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An Introduction to Pediatric Ophthalmology

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INTRODUCTION

Pediatric ophthalmology can feel like a completely different world compared to its adult counterpart. Children can sometimes be extremely challenging to examine and pediatric ophthalmologists examine a wide variety of age groups, including the non-verbal infant, the animated toddler, the adolescent, and the nonverbal adult. Careful exams are important at each stage of development because visual development is most critical in the first decade of life. Each patient group requires a distinctive expertise to perform an effective eye exam.

This review gives a brief introduction to some topics central to pediatric ophthalmology, including common clinical tests, common pathology, and some "can't miss" diagnoses.

AMBLYOPIA

Amblyopia is a broad term used to describe abnormal development of the visual pathways and visual cortex, resulting from disruption of equalness/clarity of images transmitted to the brain during maturation[1]. The ultimate result is permanent vision loss. It is estimated that 30% of the volume of the cerebral cortex is involved in visual processing[2]. Visual pathways within the CNS undergo major development and maturation from birth through the first 8-10 years[1]. After this point, the visual system becomes significantly less plastic. Any disruptions to vision during this period can have permanent consequences. Studies by Hubel and Wiesel in rhesus monkeys and kittens have shown significant atrophy of the lateral geniculate nucleus and striate cortex of the occipital lobe following experimental visual deprivation (ie: patching/sewing one eye shut) during visual maturation[3].

Because development of good visual acuity depends on the quality (clearness) and sameness of the images that the eyes present to the brain, any chronic disruption to those images during the critical period can cause amblyopia. This is because either the brain does not receive a clear image from the eye/s or it actively suppresses vision development from one eye to reduce the confusion of having two different or

unequal images. Causes are categorized as refractive, strabismic, or deprivation; a child may suffer from one or more cause. The most common causes are uncorrected high or unequal ("anisometropic") refractive error and strabismus[4]. Light-obstructing opacities (eg: congenital cataracts, corneal scars), retina disease, and optic nerve pathology are also important, albeit less common, causes of amblyopia. An easy way to remember the underlying causes of amblyopia is through the mnemonic SOS – Spectacles (refractive error), Obstruction (cataracts, corneal opacities, retinal or optic nerve pathology) and Strabismus. Treatment for amblyopia entails identification and correction of the underlying visual deficit as well as occlusion of the "good eye"[5] . Examples of this are performing careful visual assessment on children and providing glasses to correct refractive error or surgery to correct strabismus error.

Patching is used to treat amblyopia by applying the patch to the better seeing eye. This forces the brain to rely only on the image transmitted by the poorly seeing eye in order to develop the vision from that eye. Patching relies on the principle of neurodevelopment. If one pathway receives significantly more stimulus than the other, it will develop more robust neuronal pathways, refined connections and synapses. It is somewhat akin to one competitive weightlifter training one arm 4 hours per day and the other arm 30 minutes per day. The arm that trained longer will undoubtedly be stronger and have gained more muscle. However, by patching, the normal eye is penalized, and the amblyopic eye is allowed to "catch up." An opaque material placed over the eye for a few hours per day usually accomplishes this goal but pharmacologic blurring with cycloplegic drops like atropine are also sometimes used[5]

.

They key to treating amblyopia is early identification of poor visual acuity or acuities and prompt intervention. Vision screening on a routine basis is recommended to prevent the development of amblyopia[7] . While the severity of amblyopia depends on the type of deprivation, degree of deprivation, and developmental stage, some degree of amblyopia may develop in as little as 4-weeks of viewing disrupted images[8] . If identified early, the prognosis is very good and lasting visual deficits will be reversed or mild; however, if visual deficits are not identified early or treatment is delayed, there can be permanent vision loss that cannot be corrected with any medical or surgical intervention on the eye or brain, as the pathology is in the brain itself.

Figure 1 What is amblyopia (lazy eye)? Image source: [6]

STRABISMUS AND EXTRAOCULAR MUSCLES

In contrast to certain mammals like horses, human eyes are aligned, and movement is synchronous and highly coordinated. This is binocular vision and is what allows a high-degree of depth perception or stereopsis. We can easily determine the location of objects relative to one another on a three-dimensional plane, even if they are millimeters apart. The importance of binocular vision in generating stereopsis can be demonstrated by pinching two coins (one with each hand) and holding them out in front of you. Trying to touch the edges together is easy with both eyes open but very difficult with one eye closed.

