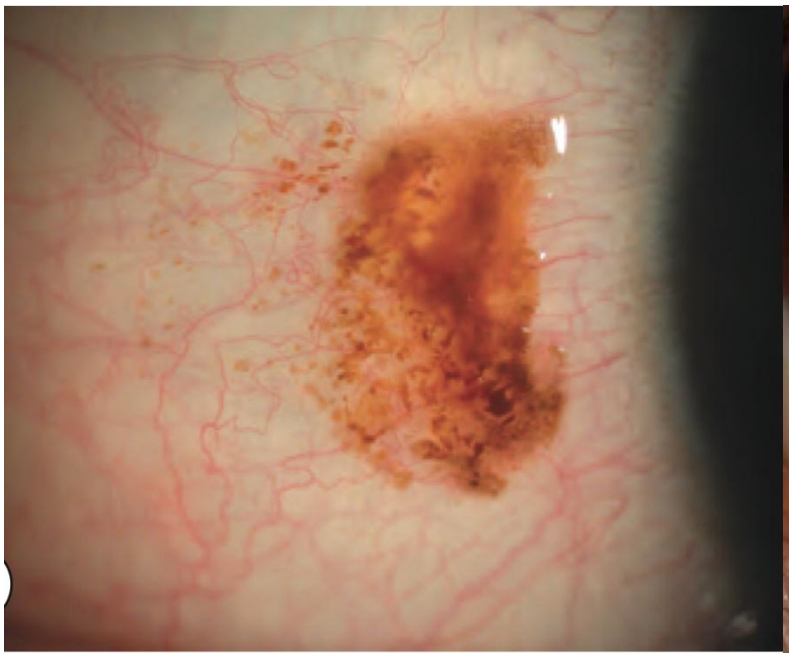
Dr.Haneen Adnan.

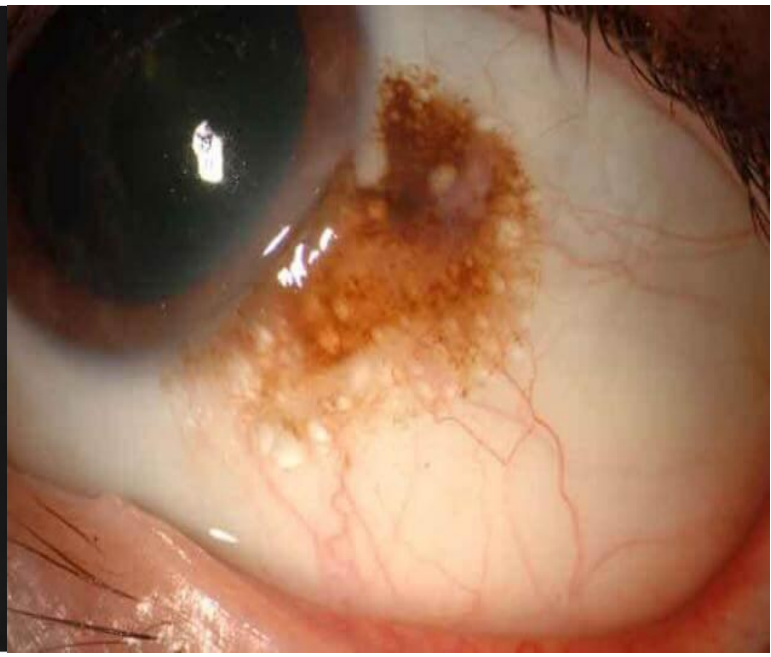
LEARNING OBJECTS

BENIGN EPIBULBAR TUMOURS

Conjunctival naevus

- It is the most common melanocytic conjunctival tumour.
- **Treatment** by excision is usually for cosmesis, irritation or a suspicion of malignancy.
- The lesion is initially noticed in the first or second decade as a solitary slightly elevated pigmented of variable size, most frequently juxtalimbal.





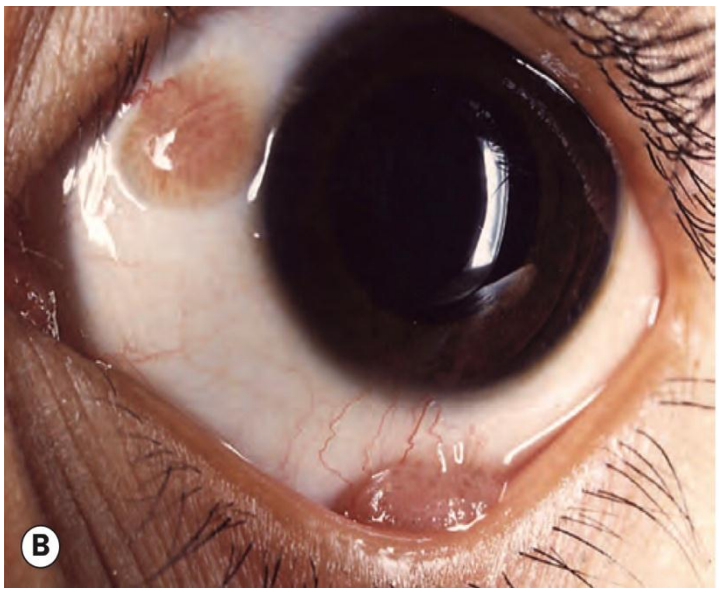
- It is mobile over the underlying sclera.

