Terminology- Greek roots: Leuko (or leuco) = “white”; coria = “pupil”
Refers to white pupil
May be associated with:
  Polycoria (multiple pupils)- rare
  Anisocoria (different size pupils)
  Corectopia (pupil out of position- not in center)
  Xanthocoria (yellow pupil- variation of white pupil)

Leukocoria may imply a serious, vision-threatening or life-threatening disorder
(For example cataract in an infant- vision-threatening due to amblyopia without rapid treatment-
or retinoblastoma- life-threatening without treatment for this cancer)

Leukocoria is a term usually used for preverbal children where the white appearance to the pupil area is a sign that something may be wrong- even though the child cannot tell us so

TRIAGE -> leukocoria should be seen urgently (within one week)
Scenarios in which leukocoria presents:
1) Family or PCP sees white pupil directly - highly likely that something is wrong
2) Pupil looks white in a photograph (the normal red reflex looks white) - more likely to be an aberration, especially if not consistent, but still could be a tumor
3) Family sees a white reflection from inside the eye on occasion - must rule out a tumor
All 3 scenarios require urgent evaluation

In the same category as leukocoria:
“No red reflex” or abnormal red reflex on PCP exam
   ➔ Urgent evaluation

PCP will use direct ophthalmoscope to do eye screening in infants and young children-
typically PCP cannot assess alignment in young infant or even optotype acuity in preverbal child-
but can look at the red reflex-
vision screening usually done by PCP at newborn exam in hospital and then at well-baby exams at age 2, 4, 6, and 12 months

Direct illumination of the pupil looks at light reflecting directly from the object - so a white cataract looks white
Retroillumination looks at light reflecting off of the retina and coming back out through the pupil - whatever blocks the light forms a silhouette - so a cataract looks black
Direct and retroillumination both useful methods for doing slit lamp photographs;
can also see corneal opacities with retroillumination using the red reflex (for example, keratic precipitates)

Retroillumination can also be used with the direct ophthalmoscope
   - look for alterations in the red reflex
   - suitable test for technicians using the direct ophthalmoscope

Bruckner test
   - looking at the red reflex of both eyes simultaneously
   - using the direct ophthalmoscope at arm’s length
   - can compare the red reflex of both eyes
Refractive error may add a white crescent to the red reflex, creating a partial white pupil reflex- may be a cause of leukocoria seen by PCP
- hyperopia -> superior crescent
- myopia -> inferior crescent

Bruckner test also shows
- cataract (dark silhouette may correlate with white cataract)
- eye misalignment
  if misalignment of sufficient magnitude, can see whitish reflection from optic nerve head

Causes of white pupil
- corneal opacity
- persistent pupillary membrane
- cataract
- fundus lesions
  - retinoblastoma
  - ocular Toxocara infestation Coats disease
  - advanced cicatricial retinopathy of prematurity
  - end-stage retinal detachment
  - fundus coloboma
  - endophthalmitis
  - toxoplasmosis

Corneal opacity
Many uncommon causes
Acronym: STUMPED
- Sclerocornea
- Tears in Descemet’s membrane (congenital glaucoma or forceps injury)
- Ulcer
- Metabolic (mucopolysaccharidosis)
- Posterior corneal defects
- Endothelial dystrophy
- Dermoid on cornea
Leukocoria

Cataract
- may be due to congenital defect in lens, hereditary, or metabolic
- nuclear cataract without family history requires metabolic work-up
- if large enough, may cause irreversible amblyopia and nystagmus- urgent evaluation
- significant unilateral cataract needs removal before one month of age
- significant bilateral cataract needs removal before 2 months of age
ONE REASON TO TAKE LEUKOCORIA SERIOUSLY

Cataract types
- anterior polar- usually small, affect vision through anisometropia and amblyopia, not directly
- nuclear or zonular- may be hereditary or metabolic in etiology- may be visually significant- high incidence of glaucoma
- cortical- Down syndrome
- posterior subcapsular- steroid use, post radiation
- posterior lenticous- unilateral- bowing of posterior lens capsule with gradual opacification
- PFV (persistent fetal vasculature)- also called PHPV (persistent hyperplastic primary vitreous)- unilateral- born with membrane adjacent to lens with variably dense cataract

Retinoblastoma- malignant tumor of the eye
- seen in children only, usually young children
- tumor is white and may give a white pupil reflex
ONE REASON TO TAKE LEUKOCORIA SERIOUSLY

Retinoblastoma
- relatively rare- 1 in 15,000 births- 11% of cancer in the first year of life
- may have a hereditary predisposition
- may be unilateral or bilateral- vision both eyes at risk
- if diagnosed early enough, often one eye may have good vision
- treatment for smaller tumors includes laser photocoagulation, cryotherapy, or chemotherapy
- advanced tumors may require enucleation- importance of early diagnosis
Toxocariasis
- disease caused by larvae of roundworms, *Toxocara canis* and *Toxocara cati*
- found in dog and cat feces
- microscopic larva enters eye (usually unilateral)
- associated with uveitis, vitritis
- forms of ocular toxocariasis: peripheral retinal granuloma (may cause central retinal fold), central retinal granuloma, endophthalmitis
- cloudy vitreous causes leukocoria
- treatment- steroids, sometimes vitrectomy

Coats Disease
- leaky telangiectatic retinal blood vessels cause exudates to form under the retina
- exudative retinal detachment looks white or yellow in pupil reflex
- unilateral in 90%
- occurs mostly in children
- treatment may include cryotherapy

Advanced cicatricial ROP (retinopathy of prematurity)
- when unfortunately retinal detachment occurs, a white membrane may form behind the pupil
- usually inoperable at this stage

End-stage retinal detachment-may form a retropupillary membrane

Coloboma of fundus
- congenital failure of closure of embryonic fissure of eye
- creates a gap in the retina and choroid
- may be associated with an iris coloboma (tear drop-shaped pupil)
- may be hereditary
- may be bilateral
- size and location of the coloboma determine visual potential
- coloboma is usually white in color

Endophthalmitis
- infection inside the vitreous cavity
- vitreous turns white
- often a red eye
- may require an urgent vitrectomy with intraocular injection of antibiotics
Toxoplasmosis
-infestation by *Toxoplasma gondii* — single-celled parasite in cat feces
-usually systemically asymptomatic unless newborn or immunocompromised
-if acquired during pregnancy, may cause white-yellow macular scars in child
-if dormant in retina, may reactivate causing white, fluffy lesion and iritis/vitritis

White Reflex in Photograph
May be optic disc reflection, but may be one of the above!
Schedule an eye Evaluation, and check it out to be sure!