

Tumors of posterior segment

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2022



Precancerous and cancerous lesions of the posterior segment of the eye



Retinoblastoma



Genetic



If parent:	has bilateral retinoblastoma				has unilateral retinoblastoma				is unaffected			
Chance of offspring having retinoblastoma	45% affected		55% unaffected		7%–15% affected		85%–93% unaffected		<<1% affected		99% unaffected	
Laterality	85% bilateral		15% unilateral		85% bilateral		15% unilateral		33% bilateral		67% unilateral	
Focality	100% multi-focal		96% multi-focal, 4% uni-focal		100% multi-focal		96% multi-focal, 4% uni-focal		100% multi-focal		15% multi-focal, 85% uni-focal	
Chance of next sibling having retinoblastoma	45%		45%		45%		45%		5%*		<1%*	
	45%		45%		45%		7%–15%		<1%*		<1	
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Diagnosis



Table 19-1 Presenting Signs and Symptoms of Retinoblastoma

Among Patients <5 Years	Among Patients ≥5 Years
Leukocoria (most common)	Leukocoria (35%)
Strabismus (≈20%)	Decreased vision (35%)
Ocular inflammation (≈5%)	Strabismus (15%)
Pseudohypopyon	Floaters (5%)
Hyphema	Pain (5%)
Iris heterochromia	
Spontaneous globe perforation	
Proptosis	
Cataract	
Glaucoma	
Nystagmus	
Tearing	
Anisocoria	



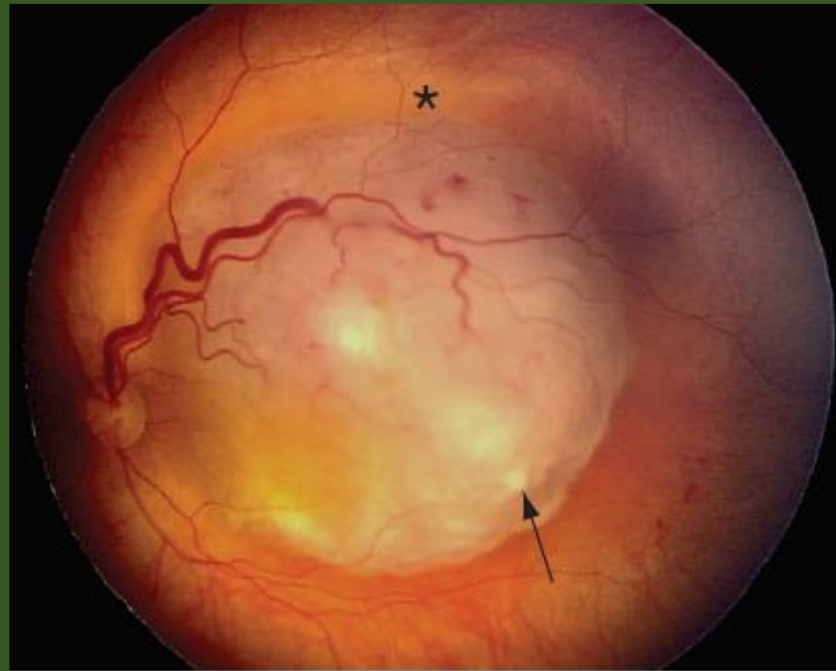


Figure 19-5 Retinoblastoma. Note the dilated retinal blood vessels, foci of calcification (arrow), and cuff of subretinal fluid (asterisk).
(Courtesy of Matthew W. Wilson, MD.)



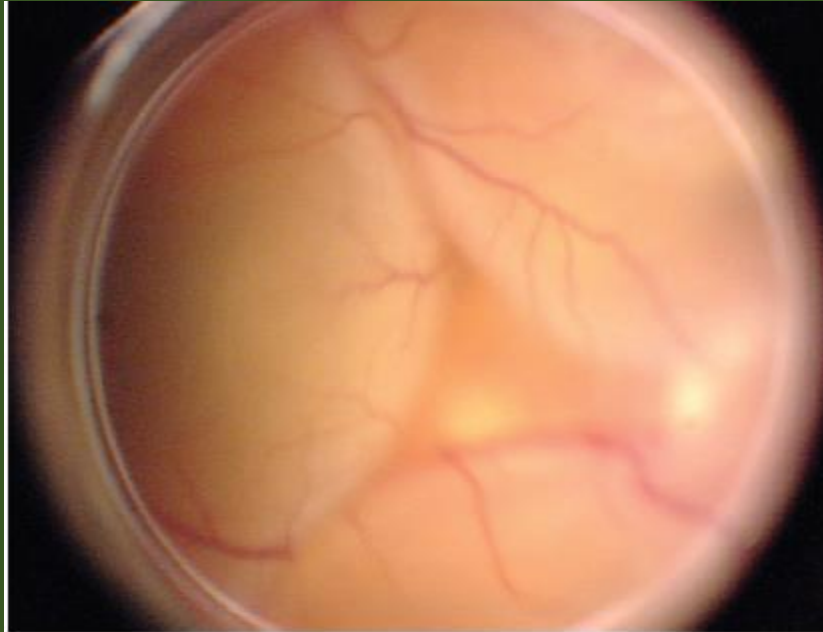


Figure 19-6 Retinoblastoma. Complete exudative detachment obscures tumor visualization. Note normal-appearing retinal vessels, as opposed to those found in Coats disease. *(Courtesy of Matthew W. Wilson, MD.)*





Figure 19-7 Retinoblastoma. Large endophytic tumor with extensive vitreous seeding (arrows). (Courtesy of Matthew W. Wilson, MD.)



