



MÀSTER UNIVERSITARI EN OPTOMETRIA I CIÈNCIES DE LA VISIÓ

TREBALL FINAL DE MÀSTER

ÚLTIMOS RETOS EN EL TRATAMIENTO DE LA CATARATA CONGÉNITA

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25 DE MAIG 2022



APPRECIATIONS

First and foremost, I would like to praise and thank God, the almighty, who has granted me countless blessings, and knowledge, so that I have been finally able to accomplish this thesis.

I would like to thank my supervisor Dr. José Lamarca Mateu for all his support, efforts, and guidance throughout my study and thesis.

I also am very grateful to all the academic management team, especially, María Calvet for her continuous support and help during this process. A special thanks to Maria Consuelo Varon Puentes for her endless support and her great help and guidance that helped me to choose this thesis topic.

I am very grateful for all my colleagues, especially Sofia Vitucci and who I have met during this amazing journey, and for every single thing that they have taught me and helped me in.

Last, but not least, my deepest gratitude goes to my family. Both starting and finishing my degree would have been impossible without the unconditional love and support they have shown me throughout all of these years.



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RESUM

La descripció de la cataracta congènita: és una opacitat de la lent ocular que apareix en el part o durant la primera infància, és una malaltia rara i sovint desconeguda, però a excepció d'altres raons associades amb ella que inclouen: genètica, metabòlica, infeccions, ús de corticoesteroides, trauma, uveïtis, així com anomalies oculars.

L'objectiu i els mètodes d'aquest treball: és investigar diversos articles sobre els últims reptes del tractament de la cataracta congènita i quins tipus d'enfocaments es poden utilitzar per curar-los i per a l'ambliopia que es produeix a causa de la cataracta.

La conclusió: malgrat l'avanç de les tècniques quirúrgiques per als nens molt millor del que ho va ser fa anys, així que els reptes com la mida de l'ull, l'edat, la selecció d'exàmens de l'OL, l'horari de cirurgia i les complicacions, així com la remodelació de la visió post-cirúrgica; encara existeixen, però sobretot la rehabilitació de la visió i el tractament amblòpic per a les condicions post-cirúrgiques, i per a aquests casos menors, han de ser seguits periòdicament.

Paraules clau: cataracta congènita, tractament, rehabilitació visual



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RESUMEN

La descripción de la catarata congénita: es una opacidad del cristalino que aparece en el parto o durante la primera infancia, es una enfermedad rara y muchas veces de causa desconocida, pero salvo por otras razones asociadas a ella que incluyen; genéticas, metabólicas, infecciones, uso de corticoides, traumatismos, uveítis, así como anomalías oculares.

El objetivo y los métodos de este trabajo: es investigar varios artículos sobre los últimos desafíos del tratamiento de la catarata congénita y qué tipos de enfoques se pueden utilizar para curarlos y para la ambliopía que se produce debido a la catarata.

La conclusión: a pesar del avance en las técnicas quirúrgicas para niños mucho mejor que hace años, los desafíos como el tamaño del ojo, la edad, la selección de exámenes de LIO, el cronograma de cirugía y las complicaciones, así como la rehabilitación de la visión posquirúrgica; aún existen, pero sobre todo la rehabilitación de la visión y el tratamiento ambliópico para condiciones postquirúrgicas, y para aquellos casos menores, deben ser objeto de un seguimiento periódico.

Las palabras clave: catarata congénita, tratamiento, rehabilitación visual



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ABSTRACT

The description of congenital cataract: is an opaqueness of eye lens that appears at childbirth or during early childhood, It is a rare disease and often unknown cause, but except for other reasons associated with it that include; genetic, metabolic, infections, corticosteroids usage, trauma, uveitis, as well as ocular anomalies.

The goal and the methods of this work: is to research various articles on the latest challenges of treating congenital cataract and what kinds of approaches can be used for curing them and for the amblyopia that is produced due to cataract.

The conclusion: in spite of the advancement in surgical techniques for children much better than it was years ago, so the challenges such as eye size, age, selection of IOL examinations, surgery schedule, and complications, as well as vision rehab post-surgical; still exist, but most of all the vision rehabilitation and amblyopic treatment for post-surgical conditions, and for those minor cases, they must be followed up periodically.

The Key words: congenital cataract, treatment, visual rehabilitation

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1. INTRODUCTION:

1.1 CRYSTALLINE LENS

1.1.1.DEVELOPMENT

At the twenty-eighth day of gestation, the surface ectoderm becomes thicker and interacts with the optic vesicle to generate lens placode. (Augusteyn, 2010).