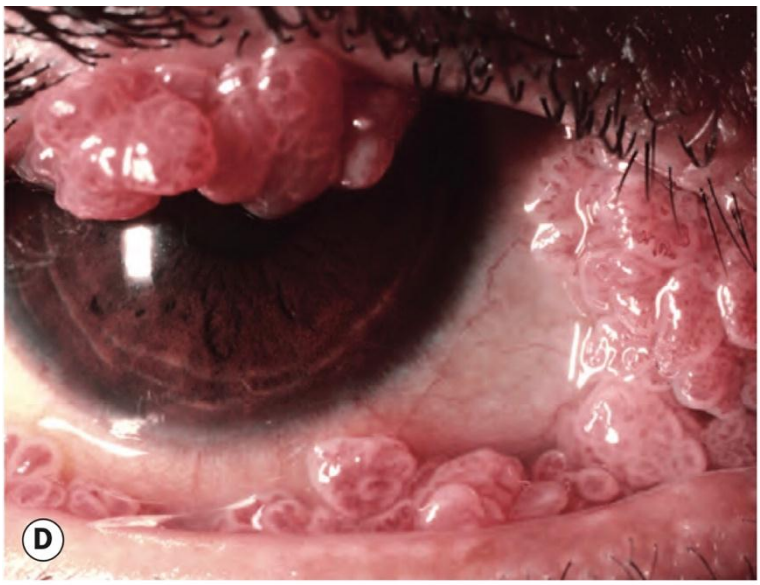
Conjunctival nevus: left pic, appearance ; right pic, contain small cysts.

Conjunctival papilloma

- strongly associated with human papillomavirus infection, especially types 6 and 11.
- The Lesions are sessile (flat) or pedunculated.
- They are usually solitary but may be multiple.
- Large lesions may cause irritation, interfere with lid closure or encroach onto the cornea.

- Small lesions may resolve spontaneously. Large lesions are



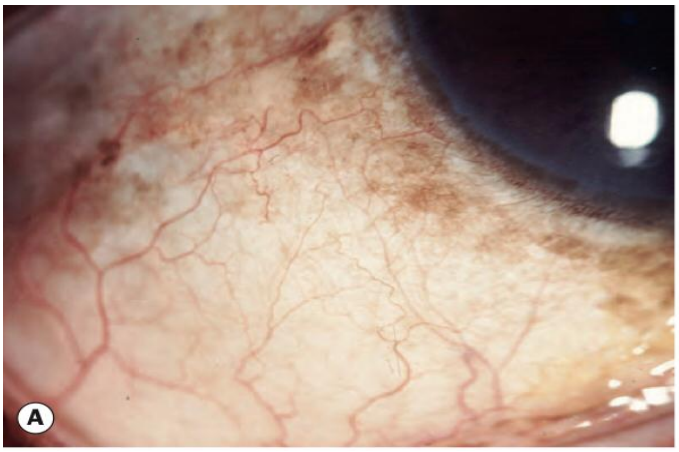


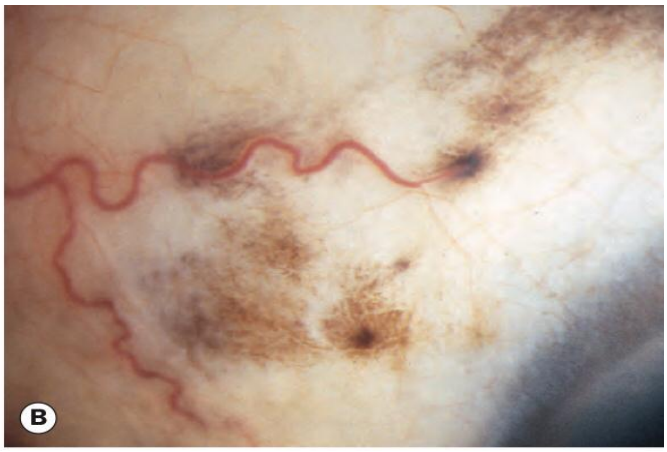
treated by excision.

Conjunctival papilloma:B.sessile. D.pedunculated.

Benign melanosis

- It is a normal variant, more common in dark-skinned individuals.
- It may have a protective effect against neoplasia.
- Both eyes are affected but involvement may be asymmetrical.
- Areas of flat, brownish pigmentation are often concentrated at the limbus and around perforating branches of vessels as





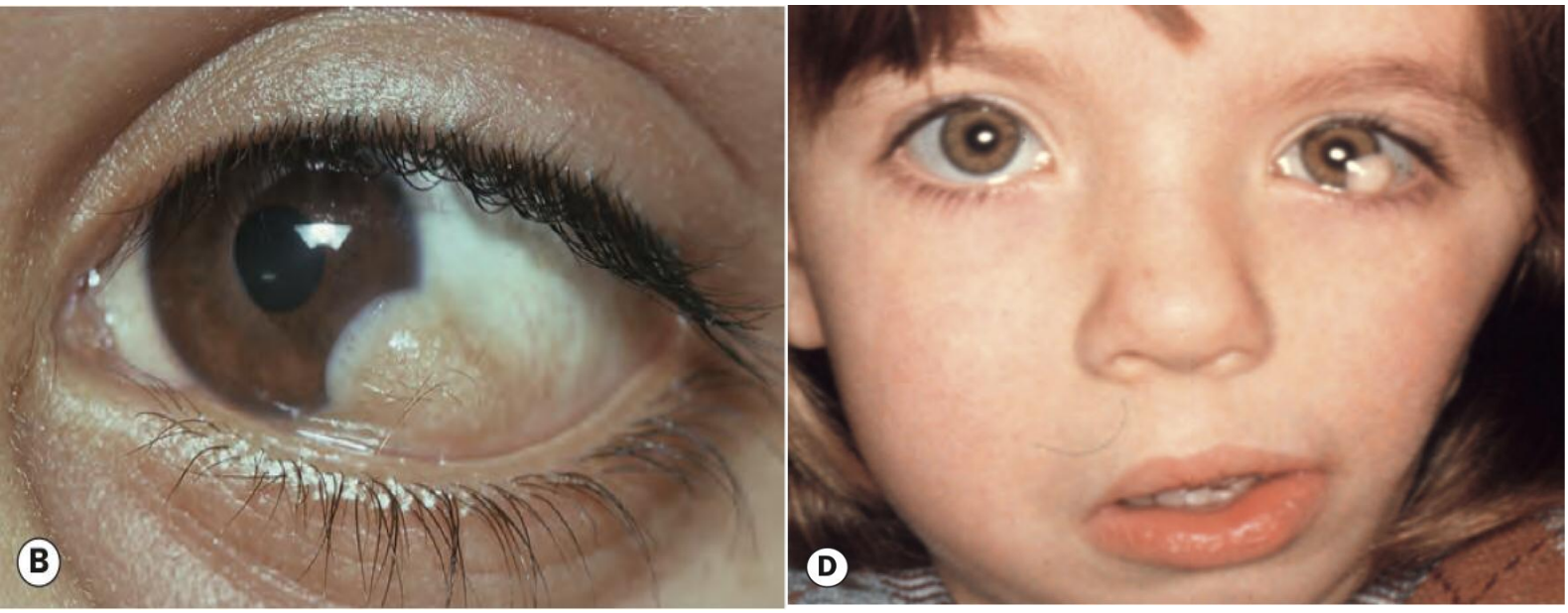
they enter the sclera.