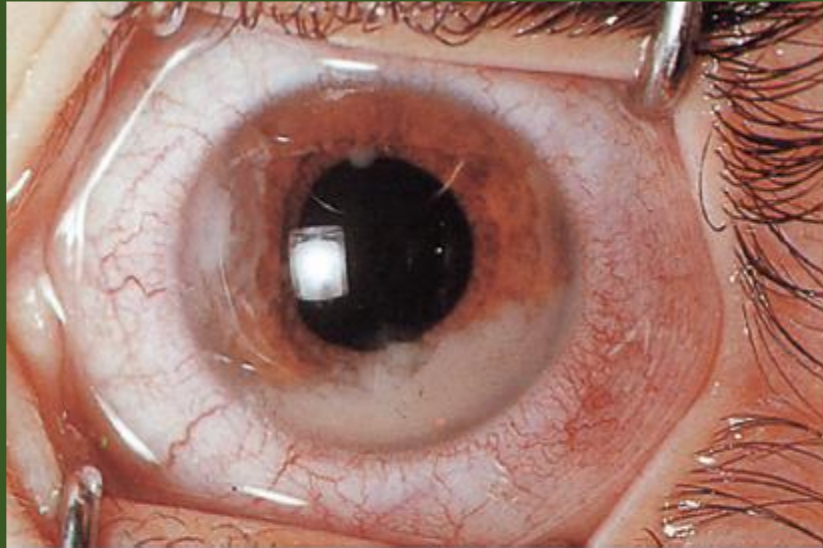


Figure 19-8 Retinoblastoma, clinical appearance. Pseudohypopyon resulting from migration of tumor cells into the anterior chamber.



Diffuse infiltrating Rb



Imaging



Metastasis



Differential Diagnosis



Table 19-2 Differential Diagnosis of Retinoblastoma

- Astrocytic hamartoma
- Coats disease
- Coloboma of choroid or optic nerve head
- Congenital retinal fold
- Organizing vitreous hemorrhage
- Persistent fetal vasculature
- Posterior cataract
- Retinal dysplasia
- Retinopathy of prematurity
- Toxocariasis (larval granuloma)
- Uveitis



Classification



Reese – Ellsworth classification



International classification (ABC system)



Table 19-3 International Classification for Intraocular Retinoblastoma (ABC System)

Group A	Small tumors (<3 mm) confined to the retina; >3 mm from the fovea; >1.5 mm from the optic nerve head
Group B	Tumors (>3 mm) confined to the retina in any location, with clear subretinal fluid up to 5 mm from the tumor margin
Group C	Localized vitreous and/or subretinal seeding
Group D	Diffuse vitreous and/or subretinal seeding
Group E	No visual potential or presence of 1 or more of the following: <ul style="list-style-type: none">• tumor in the anterior segment• tumor in or on the ciliary body• neovascular glaucoma• vitreous hemorrhage obscuring the tumor or significant hyphema• phthisical or prephthisical eye• orbital cellulitis-like presentation



Treatment



Enucleation



Chemotherapy



Laser therapy and TTP



Cryotherapy



External Beam Radiation



Plaque radiation (Brachy therapy)