Strabismus is any misalignment of the eyes relative to one another, i.e. the eyes don't fixate together on the same target[9] . Much like the discussion on amblyopia, strabismus is a broad term and can be due to a number of underlying reasons. This includes weakness or palsy of the cranial nerves involved in eye movement (III, IV and VI), failure of the cerebral mechanisms that maintain eye alignment, or pathology involving the eye muscles themselves[9] .

Table 1 When discussing strabismus, it is important to review the muscles involved in eye movement so they can be consistently documented during examination. The extraocular muscles, their corresponding cranial nerve and action on the eye are summarized below. Remember the mnemonic "LR6, SO4, AO3" to help keep these straight: Lateral Rectus is innervated by CN6, Superior Oblique is innervated by CN4, All Other Extraocular Muscles are innervated by CN3.

Muscle	Innervation	Action
Superior Rectus	CN 3 (Oculomotor)	Elevate, Adduct, Intort
Medial Rectus	CN 3 (Oculomotor)	Adduct
Inferior Rectus	CN 3 (Oculomotor)	Depress, Adduct, Extort
Inferior Oblique	CN 3 (Oculomotor)	Extort, Elevate, Adbduct,
Superior Oblique	CN 4 (Trochlear)	Intort, Depress, Abduct
Lateral Rectus	CN 6 (Abducens)	Abduct

Understanding the actions of eye muscles requires an interpretation of eye movement in the field of action of each muscle (this is what we do when watching the eyes move in the "H" and measuring alignment in all fields of gaze).

It is important to describe strabismus using standard terminology. Strabismus is misalignment of the eyes relative to one another. When the eyes are in alignment, we say that patient is "orthophoric," or simply "ortho." (Remember ortho = straight like orthopedics = straight bones). When the eyes are misaligned, one eye can be deviated inward ("esophoria/tropia," or "eso"), outward ("exophoria/tropia," or "exo"), upwards ("hyperphoria/tropia," or "hyper"), or downwards ("hypophoria/tropia," or "hypo"). Strabismus is further described as a "tropia" or "phoria." A tropia is a manifest deviation, meaning that it is usually visible at all times (there is an exception, that of an intermittent tropia, where the eye deviates under decompensated conditions). In contrast, a phoria is a latent deviation: present only under monocular conditions. When the brain is seeing through monocular conditions, fusion of the two images (one from each eye) is disrupted. Normally we are seeing under binocular conditions, so a phoria is far less significant than a tropia. Thus

we would describe an eye that constantly deviated inwards as an esotropia and an eye that drifts outwards only under monocular conditions an exophoria[9] .