Benign melanosis.(A)Juxtalimbal involvement (B)at site of perforating vessel and nerve

Limbal dermoid

- It is a choristoma (a mass of histologically normal tissue in an abnormal location) consisting of a mass of collagenous tissue containing dermal elements, which is covered by stratified squamous epithelium.
- **Presentation** is in early childhood, with a smooth, yellowish, subconjunctival mass commonly located at the inferotemporal limbus, often with protruding hair.
- **Treatment** is indicated for cosmesis, chronic irritation, and amblyopia from astigmatism or involvement of the visual axis. A small dermoid can undergo simple excision.

- **Systemic associations** :Goldenhar syndrome characterised by hypoplasia of the malar, maxillary and mandibular regions, preauricular and facial skin tags.

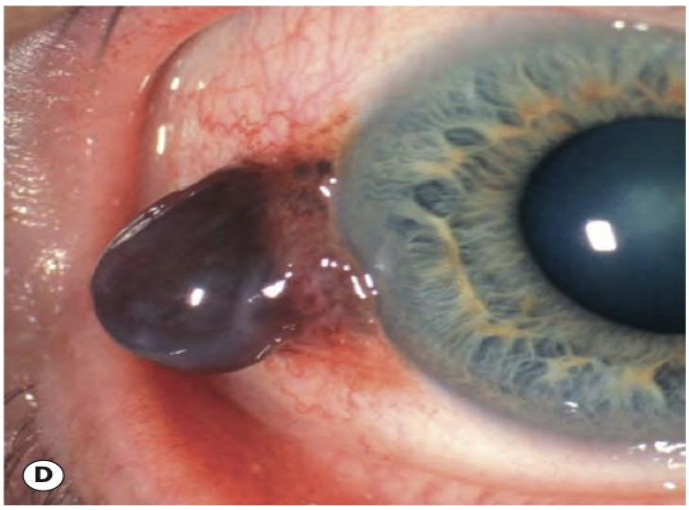


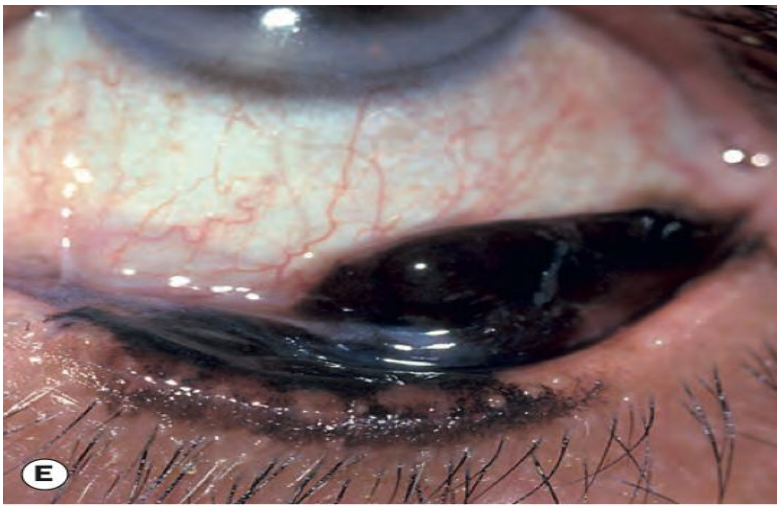
(B) typical lesion with protruding hairs; (D) Goldenhar syndrome

MALIGNANT EPIBULBAR TUMOURS

Conjunctival melanoma

- Conjunctival melanoma is an uncommon but serious condition.
- Presentation is often in the sixth decade.
- Metastasis occurs particularly to regional lymph nodes, lung, brain and liver.
- **Appearance**. A black vascularized nodule that may be fixed to the episclera. The limbus is a common site, but a melanoma may arise anywhere in the conjunctiva.
- **Treatment** by excision with a wide margin. Adjunctive radiotherapy or Cryotherapy to the bed and surrounding tissue.