Melanocytic tumors

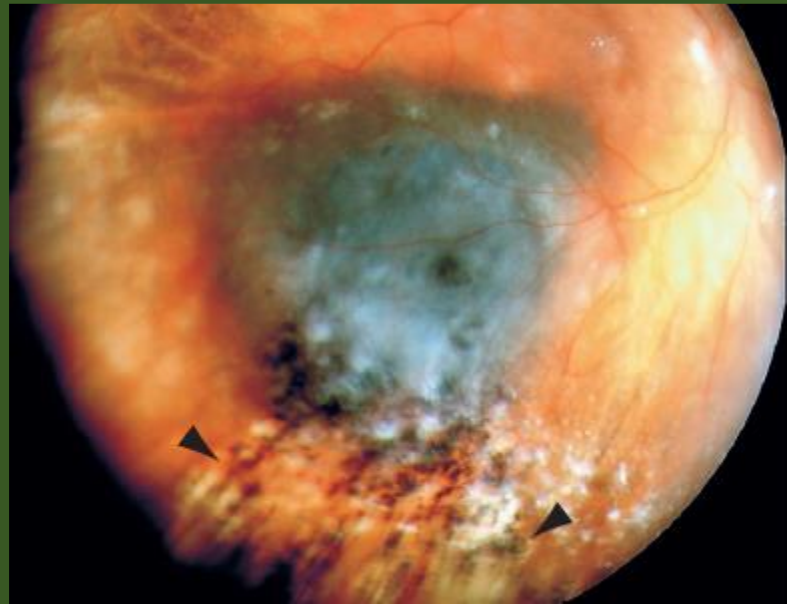


Choroidal NEVUS







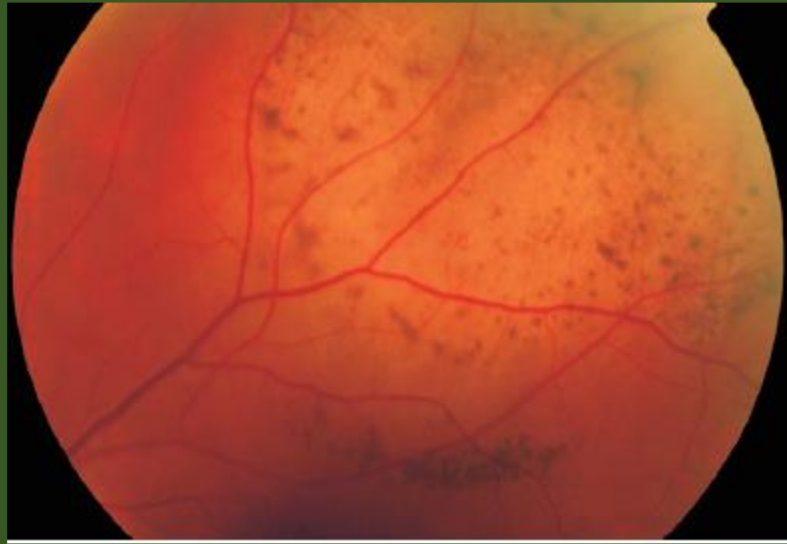


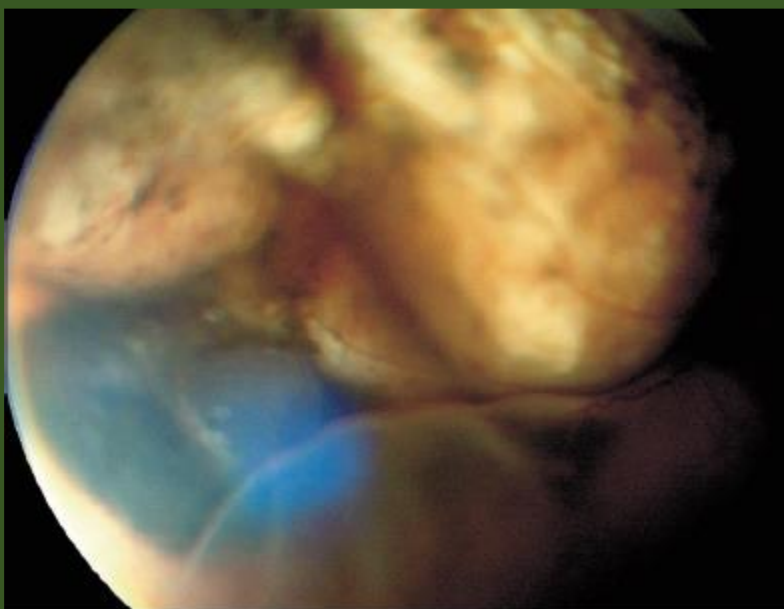
Choroidal melanoma

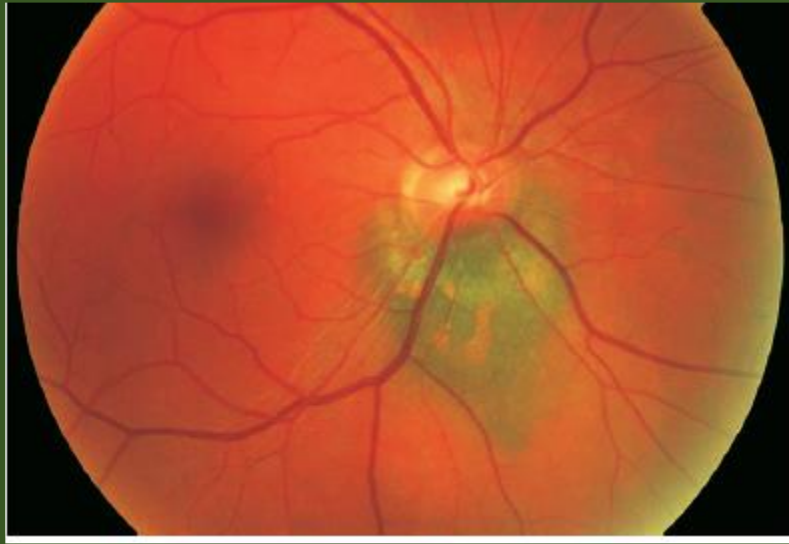


Clinical manifestations















Diagnosis

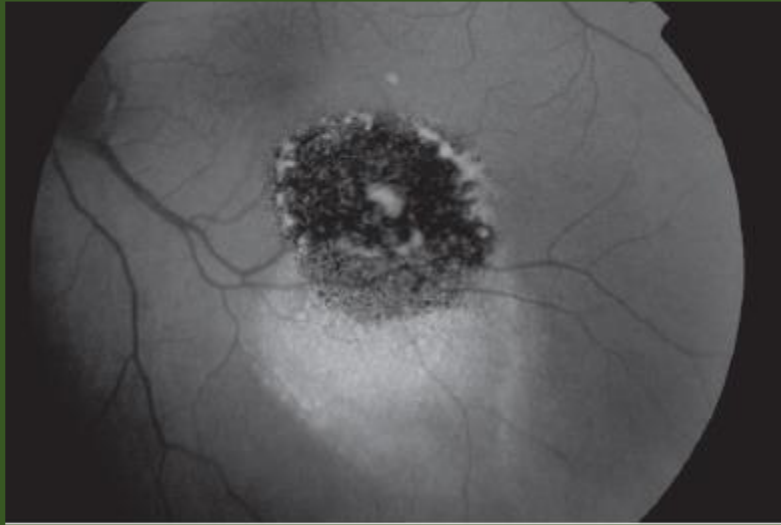


Fundus photography



FAF



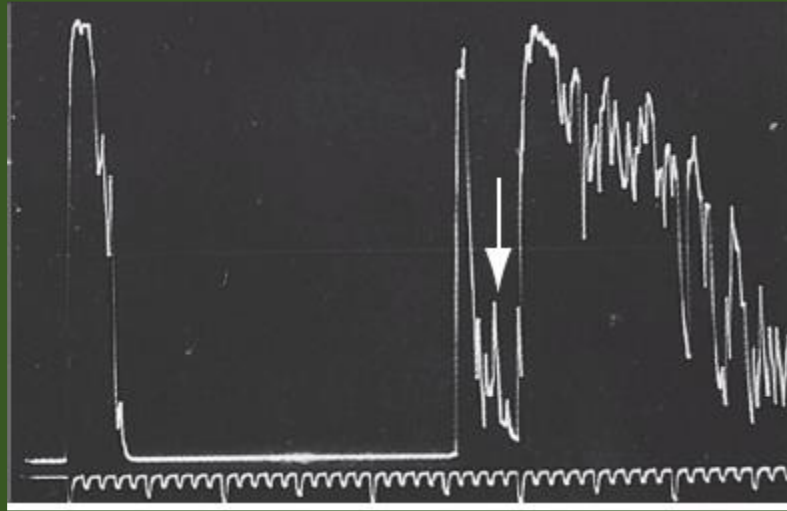


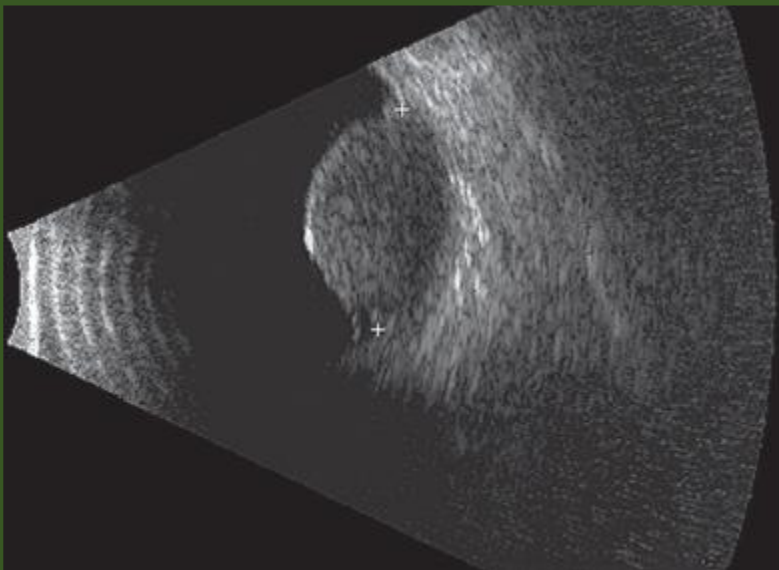
OCT

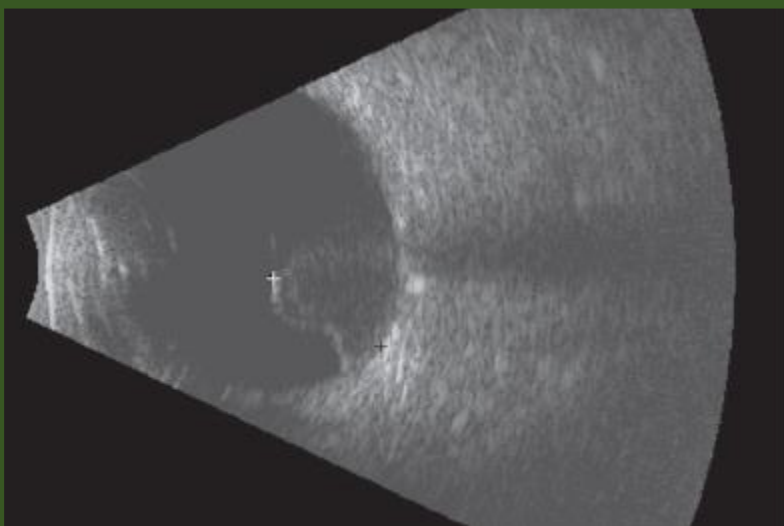


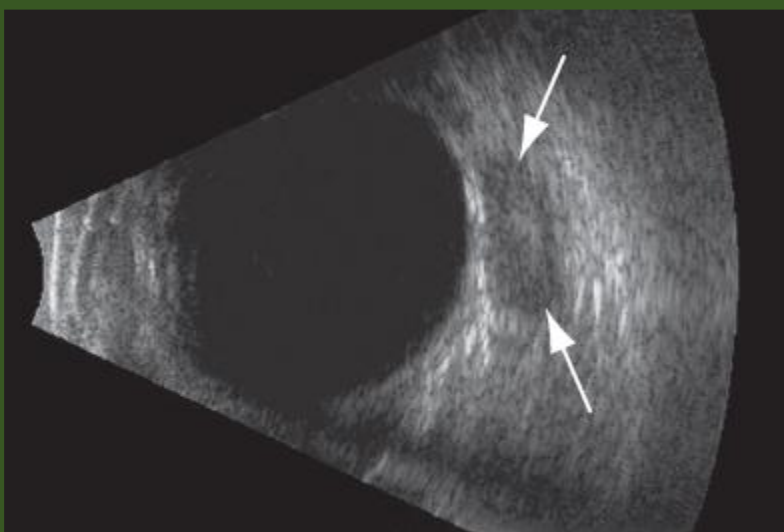
Ultrasonography











