PEDIATRIC EYE CONDITIONS NOT TO BE MISSED**

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2 CHILDREN LATER…

INFOCUS FAMILY EYECARE


BEVEL
INFOCUS
FAMILY EYECARE

VISION IS LEARNED

- Most critical stages of vision development occur in the first year of life.
- Undetected vision problems can lead to permanent vision impairment.

VISUAL DEVELOPMENT

- Vision development is an important part of child development
- Early child development is critical
  - Most survival skills are based on visual clues:
  - 1st year - 50% of what we need to know for life is learned
  - 2nd year- another 25% is learned
  - 3rd year and beyond- only 25% of life's survival skills are added

EYE CARE

- Pediatric clinical care guidelines
  - Vision screening part of well exam
  - Red pupil reflex and eye alignment
- Studies show pediatric population do not consistently receive vision screening during clinical visit

AOA STUDY:**

- Children 3-5 years of age seen by pediatricians
  - 66% received a vision screening
- At age 3
  - 40% were screened
- Only 14% of all children have had an eye exam prior to entering school.

SCREENINGS VS EXAMS

- Comprehensive eye exams and vision screening programs should NOT be confused.
  - Each has different purpose, generates different results
  - Examinations are necessary even if screenings are done periodically
    - Good acuity does not guarantee healthy eyes

EXAM FREQUENCY

An initial comprehensive eye and vision examination should take place:

- Between the ages of 6-12 months
  - Again at age 3
- Before a child starts school
- Annually thereafter

InFant SEE

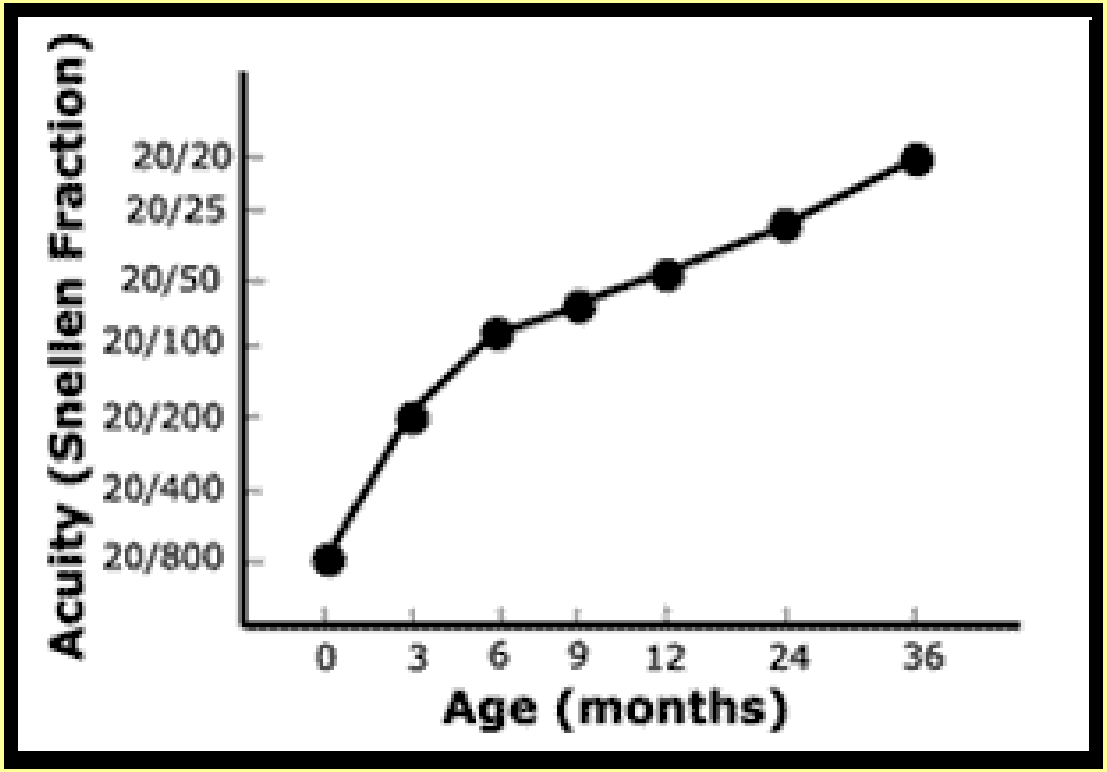
One-time no-cost public health program provides eye care for infants

- **For infants 6-12 months of age**
- **Complements pediatrician well-baby vision screenings**
- **No cost regardless of income**

NORMAL VISUAL DEVELOPMENT

- Birth to 6 weeks
  - Stares at Surroundings
  - Hold gaze on bright objects momentarily
  - Eyes and Head move together
  - Blinks at camera flash
  - One eye may turn in at times

ACUITY PROGRESSION



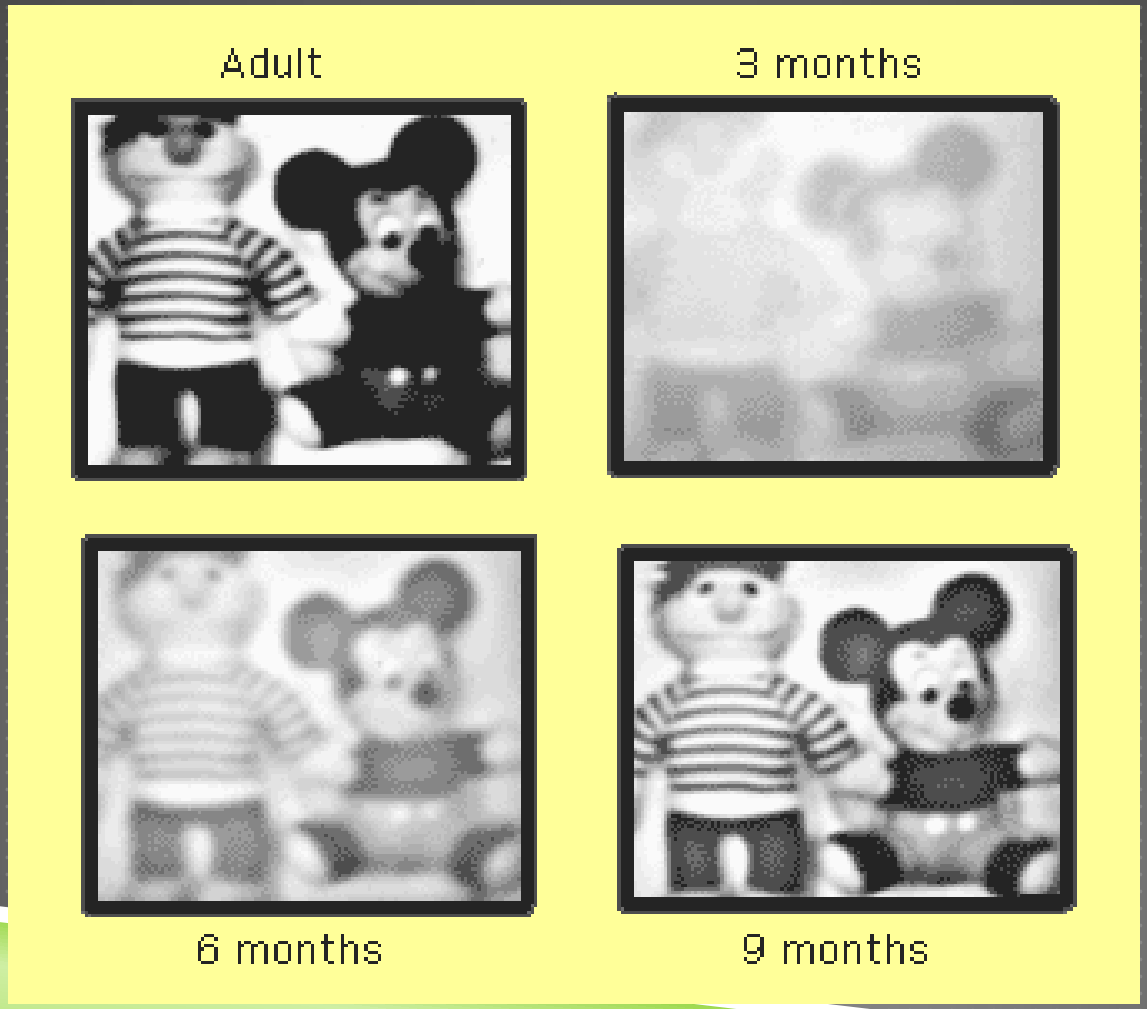
NORMAL VISUAL DEVELOPMENT

- 2-4 Months (20/200)
  - Eyes move with less head movement
  - Follow moving objects or people
  - Watches mom & dad's face when talking
- 4-6 months (20/100)
  - Eyes move actively
  - Inspection of surroundings
  - Watches distance objects

NORMAL VISUAL DEVELOPMENT

- 6-9 months (20/80)
  - Eyes turn inward while inspecting near objects
  - Watches activities for longer periods of time
  - Good alignment
- 9-12 Months (20/50)
  - Moves toward toy
  - Looks around room
  - Visually responds to smiles and voices

VISUAL EXAMPLES



NORMAL VISUAL DEVELOPMENT

- 12-18 Months (20/40)
  - Interested in simple pictures
  - Often holds objects very close to inspect
  - Points to objects or people
  - Identifies pictures in books
- 18-24 Months (20/25)
  - Visually inspects without needing to touch
  - Smiles when views favorite object/people
  - Likes to watch moving objects

NORMAL VISUAL DEVELOPMENT

- 24-36 months (20/20)
  - Watches own hand while scribbling
  - Watches and imitates other children
  - Can now keep coloring on the paper
  - "Reads" pictures in books