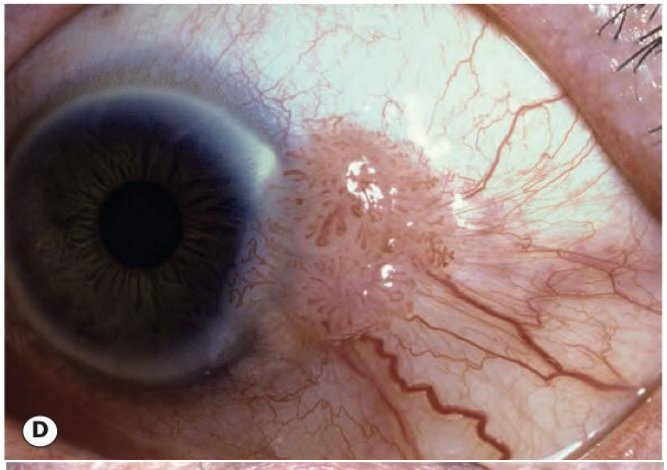


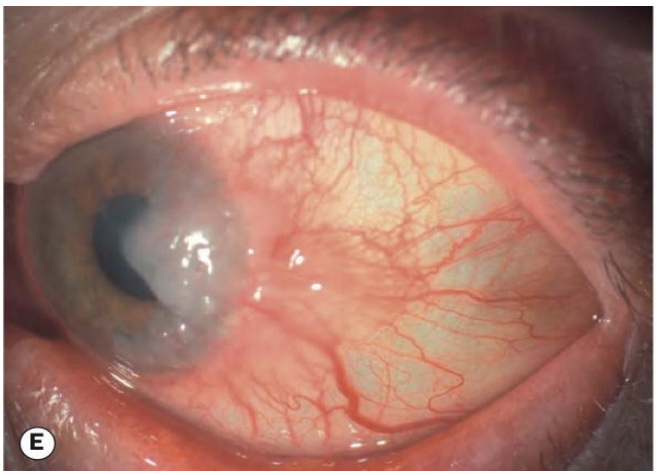


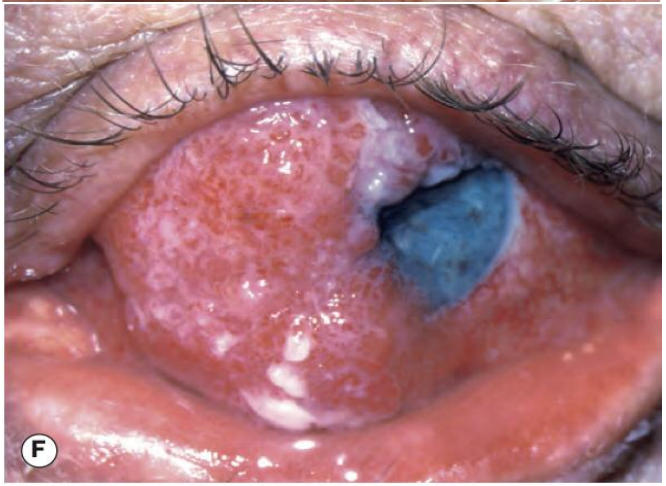
(D) conjunctival melanoma with vascularisation (E) melanoma extending onto the eyelid margin;

Ocular surface squamous neoplasia

- Are a spectrum of slowly progressive malignant epithelial lesions of the conjunctiva and cornea.
- Older adults are usually affected.
- **Risk factors** include ultraviolet light exposure, a pale complexion, smoking, acquired immunodeficiency syndrome (AIDS) and xeroderma pigmentosum. Human papilloma virus infection (especially type 16) has been implicated in some cases.
- Metastatic disease is rare.
- **Diagnosis**
    - Most tend to develop within the interpalpebral fissure, particularly at the limbus, although any part of the conjunctiva or cornea may be involved. The lesion may appear fleshy, gelatinous, leukoplakic or papillomatous, superficial or feeder vessels may be prominent. Intraocular extension is uncommon.
- **Treatment**
    - Excision with 2–4 mm margins. Adjunctive measures reduce recurrence and include cryotherapy, topical chemotherapy.
    - Topical chemotherapy may also be employed as a







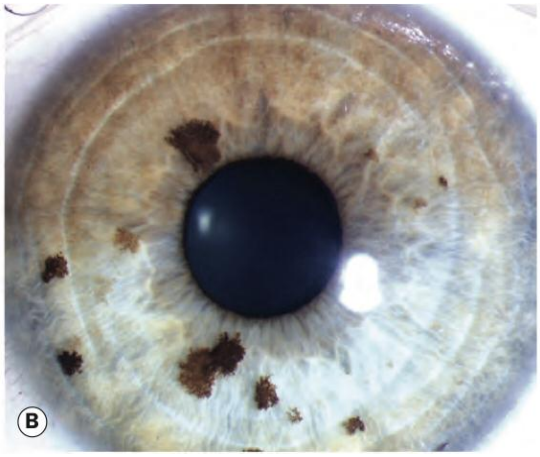
primary modality.

(D) gelatinous lesion; (E) leukoplakic lesion; (F) extensive lesion in patient with AIDS

IRIS TUMOURS

Iris naevus

- circumscribed flat pigmented lesion.
- The malignant transformation rate is up to 8% over 15 years.
- **Observation** should be lifelong and involve documentation by



slit lamp examination and photography.