Differential diagnosis



Choroidal NEVUS



- **Melanocytoma of CB or ONH**
- **CHRPE**
- **AMD with CNV**
- **Hemorrhagic choroidal detachments**
- **Choroidal osteomas**
- **Choroidal hemangiomas**
- **Varix of vortex vein**
- **Intraocular metastasis**



Classification



Table 17-2 AJCC Staging of Ciliary Body and Choroidal (Posterior) Melanoma

Stage	Percentage of Patients	5-Year Survival	10-Year Survival
Stage I	21%–32%	96%–97%	88%–94%
Stage IIA	31%–34%	89%–98%	80%–84%
Stage IIB	22%–23%	79%–81%	67%–70%
Stage IIIA	9%–17%	66%–67%	45%–60%
Stage IIIB	3%–8%	45%–50%	27%–50%
Stage IIIC	1%	25%–26%	0%–10%
Stage IV (metastasis)	<2%	<5%	<1%



Metastatic evaluation



Table 17-3 Clinical Evaluation of Metastatic Uveal Melanoma

Liver imaging—ultrasonography in routine evaluation

Liver function tests

Chest x-ray

If any of the above are abnormal:

Triphasic liver CT

PET-CT of the abdomen/chest

MRI of the abdomen

Treatment



Observation



Enucleation



Brachytherapy with radioactive plaque



- Charge – particle radiation
- TTP and PDT
- Surgical excision
- Immuno therapy
- Exenteration



