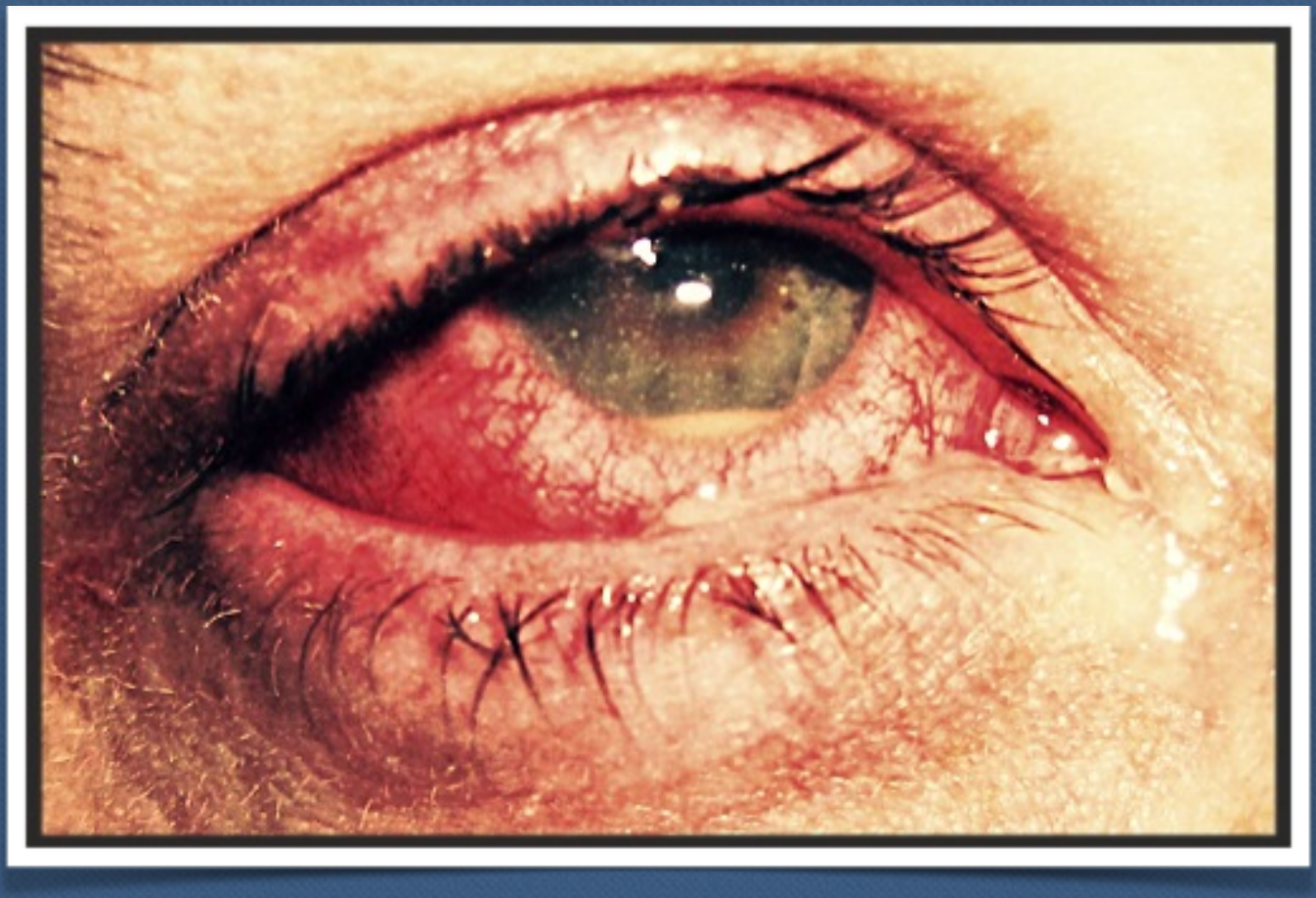
# QUICK UVEITIS



## Quick guide lines

*The most important information you need to know about managing your patients with Uveitis*

Ameen Marashi

بسم الله الرحمن الرحيم

# INTRODUCTION

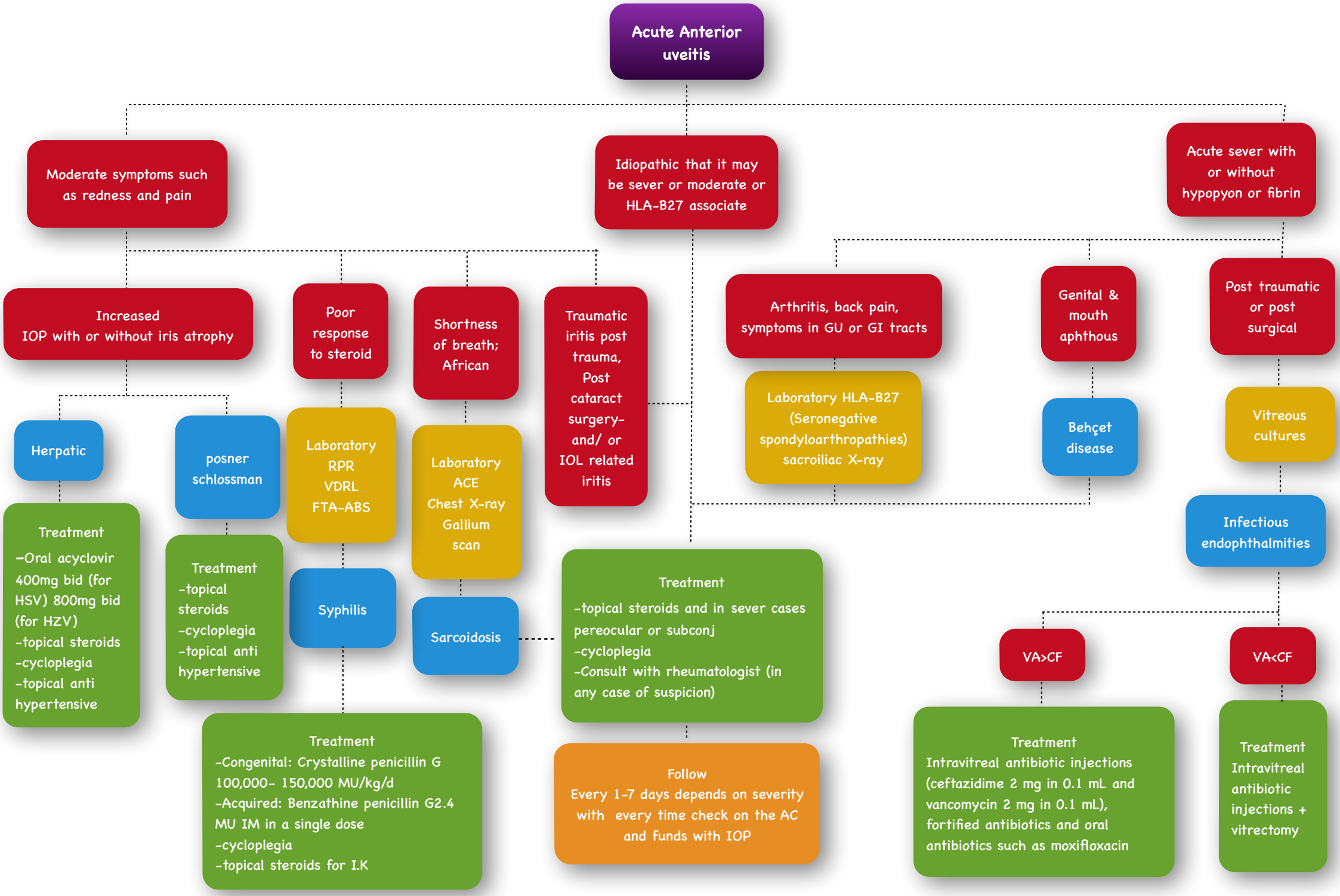
This book is written as algorithms and text configuration, where algorithms are made to help us doctors to easy diagnose and treat patients with most common cases of uveitis where text is made to give a summary explanations about diseases that are in the algorithms.

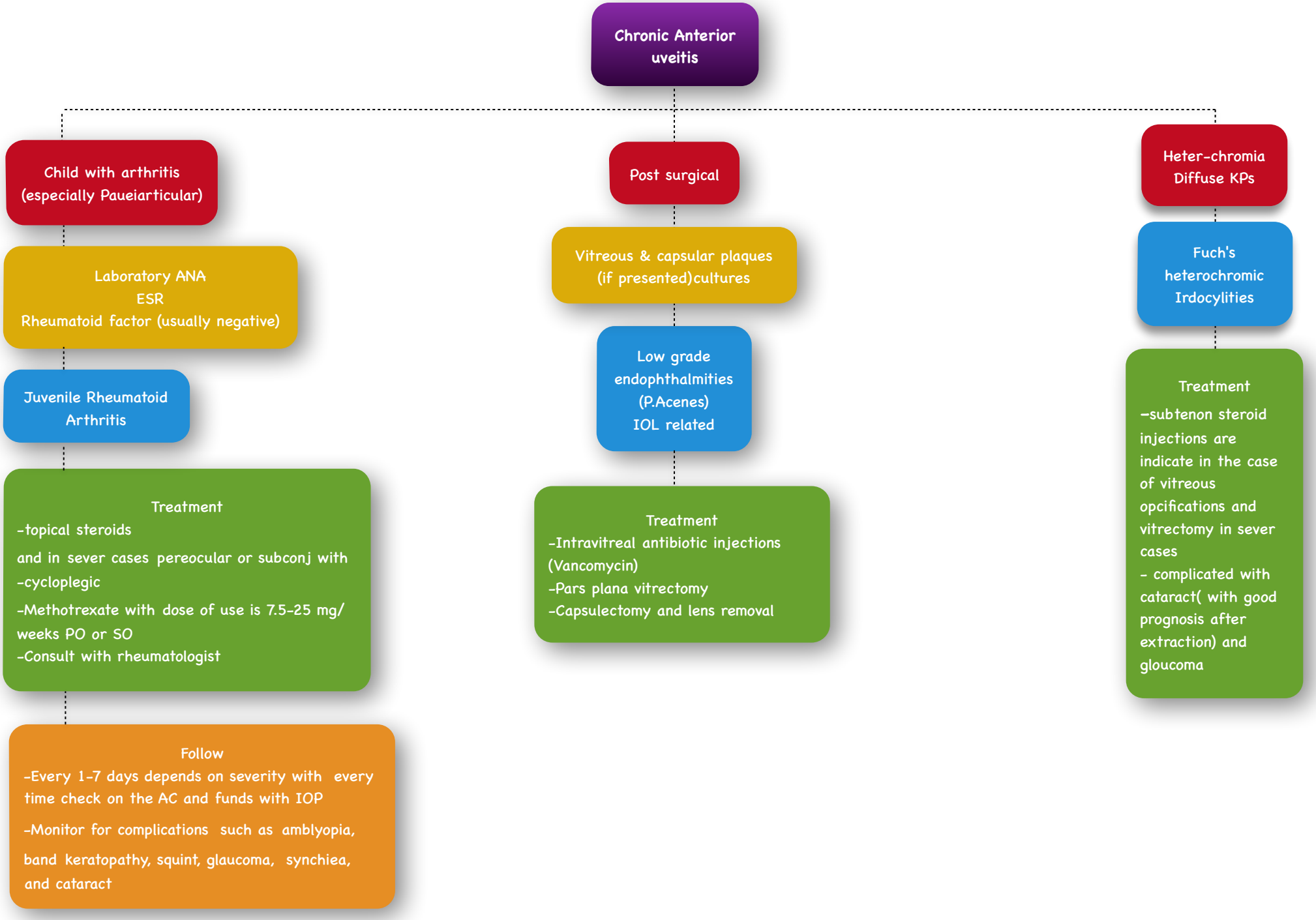
This book is divided into chapters such as principles of treatment infectious, non infectious inflammatory, masqueraded diseases, and algorithms.

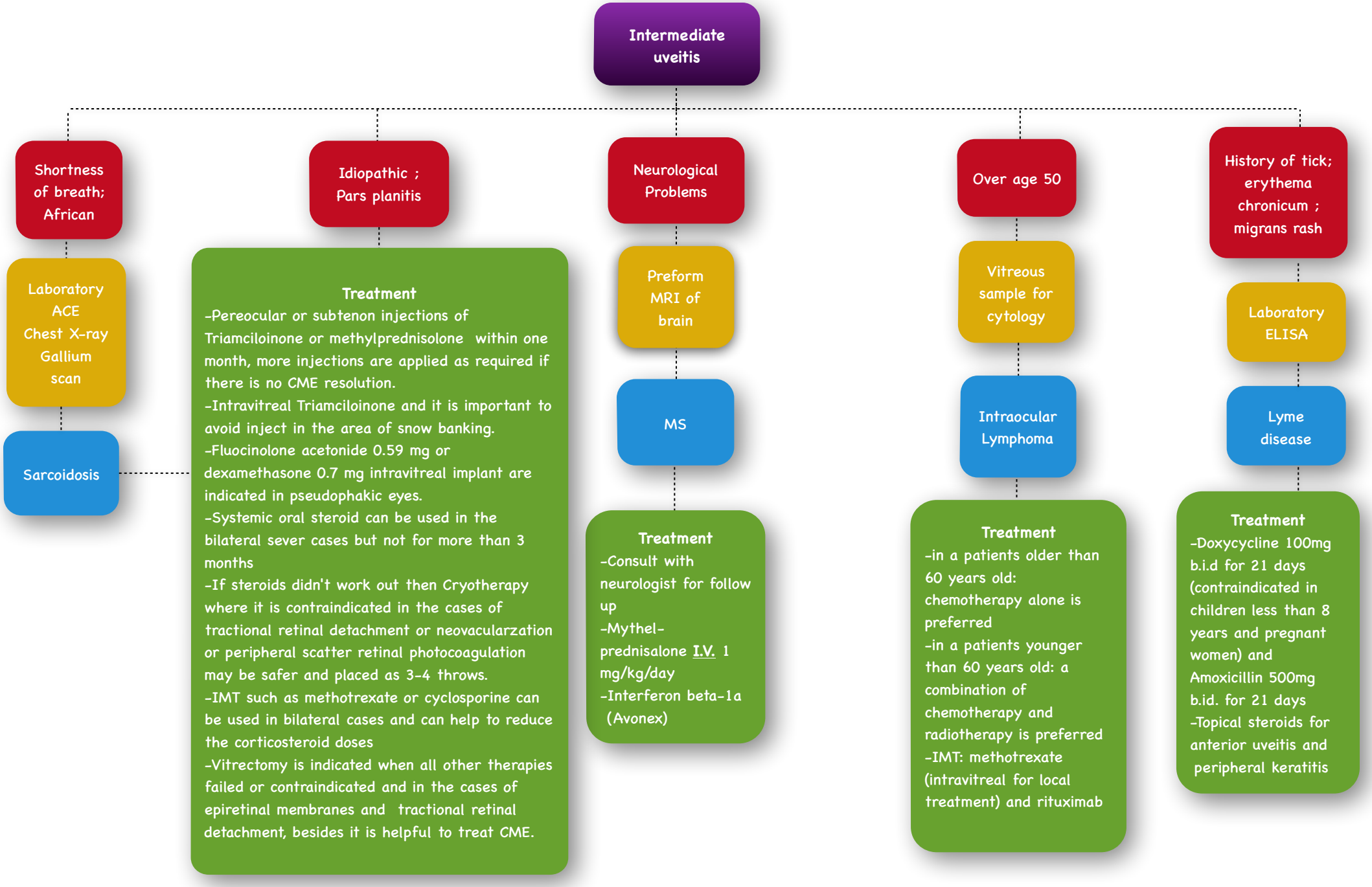
As you noticed there is no pictures in the text chapters because I don't have pictures of uveitis cases that are mine and I don't have any attention to violate any copy rights.