B:numerous small naevi

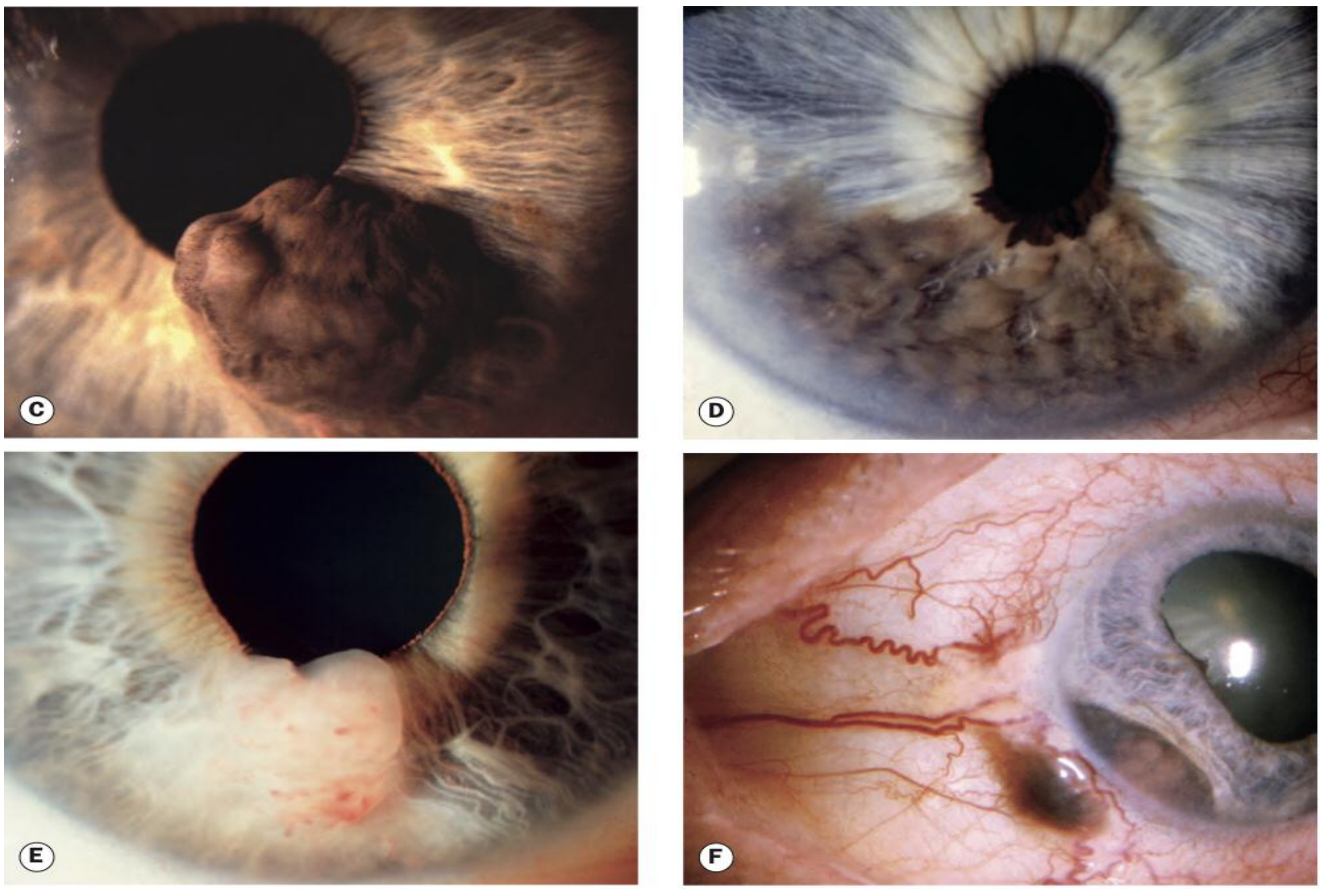
Iris melanoma

- About 8% of uveal melanomas arise in the iris.
- The prognosis is comparatively good.
- Conditions associated with or predisposing to uveal melanomas include fair skin and lighter iris colour, iris or choroidal naevus. Chronic sunlight exposure and arc welding are environmental risk factors.
- Presentation is typically in middle age.
- **Diagnosis**

- A pigmented nodule, typically located in the inferior half of the iris and often associated with surface blood vessels.
- Pupillary distortion, and occasionally localized cataract may be seen.
- A non-pigmented melanoma may occasionally be seen.
- Growth is usually slow.
- The angle may be infiltrated.
- Fine-needle aspiration biopsy may be employed prior to major surgical intervention.

➢ **Treatment**

- Sector iridectomy for small tumours.
- Radiotherapy.
- Enucleation may be required for diffusely growing tumours if radiotherapy is not possible.



(C) nodular melanoma; (D) extensive tumour with angle invasion (E) amelanotic tumour; (F) angle invasion and extrascleral extension

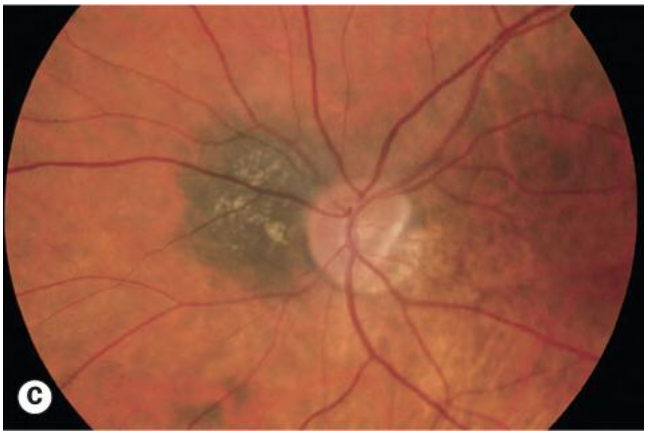
TUMOURS OF THE CHOROID

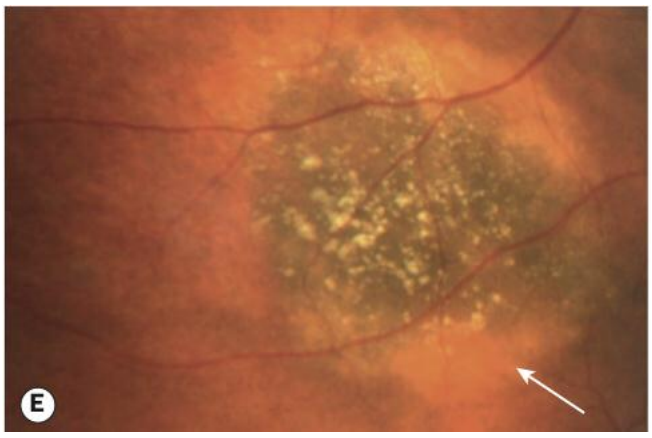
Choroidal naevus

- Growth occurs mainly during the pre-pubertal years and is extremely rare in adulthood. For this reason, detectable growth should raise a suspicion of malignancy.
- The risk of malignant transformation is up to 1%.
- **Diagnosis**
    - Usually brown lesion with indistinct feathery margins . Overlying drusen are typical. A depigmented halo is very common.
    - **Features suspicious of early melanoma**

1. The presence of overlying orange pigment.
2. The presence of associated sub-retinal fluid.
3. Acoustic hollowness on ultrasonography.
4. Symptoms such as photopsia, blurred vision.
5. Thickness greater than 2 mm.
6. Absence of drusen.
7. Absence of a halo.

- **Treatment** Baseline fundus photography and ultrasonography





or OCT, with indefinite regular review.