PREVALENCE OF EYE PROBLEMS OR DISEASE

- 1 in 10
  - At risk from Undiagnosed Vision Problem
- 1 in 30
  - Will be affected by Amblyopia
    - Leading cause of vision loss in people under 45
- 1 in 25
  - Will develop Strabismus
    - Risk factor for amblyopia

PREVALENCE OF EYE PROBLEMS OR DISEASE

- 1 in 33
  - Will have significant refractive error
    - Near sighted, Far sighted, Astigmatism
- 1 in 100
  - Will have evidence of eye disease
    - Glaucoma, Cataract, Retinopathy, etc
- 1 in 20,000
  - Will have Retinoblastoma
    - Most common intraocular malignancy in childhood

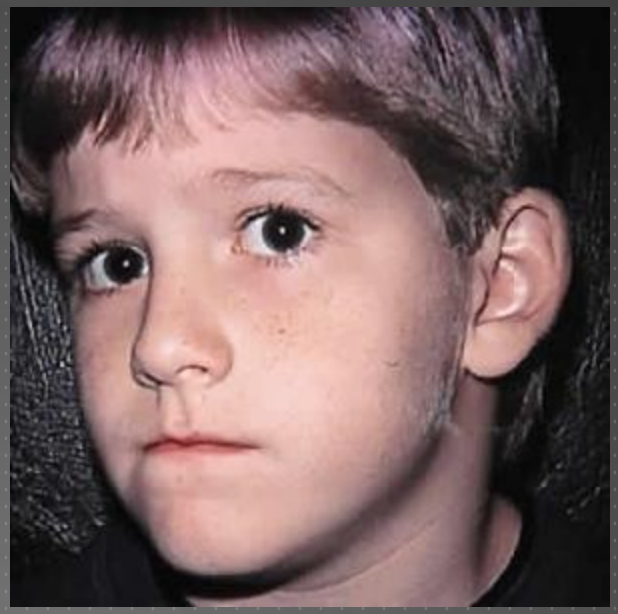
STRABISMUS

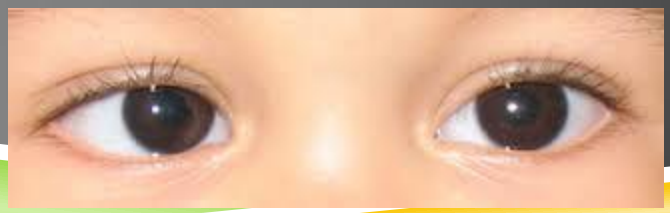
- Misalignment of eyes
- Normal in newborns
- Development:
  - Infants begin life with exodeviation
  - Disappears between 2-4 months of age

EXAMINATION

- **Parental assessment**
- Head Tilt
  - Cranial nerve IV Pasly
- Eye drift when tired/day dreaming
- Diplopia

- **Pediatrician/Medical Exam**
- Corneal light reflex
- Cover Test
- Ocular motility





CONGENTIAL ESOTROPIA

- Most common strabismus in children
- Occurs when esotropia persists>4 mos of age
- Incidence is 1/100
  - Higher in patients with neurological disorders
- 25-40% will develop amblyopia
- Most will alternate fixation

Fig. 1 Large-angle infantile-onset esotropia.

TREATMENT

- **Treatment**:
  - Equalize vision OD/OS with occlusion therapy
  - Cycloplegic drops are an alternative
  - Consider refractive tx if rx >+2.00D
  - Surgery indicated prior to 24 mos
    - 93% Fusion
    - >24 mos: 31% Fusion
- **Outcome:**
  - Excellent acuity in each eye
  - Normal esthetic appearance
  - Peripheral fusion
    - Binocular vison unattainable
  - Success rates >90% if tx <24 mos old

ACCOMMODATIVE ESOTROPIA

- High hyperopia >+4.50D
- Anisometropia if less than +3.00D

- Signs/Symptoms
  - Onset 6 mos to 7 years of age (30 mos average)
  - Worsens at end of day, fatigue, ill, "day dreaming"
  - Greater with near fixation

ACCOM ESO TX

- Full cycloplegic rx with bifocal
- Re-evaluate at 1 mos
- Repeat cycloplegic refraction q 6 mos

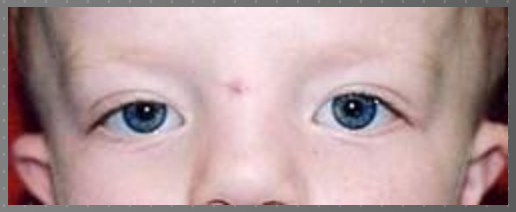
- Prognosis
  - At age 8-9 patient bifocal no longer indicated
  - Early teens may no longer need rx

Figure 3: Well fitting bifocal.

PSEUDOSTRABISMUS

- Epicanthal folds can mimic esotropia
  - Extend over the nasal sclera in children

- Hypertelorism can mimic exotropia



EXOTROPIA

- Outward turning of the eyes
- Less common than esotropia
  - More prevalent in Middle East, Asia and Africa
- Family history in 28% of patients
- Often Intermittment but progressive in up to 75%
  - Exophoria
  - Intermittent Exotropia
  - Constant Exotropia

EXOTROPIA EXAMINATION

- Onset 18 mos – 4 years
- Visible with distance viewing
  - Cover Test
- Rubbing eyes
- Aversion to bright lights
- Can be difficult to detect

- Treatment
  - Vision Therapy very effective
  - Prism
  - Overcorrection minus lenses

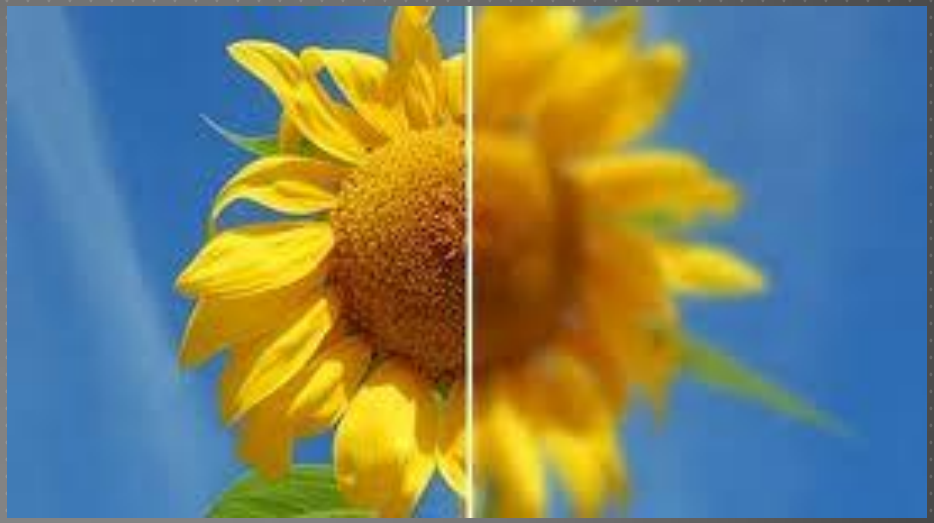
SURGICAL INTERVENTION

- Surgery indicated only when non surgical approaches fail
- Success rates
  - 60-90% for functional success
  - 70-95% for cosmetic success


EPIC FACTZ #8
HAVING EYE CONTACT FOR MORE THAN 6 SECONDS WITHOUT LOOKING AWAY OR BLINKING REVEALS A DESIRE FOR EITHER SEX OR MURDER.
more awesome pictures at: THEMETAPICTURE.COM

AMBLYOPIA

- Developmental defect that occurs in the visual pathway of the brain
- Loss of acuity in an eye
- Loss of contrast sensitivity and detail perception
- Most common cause of visual impairment in children and young adults
  - Strabismic Amblyopia
    - Monocular fixation
  - Deprivation Amblyopia
    - Cataract, corneal opacity
  - Refractive Amblyopia
    - High refractive error
    - Unequal refractive error