Angiomatous Tumors

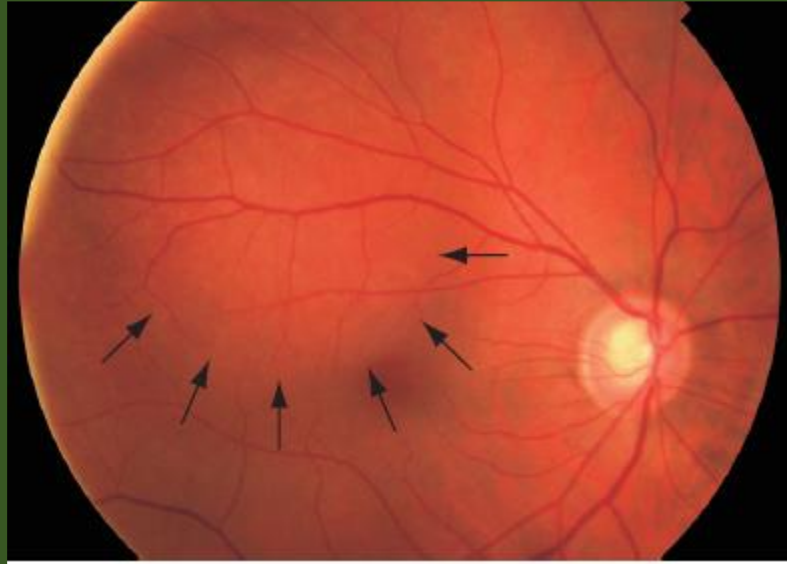


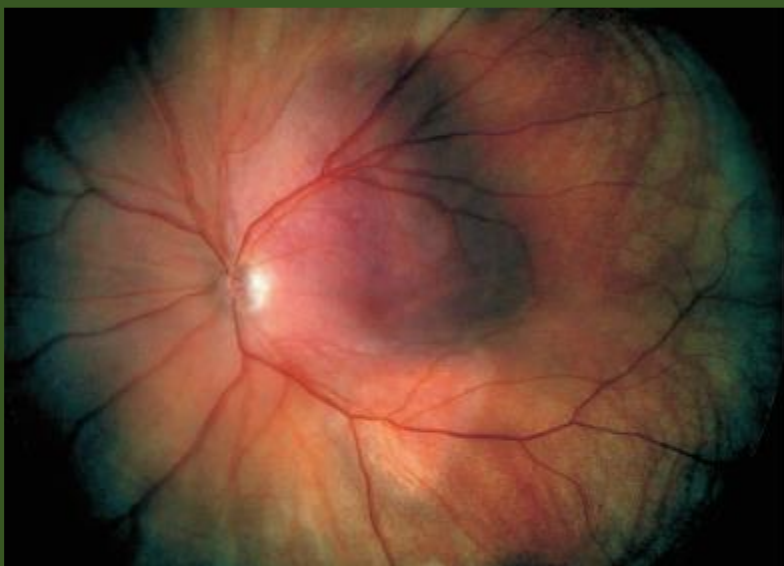
Choroidal hemangiomas



Cirumscribed C.H.

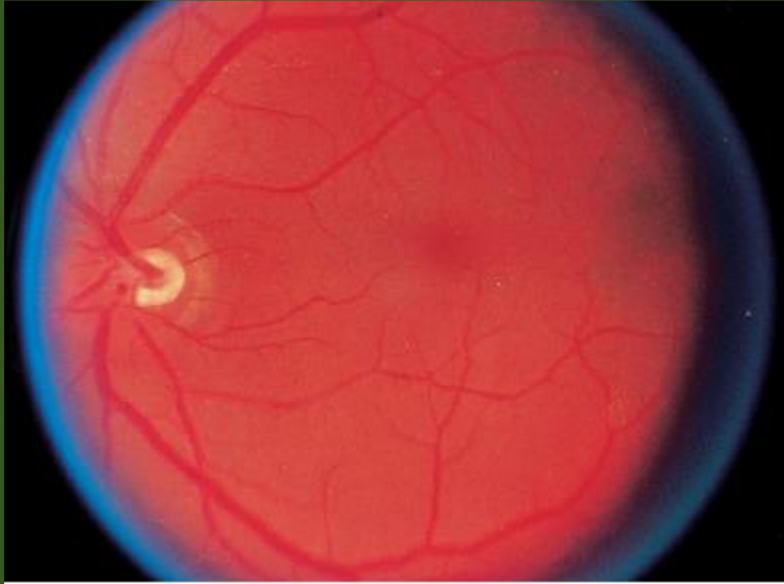


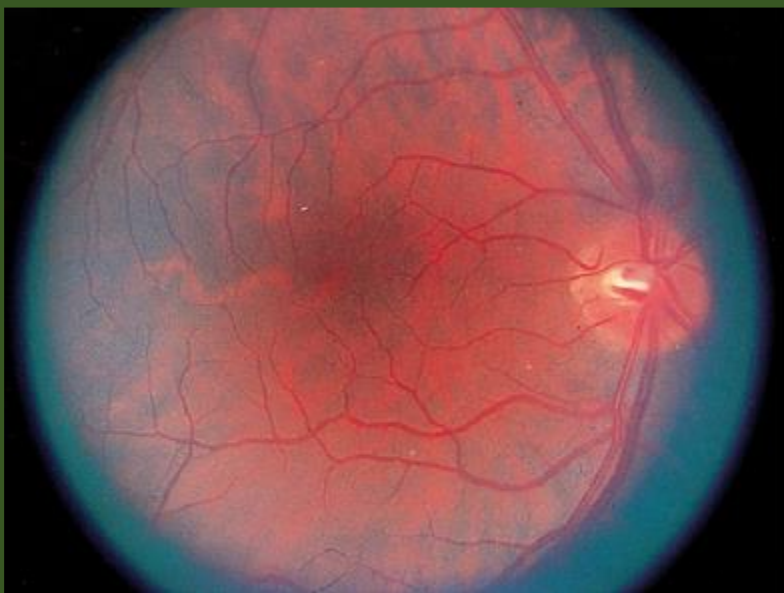




Diffuse C.H.