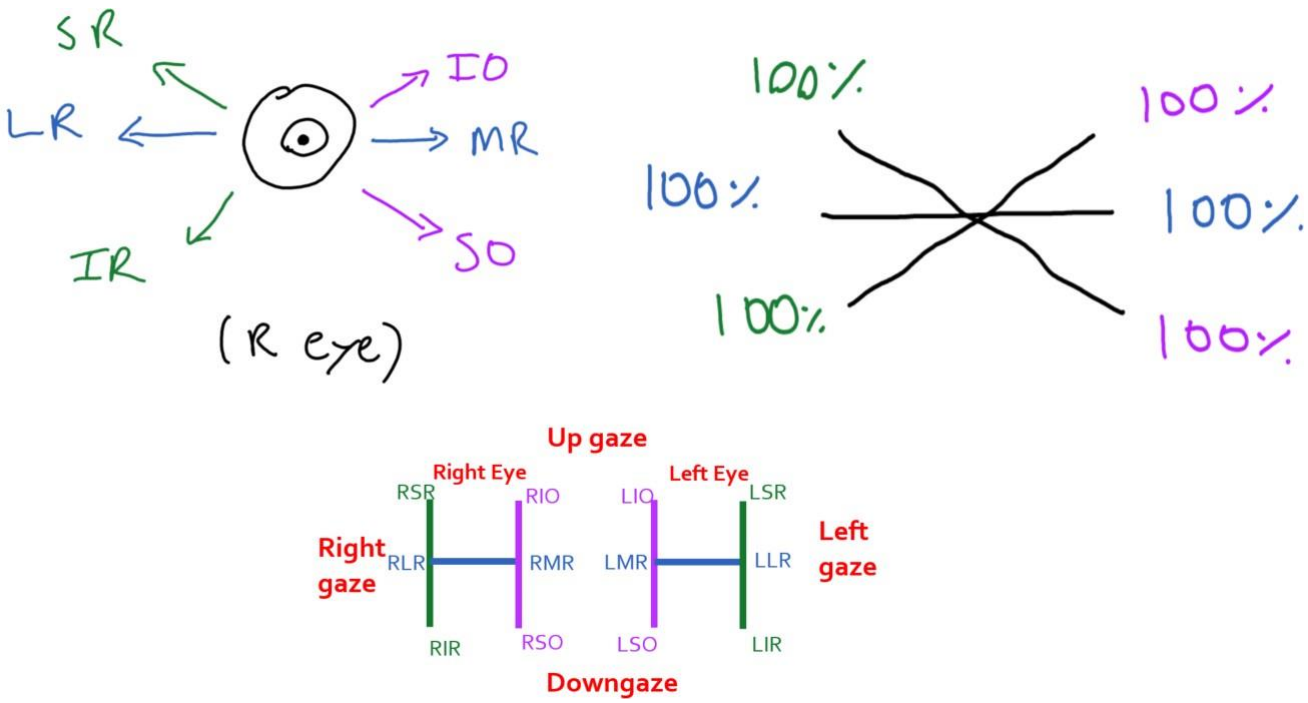

Figure 2 Field of Action of each ExtraocularMuscle: The gaze direction where one muscle isthe primary mover of the eye. eg: adductionof either eye is the field of action for the medial rectus of either eye

(another way to say this is: left gaze for the right eye is the field of action for the the right eye's medial rectus and right gaze for the left eye is the field o faction for the left medial rectus)
eg: down and left gaze is the field of action for the right superior oblique

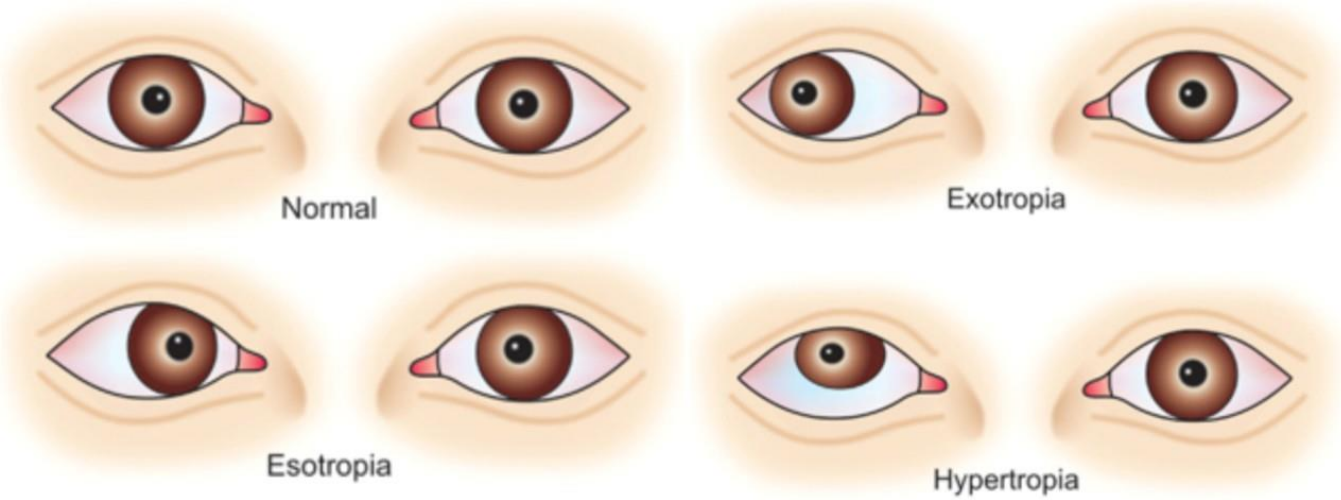

Figure 3 Illustration of eye deviations. Image source: [10]

Two tests are commonly used to identify a strabismus: the cover-uncover test and the alternate cover test (or cross-cover test). These tests assess strabismus by covering one eye or the other to see which eye the brain is using at a given moment, and which direct the deviating eye is going. The example below illustrates the cross-cover test being used to detect a left exotropia.