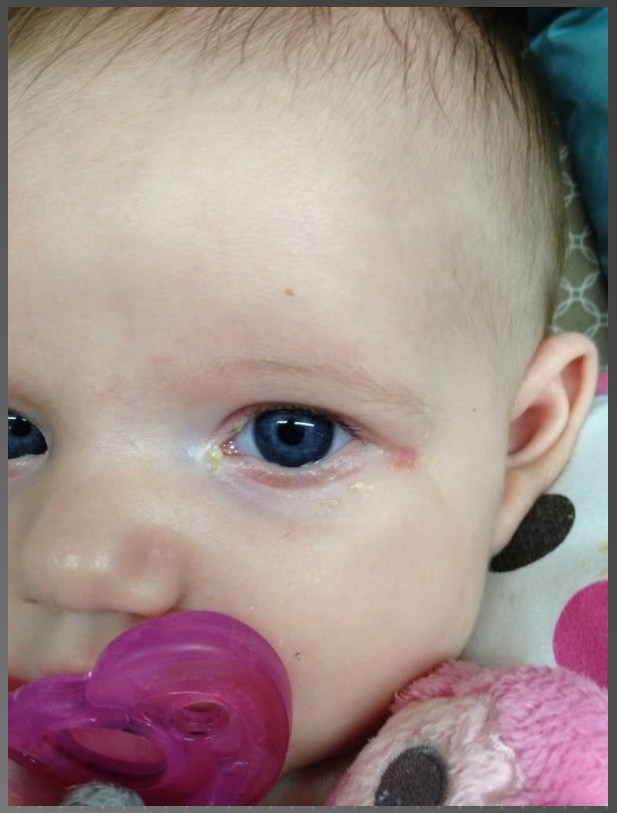


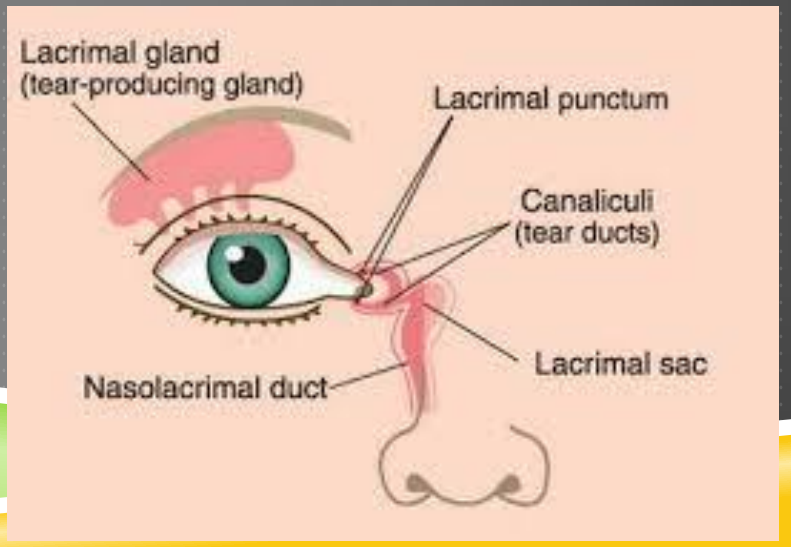
AMBLYOPIA

- Incidence 3/100
- Preventable if detected early
- Treatment:
  - Patching
  - Atropine
- Age is key: Better results if tx < 7 years old
  - Age 7-13: acuity improved but to a lesser degree
- Originally 6+ hours/day
  - New studies show adequate at 2 hours/day

BLOCKED TEAR DUCT

- Common in infants and children: up to 1/3 of newborns
- Signs:
  - Excessive watering of the eyes
  - Tears running down the cheek
  - Unilateral or bilateral (30%)
- Pathophysiology
  - Tears are produced and are drained out of the eye
    - Pass through tear duct
  - Narrowing of the duct
  - Blockage
  - Incomplete development



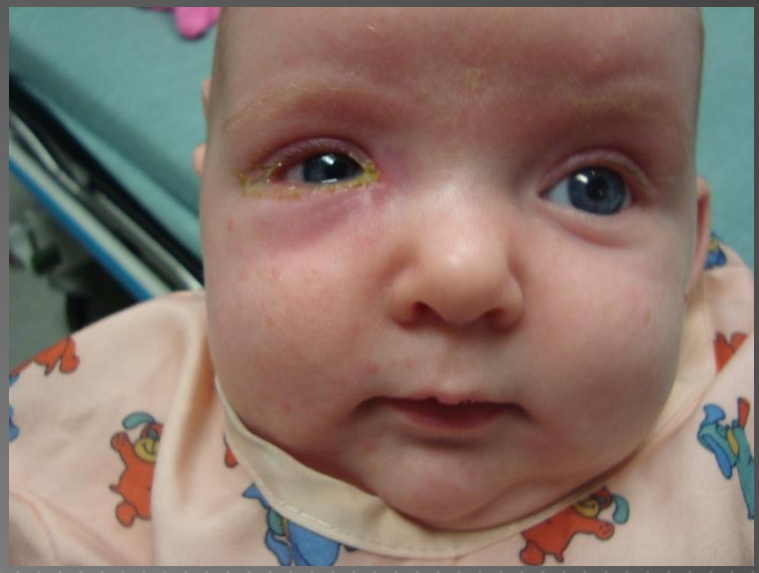


BLOCKED TEAR DUCT

- Treatment:
  - Warm compresses and lid massage 2-3x/day
  - Reassurance
- Vast majority open at 1 year
- If not, additional procedures necessary
  - Probing procedure
  - Tube/Stent placed in duct

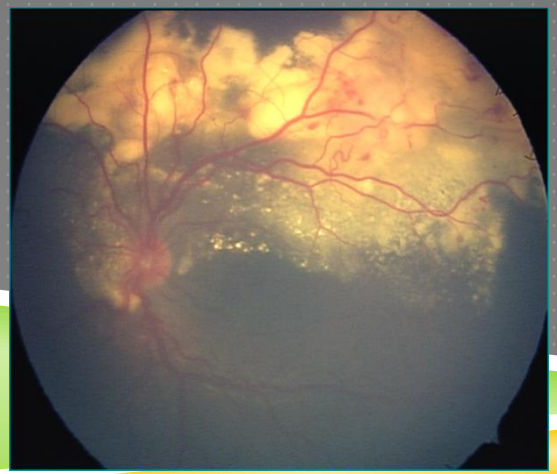
BLOCKED TEAR DUCT COMPLICATIONS

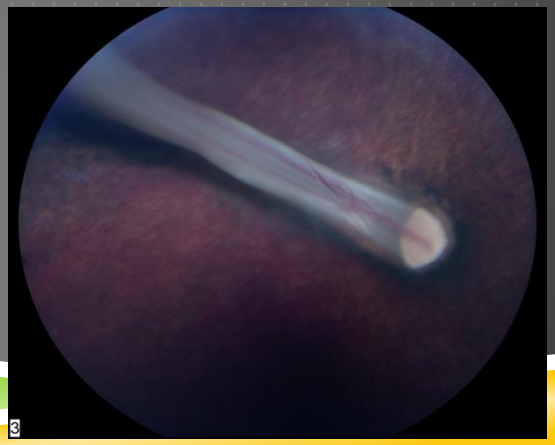
- More prone to infection
- Signs
  - Inflammation
  - Redness
  - Mucus/pus
  - Nodule



LEUKOCORIA

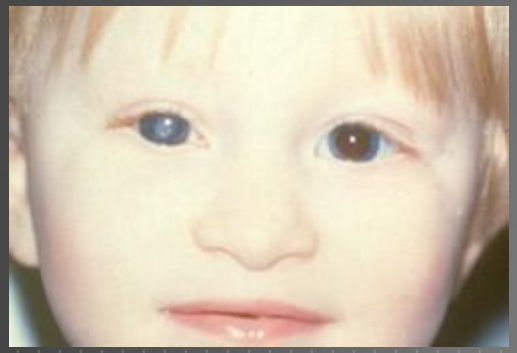
- Differential Dx:
  - Retinoblastoma
  - Toxocariasis
  - Coats Disease
  - PHPV (persistent hyperplastic primary vitreous)
  - Congenital Cataract
  - ROP (retinopathy of prematurity)

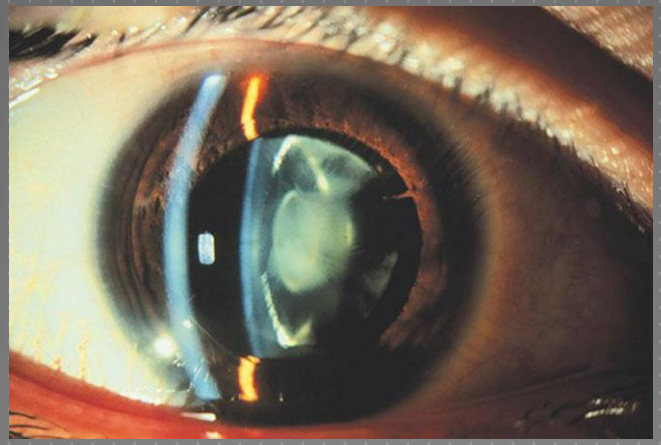


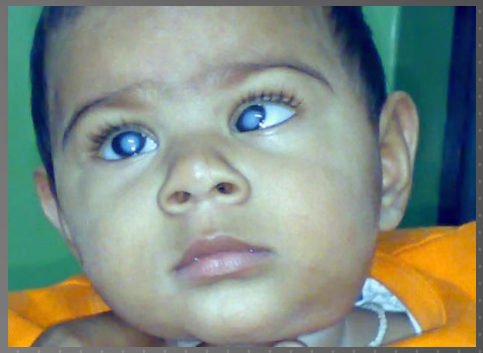


CONGENITAL CATARACT

- Opacification of Crystalline Lens
  - Unilateral or Bilateral
- Usually Diagnosed at Birth







CONGENITAL CATARACT

- Visual Morbidity
  - Due to Deprivation Amblyopia
  - Refractive Amblyopia
  - Glaucoma
    - 10% post surgical removal
  - Retinal Detachment
- Incidence
  - 1.2-6 cases per 10,000
  - International = Unknown: much higher

CONGENITAL CATARACT

- Unilateral usually Isolated Incidence
  - Associated with Ocular Abnormalities
    - Persistant hyperplastic primary vitreous
    - Trauma
    - Interuterine Infection (Rubella)
- Bilateral Associated with Systemic Disease (60%)
  - Require full Metabolic, Infectious, Systemic Workup
  - Common Causes:
    - Hypoglycemia
    - Trisomy (Downs, Edward, Patau Syndromes)
    - Infectious (Toxoplasmosis, CMV, HSV, Rubella)
    - Prematurity

CONGENITAL CATARACT FINDINGS

- Found at Birth typically
  - Not all
    - Static vs Progressive
    - Progressive might not be found until later in life (Better Prognosis)
- Not all Visually Significant
  - Central more likely significant
  - Larger than 3mm