(C)naevus with overlying drusen; (E) naevus with overlying drusen and distinct halo (arrow);

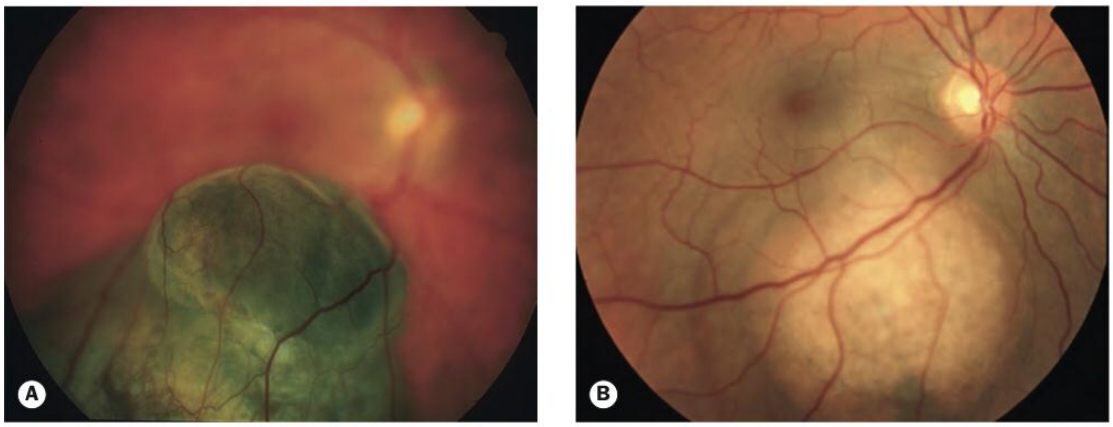
Choroidal melanoma

- the most common primary intraocular malignancy in adults and accounts for 80% of all uveal melanomas, but it is still relatively uncommon.
- Predisposing factors are as for iris melanoma (above).
- Presentation peaks at around the age of 60 years.
- Symptoms are often absent, with a tumour detected by chance on routine fundus examination.
- **Signs**
  - A solitary elevated sub-retinal grey-brown or dome-shaped mass.
  - About 60% are located within 3 mm of the optic disc or fovea.
  - Clumps of overlying orange pigment are common
  - If the tumour breaks through the Bruch membrane it acquires a 'collar stud' appearance.
  - Associated sub-retinal fluid are common.
- **Investigation**
- Examination is sufficient for diagnosis in the majority of cases.
- FA The most common findings are an intrinsic tumour ('dual') circulation.
- Ultrasound is used to measure lesion dimensions. 'acoustic hollowing' is typical and is due to the greater tissue homogeneity in this region. A 'collar stud' configuration is almost pathognomonic when present.
- OCT measures dimensions and may demonstrate associated sub-retinal fluid.

- Magnetic resonance imaging (MRI) is useful to demonstrate extra-ocular extension.
- Systemic investigation is directed principally towards detecting metastatic spread. Liver function testing and ultrasonography are mainstays. Chest radiography rarely shows lung secondaries in the absence of liver disease.