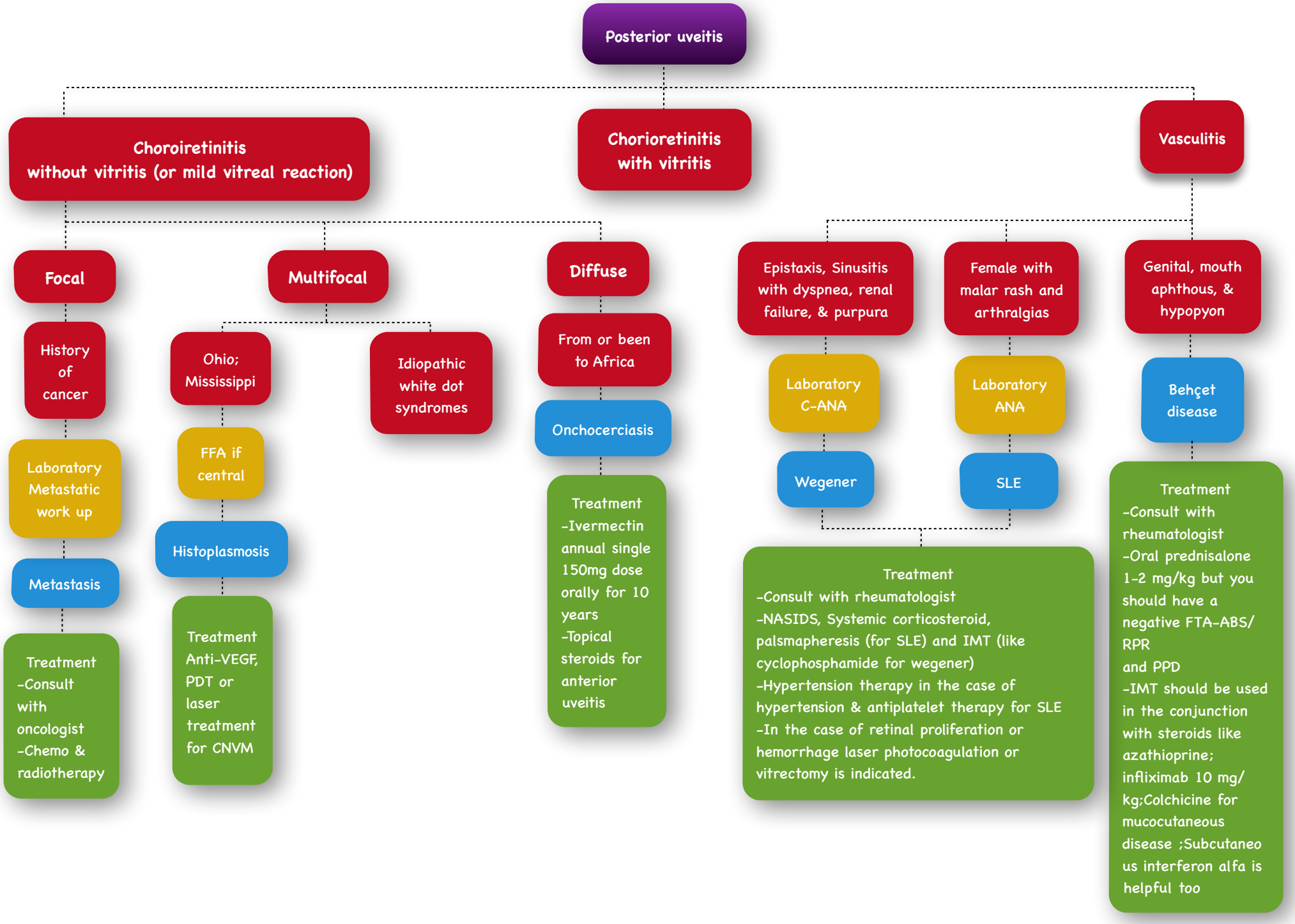
This book is made to be free of charge as a PDF file as you notice there is no page numbers so if you want to navigate threw chapters you have to choose subjects from bookmarks in your PDF reader.

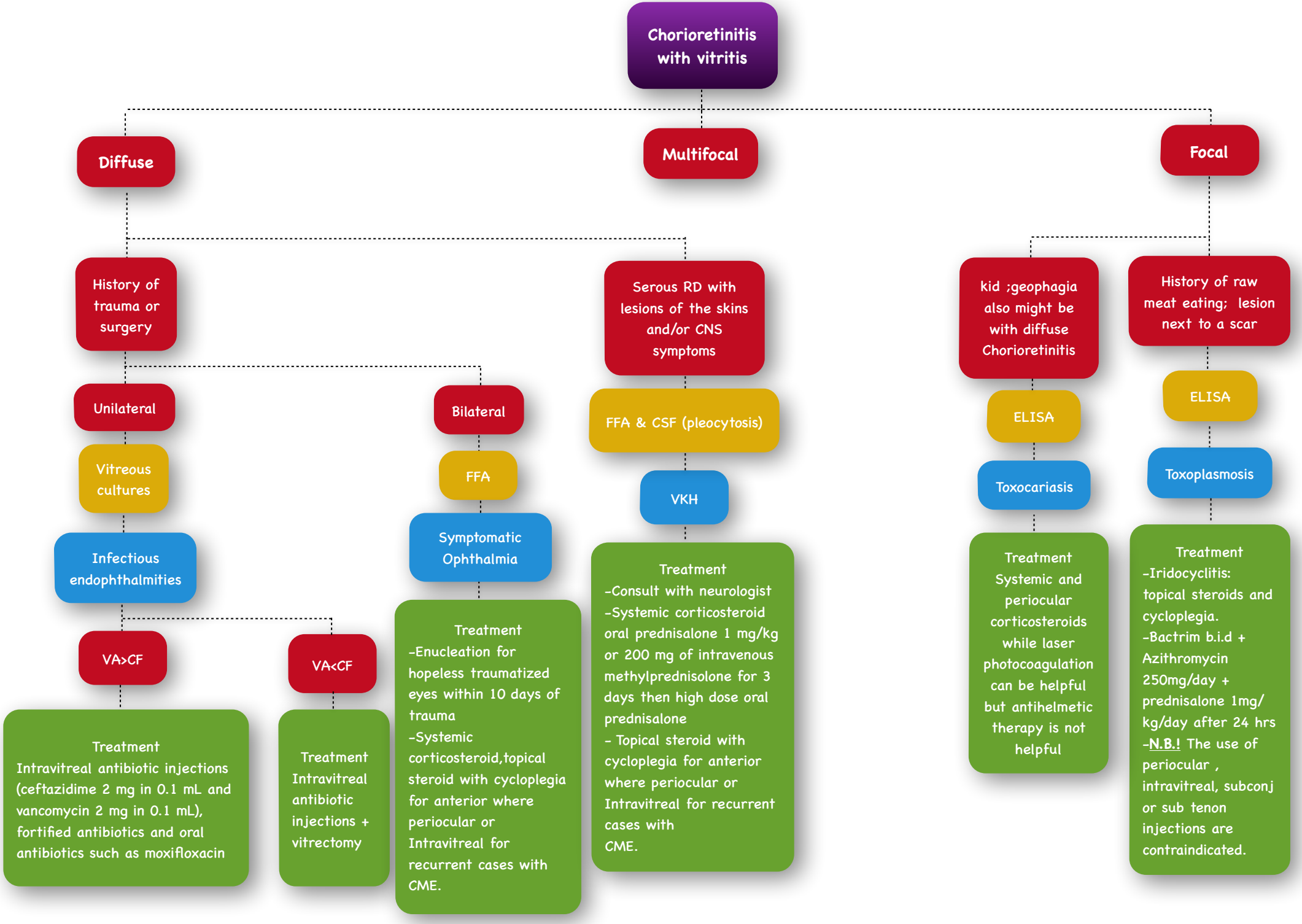
Ameen Marashi, M.D.


Acute Anterior uveitis
Moderate symptoms such as redness and pain
Idiopathic that it may be sever or moderate or HLA-B27 associate
Acute sever with or without hypopyon or fibrin
Increased IOP with or without iris atrophy
Poor response to steroid
Shortness of breath; African
Traumatic iritis post trauma, Post cataract surgery- and/ or IOL related iritis
Arthritis, back pain, symptoms in GU or GI tracts
Genital & mouth aphthous
Post traumatic or post surgical
Herpatic
posner schlossman
Laboratory RPR VDRL FTA-ABS
Laboratory ACE Chest X-ray Gallium scan
Laboratory HLA-B27 (Seronegative spondyloarthropathies) sacroiliac X-ray
Behçet disease
Vitreous cultures
Treatment -Oral acyclovir 400mg bid (for HSV) 800mg bid (for HZV) -topical steroids -cycloplegia -topical anti hypertensive
Treatment -topical steroids -cycloplegia -topical anti hypertensive
Syphilis
Sarcoidosis
Treatment -topical steroids and in sever cases pereocular or subconj -cycloplegia -Consult with rheumatologist (in any case of suspicion)
Infectious endophthalmities
Treatment -Congenital: Crystalline penicillin G 100,000- 150,000 MU/kg/d -Acquired: Benzathine penicillin G2.4 MU IM in a single dose -cycloplegia -topical steroids for I.K
Follow Every 1-7 days depends on severity with every time check on the AC and funds with IOP
VA>CF
VA<CF
Treatment Intravitreal antibiotic injections (ceftazidime 2 mg in 0.1 mL and vancomycin 2 mg in 0.1 mL), fortified antibiotics and oral antibiotics such as moxifloxacin
Treatment Intravitreal antibiotic injections + vitrectomy

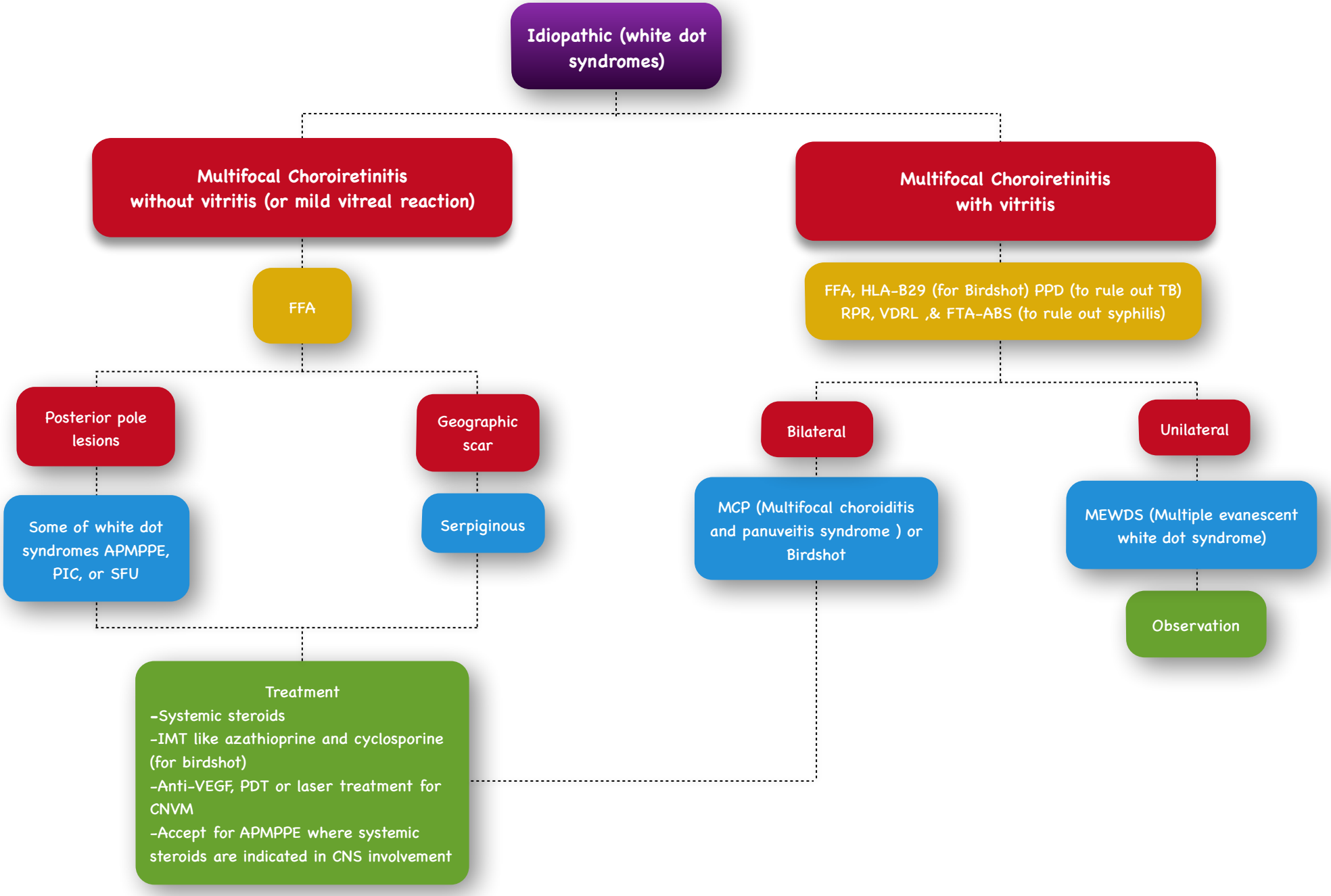

Chronic Anterior uveitis
Child with arthritis (especially Paueiarticular)
Laboratory ANA
ESR
Rheumatoid factor (usually negative)
Juvenile Rheumatoid Arthritis
Treatment
-topical steroids
and in sever cases pereocular or subconj with
-cycloplegic
-Methotrexate with dose of use is 7.5-25 mg/ weeks PO or SO
-Consult with rheumatologist
Follow
-Every 1-7 days depends on severity with every time check on the AC and funds with IOP
-Monitor for complications such as amblyopia, band keratopathy, squint, glaucoma, synchiea, and cataract
Post surgical
Vitreous & capsular plaques (if presented)cultures
Low grade endophthalmities (P.Acenes)
IOL related
Treatment
-Intravitreal antibiotic injections (Vancomycin)
-Pars plana vitrectomy
-Capsulectomy and lens removal
Heter-chromia
Diffuse KPs
Fuch's heterochromic Irdocylities
Treatment
-subtenon steroid injections are indicate in the case of vitreous opcifications and vitrectomy in sever cases
- complicated with cataract( with good prognosis after extraction) and gloucoma