CONGENITAL CATARACT FINDINGS

- Hallmark Sign
  - Irregular Red Reflex
- Leukocoria (white pupil)
  - Haider et al. 2008 study.
  - 60% presenting with Leukokoria had cataract
    - 18% Unilateral, 42% Bilateral
    - Other causes
      - Retinoblastoma 18%
      - Retinal Detachment 4.2%
      - PHPV 4.2%
      - Coat's 4.2%

TREATMENTS FOR CATARACT

- Tx is Directed at the Prevention of Amblyopia
- Surgical removal
  - Aphakia
    - Need for CL's
  - IOL
    - Increases complication rates

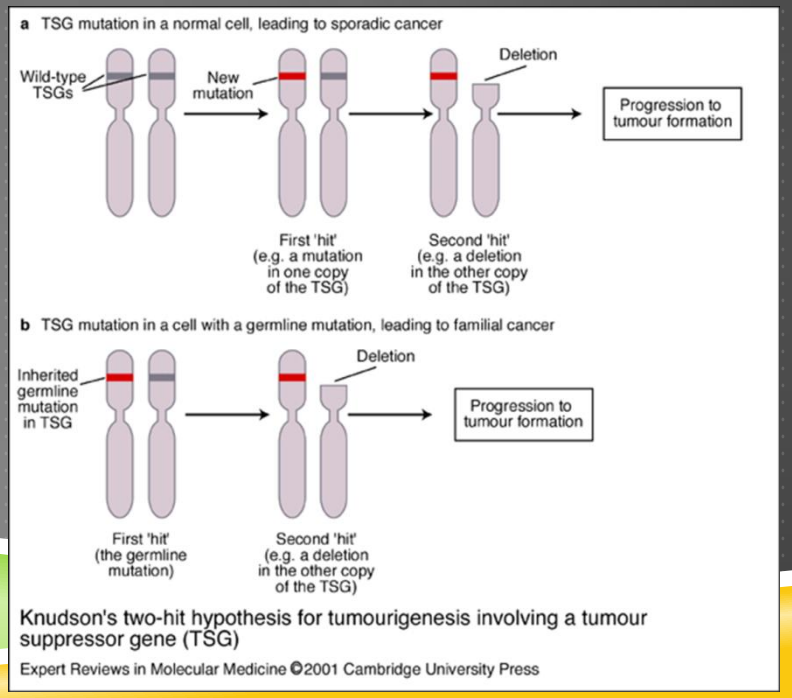
RETINOBLASTOMA

- Most common intraocular malignancy in childhood
- Incidence: 1 in 14-20,000 live births
- 250-500 new cases diagnosed each year in U.S.
- No sex or race predilection
- Diagnosed before age 3 in 90% of patients

RETINOBLASTOMA

- **RB gene** a.k.a. **Tumor Supressor Gene** is found on the long arm of chromosome 13
- Normal individuals have 2 copies of the **RB gene**
- A random mutation/deletion of the ***RB* gene** in any retinal cell completely removes the inhibition provided by the **RB protein**, and the affected cell grows into a tumor.

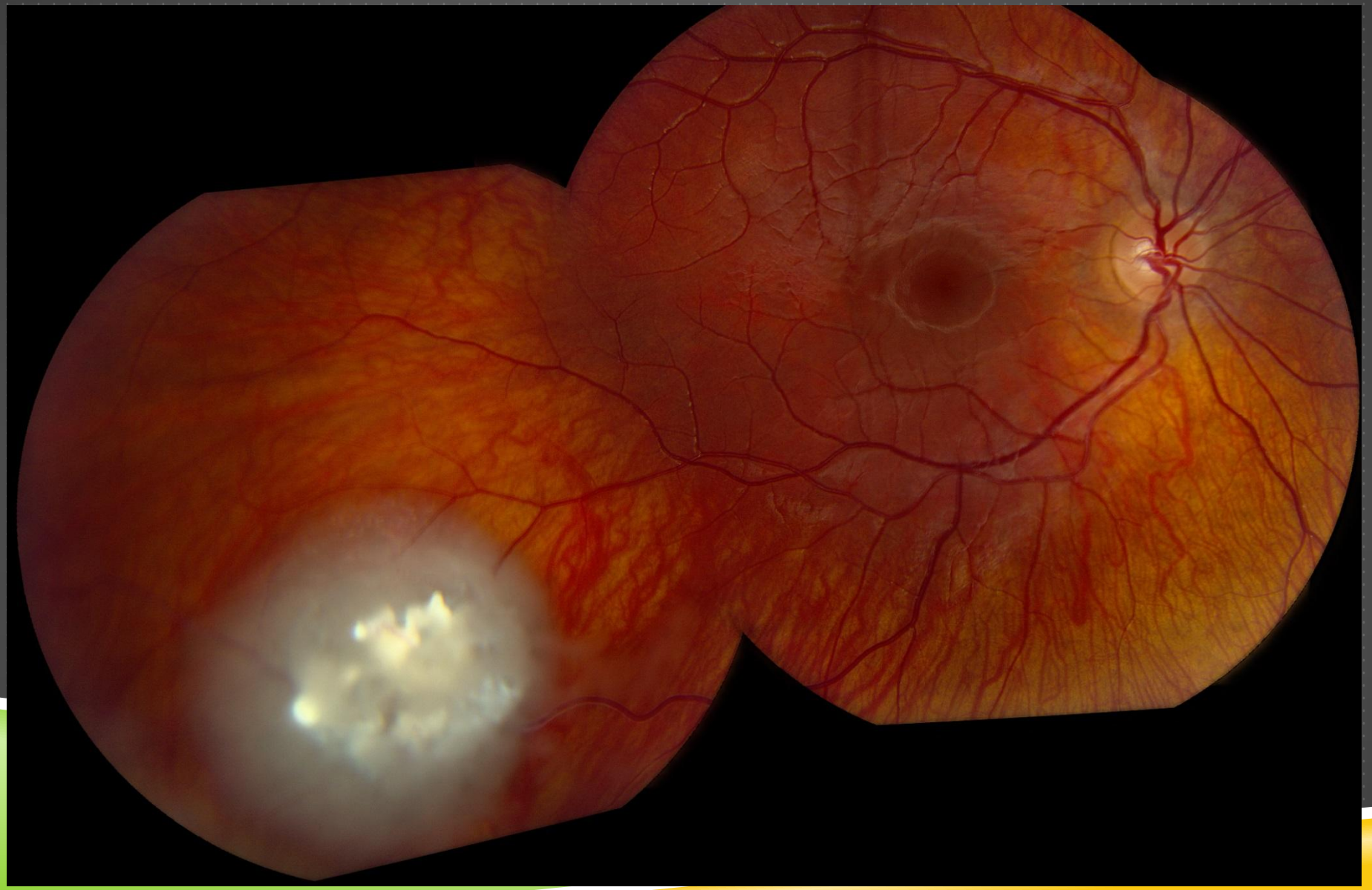
-5-10% Family Hx
-85-90% New Germline Mutation
-85% Bilateral
-15% Unilateral