The pathology responsible for the great majority of strabismus is idiopathic, likely stemming from an impaired cerebral mechanism of gaze control. A known cause is found in a minority of cases. These include Duane Syndrome (congenital absence of CN6 nucleus), Brown Syndrome (congenital superior oblique tendon tightness) and thyroid eye disease (which causes inflammation and dysfunction of the extraocular muscles). It is essential to rule out pathology that may require urgent management. For example, suspected cranial nerve palsies with complains of diplopia, headache, nystagmus, altered mental status, pupil abnormality (anisocoria, RAPD, poor constriction/dilation), or other neurological signs should prompt immediate neuroimaging[11].

Strabismus can be treated with a) glasses b) prism lenses c) surgery or d) vision therapy to manipulate the extraocular muscles.

Initially, it is essential to test the visual acuity in any child presenting with strabismus. Refractive error (hyperopia, or “far-sightedness”) can cause excess accommodation, even at distance, as the eye is working to thicken the lens and focus light on the back of the retina. If one eye has a much greater refractive error than the other (“anisometropia”), this can also lead to strabismus.

Prisms refract light in a predictable manner, similar to concave and convex lenses used for refractive error. They are used both as a diagnostic (i.e. measure the amount of deviation) and therapeutic tool (i.e. prescribed in glasses to alleviate small deviations, but have a limit as they add considerable bulk to glasses). The correct prism moves the fixation target onto the fovea of the strabismic eye.

In surgical cases, the insertion of the extra-ocular muscles upon the sclera is physically altered. By moving the muscle attachment to the eye anteriorly or posteriorly, the forces are changed to balance the muscle tension and align the eyes. For example, a recession procedure can be performed on the rectus muscles to correct esotropia or exotropia. Finally, vision therapy (i.e. eye exercises) has a role in treating a very specific type of strabismus called convergence insufficiency, which affect the ability to pull the eyes

together when trying to read or look at close objects.

The bottom picture is a child before and after surgery to repair a left-sided esotropia. Surgery can change the entire appearance of the face and has a major impact on self-esteem and psychological health.

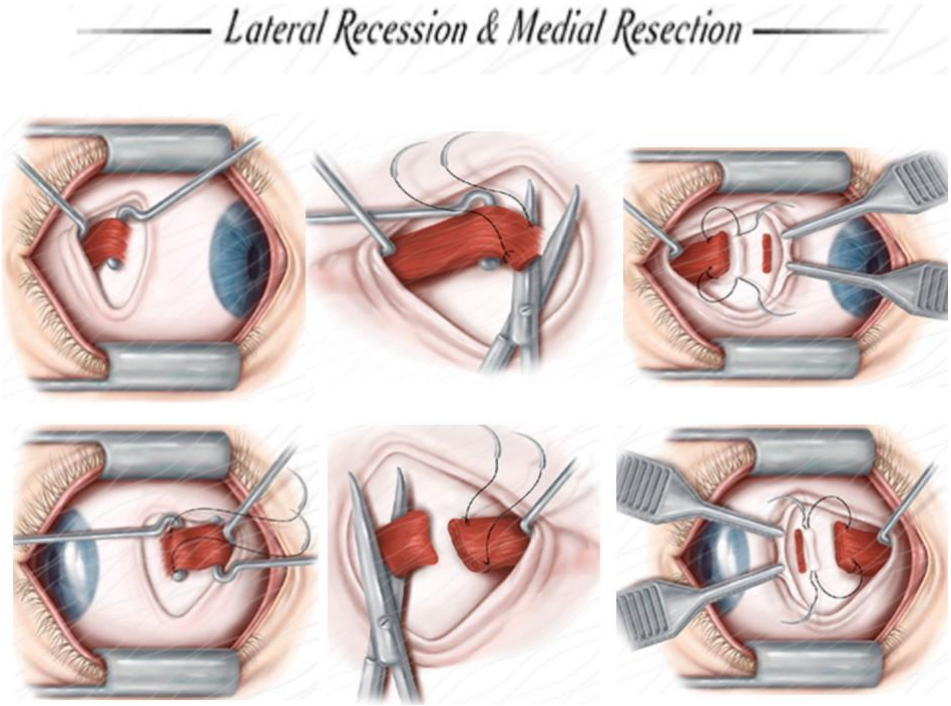

Figure 4 Image source: [12]