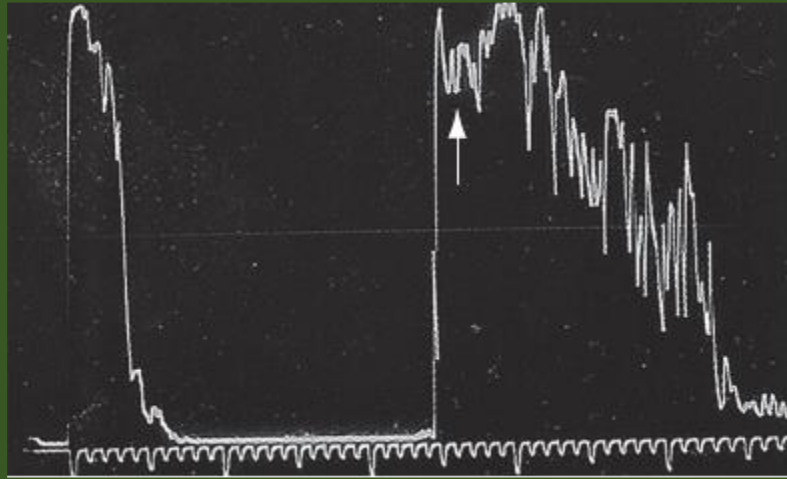


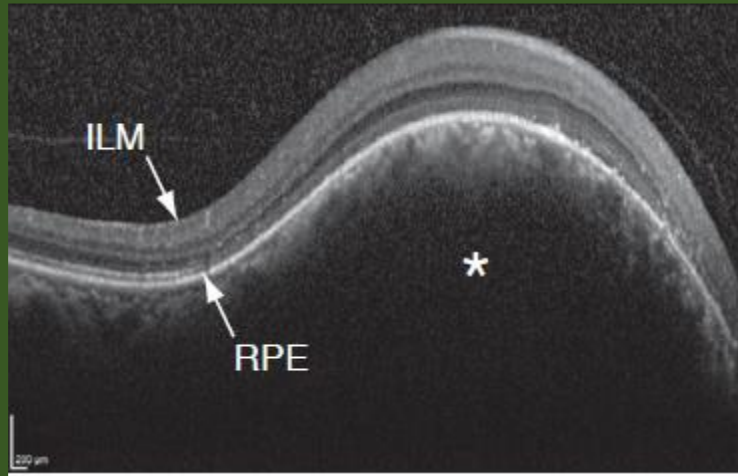


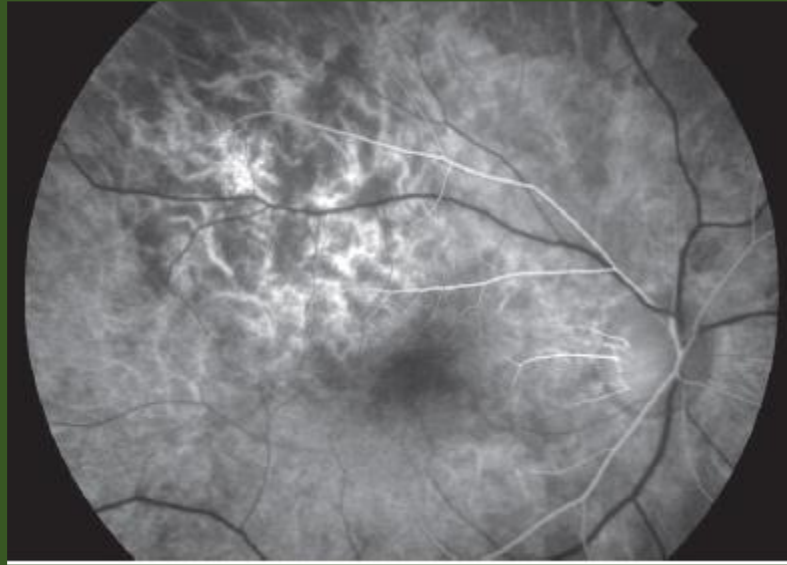


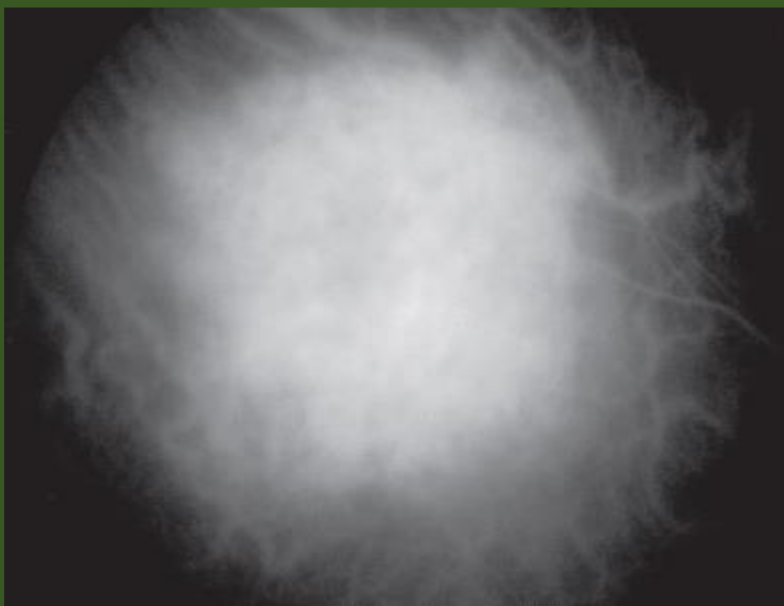
Diagnosis











Treatment