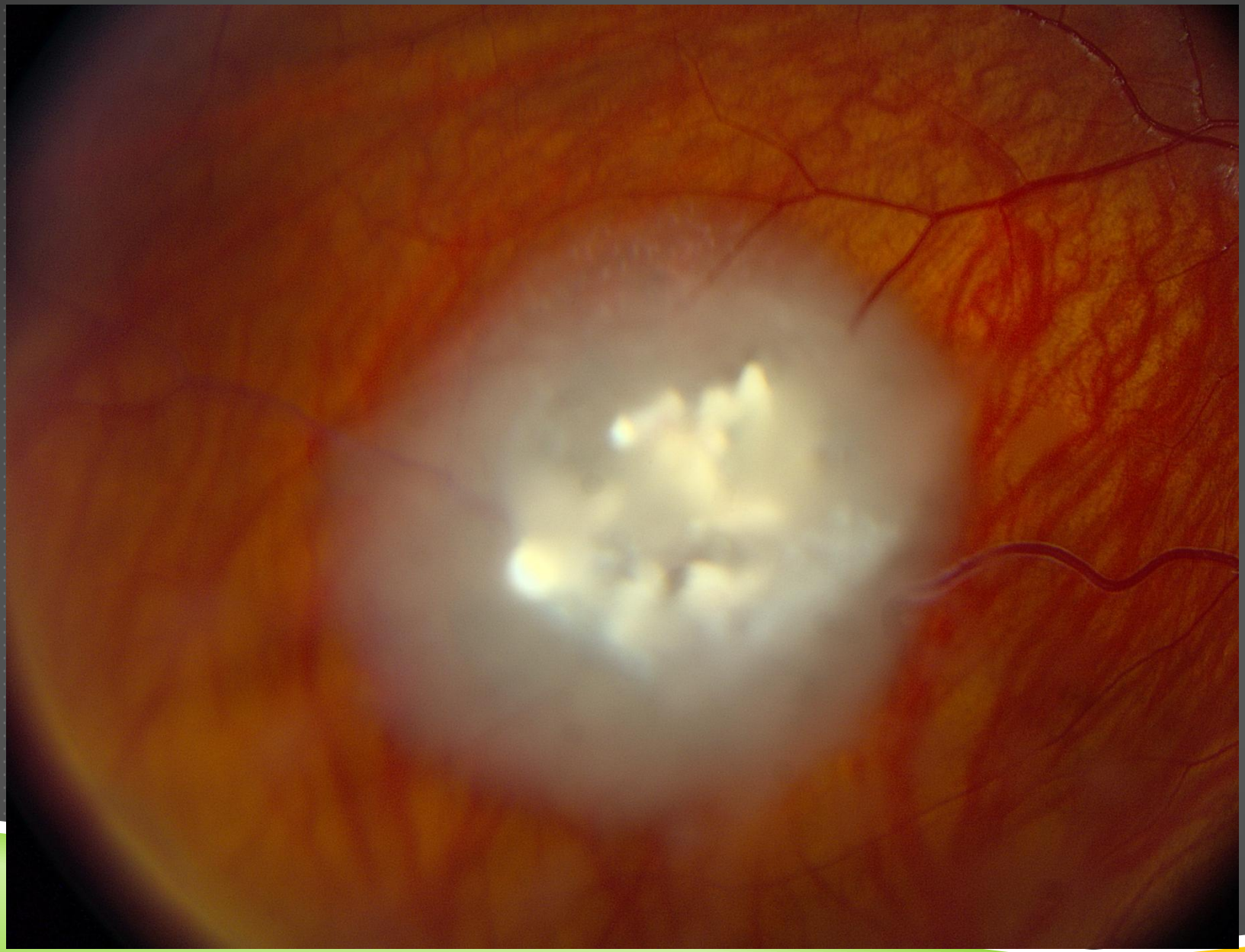


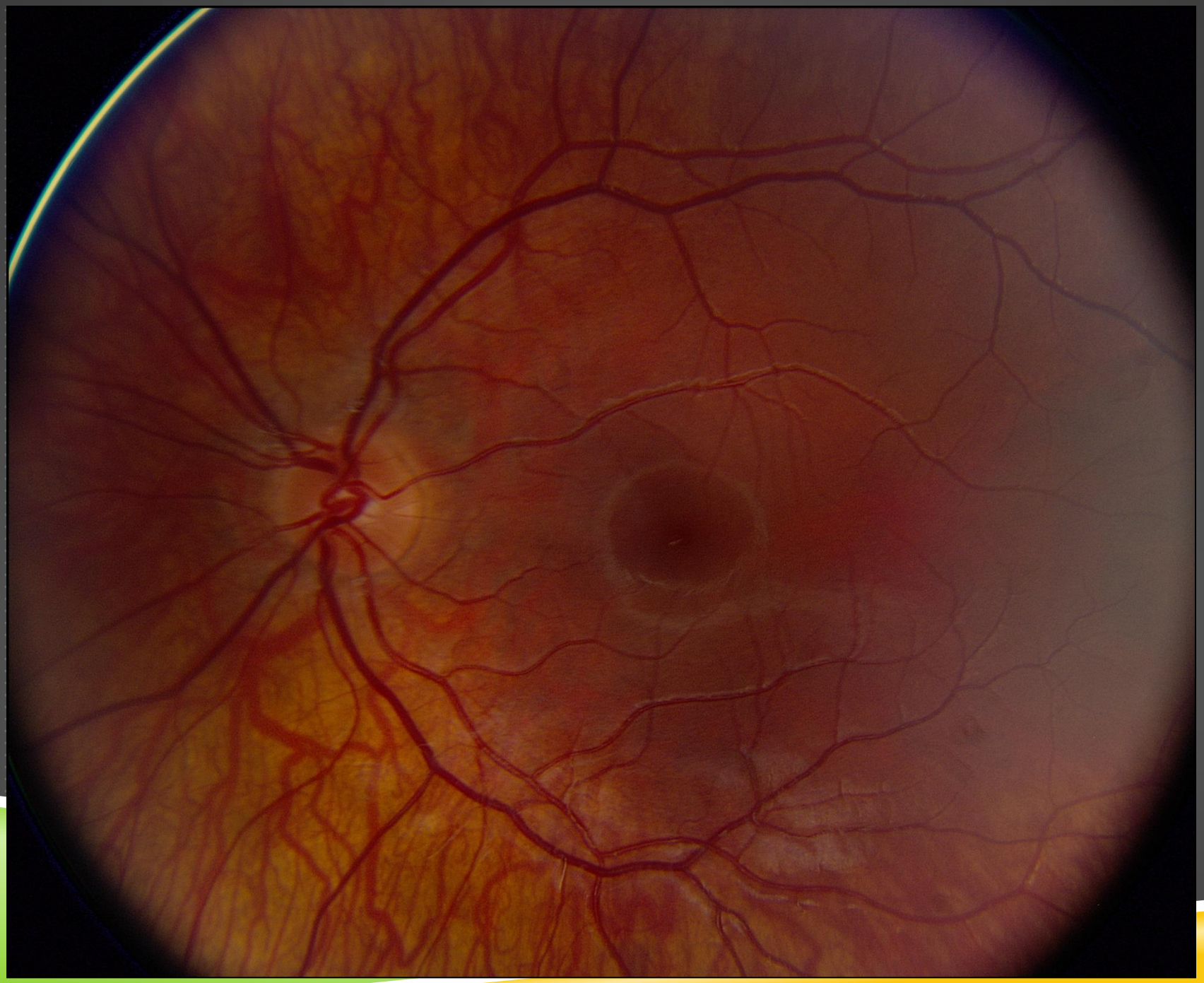
Knudson's two-hit hypothesis for tumourigenesis involving a tumour suppressor gene (TSG)

PATHOGENESIS & GENETICS

- **Somatic Mutations:**
  - 60 %
  - Mutation confined to retinal cell
  - Unilateral or Solitary lesion
  - No predisposition to second malignant tumors
  - Later onset- 2 years of age
- **Germline Mutations:**
  - 40%
  - Mutation in sperm or ova
  - Every cell body contains a mutation
  - Increased likelihood of Bilateral and Multiple lesions
  - Higher risk for malignant tumors



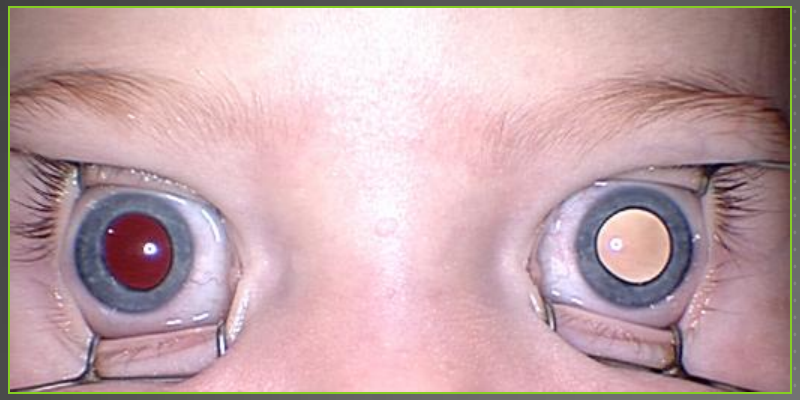




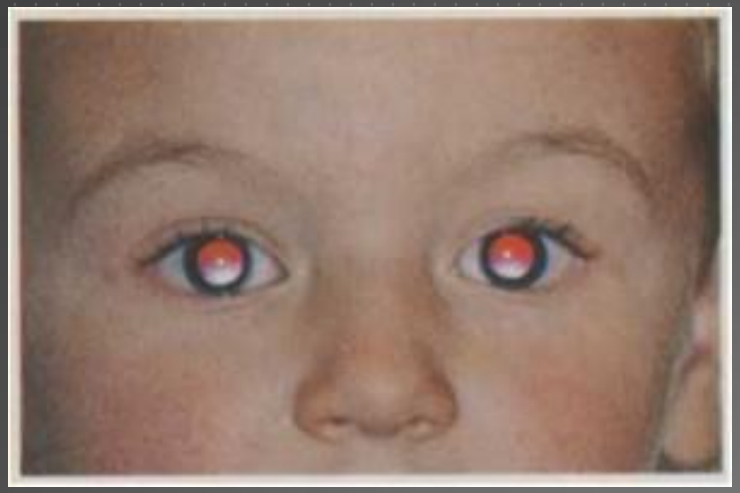
PRESENTING SIGNS

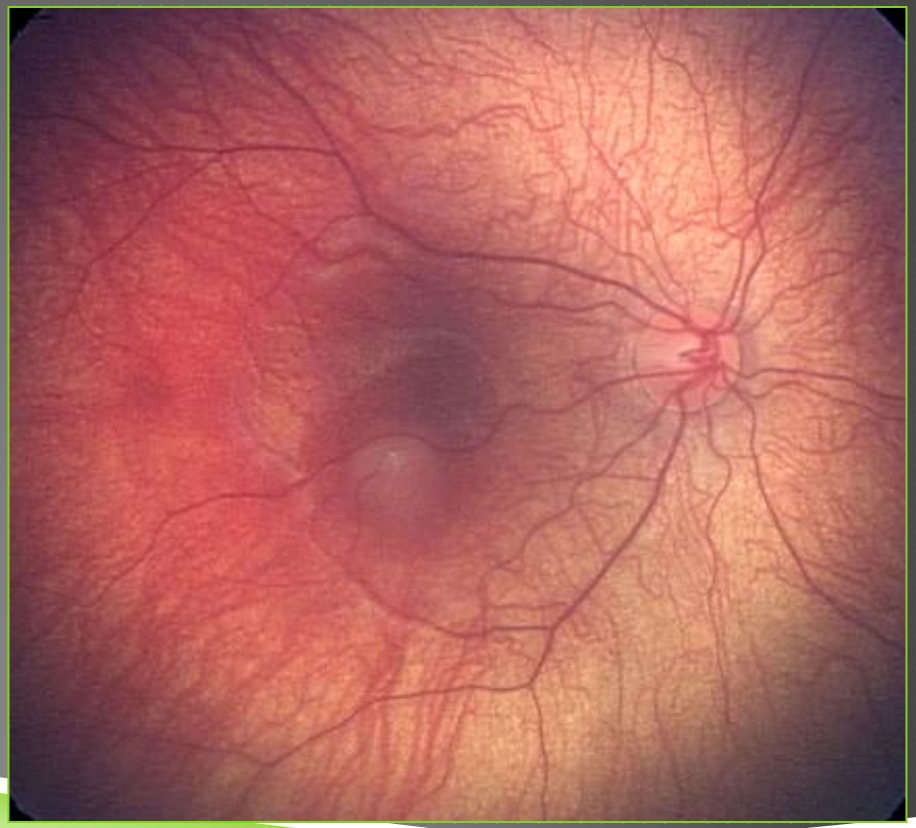
- Leukocoria (white pupil) – 50-60%
- Sensory Strabismus – 20%
- Red, painful eye – 7%