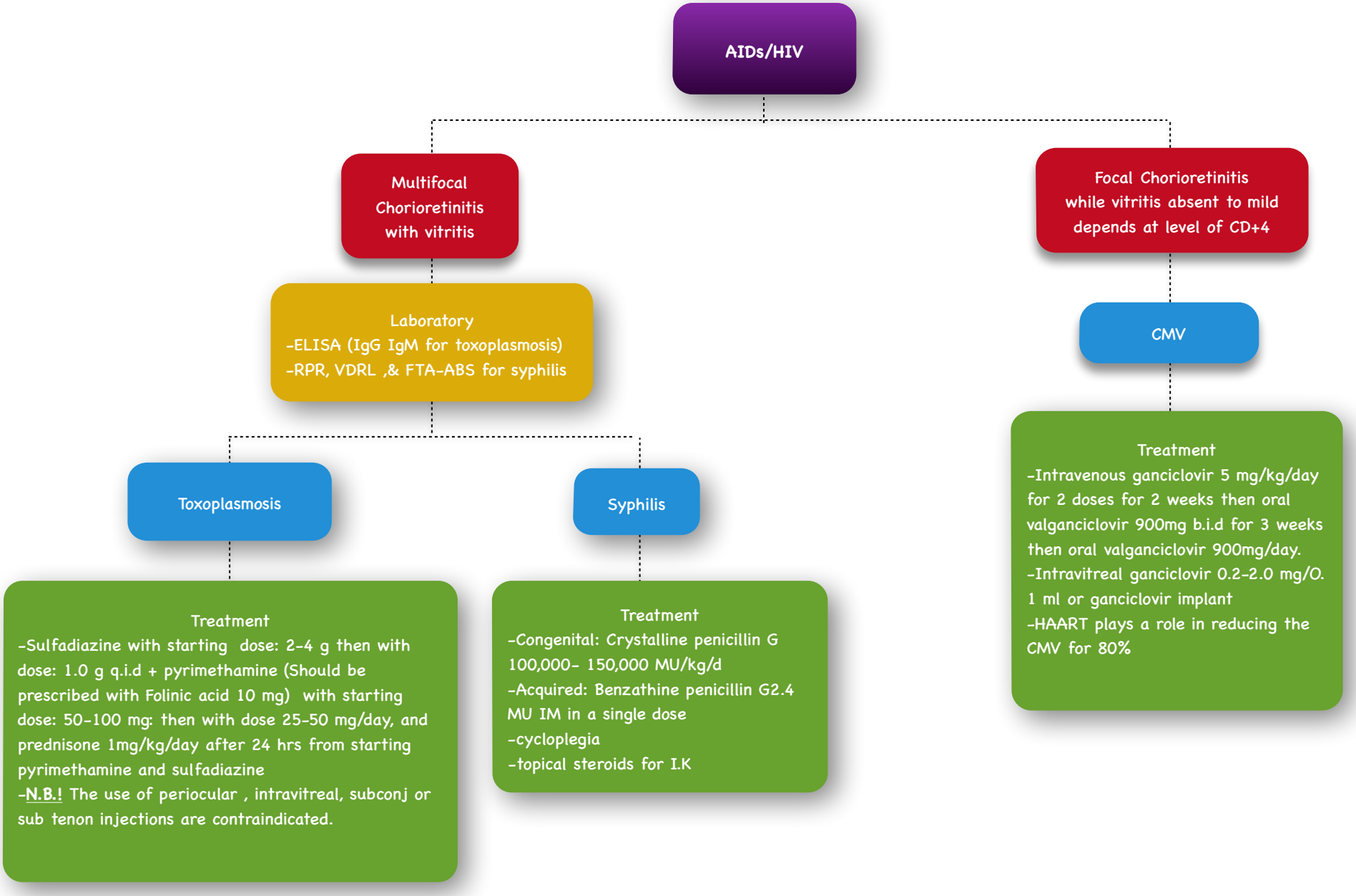

Intermediate uveitis
Shortness of breath; African
Laboratory ACE Chest X-ray Gallium scan
Sarcoidosis
Idiopathic ; Pars planitis
Treatment
-Pereocular or subtenon injections of Triamciloinone or methylprednisolone within one month, more injections are applied as required if there is no CME resolution.
-Intravitreal Triamciloinone and it is important to avoid inject in the area of snow banking.
-Fluocinolone acetonide 0.59 mg or dexamethasone 0.7 mg intravitreal implant are indicated in pseudophakic eyes.
-Systemic oral steroid can be used in the bilateral sever cases but not for more than 3 months
-If steroids didn't work out then Cryotherapy where it is contraindicated in the cases of tractional retinal detachment or neovacularzation or peripheral scatter retinal photocoagulation may be safer and placed as 3-4 throws.
-IMT such as methotrexate or cyclosporine can be used in bilateral cases and can help to reduce the corticosteroid doses
-Vitrectomy is indicated when all other therapies failed or contraindicated and in the cases of epiretinal membranes and tractional retinal detachment, besides it is helpful to treat CME.
Neurological Problems
Preform MRI of brain
MS
Treatment
-Consult with neurologist for follow up
-Mythel-prednisalone I.V. 1 mg/kg/day
-Interferon beta-1a (Avonex)
Over age 50
Vitreous sample for cytology
Intraocular Lymphoma
Treatment
-in a patients older than 60 years old: chemotherapy alone is preferred
-in a patients younger than 60 years old: a combination of chemotherapy and radiotherapy is preferred
-IMT: methotrexate (intravitreal for local treatment) and rituximab
History of tick; erythema chronicum ; migrans rash
Laboratory ELISA
Lyme disease
Treatment
-Doxycycline 100mg b.i.d for 21 days (contraindicated in children less than 8 years and pregnant women) and Amoxicillin 500mg b.id. for 21 days
-Topical steroids for anterior uveitis and peripheral keratitis


Posterior uveitis
Choroiretinitis without vitritis (or mild vitreal reaction)
Chorioretinitis with vitritis
Vasculitis
Focal
Multifocal
Diffuse
History of cancer
Laboratory Metastatic work up
Metastasis
Treatment -Consult with oncologist -Chemo & radiotherapy
Ohio; Mississippi
FFA if central
Histoplasmosis
Treatment Anti-VEGF, PDT or laser treatment for CNVM
Idiopathic white dot syndromes
From or been to Africa
Onchocerciasis
Treatment -Ivermectin annual single 150mg dose orally for 10 years -Topical steroids for anterior uveitis
Epistaxis, Sinusitis with dyspnea, renal failure, & purpura
Laboratory C-ANA
Wegener
Female with malar rash and arthralgias
Laboratory ANA
SLE
Treatment -Consult with rheumatologist -NASIDS, Systemic corticosteroid, palsmapheresis (for SLE) and IMT (like cyclophosphamide for wegener) -Hypertension therapy in the case of hypertension & antiplatelet therapy for SLE -In the case of retinal proliferation or hemorrhage laser photocoagulation or vitrectomy is indicated.
Genital, mouth aphthous, & hypopyon
Behçet disease
Treatment -Consult with rheumatologist -Oral prednisalone 1-2 mg/kg but you should have a negative FTA-ABS/ RPR and PPD -IMT should be used in the conjunction with steroids like azathioprine; infliximab 10 mg/ kg;Colchicine for mucocutaneous disease ;Subcutaneo us interferon alfa is helpful too


Chorioretinitis with vitritis
Diffuse
Multifocal
Focal
History of trauma or surgery
Unilateral
Vitreous cultures
Infectious endophthalmities
VA>CF
Treatment
Intravitreal antibiotic injections (ceftazidime 2 mg in 0.1 mL and vancomycin 2 mg in 0.1 mL), fortified antibiotics and oral antibiotics such as moxifloxacin
VA<CF
Treatment
Intravitreal antibiotic injections + vitrectomy
Bilateral
FFA
Symptomatic Ophthalmia
Treatment
-Enucleation for hopeless traumatized eyes within 10 days of trauma
-Systemic corticosteroid,topical steroid with cycloplegia for anterior where periocular or Intravitreal for recurrent cases with CME.
Serous RD with lesions of the skins and/or CNS symptoms
FFA & CSF (pleocytosis)
VKH
Treatment
-Consult with neurologist
-Systemic corticosteroid oral prednisalone 1 mg/kg or 200 mg of intravenous methylprednisolone for 3 days then high dose oral prednisalone
- Topical steroid with cycloplegia for anterior where periocular or Intravitreal for recurrent cases with CME.
kid ;geophagia also might be with diffuse Chorioretinitis
ELISA
Toxocariasis
Treatment
Systemic and periocular corticosteroids while laser photocoagulation can be helpful but antihelmetic therapy is not helpful
History of raw meat eating; lesion next to a scar
ELISA
Toxoplasmosis
Treatment
-Iridocyclitis: topical steroids and cycloplegia.
-Bactrim b.i.d + Azithromycin 250mg/day + prednisalone 1mg/kg/day after 24 hrs
-N.B.! The use of periocular , intravitreal, subconj or sub tenon injections are contraindicated.


Multifocal chorioretinitis with vitritis
Peripheral retinal necrosis
Idiopathic white dot syndromes
From Africa; visible parasite
Drugs IV, or/ & immunocompromised
Shortness of breath
Age > 50
Laboratory ELISA for HSV & HZV vitrectomy and retinoectomy
Cysticercosis
Onchocerciasis
Vitreous & blood cytology
Laboratory ACE Chest X-ray Gallium scan; PPD
Vitreous sample for cytology
ARN
PORN (immunocompromised)
Candida; Aspergillus
Sarcoidosis
TB
Intraocular Lymphoma
Treatment -Intravenous acyclovir 10 mg/kg/ day divided into 3 doses for 2 weeks then oral acyclovir 800mg for 5 times or for 3 months, and after 24-48hr start prednisone 1 mg/kg/ day. -Intravitreal ganciclovir 0.2-2.0 mg/0.1 ml with systemic antiviral therapy would rapid the resolution -Aspirin
Treatment -A combination of Systemic and intraocular antiviral therapy because PORN is resistant for I.V. treatment -HAART
Treatment photocoagulation while antihelmetic may cause inflammatory reaction
Treatment -Ivermectin annual single 150mg dose orally for 10 years -Topical steroids for anterior uveitis
Treatment -Intravitreal 5 µg amphotericin B Amphotericin B I.V. for Aspergillosis where oral fluconazole 400 mg initially then 200 mg b.i.d for candida
Treatment - Consult with rheumatologist -corticosteroid can be oral prednisalone 1 mg/kg and periocular, intravitreal injections of Triamciloinone and intravitreal implants such as fluocinolone acetonide. -methotrexate and rituximab
Treatment - Consult with TB Specialist -1st 2 months isoniazid +rifampin and pyrazinamide daily then for 4 month with isoniazid +rifampin -Topical steroids for anterior uveitis + methotrexate (intravitreal) for postrior
Treatment -in a patients older than 60 years old: chemotherapy alone is preferred -in a patients younger than 60 years old: a combination of chemotherapy and radiotherapy is preferred -IMT: methotrexate (intravitreal for local treatment) and rituximab


Idiopathic (white dot syndromes)
Multifocal Choroiretinitis without vitritis (or mild vitreal reaction)
Multifocal Choroiretinitis with vitritis
FFA
FFA, HLA-B29 (for Birdshot) PPD (to rule out TB) RPR, VDRL ,& FTA-ABS (to rule out syphilis)
Posterior pole lesions
Geographic scar
Bilateral
Unilateral
Some of white dot syndromes APMPPE, PIC, or SFU
Serpiginous
MCP (Multifocal choroiditis and panuveitis syndrome ) or Birdshot
MEWDS (Multiple evanescent white dot syndrome)
Observation
Treatment
-Systemic steroids
-IMT like azathioprine and cyclosporine (for birdshot)
-Anti-VEGF, PDT or laser treatment for CNVM
-Accept for APMPPE where systemic steroids are indicated in CNS involvement


AIDs/HIV
Multifocal Chorioretinitis with vitritis
Focal Chorioretinitis while vitritis absent to mild depends at level of CD+4
Laboratory
-ELISA (IgG IgM for toxoplasmosis)
-RPR, VDRL ,& FTA-ABS for syphilis
CMV
Toxoplasmosis
Syphilis
Treatment
-Intravenous ganciclovir 5 mg/kg/day for 2 doses for 2 weeks then oral valganciclovir 900mg b.i.d for 3 weeks then oral valganciclovir 900mg/day.
-Intravitreal ganciclovir 0.2-2.0 mg/0.1 ml or ganciclovir implant
-HAART plays a role in reducing the CMV for 80%
Treatment
-Sulfadiazine with starting dose: 2-4 g then with dose: 1.0 g q.i.d + pyrimethamine (Should be prescribed with Folinic acid 10 mg) with starting dose: 50-100 mg: then with dose 25-50 mg/day, and prednisone 1mg/kg/day after 24 hrs from starting pyrimethamine and sulfadiazine
-N.B.! The use of periocular , intravitreal, subconj or sub tenon injections are contraindicated.
Treatment
-Congenital: Crystalline penicillin G 100,000- 150,000 MU/kg/d
-Acquired: Benzathine penicillin G2.4 MU IM in a single dose
-cycloplegia
-topical steroids for I.K

# Principles of treatment

## Cycloplegic agents

The usage of Cycloplegic agents in uveitis agents is for 2 main purposes, first to break or/and prevent formation of posterior synechiae by allowing some mobility of the pupil, and second is to relieve pain and photophobia that is induced by ciliary spasm by relieving it.
The stronger the inflammation the stronger Cycloplegic agent should be used such as atropine while in weaker inflammation short acting Cycloplegic agent (such as cyclopentolate) should be used to have a faster recovery, it can be instilled in bed time to prevent difficulties with accommodation during the day.

## Nonsteroidal anti inflammatory agents (NASIDs)

Works by inhabiting COX (cyclooxygenase) which will be involved in platelet aggregation by COX-1 and mediate inflammation by COX-2, indication for NASIDs are non necrotizing anterior scleritis, chronic iridocyclitis (JIA), and CME
In other hand NASIDs may be useful in reducing the dose of topical steroids.
Complications of NASIDs are hypertension, stroke, and gastrointestinal ulcers and bleeding, and complication of topical usage of NASIDs are keratitis and rarely perforation.
Topical usage of NASIDs is limited to treat episcleritis and CME post pseudophakia and has shown no effect on noninfectious anterior uveitis.

## Corticosteroids

they are indicated for active inflammation, to prevent CME development and to reduce inflammatory reaction on retina, choroid and optic nerve, nevertheless corticosteroids induce compilations that may be accompanied by any rout of administration, besides corticosteroids are not always indicated like in Fuchs heterochromic iridocyclitis or chronic flare.
Corticosteroids should be tapered gradually if they have been used for more than 2-3 weeks to prevent disease relapse.
There are 4 routs of administration of corticosteroids, topical, periocular, systemic, and intravitreal

## Topical

Which can can be beneficial in treating anterior uveitis and some times vitreous and CME in pseudophakia.
They are used as hourly and tappered gradualy to once daly and some use ointment for night or in case of preservatives intolerance; Potential complications are rise of IOP and cataract formation
Note that difluprednate 0.05 if given q.i.d has the same effect of prednisolone for every 2 hours but has the same side effects.
Topical corticosteroids that have minimal hypertensive effect they have smaller effect on uveitis too and they are limited beneficial for a range of mild to moderate anterior uveitis.

## Periocular

It is indicated when patient has no complaint nor response to topical steroids, and when systemic steroids are contraindicated, it is indicated in patients with intermediate uveitis, posterior (espcially when asymetrical), and CME; Nevertheless they have the same side effect as oral steroids; triamcinolone acetonide 40mg and methylprednisolone can be used.
There are two ways for periocular administration subtenon and transseptal and the periocular rout of administration is contraindicated in the case of infectious uveitis (toxoplasmosis) and in necrotizing scleritis as there is a risk of scleral melt.
Complications of periocular steroid injections are glob perforation periocular hemorrhage, optic nerve damage, ptosis ( when subtenon is injected supra temporal ) , glaucoma , and steroid induced cataract.

## Systemic

Which can be oral or I.V.(methylprednisolone 1 g/day, repeated for 2 to 3 days is an option in severe disease.) which indicted when there is a chronic uveitis with vision threatening condition, in bilateral cases, as pre surgical prophylaxis in uveitic eyes and when topical or/and systemic disease needs treatment too.
Treatment shouldn't be less than one week and if the treatment last for more 3 months, it should be accompanied with immune modularity therapy IMT.
The appropriate corticosteroid ( prednisalone ) dose is 1-2 mg/kg and should be tapered every 1-2 weeks until quiescent of the disease and the aim is to have minimal dose (no more than 5-10 mg/day) to control the disease with minimal side effects, if more than 5-10 mg/day IMT should be initiated; However in sever cases such as explosive posterior of pan uveitis with noninfectious etiology, it is indicated high dose I.V. pulse methylprednisalone 1g/day in one hour for three

days followed by gradual tapper of oral prednisalone 1 mg/kg/day but this approach is accompanied with a lot of complications such as hypertension and elevated blood glucose and should be preformed in hospital.
Complications of systemic corticosteroid therapy is many such as hypertension, psychosis, diabetes, peptic ulcer ( which it is mandatory to prescribe H2 receptor blockers) , gastro intestinal reflux, osteoporosis ( which it is important to prescribe D3 vit especially in postmenopausal women ).
Finally it is desirable to consult with patient's rheumatic physician or internist before initiating systemic corticosteroid therapy.

### Intravitreal

Single parsplana intravitreal triamcinolone acetonide injection of 4mg 0,1 ml can produce sustained vision improvement for 3 to 6 months which can be accompanied with recurrence of CME after this period, it is helpful as prophylaxis after intraocular surgery in eyes with uveitis, however this injections can induce complications such as cataract, glaucoma ( 25% require medication 1-2% needs surgery), sterile endophthalmities ( which may be avoided if preservative free triamcinolone), infectious endophthalmities and rhegmatogenous retinal detachment
Parsplana fluocinolone acetonide implant is indicated for pseudophakic patients with chronic noninfectious posterior uveitis which can control uveitis for 36 weeks but there is high risk of glaucoma development; Another implant is dexamethasone 700 mg which can be effective in CME treatment.

## Immune modularity therapy (IMT)

It works by killing the rapid clones of lymphocytes that are responsible of inflammation.