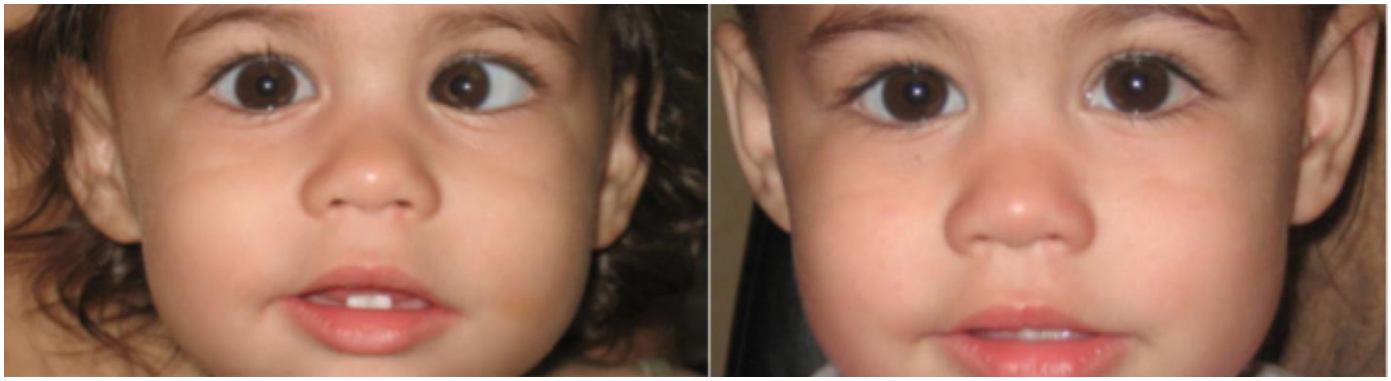

Figure 5 Before and After Surgery Photos. Image source: [13]

LEUKOCORIA

Leukocoria describes a white (instead of red) reflex ("leuko – coria" = white pupil). Pediatricians perform screening exams with direct ophthalmoscopy to evaluate the red reflex in newborns and during childhood. The differential diagnosis of leukocoria is extensive. The differential includes tumors (e.g. retinoblastoma, optic nerve glioma), anterior segment opacities (e.g. cataract, corneal scarring), congenital malformations (e.g. retinal dysplasia), vascular diseases (e.g. retinopathy of prematurity, Coats disease), inflammatory

diseases (e.g. ocular toxicariasis) and trauma (e.g. shaken baby syndrome, vitreous hemorrhage). Congenital cataract is the most common of these problems. Many of the other diagnoses, like retinal dysplasia, are rare[14].

Retinoblastoma, although also rare, is responsible for approximately half of the cases of leukocoria in infants and is the most common intraocular malignancy arising in children[16].