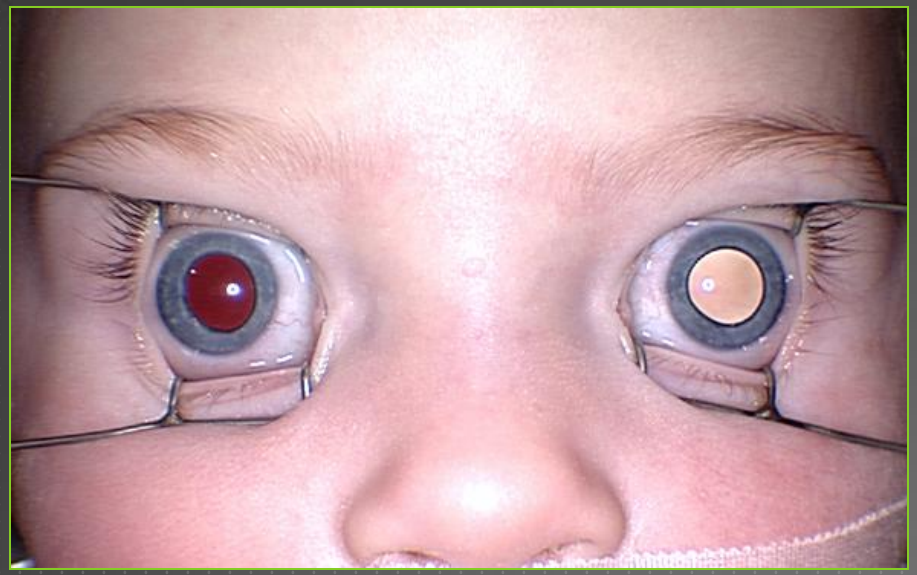
LEUKOCORIA



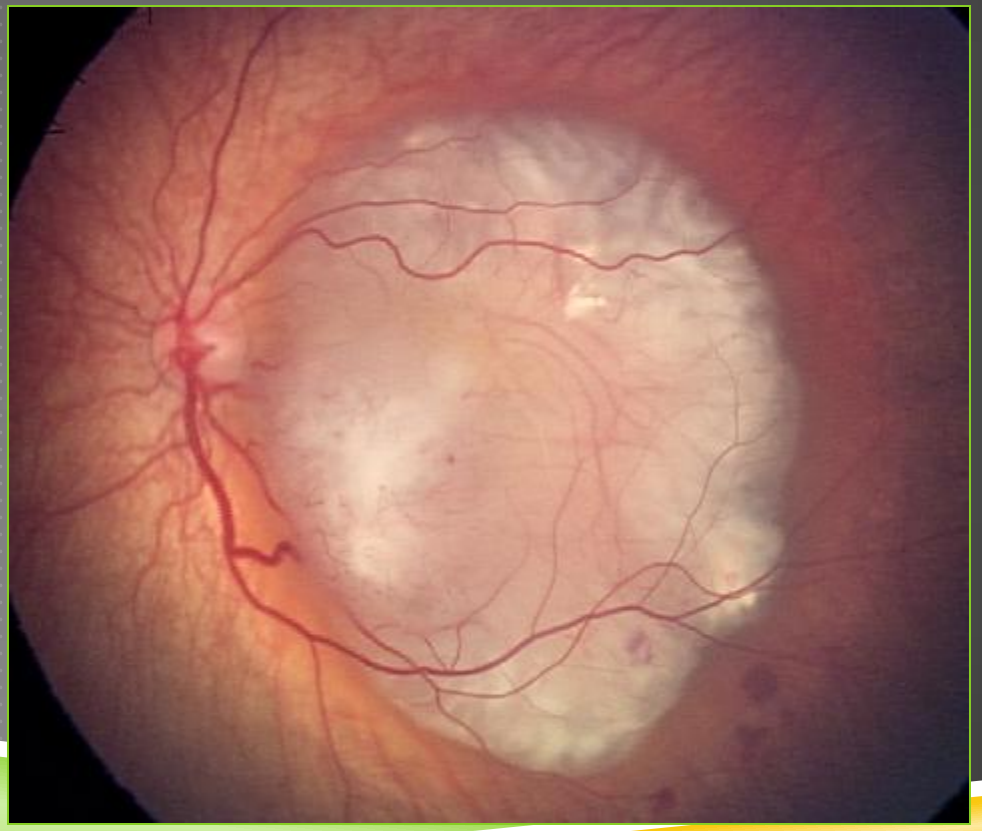
NO LEUKOCORIA WITH AN EARLY RB







LEUKOCORIA WITH LARGE RB



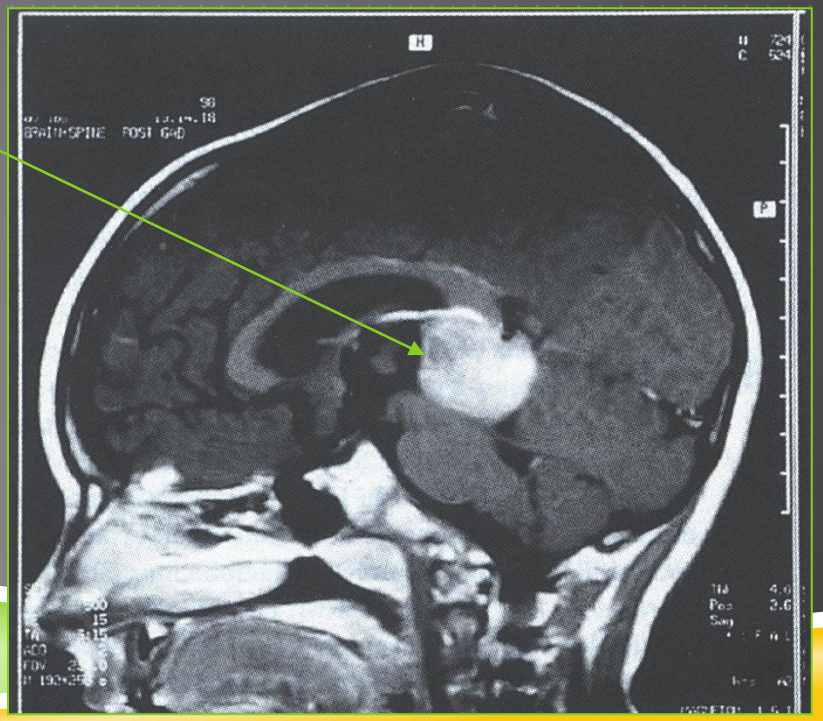
LOCAL INVASION

- Optic nerve invasion most common
- Orbital involvement → proptosis
- Enter CNS via the optic nerve and subarachnoid space
- Choroidal invasion
- Rarely extends directly through the sclera

METASTATIC DISEASE

- Primarily into the brain/spinal cord, then bones (skull), regional lymph nodes, & rarely the abdominal viscera
- Increased risk for metastases with optic nerve invasion, orbital invasion, choroidal invasion, large/advanced tumors
- Poor prognosis if metastasis occurs

- Trilateral RB:
  - Associated with bilateral RB + tumors that grow outside the eye
  - Ocular tumors occur before CNS tumors
  - Ex: Pinealomas
    - retinal-like tissue
    - rare
    - fatal (median survival 8 mo.)



RETINOBLASTOMA EVALUATION

- Good History (especially family Hx)
- Complete Ocular Exam w/ focus on:
  - VA's
  - Externals
  - SLE/Anterior Segment
  - BIO
- Exam Under Anesthesia (EUA)
- Photos
- Ultrasound
- CT scan
- MRI
- Metastatic Work-up (if suspicious lesion/location)
- Complete physical exam by Pediatrician
- Examination of siblings/parents

PROGNOSIS

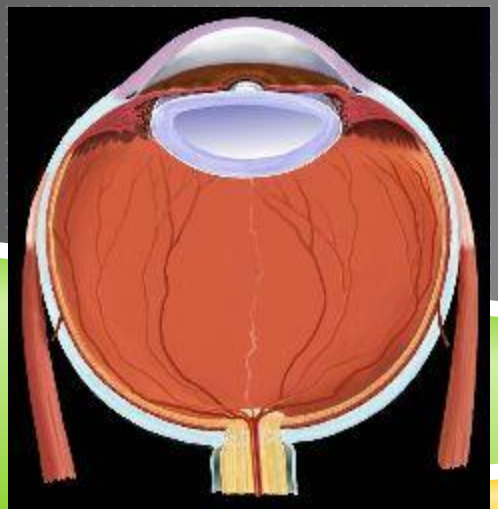
- Much better today thanks to modern technology
  - **Enucleation**
  - **Plaque radiotherapy**
  - **External beam radiotherapy**
  - **Chemotherapy**
- 85-90% survive in developed countries
- Secondary Tumor incidence:
  - 8-15% @ 20 years
  - 14-35% @ 30 years
  - 51% @ 50 years