### Indications

Vision threatening of intra ocular inflammation, reversibility of the disease, when corticosteroids are indicated for more than 3 months (more than 5-10 mg/day) inadequate of corticosteroid therapy or failure of treatment, and when corticosteroids are contraindicated due to unaccepted side effects or chronic dependence.
some times it better to start with IMT to improve the prognosis in diseases such as VKH, SO,retinal and systematic vasculitis, behçet, pars planitis, serpiginous, panuveitis, and chronic iridocyclitis

Before treating with IMT the physician should be careful about certain points such as there is no infections  nor hematologic or hepatic status which contraindicated the usage of IMT, the physician should closely follow up the patient and be able to manage any IMT induced toxicity, rgular tests should be preformed such as CBC, renal and hepstic function tests and all that should be after informed  consent.
IMT may not have effect immediately and may need to maintain the steroid dose and the tapper,  treatment should be for 6-24 months, after which gradual tapering and discontinuation of IMT should be attempted over the next 3-12 months and some times it advisable to prescribe bactrim against pneumococcus carinii
There a several types of IMT such antimetabolites, inhibitors of T-cell signaling, alkylating agents, and biologic response modifiers.

## Antimetabolites

### Azathiopirne

It alters purine metabolism the dose of use is 100-250 mg/day (2mg/kg/day) PO the dose is doubled after 2 weeks, azathiopirine is tapered and shouldn't be stopped until 1year remission and due to the drug needs weeks to be effective it is not recommended for acute conditions, it can be used to prevent ocular involvement in behçet without ocular involvement or to prevent the involvement of the contralateral eye, it can be helpful in SO, VKH, intermediate uveitis, and necrotizing scleritis how ever it is advisable to do TPMT to determine the needed dose for treatment with potential complication GI upset (some start as 50mg/day for 1 week to see if there is any GI symptoms) with hepatic toxicity, it is recommended to check CBC and hepatic function tests every 4-6 weeks

### Methotrexate

It is folate analogue, inhabits dihydrofolate reductase the dose of use is 7.5-25 mg/weeks PO or SO it is effective against JIA iridocyclitis(recommended as 1st line for children with increases dose up to 30mg), sarcoidosis, panuveities, scleritis;  potential complication GI upset with hepatic toxicity and fatigue and pneumonitis it is recommended to prescribe folate 1mg/day to reduce side effects, and to avoid alcohols.

### Mycophenolate mofetil

Inhabits purine synthesis 1-3 g/d PO can be a good alternative to Methotrexate potential complication such as diarrhea, nausea, and GI ulcers

## Inhibitors of T-cell signaling

### Cyclosporine

It inhabits nuclear factor of activated t lymphocytes activation it is a mcrolide product of fungus beauveria nivea  it is effective against Behçet, VKH, and intermediate uveitis dose is 2.0-2.5 kg/kg/day potential complications are nephrotoxicity, parestesias, GI upsets, gingival, hypertension that's BP, creatinine level and CBC should be checked monthly, care must be taken of carcinogenic effect when used in the presence of psoriasis.

## Alkylating agents

### Cyclophosphamide

It acts as cross link to DNA  dose is 1-2 mg/kg/day PO, patient should drink 2 liters of fluid a day while the treatment, it can be used to treat necrotizing scleritis, retinal vasculitis, and other refractory uveitic conditions, potential complications are hemorrhagic cystitis, sterility, and increased risk of malignancy, it is recommended to check CBC and urine analysis, it is important to stop cyclophosphamide in the case of leukocytes drop to below 2500 cell/ml, it advisable to prescribe bactrim as prophylaxis against pneumocystis pneumonia.

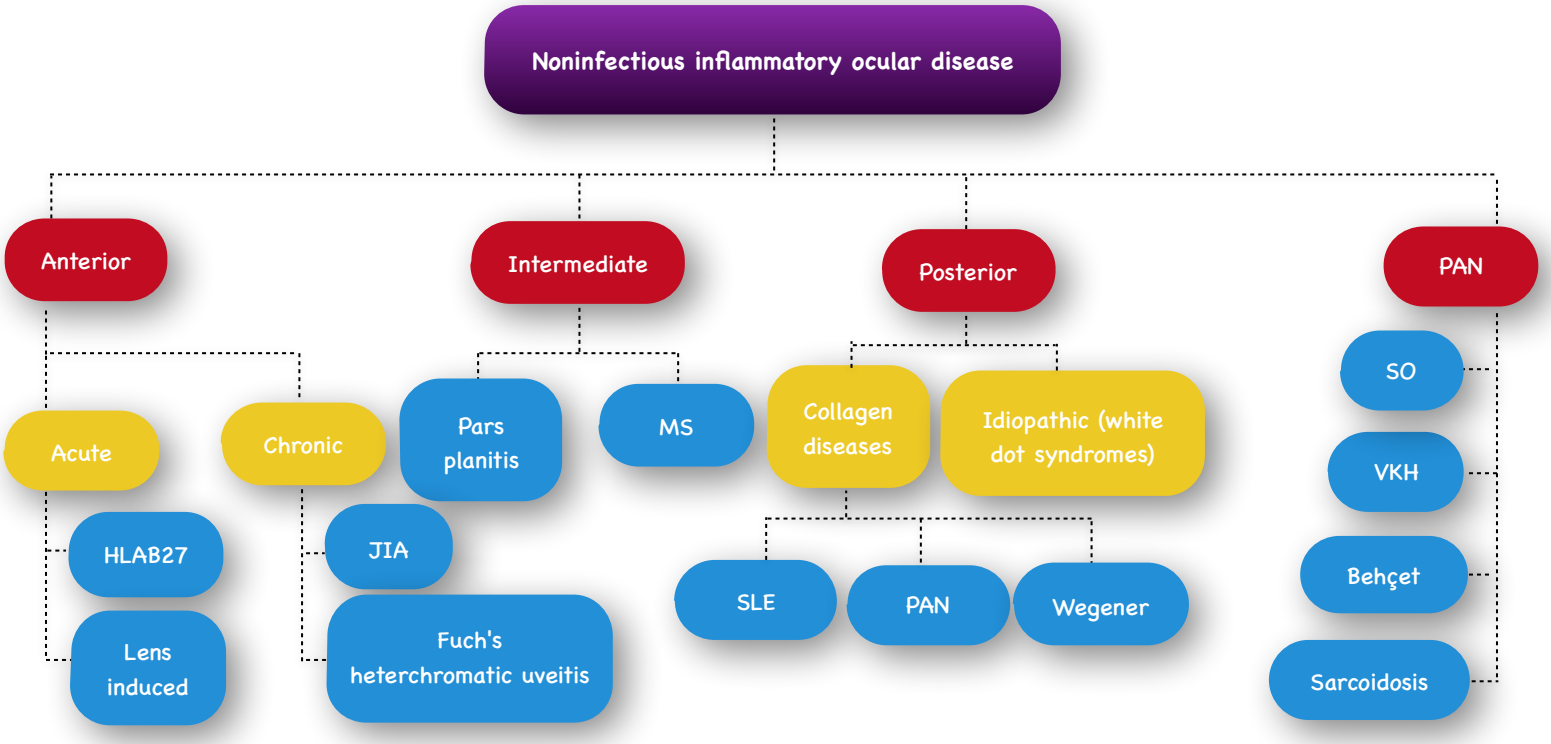
## Biologic response modifiers

### Infliximab

TNF-alpha inhibitor dose of use is 3 mg/kg/ I.V. Weeks 0,2,4 then every 6 weeks it is effective against future attacks of Behçet, sarcoidosis, VKH, HLA-B27 uveitis and idiopath uveitis, potential  complications are infusion reaction, infection such as TB ( Infliximabis contraindicated if PPD is positive), malignancies such as lymphoproliferative diseases, and auto antibodies such as SLE like.

### Adalimumab

TNF-alpha inhibitor dose of use is 40 mg q 1-2 week/s it can be given subcutaneous, it is recommended for pediatric uveitis, Behçet and panuveitis, potential complications such as headache, nausea, rash, stomach upset


Noninfectious inflammatory ocular disease
Anterior
Intermediate
Posterior
PAN
Acute
Chronic
Pars planitis
MS
Collagen diseases
Idiopathic (white dot syndromes)
SO
VKH
Behçet
Sarcoidosis
HLAB27
Lens induced
JIA
Fuch's heterchromatic uveitis
SLE
PAN
Wegener

# Noninfectious inflammatory ocular deceases

## Acute Anterior

### Acute Nongranulomatous irtis and Iridocyclitis:

### Ophthalmic manifestation

**Symptoms:** pain, redness, and photophobia
**Presentation:** unilateral up to 3 month with alternating and recreancy potentials
**Clinical features:**
Corneal: KPs (Small nongranulomatous) and edema
Anterior chamber: cell with little flare and in sever cases hypopyon and fibrin nest which may lead to seclusion membrane with iris bombe
Vitreous & retina: some vitreous cells and CME
IOP: may be elevated
**Treatment:** topical steroids and in sever cases pereocular or subconj with cycloplegic agents to relieve pain and to break synechia

### Systemic manifestation

**HLA-B27:** comes with negative rheumatoid factor.
The seronegative spondyloarthropathies include
-Ankylosing spondylitis
-Reactive arthritis syndrome
-Inflammatory bowel disease
-Psoriatic arthritis (typical cutaneous changes, ungual involvement, nail dystrophy, and terminal phaIangeal joint inflammation with association with sacroiliitis and the ophthalmic implication may be CME and papiltis
**Ankylosing spondylitis:** associated with lower back pain and stiffness after inactivity with HLA-B27 positive with onset in 3rd or 4th decade the uveitis is not correlated to the joint involvement the sacroiliac image studies od sacroilliac should be obtained which shows in the early stages osteoporosis in the juxtaarticular, then sclerosis and Ankylosing spondylitis can be complicated with Pulmonary apical fibrosis and aortitis.
Treatment with NSAIDs morning stiffness and the ESR are lower in patients taking sulfasalazine patients should be warned about possible deformity.
**Reactive arthritis syndrome:** nonspecific urethritis (prostatic fluid is negative), conjunctival inflammation follows urethritis within about 2 weeks and before arthritis (mucopurulent and

papillary punctate and subepithelial keratitis) maybe accompanied by iritis (acute nongranulomatous), and polyarthritis (oligoarticula) with onset in 3rd or 4th decade.
**Inflammatory bowel disease:** Acute iritis is with both Ulcerative colitis and Crohn disease, 20% of patients have sacroiliitis (HLA-B27 positive) while HLA-B27 negative don't

## Lens induced

Uveitis due to an immune reaction to lens material. Which occurs following traumatic or surgical damage of the lens capsule or from leakage of lens protein via lens capsule of mature or hypermature cataract.
It can be classified as phacoanaphylactic endophthalmitis, phacolytic glaucoma and phacotoxic uveitis.
**Clinical features:**
Corneal: KPs (Small or large it can be granulomatous or nongranulomatous)
Anterior chamber: little to sever reaction and in sever cases hypopyon and fibrin nest which may lead to seclusion membrane with iris bombe
Vitreous & retina: vitreous inflammation with no retinal involvement
IOP: may be elevated especially in phacolytic glaucoma due to filling of the trabecular meshwork by lens protein and lipid-laden macrophages.
**Treatment:** topical steroids and in sever cases systemic steroids with cycloplegic agents, the surgical removal is curative and indicated

## Chronic Anterior

## Juvenile idiopathic arthritis

## Ophthalmic manifestation

**Symptoms:** white eye, mild to moderate pain, with photophobia and blurred vision, May have exacerbations (with +4 cells) and remission periods
**Presentation:** develop between 5-7 years with a risk factor for females, pauciarticular, and positive ANA with negative RF and often symmetrical bilateral in 70%
**Clinical features:**
Corneal: band keratopathy with small KPs (Small nongranulomatous)
Anterior chamber: flare and cell, synchiea, and cataract with no hypopyon
Vitreous & retina: vitreous debris and CME.
Complications: amblyopia, band keratopathy, squint, glaucoma, and hypotony
**Treatment:** topical steroids ( dose should be calibrated according to cells not flare in the anterior chamber) and in sever cases pereocular or subconj with cycloplegic agents to relieve pain and to break synechia, Immune modularity therapy (IMT) such as Methotrexate with dose of use is 7.5-25 mg/weeks PO or SO but all with the conjunction with rheumatoid specialist and a pediatrics.

It is so important to fellow up the patient closely to monitor the any development of complications such as amblyopia, band keratopathy, squint, glaucoma, hypotony, synchiea, and cataract which have a high rate of complications and IOL is placed in selected cases and it is so advisable to preform surgery when the eye is without inflammation for at least three months.

## Systemic manifestation

There are three types of JIA
**Still disease:** usually in children younger than 5 years (20%) and uveitis is rare (less than 6%); Clinical features are fever, rash(maculopapular), hepatosplenomegaly, and lymphadenopathy, where joint involvement is minimal or initially absent with negative ANA.
**Polyarticular onset:** presents 40% of JIA ( females more than males) and 10% of iridocyclitis cases ( absent in patients with positive RF) with 4 or more joint involvement while rash is little or absent and ANA is positive.
**Paueiarticular onset:** presents 80-90% of JIA cases with 4 or less joints with maculopapular rush and have 2 subtypes the first type effect girls younger than 5 years and they are ANA positive (risk factor for uveitis) where chronic iridocyclitis is 25% and the second type is more common in older boys where JIA behaves like seronegative scaroilities with episodes of acute anterior uveitis and recurrent.

## Fuch's heterchromatic uveitis

**Symptoms:** none, mild blurring of vision with discomfort, and deferent iris colors
**Presentation:** 2%-3% of uveitis cases in the 3rd-4th decades with Unilatral heterchromic iridocyclitis; Parry-Romberg syndrome may be associated
**Clinical features:**
Corneal: Stellate KPs (covers the whole corneal endothelium)
Anterior chamber: Mild reaction, faint flare, diffuse iris atrophy which cause heterchromia iridis lighter in the uveitic eye (where iris will be darker in blue colored eyes) with posterior iris pigment atrophy which can be seen in retroillumination , trabecular meshwork is crossed by fine vessels (may cause bleeding during cataract extraction) with small peripheral synchiea and no posterior synchiea.
Vitreous & retina: vitreous debris.
Complications: Cataract which with good prognosis after extraction (but sometimes vitreous opacification may occur which needs vitrectomy), and glaucoma.
**Treatment:** Steroids and cycloplegic agents won't do a significant help, but the prognosis is good how ever in the case of moderate vitreous opacification subtenon steroid injection is indicated where in sever cases vitrectomy is indicated.

## Intermediate uveitis

### Idiopathic parsplanitis

**Symptoms:** Floaters, discomfort, redness and photophobia
**Presentation:** Equal between males and females with age of 5 to 40 years and bilateral in 80% classes but asymmetric and relapsing.
**Clinical features:**
Anterior chamber: Inflammation with spellover of anterior chamber cells.
Vitreous: Vitreous cells, snow balls, snow bank, parsplana exudates
Retina: phelibitis with venous sheathing and CME which the main cause of visual loss where FFA shows venal and disc leakage and CME, in the cases of opaque media US is helpful in detecting membranes and peripheral exudates.
Complications: phelibitis with venous sheathing which may cause ischemia and neovacularzation of the inferior pars planitis and may lead to hemorrhage and TRD
Also cataract may occur which can be complicated with low grade inflammation after extraction and posterior capsular opacification, it is recommended to start with oral prednisone 60 mg daily 5 days before the surgery and tapering it over the next month, it is also recommended to preform pars plana vitrectomy at the time of cataract surgery in the case of significant opacification of vitreous at the time of the surgery.
**Treatment:**
Step1: 4 pereocular or subtenon injections of Triamciloinone or methylprednisolone within one month, more injections are applied as required if there is no CME resolution, but caution with patients with glaucoma as steroids elevate IOP and also can cause cataract, it is also possible to use intravitreal Triamciloinone and it is important to avoid inject in the area of snow banking ,fluocinolone acetonide 0.59 mg or intravitreal implant dexamethasone 0.7 mg intravitreal implant are indicated in pseudophakic eyes, Systemic oral steroid can be used in the bilateral sever cases but not for more than 3 months
Step 2: If steroids didn't work out then Cryotherapy can be placed as double throw in one hour away from the snow banking manner where it is contraindicated in the cases of tractional retinal detachment or neovacularzation.
Or peripheral scatter retinal photocoagulation may be safer and placed as 3-4 throws posteriorly and not on the snow bank where it is contraindicated.
Step 3: IMT such as methotrexate or cyclosporine can be used in bilateral cases and can help to reduce the corticosteroid doses
Step 4: Vitrectomy is indicated when all other therapies failed or contraindicated and in the cases of epiretinal membranes and tractional retinal detachment, besides it is helpful to treat CME.
**Prognosis:**long term prognosis is good but it can be divided into three types of courses: the self limiting (10%), a course with remissions and exacerbations (30%) and the prolonged course (60%)

## Multiple sclerosis (MS)

**Symptoms:** Floaters, and discomfort
**Presentation:** Bilateral, between 20-50 of age and 30% of patients with MS will develop uveitis where 15% of patients with pars planitis will develop MS with HLA-DR15 assossition
**Clinical features:**
Vitreous: Mild vitreous cells.
Retina: periphelibitis not related to optic neuritis and CME is less common.
**Treatment:**
-MRI of the brain with gadolinium
-Consult with neurologist for follow up
-Mythelprednisalone **I.V.** 1 mg/kg/day
-Interferon beta-1a (Avonex)

## Deferential diagnosis

**Fuch's uveitis syndrome:** beside being unilateral, and the anterior chamber involvement, there is no CME
**Primary intraocular lymphoma:** snow balls are absent in elder age > 50 years
**Toxocariasis with peripheral granuloma:** usually unilateral with mild vitritis with snowbanking like configuration.
**Endogenous candida endophthalmitis** may be associated with snowbals.
**Amyloidosis** may presents as vitreous opacities but no vasculitis nor CME.
**Toxoplasmosis** can be with sever vitritis that can hide the focus of inflammation or appear as a light in the fog and it is so important to check for toxoplasmosis before starting any steroid therapy a specially periocular injections.
**Whipple disease** may presents with vitritis but no snowballs.