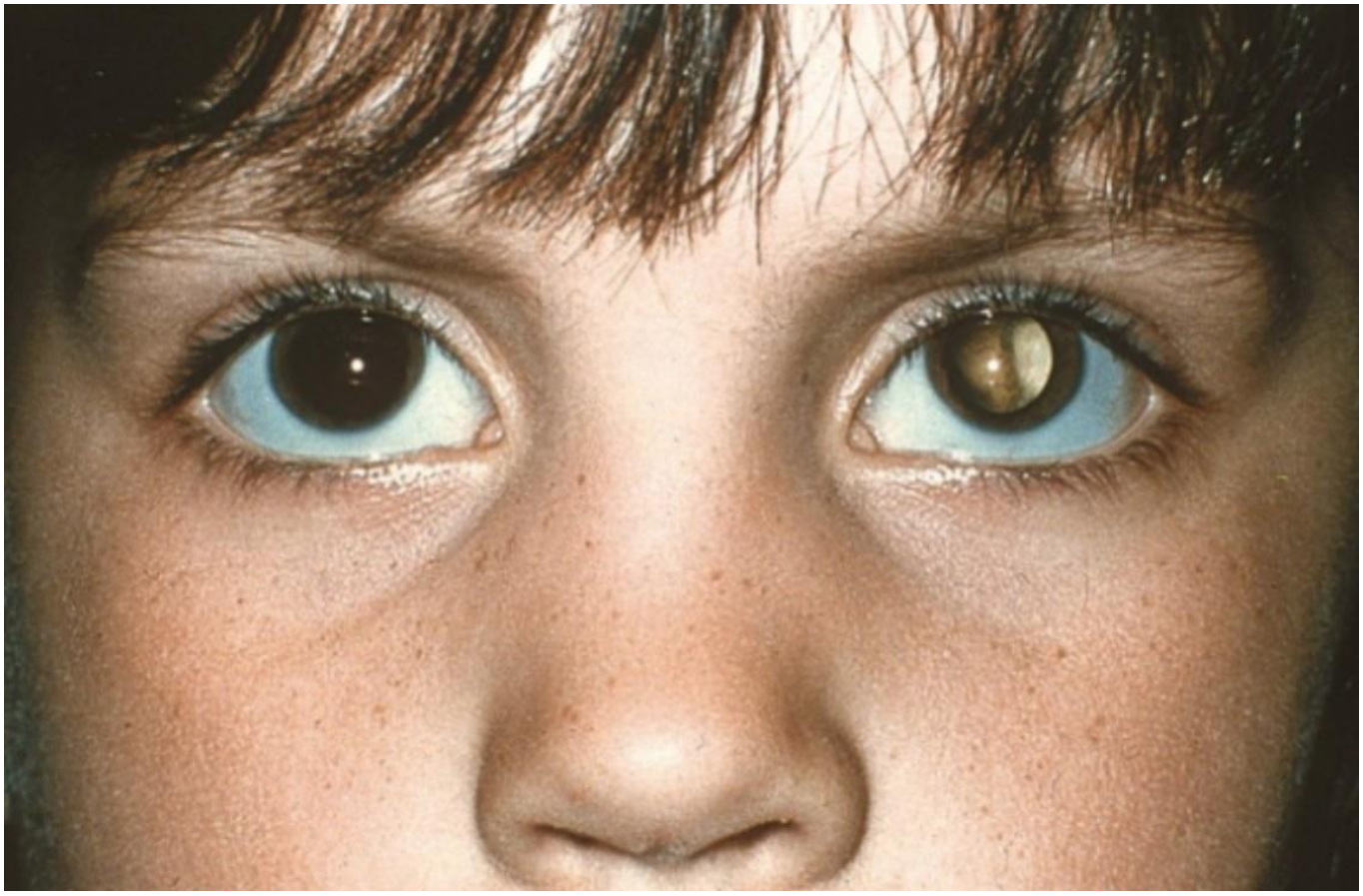

Figure 6 Leukocoria in a patient with retinoblastoma. Image source: [15]

Retinoblastoma is a malignancy arising from retinal photoreceptors. Retinoblastoma demonstrates one of the classic principles in genetics and oncology, the "two-hit hypothesis" and loss of heterozygosity. The retinoblastoma (RB1) gene codes for a tumor suppressor protein and two functioning copies are normally found in cells. In hereditary retinoblastoma, one copy of RB1 is mutated in the chromosome for the germline cells germline cells and passed to offspring while in the non-hereditary form the child is the first in their family to have a germline mutation and has the mutation in both alleles of the germline retinal cells. In hereditary retinoblastoma the child is at significant increased risk of cancer and one more "hit" is all it takes to develop an ocular neoplasm and a variety of other cancers (e.g. osteosarcoma, lung cancer, brain tumors and melanoma). In hereditary retinoblastoma, the ocular neoplasm will likely be bilateral while in the non-hereditary form it will most likely be unilateral[17]. Dilated exam will show nodular

masses arising from the retina, which can grow inwards into the vitreous and even seed the anterior chamber, leading to secondary open angle glaucoma[17] . Early diagnosis and treatment is key to determining prognosis. If detected early, the prognosis is generally very good and survival rates high. There are various treatment modalities for retinoblastoma including systemic/intraarterial/intravitreal chemotherapy and radiation therapy[18] . The patient may require an enucleation, or removal of the eye, can be used depending on extent of disease or failure of other therapies.

Retinopathy of Prematurity

Retinopathy of prematurity is a disease of abnormal development of the retinal vessels, which in its most severe form can lead to blindness. If not detected and treated, incomplete vascular development in the retina can lead to aberrant vessels that lead, ultimately to retinal detachment. It can be helpful to draw a comparison between diseases like diabetic retinopathy in order to understand retinopathy of prematurity (ROP). Although the underlying pathophysiology is different, both disease states have a common theme: ischemic retina inducing neovascularization and incompetent vessels that can induce tractional retinal detachment[19] .