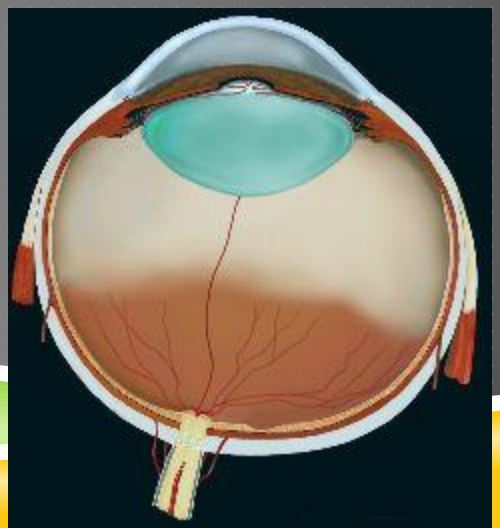
RETINOPATHY OF PREMATURITY

- Disease of retinal vasculature
  - Affects immature vasculature in eyes of premature babies
  - Can be mild with no visual defect
  - Can lead to blindness
    - Neovascularization
    - Retinal detachment
- Incidence is Rising
  - Younger and smaller babies surviving

VASCULAR DEVELOPMENT****

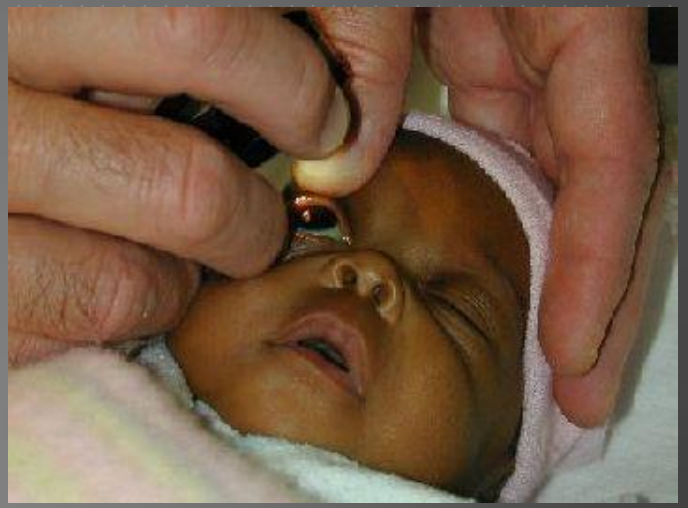
- Retinal vasculature begins in the 16th week of gestation
- Retinal vessels grow out of the optic disc as a wave of mesenchymal spindle cells
  - Mesenchymal cells develop into capillaries and eventually the mature retinal vessels
- Choroidal Vessels vascularized by 6 weeks gestation
- Nasal Retina by 32 weeks
- Temporal Retina by 40-42 weeks



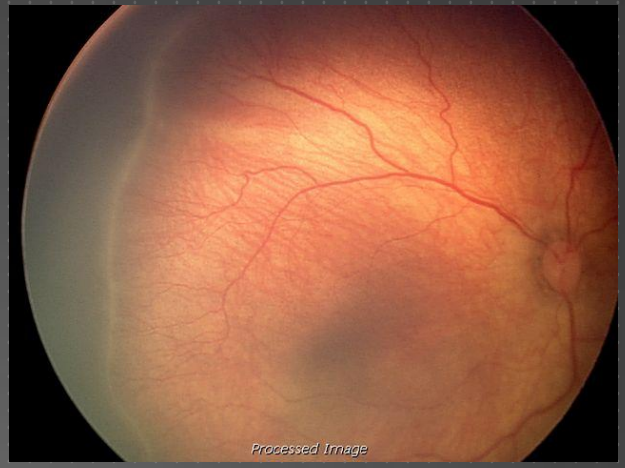


RISK FACTORS FOR ROP***

- Birth before 32 weeks gestation
  - Especially before 30 weeks
- Birth Weight less than 1500g
  - Especially less than 1250g (>50% will have evidence of ROP)
- Supplemental Oxygen therapy



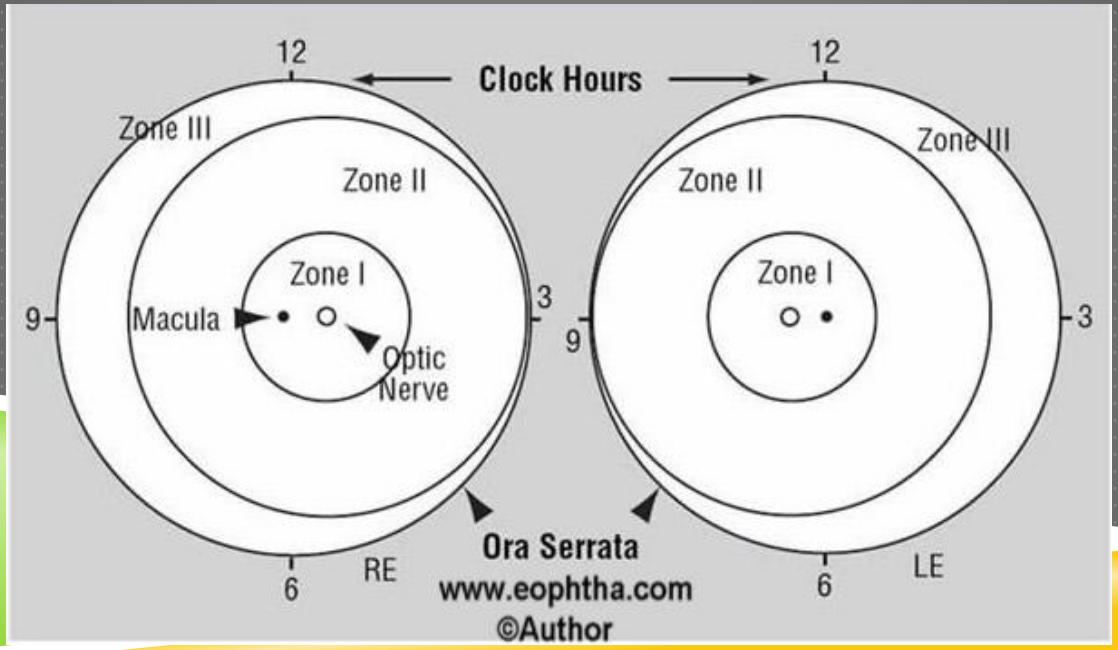
STAGES OF ROP***



- Stage I
  - Mild abnormal blood vessel growth
  - Thin demarcation line between vascular and avascular zones
- Stage II
  - Moderate abnormal blood vessel growth
  - Thickened broad demarcation
- Stage III
  - Severe abnormal blood vessel growth (Neovascularization)
- Stage IV
  - Partially detached retina
- Stage V
  - Complete retinal detachment

ZONES OF ROP***

- 3 Distinct Zones
- Zone I: 2x distance from ON to macula
- Zone II: Goes to the edge of Nasal Ora Serrata
- Zone III: Remaining sliver of Temporal Retina
  - Last to Vascularize



TREATMENT OPTIONS***

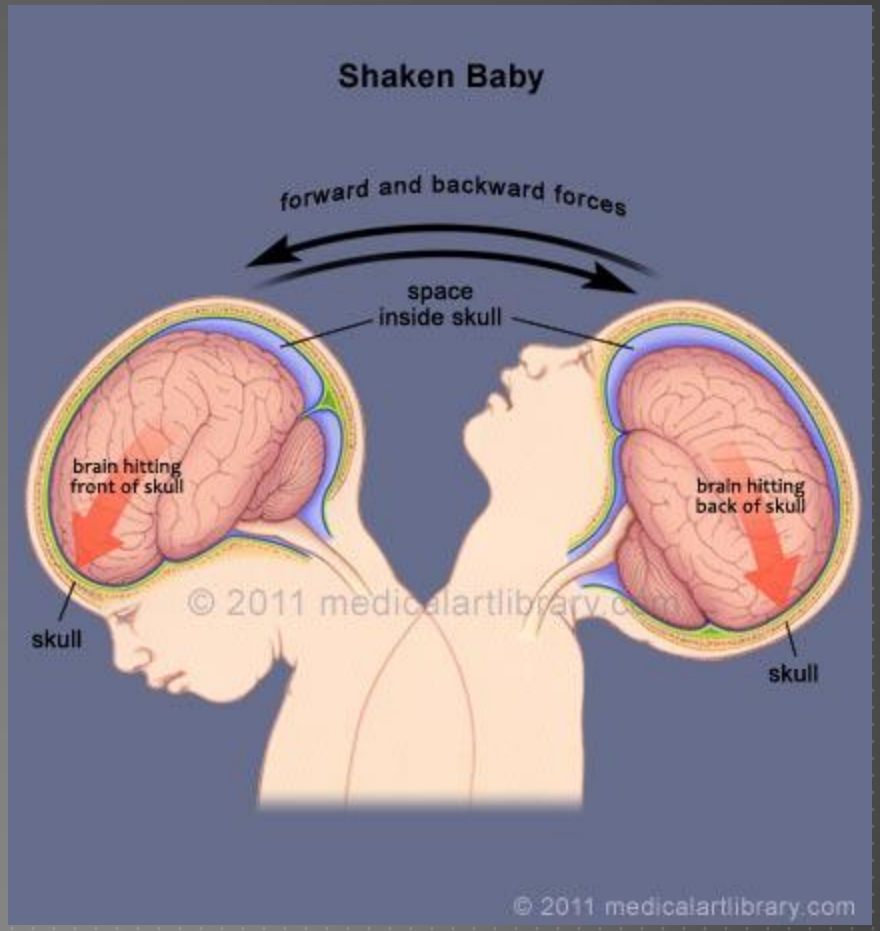
- Cryotherapy
  - Original method of Tx Since 1970s
- Laser Sx
  - Argon, Xenon, Diode
  - Lower complication rate than Cryo
- Scleral Buckling/Vitrectomy
  - Stage IV and V