## Posterior uveitis

## Collagen diseases

### Systemic lupus erythematosus (SLE)

### Ophthalmic manifestation

**Presentation:** Young Afro-American or Hispanic women are at high risk with HLA-B8, HLA-DR3 association and positive ANA in serology.
**Clinical features:**
Eyelids: Discoid lupus erythematosus in 50% of patients
External disease: Sjögren disease (20%), and scleritis
Retina: Posterior manifestations can be related to the diseases severity and however SLE posterior segment manifests in a three ways
-Cotton-wool spots and hemorrhages which is due to microangiopathy induced by SLE not due to hypertension
-Vascular occlusive disease which produce ischemia, neovacularzation and hemorrhage where in sever cases there is a CNS manifestation and antiphospholipid antibodies may be positive.
-Lupus choirodopathy which presents as serous focal detachment of retina or/and RPE due to systemic vasculitis or nephrotic hypertension (which can cause arterial narrowing, hemorrhage and optic disc edema).
Cranial nerves: palsy with optic neuropathy with retro chiasmal manifestations.
**Treatment:** It is important to Consult with rheumatologist
-NASIDS, Systemic corticosteroid, palsmapheresis and IMT
-Hypertension therapy in the case of hypertension
-Antiplatelet therapy for patients with antiphospholipid antibodies
-In the case of retinal proliferation or hemorrhage laser photocoagulation or vitrectomy is indicated.

### Systemic manifestation

The diagnosis is clinical and based on finding 4 of 11 manifestation:
Acute cutaneous diseases in 70%-80% of patients such as malar rash, discoid lupus, photosensitivity, mucosal lesions, and discoidrash
Renal disease such as nephritis, proteinuria in 50%-75%
Arthritis in 80%- 85%
Neurologic manifestation in 35% such as psychos and seizures
Raynaud phenomenon in 30%- 50%
Cardiac, hepatic and pulmonary disease.
Hematologic disorder such as lymphopenia,or thrombocytopen, hemolyticanemia,or leukopenia.

# Polyarteritis nodosa (PAN)

## Ophthalmic manifestation

**Presentation:** Focal acute or chronic necrotizing inflammation of medium-sized and small muscular arteries presents between 40-60 years with ocular manifestations (20%) and 3 times in males more than females where they are ANCA and hepatitis B surface antigen positive.
**Clinical features:**
External diseases: PUK and scleritis
Retina: Posterior manifestations can be related to the diseases severity and however PAN posterior segment manifests in a two ways
-Hypertension retinopathy cotton-wool spots and hemorrhages with macular star formation which is due to nephropathy
-Arteriolar occlusive disease and choroidal infarcts ( where Elschnig pearls are observed) with serous retinal detachment due to vasculitis
Cranial nerves: palsy with optic atrophy with homonymous hemianopia, horner syndrome and amaurosis fugax
**Treatment:** It is important to treat in the conjunction with rheumatologist
-Systemic corticosteroid reduced mortality rate to 50% , and IMT like cyclophosphamide 50% enhanced 5 years survival
-High mortality rate 90% if untreated

## Systemic manifestation

**Symptoms:** in 75% of patients arthralgia, fever, fatigue and weight loss
PAN diagnosis is conformed by finding 3 of 10 manifestations:
weight loss > 4kg
Myalgia,weakness or leg tenderness
Testicular tenderness
Livedo reticularis
Hepatitis B surface antigen positive.
Blood urea nitrogen is elevated
Polyneuropathy or Mononeuropathy
Diastolic blood pressure >90mmHg
Neutrophils are demonstrated on biopsy specimens of small or medium sized arteries
Arteriographic findings are abnormal
Ischemic bowl disease which lead to complications
Subcutaneous nodule and purpura

# Wegener granulomatosis

## Ophthalmic manifestation

**Presentation:** Triad of focal segmental glomerulonephritis, necrotizing granulomatous vasculitis of the upper and lower respiratory tract and necrotizing vasculitis of small arteries and veins, ocular involvement is 50% diagnosis is established by biopsy, chest X-ray C-reactive protein and ANCAs where C-ANCAs is specific for wegener and P-ANCAs for PAN
**Clinical features:**
Orbital: Pseudotumor, dacryocystitis and orbital cellulitis
External diseases: PUK and scleritis and anterior with intermediate uveitis
Retina:
-Cotton-wool spots and hemorrhages and retinitis
-Vaso occlusive disease like CRAO BRAO, vasculitis, neovacularzation, and
Cranial nerves: ischemic optic neuropathy
**Treatment:** It is important to Consult with rheumatologist
-Systemic corticosteroid, and IMT like cyclophosphamide
-High mortality rate within one year if untreated

## Systemic manifestation

**Symptoms:** Pulmonary symptomatology, arthritis, and sinusitis

Paranasal sinuses Involvement important clinical finding, then pulmonary involvement and renal diseases.
Renal involvement 85% manifests as glomerulonephritis carries high risk of mortality.
Involvement of the skin manifests as purpura with subcutaneous nodules and ulcers.
Neurological involvement manifests as peripheral neuropathies, and seizures, cranial neuropathies, and cerebral vasculitis.

# White dot syndromes

## Birdshot

**Presentation:** Autoimmune bilateral insidious, chronic recurrent presents in ages between 30-70 more in females with HLA-A29 association

**Symptoms:** Blurry vision , photopsias, floaters with distortion of color and night vision

**Clinical features:**

Vitreous: Vitritis

Retina: white-yellow, creamy, ovoid or irregular lesions (50-1500 micron), located posterior to the equator which do not pigment

Ancillary testing: FFA; early hypofluorescence with late subtle leak with evidence of CME, vessel or disc leakage ICG; more hypofluorescence than in FFA ERG; abnormal with reduced b-wave with time

Complications:CME, disk edema, CNV, and Vasculitis

**Treatment:** Systemic steroids and IMT such as ciclosporin

**Prognosis:** Guarded if not treated

## Acute posterior multifocal placoid pigment epitheliopathy (APMPPE)

**Presentation:** Bilateral Acute self limiting presents in ages between 20-50 where females and males are equal with viral predrome and HLA-B7 association.

**Symptoms:** Blurry vision , scotomas and photopsias

**Clinical features:**

Vitreous: No vitritis

Retina: Multifocal white-gray, lesions (within 2 disc areas), which are located in the outer retina or RPE and can get pigmented

Ancillary testing: FFA; early hypofluorescence with late hyperfluorescence and window defect ICG; hypofluorescence corresponds to have seen in FFA ERG; abnormal

Complications: Disk edema

**Treatment:** Systemic steroids if there is CNS involvement

**Prognosis:** good

## Serpiginous choroiditis

**Presentation:** Bilateral but asymmetric chronic and recurrent presents in ages between 20-60 where females and males are equal with HLA-B7 and herpes association.

**Symptoms:** Blurry vision ,and scotomas

**Clinical features:**
Mild anterior uveitis
Vitreous: No to mild vitritis
Retina: Geographic yellow-gray lesions extends from peripapillary (in 1/3 of patients it may start from the periphery) to macular area which associated with RPE and choroicapillary atrophy ( inactive lesions) and which is awakes (active lesions for several months) from the peripheral edges and continue it's extension centripetally.
Ancillary testing: FFA; early hypofluorescence with late staining and leack from the border and CNVM if present ICG; hypofluorescence then late staining more than seen in FFA ERG; normal
Complications: RPE pigmentation, molting with choroicapillary atrophy and a risk of CNV and sub retinal fibrosis
**Treatment:** Systemic steroids, IMT like azathioprine, Anti-VEGF, PDT or laser treatment for CNVM
**Prognosis:** Guarded

## Multifocal choroiditis and panuveitis syndrome (MCP)

**Presentation:** Bilateral asymmetrical chronic and recurrent presents in ages between 9-69 where myopic females more than males.
**Symptoms:** Blurry vision , metamorphopsia, floaters, scotomas and photopsias
**Clinical features:**
Iridocyclitis
Vitreous: vitritis
Retina: Multifocal white-yellow chorioretinal oval lesions (50-200 micron), in clumbs or linear streaks in a punch out scars configuration.
Ancillary testing: FFA; early hypofluorescence from blockage with late staining and leakage from CNVM or CME if presented ICG; multi hypofluorescence around the optic disc more than is seen in FFA ERG; Normal
Complications: Disk edema with peripapilary atrophy with CME and CNVM
**Treatment:** Systemic steroids, IMT, Anti-VEGF, PDT or laser treatment for CNVM
**Prognosis:** Guarded

## Punctate inner choroiditis (PIC)

**Presentation:** Bilateral acute and self limiting presents in ages between 29-40 where females (myopic) more than males.
**Symptoms:** Metamorphopsia, scotomas and photopsias
**Clinical features:**
Vitreous: No vitritis

Retina: White-yellow chorioretinal lesions in the posterior pole with fuzzy borders which got pigmented within weeks from the acute attack.
Ancillary testing: FFA; early hyperfluorescence with late leakage and leakage from CNVM if presented ICG; multi hypofluorescence around the optic disc corresponds that seen in FFA ERG; Normal Visual field; enlargement of the blind spot.

Complications: Serous retinal detachment over the lesion and CNVM
**Treatment:** Systemic steroids or observation, IMT, Anti-VEGF, PDT or laser treatment for CNVM
**Prognosis:** good if no CNVM

## Subretinal fibrosis and uveitis syndrome (SFU)

**Presentation:** Bilateral asymmetric chronic recurrent autoimmune presents in ages between 29-40 where in females only.
**Symptoms:** Decreased and blurred vision
**Clinical features:**
Vitreous: Mild vitritis
Retina: White-yellow subretinal lesions that may coalesce (50-500 micron) in the posterior pole to the mid periphery with RPE hypertrophy and atrophy and scar formation
Ancillary testing: FFA; early hypo and molted hyperfluorescence and window defect with late satinig from edges, leakage and leakage from CNVM or CME if presented
Complications: Serous neurosensory retinal detachment over the lesion, CME and CNVM
**Treatment:** Systemic steroids, IMT, Anti-VEGF, PDT or laser treatment for CNVM
**Prognosis:** Guarded

## Multiple evanescent white dot syndrome (MEWDS)

**Presentation:** Unilateral, acute self limiting autoimmune presents in ages between 10-47 where females more than males and viral etiology is assumed.
**Symptoms:** Decreased and blurred vision with scotomas and photopsias.
**Clinical features:**
Iridocyclitis and myopia
Vitreous: Vitritis
Retina: White-orange lesions (100-200 micron) in the posterior pole to mid periphery between RPE and outer retina which fades within weeks with macular granulaty
Ancillary testing: FFA; early hyperfluorescence and wreath like configuration with late staining ICG; hypofluorescence more than seen in FFA ERG; reduced a wave Visual Field; enlarged blind spot and para central scotoma.
Complications: vinous sheathing and optic disc edema

**Treatment:** observation
**Prognosis:** very good

## Pan uveitis

### Symptomatic ophthalmia (SO):

### Ophthalmic manifestation

**Presentation:** Asymmetric bilateral, non-necrotizing, granulomatous panuveitis may occur after penetrating trauma especially if there is prolapse of uvea or surgery to injured eye (more inflamed) and after a period the second uninjured eye (from 3 months to 1 year) will be involved, vitrectomy and age are high risks.
**Symptoms:** Photophobia, pain, blurred vision (near more than far) and mild redness
**Clinical features:**
Anterior chamber: Diffuse granulomatous mouton fat KPs with iris thinking and nodules and posterior & peripheral anterior synchiea
Vitreous: Vitritis which can be moderate to sever
Retina:
-Dalen-Fuchs nodules (sub RPE yellow white choroidial lesions) which is located in the med equator ( can be located in peripapilary) which can be confluent and may produce scar at the macula.
-Exudative retinal detachment can be seen
-Sunset glow like VKH
-FFA; hyperfluorescence with pooling due to neurosensory detachment with leakage from the RPE where Dalen-Fuchs shows early hypofluorescence and then hyperfluorescence
-B-scan shows choroidial thinking
Complications: Elevated IOP due to trabeculitis or low IOP due to ciliary shutdown, optic disc atrophy cataract, CME, and CNVM
**Treatment:**
-It is important to consider Enucleation for vision hopeless sever traumatized eyes within 10 days of trauma
-Systemic corticosteroid, where topical corticosteroid with cycloplegia are used for anterior and periocular for recurrent cases with CME, intravitreal injections of Triamciloinone and intravitreal implants such as fluocinolone acetonide 0.59 mg and dexamethasone 0.7 mg can be used in the case of contraindications of systemic steroids.
-IMT like azathioprine
-**Follow up** initially every 1-7 days then every month steroids should be maintained for 3-6 months after all signs of inflammation has resolved
-**Prognosis** is Good if treated (60% 20/40)

### Systemic manifestation

CSF: pleocltosis, alopecia, vitiligo, poliosis, sensory neural hearing disturbance

## Diferantial diagnosis

-VKH syndrome :no history of ocular surgery or trauma.
-Phacoanaphylactic endophthalmitis: Severe reaction in anterior chamber as a result of injury to the lens antrior capsule.
-Syphilis: Positive FTA-ABS.
-Tuberculosis: Positive PPD and chest X-ray
-Sarcoidosis: elevated ACE and chest X-ray

## Vogl-Koyanagi-Harada Syndrome (VKH):

## Ophthalmic manifestation

**Presentation:** Asymmetric bilateral, chronic, autoimmune granulomatous panuveitis associated with cutaneous, auditory , and neurologic disorders especially Hispanic and middle Eastern (ethnics which are darkly pigmented) with HLA-DR4 or HLA-DR1 association (it is thought that T lymphocytes attack melanocytes of all organ tissues)
**Symptoms:** Blurred vision, photophobia, pain and redness
**Clinical features:**
Anterior chamber: Bilateral diffuse granulomatous mouton fat KPs with flare and cells, nodules and shallow AC due to ciliary body edema posterior & peripheral anterior synchiea and iris atrophy in recurrent and chronic stage
Vitreous: Vitritis with opacities
Retina:(harada)
-Peripapillary thickening of the choroid with elevation of the retinal choroidal layer with papilitis
-Focal serous retinal detachment which can coalesces and form a bolus detachment
-Edema and hyperemia and of the optic disc
-Sunset glow appearance in the retinal after serous retinal detachment resolution in convalescent stage and retinal focal depigmentation is presented in this stage too.
-FFA; hyperfluorescence with pooling due to neurosensory detachment with leakage from the RPE where in the convalescent and chronic stage we see window defects due to RPE atrophy
-B-scan shows choroidial thinking in the peripapilary area to the equator
-OCT; helpful to monitor serious detachments
Complications: Elevated IOP or low IOP due to ciliary shutdown, posterior subcapsular cataract, sub retinal fibrosis, and CNVM
**Treatment:** It is important to consult with neurologist and interest
-Systemic corticosteroid can be oral prednisalone 1 mg/kg or 200 mg of intravenous methylprednisolone for 3 days then high dose oral prednisalone , where topical corticosteroid with

cycloplegia are used for anterior and periocular for recurrent cases with CME, intravitreal injections of Triamciloinone and intravitreal implants such as fluocinolone acetonide 0.59 mg and dexamethasone 0.7 mg can be used in the case of contraindications of systemic steroids.
-IMT like azathioprine
-**Follow up** initially every 1-7 days then every month steroids should be maintained for 3-6 months after all signs of inflammation has resolved
-**Prognosis** is fair if treated (70% 20/40)