The retinal blood supply develops from endothelial cell precursors starting at the optic disc at approximately 14-weeks gestation. The macular vessels complete their development at approximately 22-weeks gestation, and the blood supply of the peripheral retina is the last to develop, becoming mature postnasally. This is accomplished via a complex and coordinated orchestra of cellular signaling involving vascular endothelial growth factor (VEGF). When infants are born prematurely, the process of retinal vascularization is incomplete. The vessels can then develop normally over the next few weeks, or they can begin to develop abnormally. As a result, the peripheral retina may remain avascular at birth, and this in turn can induce neovascularization and vessels to grow into the vitreous. This is termed extraretinal fibrovascular proliferation and can induce vitreous hemorrhage and tractional retinal detachment, in a manner similar to what occurs in proliferative diabetic retinopathy. The severity of ROP is partially based on the extent of this extraretinal fibrovascular proliferation. A classification system was developed in the 1980s to standardize clinical descriptions of ROP and describes zones of the retina where ROP is found. The drawing below shows the different retinal zones; the picture shows the fibrovascular ridges and traction bands that can appear as ROP affects the retina[19] .

The greatest risk factors for ROP is short gestational age and low birth weight. Infants born before 30 weeks or a birth weight of 1500 grams or less should also be screened for ROP with dilated fundus exams.

The need for supplemental oxygen is also a risk factor for developing ROP, as well as an eventful NICU course (sepsis, etc). The mechanism is incompletely understood, but supplemental oxygen may induce vascular injury in premature retinal vessels. Thus, infants born greater than 30 weeks and greater than 1500 grams may still need to be screened if they received supplemental oxygen as neonates[19] .

Much like diabetes, the mainstay of treatment for ROP involves disruption of the angiogenic signaling responsible for new vessel growth. This includes ablation of the peripheral retina with laser photocoagulation. Intravitreal injection of VEGF inhibitors like bevacizumab are also used, although optimal dose and longterm outcomes are still being investigated[20] .