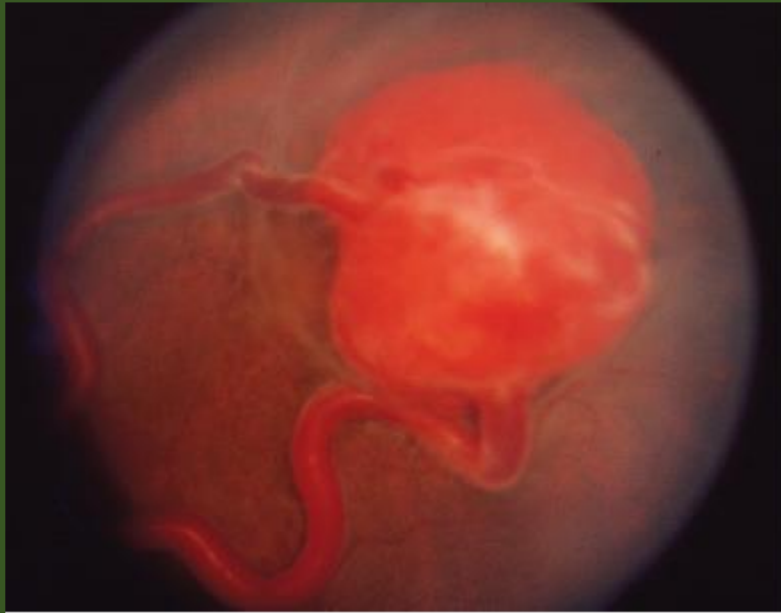
Retinal Angiomas

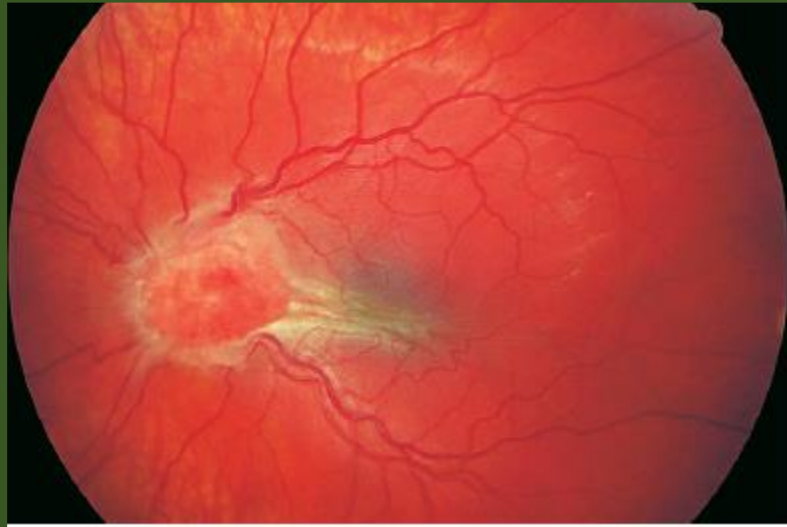


Hemangioblastoma







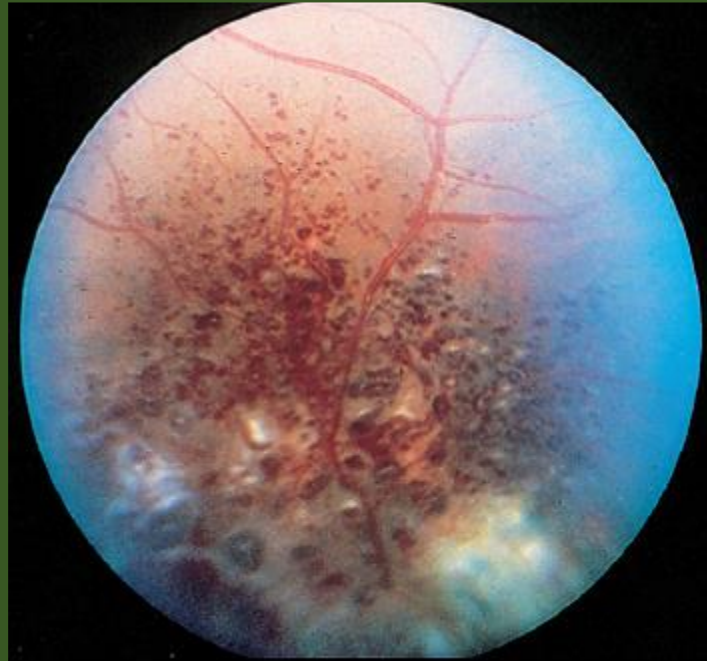


Treatment



Cavernous Hemangioma







The End