## Systemic manifestation

Stages of VKH syndrome:
Prodromal: Flulike presentation with, hypersensitivity of the hair and skin, focal neurologic signs, cranial neuropathies, hemiparesis, meningitis with stiffness of the nick and headache, CSF fluid analysis reveals lymphocytic pleocytosis and normal glucose levels , auditory disorders (75%) & tinnitus
Acute uveitic: It starts after 2 days of neurological onset with blurred vision check the ophthalmic manifestation above
Convalescent: alopecia, vitiligo, and poliosis with sunset glow and pigmentation of the retina
Chronic recurrent: check the ophthalmic manifestation above

## Behçet Disease:

## Ophthalmic manifestation

**Presentation:** Bilateral ,chronic, relapsing, nongranulomatous panuveities with occlusive systemic vasculitis presented in male and females equally where more in males in ocular disease, especially in eastern Mediterranean & Asians between 25-35 of age it can be sporadic (most of cases) or familial with positive pathergy (Behçetine) test
**Symptoms:** Blurred vision, photophobia, pain, floaters and redness
**Clinical features:**
External diseases: Ulcers in the conjunctiva, episcleritis, scleritis, and ring opacities in cornea.
Anterior chamber: Bilateral nongranulomatous KPs with mobile hypopyon iris bombè posterior synchiea
Vitreous: Vitritis
Retina:
-Necrotizing retinal vasculitis ( veins and artery), BRVO, or/and BRAO with vascular sheathing, retinal ischemia, multi focal white chalky retinitis and papilties due to vasculitis of the optic disc vessels.
-FFA; Vascular dilatation with retinal ischemia, and leak from CME and neovacularzation
-B-scan shows choroidial thinking in the peripapilary area to the equator
-OCT; helpful to monitor serious detachments

Complications: Cataract, conjunctivitis, conjunctival ulcers, scleritis, episclritis, angle closure, neovascular glaucoma, ophthalmoplegia, retinal neovacularzation and optic disc atrophy.

**Treatment:** It is important to consult with rheumatologist

-Systemic corticosteroid can be oral prednisalone 1-2 mg/kg for acute attacks but patients will be resistant to it eventually ,before using steroids it is recommended to have a negative FTA-ABS/RPR and PPD

-IMT should be used in the conjunction with steroids like azathioprine which is useful in preserving vision ; infliximab 10 mg/kg which is effective in controlling relapses, help to steroid tapering and vision threatening posterior uveitis and Colchicine for mucocutaneous disease ;Subcutaneous interferon alfa is helpful too

-**Prognosis** is guarded if treated (20/200)

## Systemic manifestation

Types of Beheçet Disease

Complete (4 major criteria)

Incomplete (3 major criteria or ocular involvement with 1 other major criterion)

Suspect (2 major criteria with no ocular involvement)

Possible (1 major criterion)

Major criteria

Recurrent oral big or small aphthous ulcers (white with red borders) which causes pain and discomfort and heal without significant scar.

Ocular inflammatory disease

Skin lesions (erythema nodosum, folliculitis, acneiform pustules)

Recurrent genital ulcers

Arthritis large joints, as knees

Minor criteria arthritis

Systemic vasculitis or associated complications

Neuropsychiatric symptoms

Cardiac manifestation

Epididymitis

Gastrointestinal ulceration

## Diferential diagnosis

-Hypopyon may occur in seronegative spondyloarthropathies, but the acute antroir uveitis isn't bilateral simultaneously where hypopyon won't be mobile because of it's assosition with fibrinous exudate where in Behçet bilateral antrior uveitis is usauly occur simultaneously with mobile hypopyon which moves with gravity when ever patient moves his head.

-Retinal vasculitis manifests in sarcoidosis but, vasculitis in sarcoidosis affect veins in segmental way and occlusion happens rarly in contrast to Behçet where usualy vasculitis occur in both veins and arteries, where in a difuse manner, and more occlusions with vitritis.
-Retinal infiltrates in Behçet may look like the infiltrates which we see in viral retinitis (which they finally coalesce) like as acute retinal necrosis.
Multiple retinal infiltrates in Behçet may look like in idiopathic acute multifocal retinitis but course and prognosis is better.

## Sarcoidosis:

### Ophthalmic manifestation

**Presentation:** Idiopathic, chronic bilateral, non-caseating granulomatous panuveities (50% of systemic sarcoidosis) with intrathoracic enlargement of parotid gland, arthralgia, and fever manifestations presented in male and females equally, especially in northern European countries & Afro American between 20-50 of age (pediatric manifestation would be atypical presentation with articular and cutaneous associations); Risk factors include with mycobacterial infection, family history and HLA-DRB1 and the diagnosis is conformed by chest X-ray, elevated ACE, positive gallium scanning and negative mantoux test
**Symptoms:** Blurred vision, photophobia, pain, floaters and redness
**Clinical features:**
Orbit and adnexa: eyelid granuloma and lacrimal gland infiltrations
External diseases: Conjunctival nodules, nummular and interstitial keratitis and band keratopathy
Anterior chamber: Bilateral granulomatous iridocyclitis with mutton-fat KPs with mobile hypopyon iris nodules (Koeppe and Busacca), bombé, peripheral anterior & posterior synchiea and angle closure.
Vitreous: Vitritis (snowballs) or vitreal opacity in linearly fashion
Retina:
-Granulomatous nodules with in the retina and choroid located either posteriorly, peripherally or even on the optic disc (that doesn't cause visual loss) which may confluent to produce amebic like or in rare cases solitary nodule
-Perivascular sheathing with diffuse or segmental granular called candle-waxdrippings with the risk of CME, CRVO, BRVO, and retinal ischemia.
Complications: Cataract, conjunctivitis, conjunctival ulcers, scleritis, episclritis, angle closure, neovascular glaucoma, ophthalmoplegia, CNVM, retinal neovacularzation, vitreous hemorrhage and optic disc edema.
**Treatment:** It is important to consult with rheumatologist
-Systemic corticosteroid can be oral prednisalone 1 mg/kg (40-80 mg/day), where topical corticosteroid with cycloplegia are used for anterior uveitis and periocular, intravitreal injections of Triamciloinone and intravitreal implants such as fluocinolone acetonide 0.59 mg and dexamethasone 0.7 mg can be used in the case of contraindications of systemic steroids.

-IMT like methotrexate, infliximab

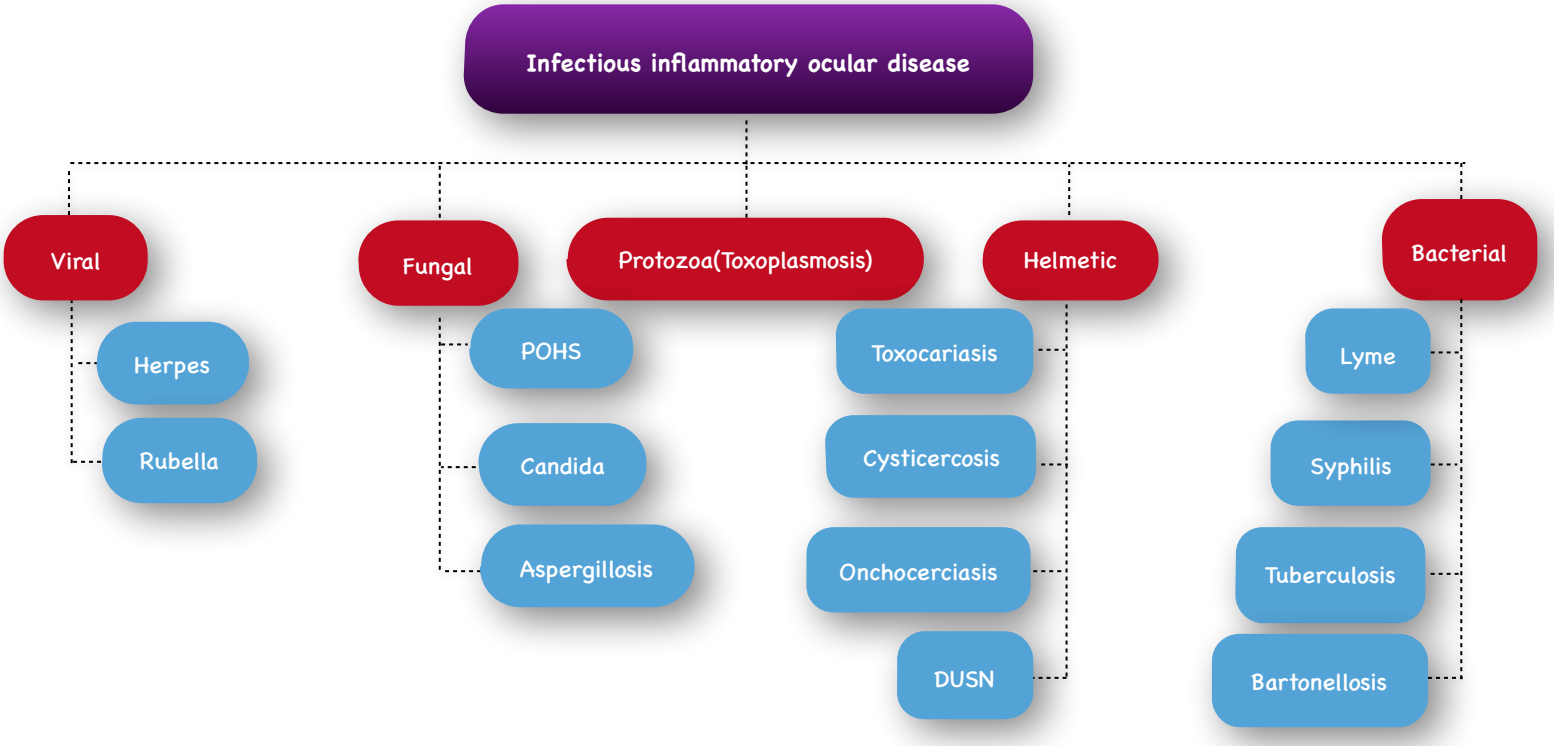
## Systemic manifestation

Löfgren syndrome consists of bilateral hilar adenopathy, arthropathy, erthema nodosum, and iritis and can be treated with systemic corticosteroid.

Heerfordt syndrome is presented as facial nerve palsy, parotitis, uveitis, and fever

## Differential Diagnosis

-Mutton-fat KP and nodules on iris include,tuberculosis (+ mantoux test), syphilis (+ VDRL & FTA-ABS) , lens-induced uveitis, and sympathetic ophthalmia(trauma history).
-Intermediate uveitis MS (neurological manifestation; MRI).
-Posterior uveitis with multiple chorioretinal lesions may be from birdshot retinochoroidopathy (white-yellow, creamy, lesions, which do not pigment FFA; early hypofluorescence with late subtle leak) , intraocular lymphoma (MRI), syphilis, sympathetic ophthalmia,VKH syndrome (CSF fluid analysis reveals lymphocytic pleocytosis and normal glucose levels , auditory disorders (75%) & tinnitus alopecia, vitiligo, and poliosis with sunset glow and pigmentation of the retina)


Infectious inflammatory ocular disease
Viral
Herpes
Rubella
Fungal
POHS
Candida
Aspergillosis
Protozoa(Toxoplasmosis)
Helmetic
Toxocariasis
Cysticercosis
Onchocerciasis
DUSN
Bacterial
Lyme
Syphilis
Tuberculosis
Bartonellosis

# Infectious inflammatory ocular deceases

## Viral

## Herps

### HSV & HZV

### Ophthalmic manifestation

**Symptoms:** pain, redness, and photophobia

**Presentation:** Acute, self limiting, mild, bilateral, nongranulomatous iridocyclitis and/or retinitis with or without keratitis or cutaneous vesicles, however chronic inflammation may occur.

**Clinical features:**

Corneal: stellate KPs in a diffuse pattern or in the inferior cornea (arlt triangle), herpatic keratopathy, corneal sensation is decreased and neurotrophic keratitis.

Anterior chamber: cells and hyphema with iris atrophy which can be sectritial (HZV) or patchy (HSV) and posterior synechiae manifests with ARN.

Vitreous & retina:

-ARN ( acute retinal necrosis): it usually occurs in healthy individuals which but it may occur in children and immune compromised patients too, and may manifest without prodrome or follow herpatic cutaneous or encephalic manifestations, it starts as unilateral then the fellow eye will be involved from 6 weeks to 26 years where ARN occurs in males more than females, ARN manifests in the conjunction of panuveities including **vitritis** , vascular occlusion with involvement of arteriolars, and peripheral peripheral multifocal retinitis (posterior pole is unaffected till late stages) in yellow-white color which in the beginning they look like scalloped separated then eventually got coalesce in a confluent 360 degree then progress to full thickness retinal necrosis with hemorrhages and vasculitis including veins and arteries, diagnosis is conformed with ELISA (titers for HSV HZV) from vitreous cultures, it is important to note the patines with ARN are at risk of encephalitis and meningitis.

-PORN (progressive outer retinal necrosis): it usually occurs in immune compromised patients (AIDS CD +4 less than 50 micro L) and presented as bilateral (70%) multifocal retinitis in yellow-white color but in the posterior pole with **NO Vitritis** (vitritis occurs in extensive necrosis of the retina)

-Non necrotizing herpetic retinitis: in children it appears as bilateral diffuse hemorrhage and acute retinochoroiditis while in adults it appears as vasculitits and chronic choroiditis

IOP: Elevated (50-60) due to trabeculitis

Complications: Tractional, rhegmatogenous (which is repaired with parsplana vitreoctomy but prophylaxis by laser photocoagulation is important ), and exudative retinal detachments

**Treatment:**
Anterior chamber: topical steroids (prolonged treatment with tapering) and cycloplegia, while topical antiviral are used in the presentation of denretic ulcers, but systemic antiviral like acyclovir 400mg bid (for HSV) 800mg bid (for HZV) would be beneficial
ARN:
-Intravenous acyclovir 10 mg/kg/day divided into 3 doses for 2 weeks then oral acyclovir 800mg for 5 times or oral valacyclovir 1 g t.i.d or oral famciclovir 500 mg t.i.d for 3 months, and after 24-48hr start prednisone 1 mg/kg/day.
-Intravitreal ganciclovir 0.2-2.0 mg/0.1 ml or foscarnet 1.2-2.4 mg/0,1 mL with systemic antiviral therapy would rapid the resolution
-Aspirin
PORN:
-A combination of Systemic and intraocular antiviral therapy because PORN is resistant for I.V. treatment
-HAART
Non necrotizing herpetic retinitis:
Systemic corticosteroid with IMT after response switch to systemic antiviral because it is resistant to classic therapy
**Prognosis:** Guarded for ARN (less than 20/200) while poor for PORN

## Cytomegalovirus (CMV)

### Ophthalmic manifestation

**Symptoms:** asymptomatic , scotoma and rarely photophobia
**Presentation:** in neonates and immunecompramised patients (AIDS CD+4 less than 50 micro L) and manifests as CMV retinitis and diagnosis is conformed with inclusion bodies of CMV in saliva and urine, and PCR analysis from aqueous or vitreous.
**Clinical features:**
Anterior chamber: rare, unilateral with stellate KPs and sectritial iris atrophy.
Vitreous & retina:
-Classic retinitis with hemorrhages in the retina against edematous and necrotic retina from disc to vascular arcades which respect the distribution of RNFL (brushfire-like) while vitritis absent to mild depends at level of CD+4
-Granular from which presents in the periphery with no necrosis nor edema or hemorrhage but progress from it borders
-Perivascular form which occurs in immunecompetent children and resemble a frosted branch angiitis.
IOP: Elevated

Complications: Rhegmatogenous retinal detachment from posterior giant retinal breaks which is repaired with parsplana vitreoctomy but prophylaxis by laser photocoagulation is important, CME, epiretinal membrane and RPE atrophy

**Treatment:**

-Intravenous ganciclovir ( which may be resistant) 5 mg/kg/day divided into 2 doses for 2 weeks then oral valganciclovir 900mg b.i.d for 3 weeks then oral valganciclovir 900mg/day.
-Intravitreal ganciclovir 0.2-2.0 mg/0.1 ml or ganciclovir implant which maintain dose for 8 months.
-HAART for long term and HAART plays a role in reducing the CMV retinitis up to 80%

**Prognosis:**

Regression is good if treatment started initially

## Systemic manifestation

Thrombocytopenia, Fever, pneumonitis, hepatosplenomegy, and anemia.