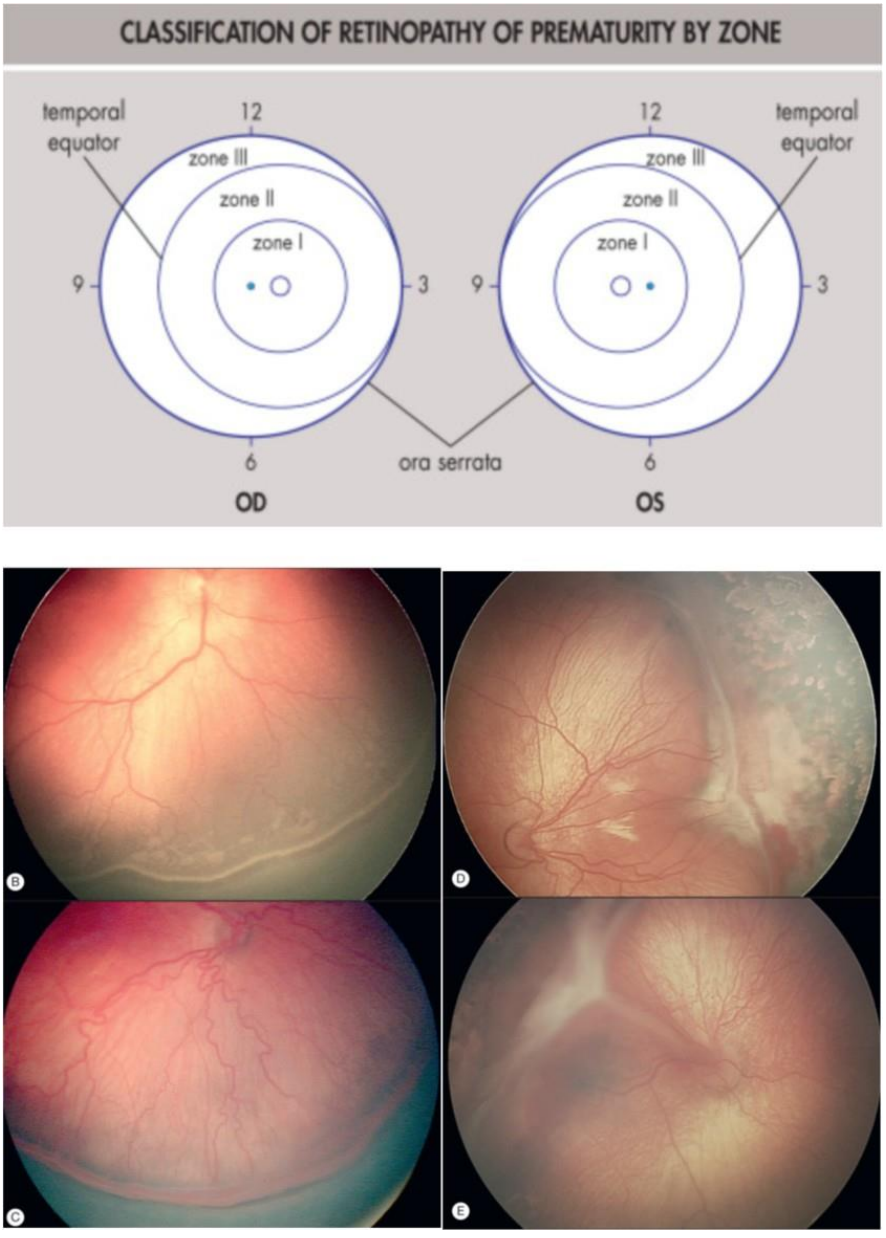

Figure 7 Image source: [19]

VI: Refractive Error

It is important to screen and correct children for refractive error to prevent amblyopia (i.e. assess the need for glasses to ensure clear vision in both eyes). Refraction is the bending of light through a medium. When an eye has a refractive error, the eye is unable to bend light so that the image on which it's fixating lands on the fovea, hence, the eyes need glasses to see clearly. The phoropter (i.e. the machine with multiple lenses placed in front of the patients face to measure glasses) is used to check glasses prescription in adults and older children. However, the machine relies on verbal communication – "I can see it" or "I can't!" This can work for children who are verbal, even if they can't discern letters (there are pediatric optotypes such as LEA or Allen symbols). However, in children who have not yet learned to speak, the phoropter cannot be used. Instead, pediatric ophthalmologists use a series of handheld lenses and a device called a retinoscope to perform retinoscopy, an objective measure of refractive error[21] .

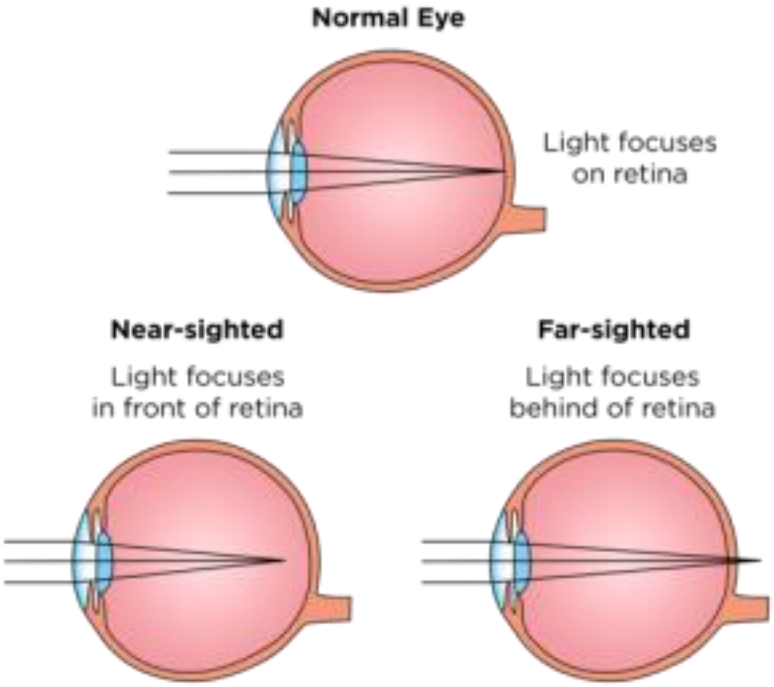

Figure 8 Myopia, Hyperopia, Presbyopia and Astigmatism. Image Source: [22]

By using retinoscopy, the pediatric ophthalmologist can determine if there is enough refractive error to warrant glasses. Myopia (near-sightedness), hyperopia (far-sightedness) and astigmatism (refractive error at an axis) can all be diagnosed with retinoscopy and the proper spectacles can be prescribed. This, in turn, can have an enormous impact on the entire development of the child, and their ability to learn. The specific nuances of retinoscopy are beyond the scope of this review.

VII: Conclusion

Pediatric ophthalmology can be an incredibly rewarding career by impacting the lives of children and their families. This review provides a brief introduction to some important introductory topics but there are many more exam techniques, diseases, syndromes and interplay with systemic pathology not discussed in this review. The author hopes the reader finds this information interesting, useful and maybe even inspires them to explore some more!

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