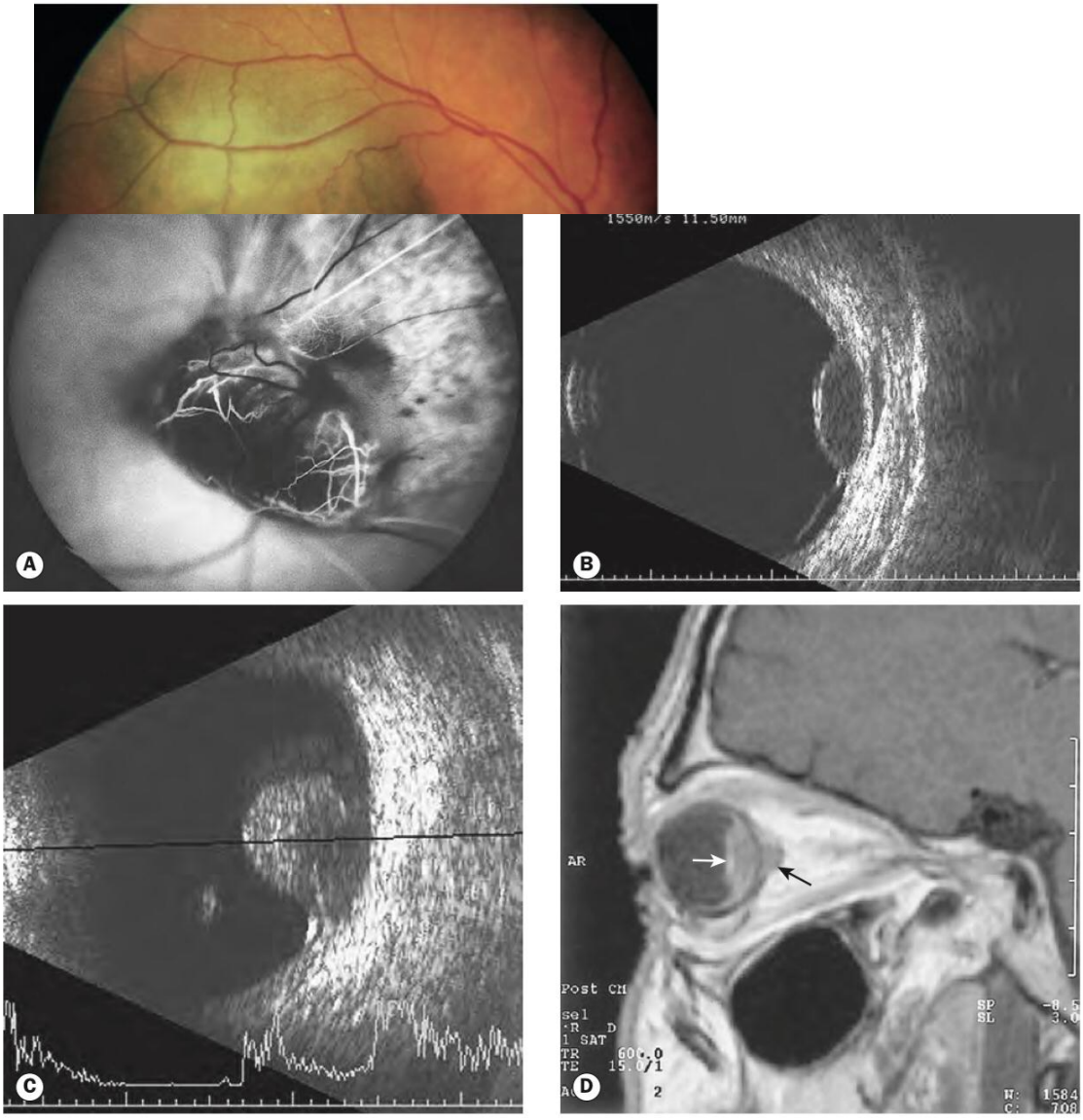
➢ **Treatment**

1. Brachytherapy (episcleral plaque radiotherapy).
2. External beam radiotherapy.
3. Enucleation.

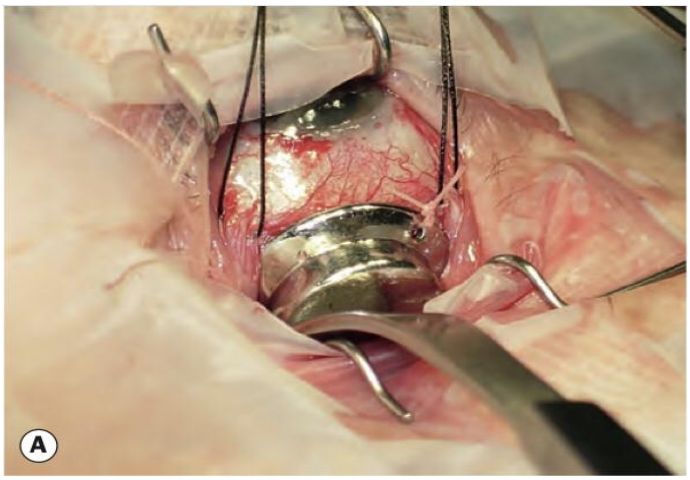


(A) Typical pigmented melanoma; (B) amelanotic lesion;

Imaging of choroidal melanoma. (A) FA of a 'collar stud' tumour showing a 'dual circulation'; (B) B-scan of a dome-shaped tumour; (C) B-scan of a 'collar stud' tumour; (D)MRI showing a choroidal melanoma (white arrow) and extra ocular extension (black arrow)



(D) overlying orange pigment (arrow)



Brachytherapy for choroidal melanoma.

NEURAL RETINAL TUMOURS

Retinoblastoma

Retinoblastoma is rare, but is the most common primary intraocular malignancy of childhood. Survival rates are over 95% in specialized centres, with preservation of vision in a majority of eyes, but are much lower in the developing world. Optic nerve invasion may occur, with spread of tumour along the subarachnoid space to the brain. Metastatic spread is to regional nodes, lung, brain and bone.

Genetics

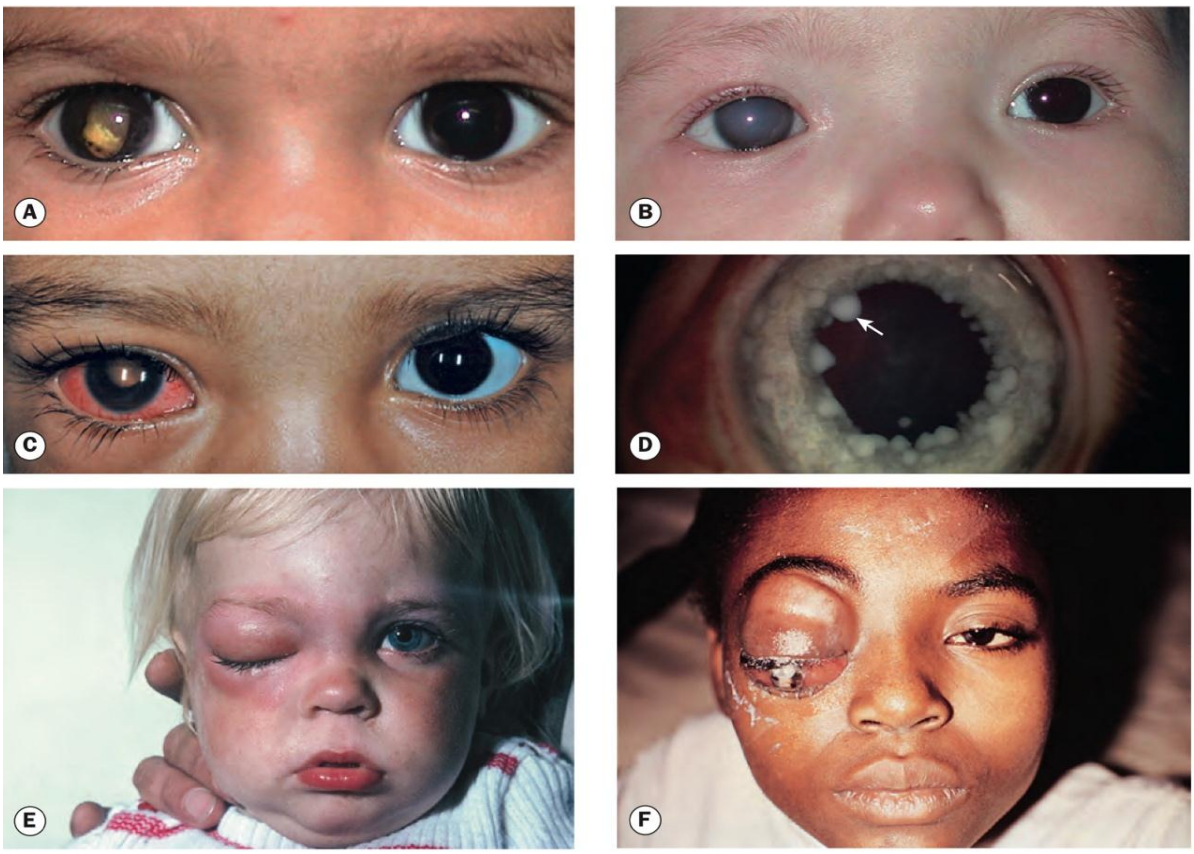
The tumour suppressor gene in which mutations predisposing to retinoblastoma occur is RB1.