## Deferential diagnosis

| Signs | PORN | ARN | CMV | Toxoplasmosis |
|---|---|---|---|---|
| Retinal hemorrhages | + | ++ | +++ | - |
| Vitritis | - | +++ | - | +++ |
| Pain | - | +++ | - | ++ |
| Immune status | Compromised | Competent | Compromised | Might be in both |
| Clinical features | Multifocal retinitis in yellow-white color but in the posterior pole | Peripheral multifocal retinitis which looks like which eventually coalesce and progress to retinal necrosis and vasculitis and vascular occlusion. | Retina hemorrhages against edematous and necrotic retina from disc to vascular arcades (brushfire-like) | Chorioretinitis with smooth edges (headlight in the fog) |

# Rubella

## Ophthalmic manifestation

**Symptoms:** vision decreased acutely in the acquired form
**Presentation:** As congenital due to rubella togavirus fetal infection in the first trimester (1st 10 days) or 3rd trimester (last month) with systemic manifestations such as cardiac, deafness and ocular or rubella can be presented as acquired (German measles) which manifests as fever, malaise, rash, ocular, and lymphadenopathy.
**Clinical features:**
Anterior chamber: it might be implicated in Fuch's heterchromatic uveitis and may develop iris atrophy
Vitreous & retina:
-Congenital: Pigmentary retinopathy in a salt and pepper configuration but the optic disc and vessels are normal
-Acquired: multifocal chorioretinitis with bolus exudative retinal detachment which resolves spontaneously and preretinal vitritis
IOP: Elevated
Complications:
Congenital: Corneal clouding, strabismus, cataract which it's material contains virus which are liberated after extraction, microphthalmia, glaucoma, and CNVM
Acquired: RPE atrophy

**Treatment:** supportive and systemic corticosteroid can be used in post vaccination optic neuritis and retinitis.
**Prognosis:** good in congenital

## Systemic manifestation

Congenital: cardiac (pulmonic stenosis and interventricular septal defects), deafness, and high risk of diabetes.

Acquired: fever, malaise, rash (which manifests in face, hands and feet then cover the whole body within 24 hrs and then disappears in the 3rd day as erlthematous maculopapular) and lymphadenopathy

**Deferential diagnosis** With TORCHES syndrome, CMV, HZV, HSV, post vaccination encephalitis and mumps

## Fungal

### Presumed Ocular Histoplasmosis Syndrome (POHS)

### Ophthalmic manifestation

**Symptoms:** Asymptomatic but central vision is decreased and metamorphopsia in macular involvement (mean age is 42)
**Presentation:** POHS is found in endemic areas like Ohio and Mississippi and non endemic areas like U.K. with no serologic evidence but DNA of H.capsulaturn was found in the chronic choroidal scars, nevertheless females and males are equally effected with mean age is adolescence and POHS is presented in triad peripapillary pigment changes, histospots (multiple atrophic choroidal scars), and a maculopathy
**Clinical features:**
Vitreous & retina:
-Peripapillary pigment changes (diffuse or focal), Histo spots ( 200 micro m) which presented at the center (risk of CNVM development) or periphery which they look like punch out and discrete in linear streak pattern (5%)
-CNVM macular involvement can cause sub retinal yellow-green membrane with exudative and hemorrhagic detachment and surrounded with pigmentation, CNVM may lead to disciform scar
-**No vitritis**
-FFA early hypofluorescence with late hyperfluorescence where CNVM hyperfluorescence then leak in late stages.
Complications:
CNVM, and huge exudative and hemorrhagic detachment.

**Treatment:** All patients should be taught how to use Amsler grid
Early: systemic or periocular corticosteroid can be used and patients with no CNVM should be examined every 6 months
CNVM: laser photocoagulation for extra foveal but for foveal Anti-VGEF or PDT with or without intravitreal steroids are indicated, sub macular surgery to remove CNVM may have short term visual improvement and it is indicated for patients with vision <20/100

**Prognosis:** poor if CNVM (which may has spontaneous resolution in 12%) occurs (20/100)

## Differential Diagnosis

ARMD: patients are older and there would be a macular drusen not histo spot.

Angioid streaks: Red, brown, lines radiating from the optic disc and CNVM may occur.

High myopia: if atrophic spots are seen they usually confined to the posterior pole and more white and the myopic crescent is on the temporal side of the disc with pigment on the outer edge of a rim while in POHS is in the inner

Multiple evanescent white dot syndrome (MEWDS): White-orange lesions (100-200 micron) in the posterior pole to mid periphery between RPE and outer retina which fades within weeks with macular granulaty with some vitritis; FFA; early hyperfluorescence and wreath like configuration with late staining ; Visual Field; enlarged blind spot and para central scotoma and with spontaneous resolution

Multifocal choroiditis with panuveitis: Similar but in MCP there would vitritis with AC cells

Old toxoplasmosis: which would be accompanied with some cells in the vitreous and chorioretinal lesion in white color.

## Candida

**Symptoms:** Bilateral, Vision is decreased , floaters & pain
**Presentation:** Candida albicans is imperfect yeast fungus can be found in the skin, mouth, and vagina risk factors include IV drug abuse, immune-compromised patients and malignancies.
**Clinical features:**
Vitreous & retina:
-Multifocal yellow white retinitis ( disc diameter and larger )
-**Vitritis** which can cause cotton balls that may be in strings of pearls configuration
Complications:
Retinal necrosis with tractional detachment.

**Treatment:**
-Intravitreal 5 µg amphotericin B
-Oral fluconazole 400 mg initially then 200 mg b.i.d
-Close follow up

## Aspergillosis

**Symptoms:** Bilateral, Vision is decreased rapidly , floaters & extensive pain
**Presentation:** Aspergillosis is filament fungus can be caused as endogenous endopthalmitis risk factors include IV drug immune-compromised patients and chronic pulmonary disease.
**Clinical features:**
Vitreous & retina:
-Confluent yellow sub retinal infiltrates may cause retinal hypopyon and hemorrhages
-**Vitritis** and anterior chamber hypopyon
Complications:
Retinal necrosis with tractional detachment.

**Treatment:**
-Intravitreal 5 µg amphotericin B
-amphotericin B I.V.
-Close follow up treatment may be for more than 4 weeks

# Protozoa

## Toxoplasmosis

## Ophthalmic manifestation

**Symptoms:** unilateral floater with hazy blurred vision, redness, and photophobia while pain is presented in the case of iridocyclitis.

**Presentation:**

-Congenital: retinochoroiditis, intracranial calcification, and hydrocephalus

-Acquired: presented as 2/3 of ocular toxoplasmosis and presented in all age groups which can be progressive, recurrent (within 3 years in 50%), self limiting in patients with good immune stats.

-Caused by parasite Toxoplasma gondii where cats are host and human is infected from eating raw contaminated meat, or contact with cats faces contaminated with Toxoplasma gondii cysts (water, fruit, & goat milk) , transplacental transmission, throw skin and contaminated blood and organ transplant

-Diagnosis is conformed by serological testing by indirect fluorescent antibody or ELISA IgG (support the diagnosis & IgM (during acute phase and in new born but IgA is more helpful in new borns)

**Clinical features:**

Anterior chamber: granulomatous reaction which could be mild to moderate as spillover and might have the features of Fuch's.

Vitreous & retina:

-Congenital: bilateral (85%) chronic retinochoroiditis in the macula or posterior pole.

-Acquired: **Vitritis** over focal white retinitis (head light in the fog) next to chorioretinal pigmented scar in the posterior pole some times next to the optic disc and perivasculitis with vascular sheathing resolution occurs with in 7-8 weeks but vitritis may presets longer.

-Atypical presentation: As retinal detachment, unilateral retinitis pigmentosa, neuroretinitis and punctate outer retinal toxoplasmosis (deep retinal lesions with absent of vitritis)

IOP: Elevated in 20% of acute cases which is treated with antiglaucoma medication

Complications: Macular & optic disc involvement, tractional & exudative retinal detachment, RPE atrophy, CME, and CNVM

**Treatment:**

-Indication:

1)Disease persistence for more than a month

2)Multiple lesions are active

3)When the size of lesion is more than one disc size

4)VA is decreased

5)Vitirits which is moderate to severe

6)When fovea or optic nerve are threatened by lesion

-Iridocyclitis is treated with topical steroids and cycloplegia.
-Bactrim (trimethoprim-sulfamethoxazole 160 mg/80 mg) b.i.d + Azithromycin 250mg/day + prednisalone 1mg/kg/day after 24 hrs from starting bactrim and Azithromycin
-Or sulfadiazine with starting dose: 2-4 g then with dose: 1.0 g q.i.d + pyrimethamine (Should be prescribed with Folinic acid 10 mg) with starting dose: 50-100 mg: then with dose 25-50 mg/day, and prednisone 1mg/kg/day after 24 hrs from starting pyrimethamine and sulfadiazine
-Newborn with congenital variant: pyrimethamine and sulfadiazine for 1 year with pediatric consultation
-Pregnant: Spiramycin 400mg t.i.d or Azithromycin.
-HIV/immunecompramised: atovaquone with pyrimethamine and sulfadiazine
-Bactrim 1 tab every 3 days may reduce reactivation and this can work for AIDS patients as well
- Vitrectomy is saved for non clearing vitreous and RDs
-**N.B.!** The use of periocular , intravitreal, subconj or sub tenon injections are contraindicated.

**Prognosis:** good if macula is intact

## Systemic manifestation

-Congenital: retinochoroiditis (the congenital form occur as a reseat of 2 trimester infection), intracranial calcification, and hydrocephalus

-Acquired: presentation in immunecompetent is usually sub clinical but it can resemble rickettsial infection, lymphoadenopathy or meningoencephalitis where in immunecompramised it can be life threatening with intracerebral lesions.

## Deferential diagnosis

-Toxocariasis: In children with history of giophagia with no Chorioretinal scars.
-Acute retinal necrosis (please check the viral infection above)
-Syphilis, and tuberculosis.

## Helmetic

### Toxocariasis

### Ophthalmic manifestation

**Symptoms:** unilateral (rarely bilateral) floater with vision down, pain, leukocoria, strabismus and photophobia.
**Presentation:**
-Presented in young adults or children which are from white nonhispanic descents where males and females are equally effected
-Caused by roundworm parasites Toxocara canis (dog) or Toxocara cati (cat) where intestine of dogs or cats are host and human is infected from geophagia or oral-face rout
-Diagnosis is conformed by ELISA
**Clinical features:**
Anterior chamber: inflammation which is nongranulomatous with posterior synechiae
Vitreous & retina:
Vitreous inflammation which can be moderate to severe with chronic endophthalmitis and cystic membranes (25% where most of cases are between 2-9), peripheral granuloma (50% where most of cases are in adulthood) that is presented as strands of connective tissue, and localized macular granuloma (25% where most of cases are between 6-14) which can be peripapilary and presented as white elevation with little reaction and about 2 discs diameter
-Atypical presentation: presented as parsplanitis with exudative granulomatous inflamtion in the periphery
Complications: Tractional membrane on the macula & optic disc, tractional & rhegmategenous retinal detachment which treated with parsplana vitrectomy, CME, and CNVM
**Treatment:** systemic and periocular corticosteroids while laser photocoagulation can be helpful but antihelmetic therapy is not helpful
**Prognosis:** poor in the case of chronic endophthalmities

### Systemic manifestation

Presented as visceral larva migrans with features such as fever, pneumonitis, and hepatosplenomegaly where blood tests shows leucocytosis and eosinophilia.

## Deferential diagnosis

-Toxoplasmosis: no history of giophagia with chorioretinal scars.
-Retinoblastoma: which may be presented with classification in C.T.
-FEVR (familial exudative vitreoretinopathy): absence of intra ocular inflammation and with family history
-ROP: history of premature birth with low wight, and history of oxygenation
-Endopthalmitis: history of trauma or surgery

# Cysticercosis

## Ophthalmic manifestation

**Symptoms:** floater with vision depends on larvae location, pain, and photophobia.
**Presentation:**
-Presented in young adults (10-30) endemic zones such as Mexico, India, south east Asia and Africa where males and females are equally effected which can be combined with neural cysticercosis in 40% of cases.
-Caused by tapeworm cestode Taenia solium where human is infected by ingestion of tapeworm by eating or drinking contaminated water or food and after egg maturation the larvae travels from intestine mucosa to choroid and sub retinal space hematogenously
-Diagnosis is conformed by ELISA
**Clinical features:**
Anterior chamber and adnexa: involvement of cysticercosis may be in the anterior chamber granulomatous inflammatory reaction with motile cysticercus orbit or eyelids
Vitreous & retina:
Vitreous inflammation with larvae of 2-6 discs size can be seen in sub retinal space which is translucent, spherical and contains head (motile reacts to light) accompanied with exudative retinal detachments and RPE atrophy where BSCAN can be helpful in diagnosis
Complications: exudative retinal detachments, RPE atrophy, blindness, and phthisis
**Treatment:** Laser photocoagulation can be helpful but antihelmetic therapy like albendazole and praziquantel are not helpful as in cerebral cysticercosis because it can cause larvae death will cause more ocular inflammatory reaction, while systemic and periocular corticosteroids and parsplana vitrectomy is more helpful
**Prognosis:** poor if not treated

## Systemic manifestation

cerebral cysticercosis usually accompanied with ocular form and can cause epileptiforn seizures and lung & muscles involvement with calcified cysts seen in chest X-rays (for lung involvement).

## Deferential diagnosis

-Toxoplasmosis: no history of giophagia with chorioretinal scars.
-Retinoblastoma: which may be presented with classification in C.T.
-FEVR (familial exudative vitreoretinopathy): absence of intra ocular inflammation and with family history
-ROP: history of premature birth with low wight, and history of oxygenation
-Endopthalmitis: history of trauma or surgery

# Onchocerciasis

## Ophthalmic manifestation

**Presentation:**
-Presented in young adults (15 years) endemic zones such as middle and south americans and sub Saharan Africa  where males and females are equally effected which can be combined with neural cysticercosis in 40% of cases.
-Caused by filarial parasite Onchocerca volvulus where human is infected by black fly and called river blindness the microfilariae travels from subcutaneous after maturation to the eye by direct invasion or hematogenously
**Clinical features:**
External disease: live microfilariae is seen on the cornea while dead one can cause opacity due to punctate stromal inflammation
Anterior chamber: anterior chamber inflammatory reaction with motile microfilariae and synechiae
Vitreous & retina:
RPE disruption in early stages then RPE focal atrophy and later chorioretinal atrophy in the posterior pole
Complications: Glaucoma, cataract, and optic nerve atrophy
**Treatment:** ivermectin is drug of choice, it can slow the progression of chorioretinal lesions but it can't stop it nor is effective on adult worms and it is advisable to take an annual single 150mg dose orally and repeated for 10 years where topical steroids are used for anterior uveitis

## Systemic manifestation

Early presentation is maculopapular rash then leopard skin (hypo hyper pigmentation) then Lazard skin (think need skin) then onchocercomas (sub cutaneous nodules) then lymphoadenopathy

# Diffuse Unilateral Subacute Neuroretinitis (DUSN)

## Ophthalmic manifestation

**Symptoms:** Unilateral decreased VA.
**Presentation:**
In a healthy young adults (14 years) caused by 2 nematodes different in size, the dog hookworm (Alleylosoma eaninum) 400- 1000 micro.m, endemic zones such as southeastern US, Barazil and Caribbean islands or the raccoon roundworm (Baylisasearis proeyonis), 1500-2000 micro.m and has been found in the northern midwestern Canada and US.
**Clinical features:**
Vitreous & retina:
-Early stages: vitreous inflammation with swelling optic disc, vasculitis, multi focal white gray disc size lesions accompanied with exudative retinal detachments where motile sub retinal nematodes can be seen.
-Late stages: optic disc and RPE atrophy, arteriolal narrowing and sub retinal scaring.
**Treatment:** Laser photocoagulation can be helpful but antihelmetic therapy like thiabendazole 22 mg/kg b.id for 2-4 days or with albendazole 200mg b.i.d for 1 month are helpful too while systemic and periocular corticosteroids helpful to control inflammation are helpful as well

## Systemic manifestation

neural larvae migrans as neurological features

## Deferential diagnosis

-Early stages: Sarcoidosis and white dots syndrome (MEWDS, MCP, and APMPPE)
-Late stages: Retinitis pigmentosa, post traumatic uveitis, toxic retinopathy and vascular occlusive disease

## Bacterial

### Lyme

### Ophthalmic manifestation

**Presentation:** in 2 peaks of ages 5-14 & 50-59 especially between April and May in Northeast and West of the US while men are effected little more than women, Lyme disease is caused by spirochete Borrelia burgdorferi which is transmitted via ticks bite where animal debo would be cows and deers, where diagnosis is conformed by ELISA

**Clinical features:**

External diseases: Follicular conjunctivitis is presented in the 1st stage while peripheral keratitis in 3rd stage where episcleritis, and scleritis may occur too.