FOLLOW UP CARE***

- Need continued monitoring until active disease subsides
- After Vascularization complete
  - F/U Care q 6 months until age 3
    - 20% Will develop Strabismus and refractive error
  - Annual exams ongoing
    - Up to 10% develop Glaucoma

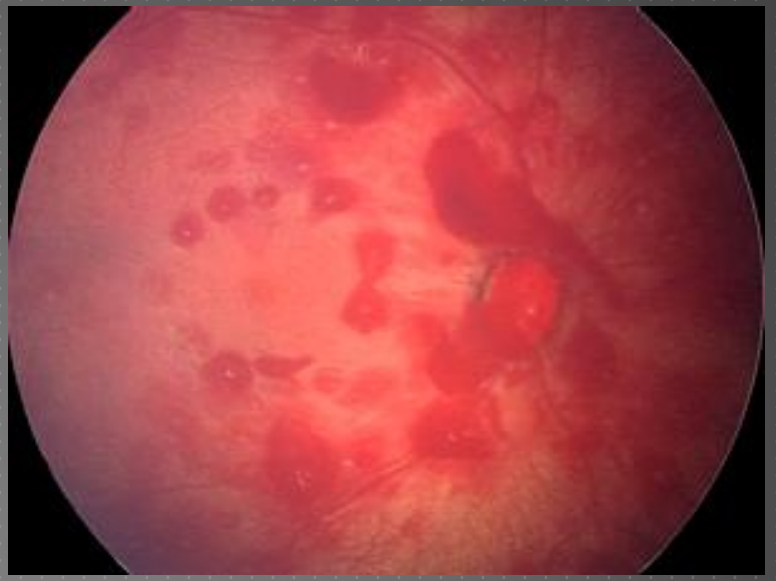
SHAKEN BABY SYNDROME

- Occurs between birth and 2 years of age
- Pathophysiology
  - Head is heavy and shaking causes whiplash like syndrome
  - Brain shifts back and forth causing intracranial bleeding
  - Vitreous gel inside the eye causes traction on internal limiting membrane and rupture of superficial retinal vessels



SHAKEN BABY SYNDROME

- Ocular manifestations
  - Retinal hemorrhages in posterior pole
  - Bilateral
  - Vitreous hemorrhage
  - **Few other disease will mimic this appearance
    - Birth Trauma
    - Viral retinitis
    - Leukemia
- Hallmark: Intracranial hemorrhage
- External exam: bruising, fractures



TREATMENT/OUTCOME

- Some degree of permanent visual loss likely
- Can have permanent neurological damage
- Prognosis limited

UV PROTECTION

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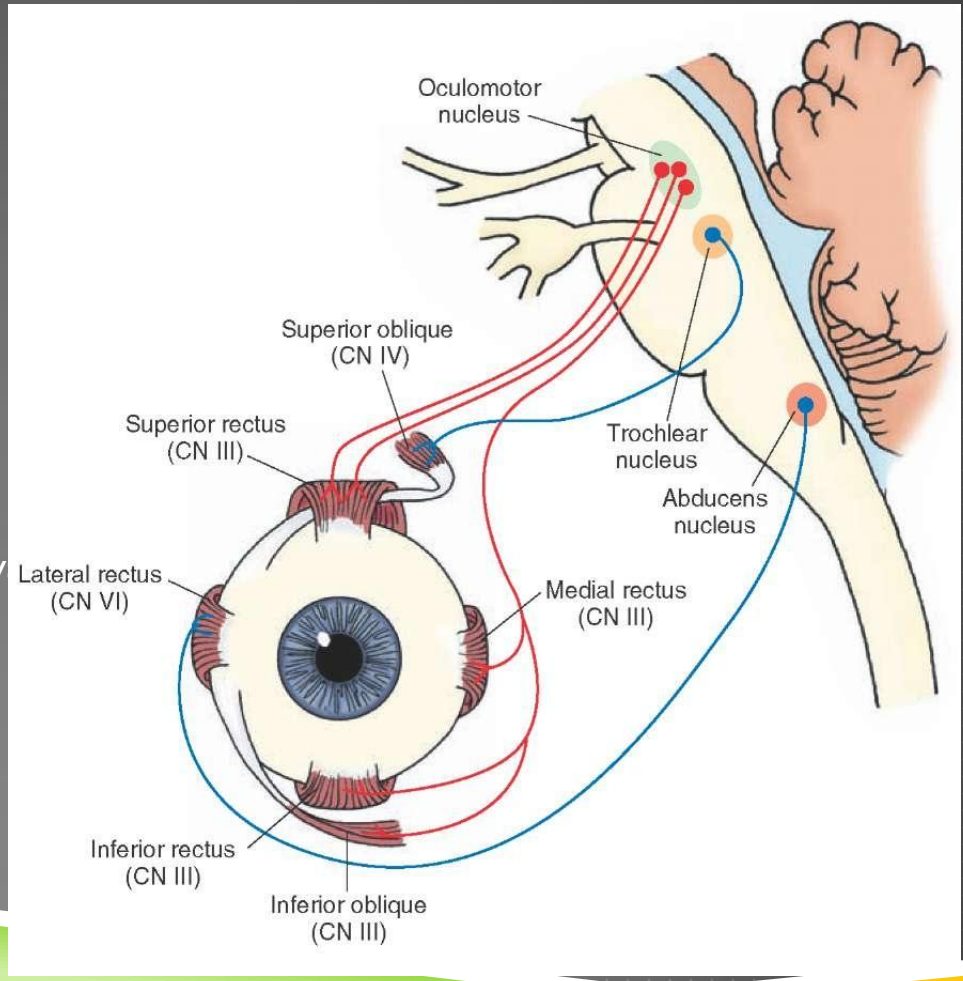
THANK YOU!

PARALYTIC STRABISUMUS

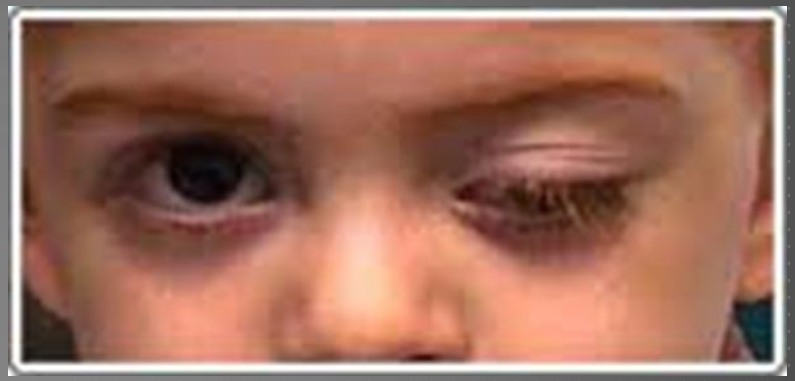
- Much less common
- Third nerve palsy
  - Congetinal or aquired (trauma)
  - Partial or complete
  - Exotropia, Hypotropia, Ptosis
  - Pupillary involvment?
    - In acquired cases, must rule out aneury



TREATMENT CN III PALSY

- Aggressive patching
- Prisms for alignment

- Surgery
  - Performed only when no/little expectation of additional recovery
  - Alignment obtained
  - Ptosis surgery



FOURTH NERVE PALSY

- Trochlear nerve
- Superior oblique muscle
  - Moves eye down and inward
- Examination
  - Head Tilt: Contralateral**
    - Hx pictures of child
  - Hypertropia and vertical diplopia
  - Difficulty reading in down gaze
  - Congenital is sporadic
- Idiopathic or trauma related
  - Very susceptible to trauma
  - Ventral exit from the brainstem -> Longest intracranial course

CN IV TREATMENT

- Low risk of amblyopia
- Treatment
  - Prisms
  - Monitor
  - If head tilt severe, may need treatment
- Surgery indicated only when non surgical approaches fail
- Success rates
  - 60-90% for functional success
  - 70-95% for cosmetic success

SIXTH NERVE PALSY

- Abducens Nerve
- **Most Common Palsy in all age groups
  - Under reported CNIV Palsy
- Innervates lateral recuts
- Congential forms uncommon

- Examination
  - Esotropia/Eye turns in in primary gaze
  - Diplopia
  - Ipsilateral face turn

Right Cranial Nerve VI Palsy

SIXTH NERVE TREATMENT

- Observation:
  - Often resolves on its own
- Prisms in limited amounts
- Surgery indicated if no improvement after 6 mos

TAKE HOME POINT…

- For acquired CN IV and VI Palsies:
  - Monitor for at least 6 mos
  - Ensure stable prior to every initiating surgical intervention

GENETIC COUNSELING

- Survivor of bilateral RB – 45% chance of producing affected child
- Suvivor of unilateral RB – 7-15% chance of producing an affected child (if the child is affected, there is an 85% chance the child will have bilateral lesions)
- Unaffected parents of child with bilateral RB – 5% chance their 2nd child will be affectd
- Unaffected parents with 2 or more affected children – 45% chance each additional child will be affected