1. ***Heritable (hereditary, germline) retinoblastoma*** *accounts for 40%. majority of these children develop bilateral and multifocal tumours. Heritable retinoblastoma patients also have a predisposition to non-ocular cancers such as pinealoblastoma, osteosarcoma, soft tissue sarcoma and melanoma.*
2. ***Non-heritable (non-hereditary, somatic) retinoblastoma*** *The tumour is unilateral, not transmissible and does not predispose the patient to second non-ocular cancers.*

Clinical features

Presentation is within the first year of life in bilateral cases and around 2 years of age if the tumour is unilateral.

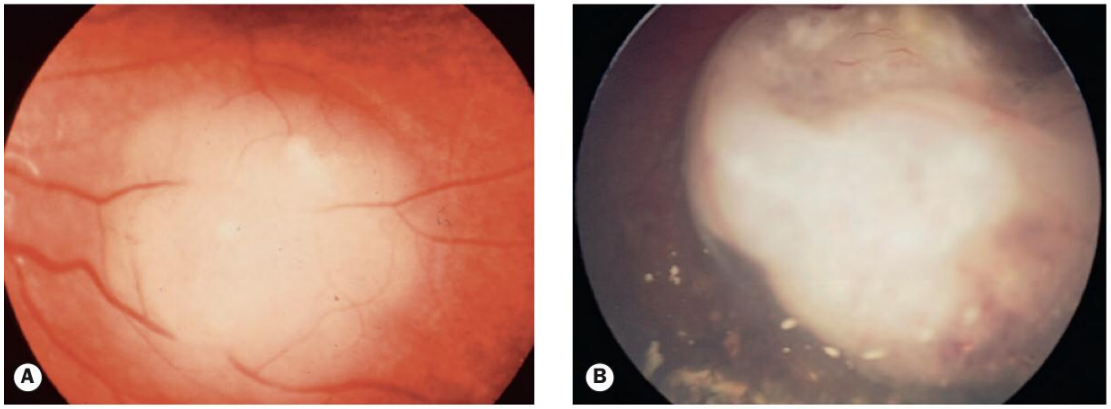
i. Leukocoria is the commonest presentation.
ii. Strabismus is the second most common.
iii. Painful red eye with secondary glaucoma.
iv. Orbital inflammation mimicking orbital or preseptal cellulitis.
v. Orbital invasion or visible extra-ocular growth may occur in



neglected cases.

Presentation of retinoblastoma. (A) Leukocoria; (B) secondary glaucoma; (C) red eye due to uveitis; (D) iris nodules (arrow); (E) orbital inflammation; (F) orbital invasion.

●**Signs**: An intraretinal tumour is a homogeneous, dome-shaped white lesion that becomes irregular, often with white flecks of calcification.



Retinoblastoma. (A) Intraretinal tumour; (B) tumour with vitreous seeding;

Investigation

• Examination under anaesthesia includes the following:

i. General examination for congenital abnormalities of the face and hands.
ii. Tonometry.
iii. Measurement of the corneal diameter.
iv. Anterior chamber examination with a hand-held slit lamp.
v. Ophthalmoscopy, documenting all findings with colour drawings or photography.
vi. Cycloplegic refraction.

• Ultrasound is used mainly to assess tumour size. It also detects calcification.

• MRI does not detect calcification but is useful for optic nerve evaluation, detection of extra ocular extension and pinealoblastoma.

• Systemic assessment includes physical examination and MRI scans of the orbit and skull. If these indicate the presence of metastatic disease then bone scans, bone marrow aspiration and lumbar puncture are also performed.

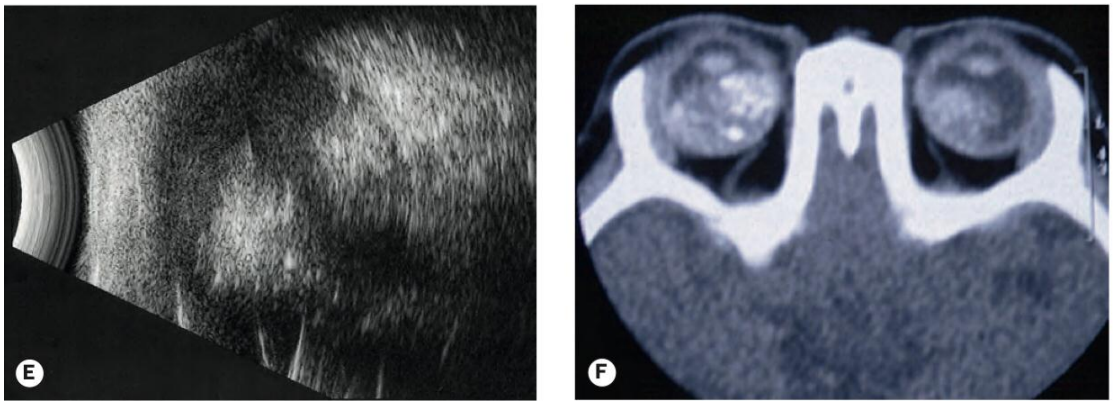
Treatment

• Chemotherapy is the mainstay of treatment in most cases and may be used in conjunction with local treatments (focal consolidation).

•Focal techniques such as diode laser and cryotherapy exert both a direct effect and probably increase susceptibility to the effects of chemotherapy.

• Enucleation is generally indicated if there is neovascular glaucoma, anterior chamber infiltration, optic nerve invasion or if a tumour occupies more than half the vitreous volume.

• Review. Careful review at frequent intervals is generally required following treatment.



(E) B-scan showing echoes from calcification; (F) axial CT showing bilateral tumours with calcification.