Anterior chamber: Granulamtouse inflammatory reaction is presented in the 2nd stage

Vitreous & retina:

Sever vitreal inflammation, neuroretinitis, paplitis, choroiditis which can be punched out in a multi focal or focal form, vasculitis, and exudative retinal detachment

Neuro Ophthalmic: 3rd, 4th, 5th and 6th cranial nerve palsies

**Treatment:**

-Prophylaxis

-Antibiotics such as Doxycycline 100mg b.i.d for 21 days (contraindicated in children less than 8 years and pregnant women) and Amoxicillin 500mg b.id. for 21 days

-Topical steroids for anterior uveitis and peripheral keratitis

### Systemic manifestation

Stage1: erythema chronicum migrans (in bull's eye configuration)

Stage : oligoarthritis where large joints like knee are involved, Bell's palsy (unilateral or bilateral), meningitis, and encephalitis are presented as neurological involvement

Stage3: chronic arthritis and shiny atrophy

# Syphilis

## Ophthalmic manifestation

**Symptoms:**
-Congenital: photophobia, and pain
-Acquired: Reduced VA, redness, photophobia, and pain
**Presentation:** In Afro-American more than in nonhispanic and it can be in congenital or in acquired form, Syphilis is caused by spirochete Treponema pallidum which is transmitted sexually and transplacentally specially in the tenth week where diagnosis is conformed by VDRL (good for monitoring therapy) , RPR, FTA-ABS (in secondary Syphilis become positive and may continue throw out life)
**Clinical features:**
External diseases:
-Congenital: interstitial keratitis which would be non ulcerative with opaque corneal appearance and innovation of blood vessels into the cornea and may involve the center then when they subside they become ghost vessels
-Acquired: interstitial keratitis and conjunctival, episclral and scleral involvement
Anterior chamber:
-Congenital: iritis which would be difficult to observe due to interstitial keratitis
-Acquired: nongranulomatous and granulomatous anterior uveitis which it could be unilateral or bilateral with iris papulosa (vaascularized papules), iris rosella, yellowish nodosa, atrophy and posterior synechiae
Vitreous & retina:
-Congenital: multifocal chorioretinitis, vasculitis (rare), optic neuritis and salt and pepper configuration which it can be mistaken with retinitis pigmentosa.
-Acquired: vitritis, multifocal and focal chorioretinitis which can be small and may become confluent which are located in the post equator as large pale yellow sub retinal placiod,focal retinitis which grow slowly and may become confluent but respond good to treatment, necrotizing retinitis which look like ARN or PORN, retinal vasculitis which can be in capillaries, varying sizes of arteries and veins , exudativeretinal detachment, papillitis, and neuroretinitis
Neuro-ophthalmic: Argyll Robertson pupil, and ocularmotor nerve palsies especially manifest in tertiary syphilic
IOP: Elevated …
Complications:
-Congenital: congenital cataract and glaucoma
-Acquired: optic disc atrophy

**Treatment:** it is important to rule out Neurosyphilis by lumbar puncture

-Congenital: Crystalline penicillin G 100,000- 150,000 MU/kg/d which is given IV as 50,000 MU/kg every 12 hours in the first 7 days of life then every 8 hours for 10 days
-Acquired (primary and secondary): Benzathine penicillin G2.4 MU IM in a single dose
-Acquired (latent and Tertiary): Benzathine penicillin G2.4 MU IM weekly for three doses
-Neurosyphilis: Aqueous penicillin G 18-24 MU is given IV as 3-4 MU every 4 hours for 10-14 days
-Patients which are allergic to penicillin can be treated with tetracycline or erythromycin 500mg three times a day for one month

**Prognosis:**
Regression is good if treatment started initially

## Systemic manifestation

-Congenital:
1)Early: Long bones, hepatosplenomegaly, dis-insertion of the abdomen, and anemia
2)Late: Neurosyphilis, cranial nerve VIII deafness, abnormal facies, Hutchinson teeth, Mulberry molars, bony changes and perforations of the hard palate, and cutaneous lesions
-Acquired:
1)Primary: a painless ulcer called chancre lasting for 2-4 weeks
2)Secondary: which appears within 6-8 weeks after chancre and presented as maculopapular rash on hands and palms, uveitis (10%), and lymphoadenopathy
3)Latent: after resolution of secondary syphilis and is detected only serologicaly
4)Tertiary: Neurosyphilis (which can be conformed by VDRL test of CSF), cardiomyopathy ,and formation of Gumma which can be found on skin and mucus membranes

## Tuberculosis

**Presentation:** classically as fever, weight loss, and night sweats where ocular involvement is presented in pulmonary and extra pulmonary tuberculosis and patients with AIDS are at high risk caused by nonmotile nonsporing aerobic fast bacillus Mycobacterium tuberculosis, which has high affinity toward highly oxygenated tissues such as lungs and choroid transmitted via droplets of aerosolize where diagnosis is conformed by PPD which is positive when more than 15 mm in imunnocompetent for 48-72 Hrs and non high risk patients more than 10 mm in high risk patients and more 5 mm in immunecompramised patients and chest X-Ray.

**Clinical features:**

External diseases: phlyctenulosis, intrastetial keratitis, infiltrates in the cornea with conjunctival, scleral involvement, Parinaud syndrome, and eyelid nodules

Anterior chamber: chronic granulamtouse inflammatory reaction with mutton fat KPs, iris nodules, and posterior synechiae.

Vitreous & retina:

-Choroid:

1)Disseminated choroiditis: multiple yellow (0.5-3.0 mm) in the posterior pole accompanied with disc edema, hemorrhages and vitritis

2)Solitary choroidal mass: 4-14 mm (shows early hyperfluorescense with late leak on FFA) presented in immunecompramised patins which comes with neuroretinal detachment and formation of macular star exudation

3)Serpiginous: like choroiditis in immunecompramised patines

-Retina: presented as imunno reaction to Mycobacterium tuberculosis, perivasculitis in 20-40 (Eales disease) and periphlebitis which presented as occlusion and non perfusion.

Complications: glaucoma, neovascularzation, CNVM, opticneuritis, subretinal abscess and tractional retinal detachment

**Treatment:**

-1st 2 months isoniazid+rifampin and pyrazinamide administered daily but in the case of any resistent add ethambutol or streptomycin then continue treatment for 4 month with isoniazid+rifampin

-Topical steroids for anterior uveitis and peripheral keratitis and oral corticosteroids can be used too

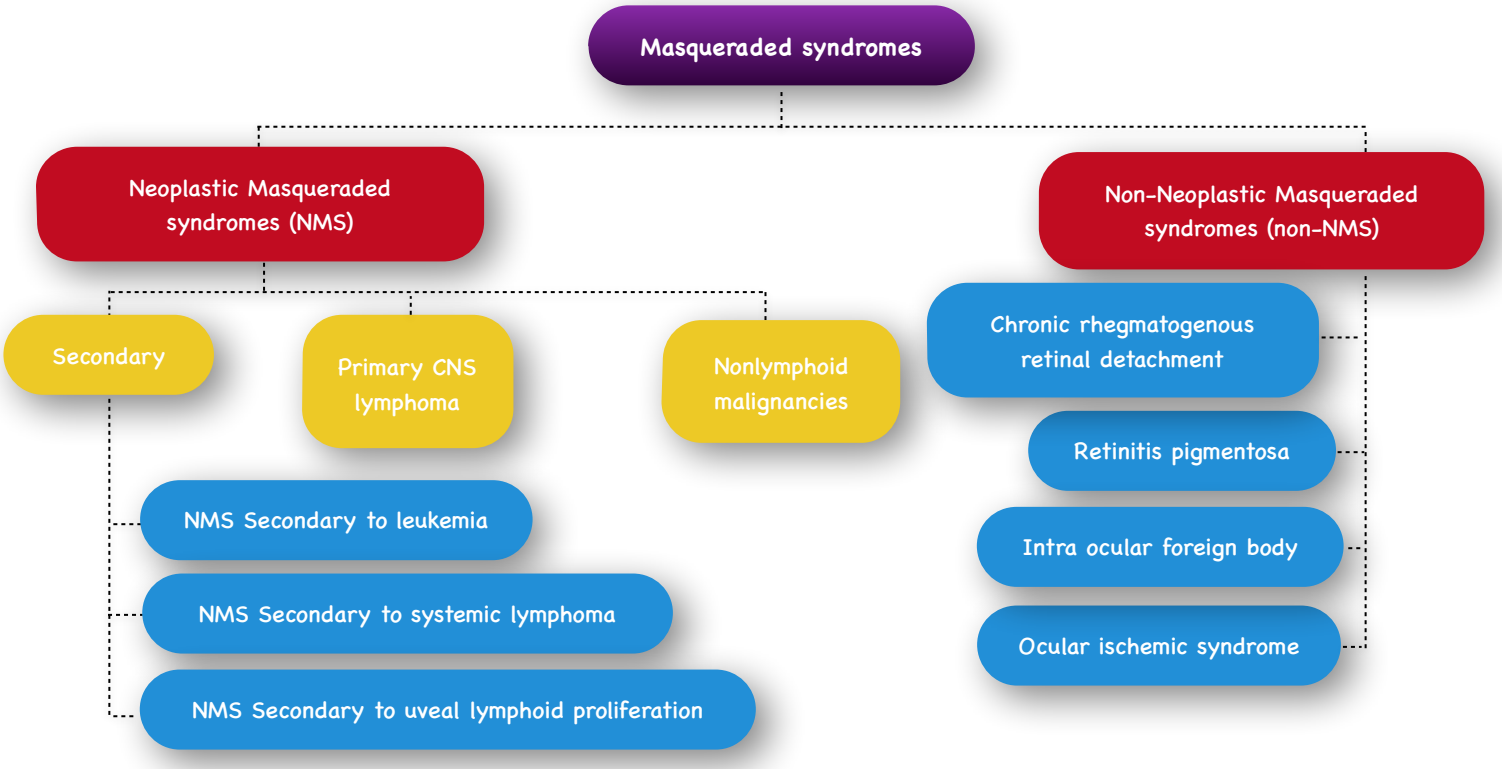
# Bartonellosis

## Ophthalmic manifestation

**Presentation:** in children > 10 years especially between fall and winter is caused by Bartonella henselae (gram negative rod) which is transmitted via scratches, licks, and bites of appearing healthy cat, which causes cat scratch disease, where diagnosis is conformed by ELISA with Western blot analysis
**Symptoms:** Visual loss
**Clinical features:**
External diseases: Granulomatous unilateral follicular granulomatous conjunctivitis is presented with regional lymphoadenopathy (Parinaud oculoglandular syndrome)
Anterior chamber: rare inflammatory reaction
Vitreous & retina:
-Unilateral or bilateral neuroretinitis presented with optic disc edema, neuroretinal detachment and then macular exudative star formation
-Focal or multifocal lesions (50-300 micro m which could be retinal or choroidal) and venous occlusion can occur too
Complications: Epiretinal membrane, inflammatory mass on optic disc, orbital abscess, and panuveities
**Treatment:**
-Antibiotics such as Doxycycline 100mg b.i.d for 2-4 weeks (contraindicated in children less than 8 years and pregnant women) with rifampin, 300 mg b.i.d or azithromycin for younger children
-IMT is indicted in the case of recurrent idiopathic neuroretinitis

## Systemic manifestation

Fluelike manifestations (fever, malaise) then cutaneous (erythematous, vesicle, papule, or pustule), lymphoadenopathy and less commonly neurological and cardiac manifestations especially in immune compromised patients


Masqueraded syndromes
Neoplastic Masqueraded syndromes (NMS)
Non-Neoplastic Masqueraded syndromes (non-NMS)
Secondary
Primary CNS lymphoma
Nonlymphoid malignancies
NMS Secondary to leukemia
NMS Secondary to systemic lymphoma
NMS Secondary to uveal lymphoid proliferation
Chronic rhegmatogenous retinal detachment
Retinitis pigmentosa
Intra ocular foreign body
Ocular ischemic syndrome

# Masqueraded syndromes

## Neoplastic Masqueraded syndromes (NMS)

### Primary

### Primary CNS lymphoma

### Ophthalmic manifestation

**Symptoms:** decreased visual acuity, photophobia and floaters
**Presentation:** it happens in non-Hodgkin lymphoma (B-lymphocytes) in 6th-7th decade where diagnosis is conformed by pars plana vitreous biopsy
**Clinical features:**
Anterior chamber: cells
Vitreous & retina:
Vitritis with sub retinal infiltrate (yellow creamy) with over lying pigment epithelial detachment with sub RPE infiltrates that may form in a ring like at the equator
Complications: exudative retinal detachment, CME, vascular occlusion and optic disc atrophy

**Treatment:**
-in a patients older than 60 years old: chemotherapy alone is preferred
-in a patients younger than 60 years old: a combination of chemotherapy and radiotherapy is preferred
-IMT: methotrexate (intravitreal for local treatment) and rituximab

**Prognosis:**
Poor the median survival is 3 months

### Systemic manifestation

Neurological involvement such as cranial nerve palsies, hemiparesis, spinal cord involvement and epileptic seizure

## Secondary

### NMS Secondary to leukemia

Presented as retinal hemorrhage, cotton wool spots, Roth spots, and neurosensory detachment if choroid is involvement
Anterior chamber involvement would be hyphema, iris heterochromia, or pseudohypopyon

### NMS Secondary to systemic lymphoma

Presented as vitritis, subretinal creamy lesions, choroiditis vasculitis and even necrotizing retinitis with anterior chamber reaction due to as spread of lymphomas hematogenously.

### NMS Secondary to uveal lymphoid proliferation

Presented as redness photophobia with salmon pink colored masses on sclera and conjunctiva, there would be creamy choroidal lesions and could be complicated in CME and glaucoma.
Treatment with radiation and periocular steroids injections.

## Nonlymphoid malignancies

### Uveal melanoma

Presented in only 5% of patients as anterior, posterior inflammation, rubeosis iridis, endophthalmitis, or panophthamitis

### Retinoblastoma

Presented in 3% of patients as pseudohypopyon with iris nodules and conjunctival chemosis

### Metastatic Tumors

Presented as bilateral multifocal choroidal lesions, retinitis( rare), vasculitis with exudative retinal detachment and anterior chamber reaction with nodules which biopsy will help in diagnosis

### Bilateral Diffuse Uveal Melanocytic Proliferation

Presented as rapid visual loss due to cataract, vitritis, multifocal choroidal and iris placoid ( pigmented or non pigmented) nodules, exudative retinal detachment, with episclritis and scleritis.

### Juvenile xanthogranuloma

Presented as yellow radish skin lesion with iris lesions and hyphema which shows foamy histiocytes and Tatoun in histology.

## Non-Neoplastic Masqueraded syndromes (non-NMS)

Such as chronic rhegmatogenous retinal detachment (some cells in the anterior chamber), retinitis pigmentosa (some vitreous cells and CME), intraocular foreign body (chronic inflammation due to toxic irritation of uveal tract specially ciliary body), and ocular ischemic syndrome (corneal edema, cells in the antroir chamber, iris atrophy, and neovascularzation of the retina and iris).

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