In the fifth week, the process of invagination occurs for optic vesicle forming an optic cup and for the middle of lens placode creating the lens pit. After that, the lens pit with its two-sided margins widen shrink and moving centrally eventually generating a slender stalk attached to the surface ectoderm. The lens pit entirely detaches from the surface ectoderm and produces a lens vesicle in the sixth week of pregnancy; that it falls into the optic cup and invade the optical vesicular later. (Liu, 2016).

The cells of the posterior vesicle extend directly to the anterior epithelial cell layer, eventually becoming the primary lens fibers filling the lens vesicle and eventually constitute the embryonic nucleus of the matured lens. Sixth to seventh week as the lens develops, secondary lens fibers (fetal nucleus) of the developing lens begin to elongate and differentiate from the epithelial cells located at the equator. By the eight weeks, at both poles (anterior and posterior) of the embryonic nucleus of the lens, this Y-shaped suture forms through the terminal ends of the secondary lens fibers adjoining each other. During lifespan, fiber cells continuously thrive after differentiation. As fiber cells go through terminal differentiation, they eliminate their nucleus and other cell organelles in order to attenuate scattering of light. (Berry et al., 2020)

1.1.2.ANATOMY, PHYSIOLOGY AND STRUCTURE

The lens performs with the cornea jointly to send the light and refract it. (Hejtmancik, 2008). It can concentrate things in various spaces hence enabling the formation of a crisp image of the thing on the retina, this process is named accommodation. The lens is elastic, translucent as well as elliptical, double convex in the form. (W. Chen et al., 2017). The curvature of the posterior surface of the lens (its apex known as a posterior pole) is more curved than the anterior surface (its apex known as an anterior pole), these two surfaces are connected by a line constituting the axis of the lens, and the equator is a margin around the lens. The lens is Located directly posterior to the iris, anterior to the vitreous body. It is held in place by Zinn zonules, which aid and bind it to the ciliary body. (Kasturi & Matalia, 2017). At birth, its weight of almost 65 mg continues to increase until 160 mg at age 10 years; after that slowly increases to reach around 250 mg at age 90 years. Contains a large percentage of proteins around 60% compared to other tissues, of its overall mass. (Hejtmancik & Shiels, 2015).

Structurally composed of: Capsule, Epithelium, Cortex and nucleus. (Aliancy & Mamalis, 1995)

1.1.2.1. THE CAPSULE

At five-six weeks of pregnancy, it can be discovered; it has a role to shape the lens throughout the accommodative variations. (Hejtmancik & Shiels, 2015). It's a basement membrane made up of type VI collagen produced by the epithelial layer, and dyes favorably into the Periodic acid-Schiff (PAS) stain. (Aliancy & Mamalis, 1995).it is clear and elastic. (W. Chen et al., 2017). In the anterior and posterior pre-equatorial zones (20 μm) the capsule is thickened more than in the region of the central posterior pole (2–4 μm) which is thinned more, and due to the posterior capsule being extremely thin that would be susceptible to rupture during the surgery of cataract. (Kasturi & Matalia, 2017). from birth, the posterior capsule is significantly thinner than the anterior lens capsule, the thickness of the capsule continues to grow lifelong. (American Academy of Ophthalmology,Jick, 2019)

1.1.2.2. THE EPITHELIUM

The Epithelium is a single-layer cuboidal cell found underneath the capsule at the lens's anterior and the equator. (Chen et al., 2017). The mitotic cells are active for differentiating more by the elongative operation, the protein rise within cells, also losing of organelles. These active cells are structured in one layer crossing the anterior capsule's posterior side. (Aliancy & Mamalis, 1995), Furthermore, this mitotic process happens in a circle surrounding the front lens, referred to as the germinative zone. The shaped new cells are transferred into the equator to separate into fibers. (Kasturi & Matalia, 2017). When the migration of the epithelial cells directly to the arc zone of the lens, and beginning the procedure of final differentiation, which results in the formation of lens fibers. As the epithelial cells extend to create lens fiber cells, which probably is the biggest morphological alteration; This alteration is tied to a significant rise in the bulk of cellular proteins in the fiber cell membrane. (American Academy of Ophthalmology,Jick, 2019).

1.1.2.3. CORTEX AND NUCLEUS

The eldest layers of the lens, the embryonic and fetal lens nuclei, were formed during embryonic development and remain at the lens's center. (American Academy of Ophthalmology,Jick, 2019)

A lens' cortex is made up of the outer fibers that have formed recently. (Kasturi & Matalia, 2017).

The cortex and the nucleus have no histological difference. (Aliancy & Mamalis, 1995)

Lens sutures are the connections at the ends of lens fibers at the front and back of the lens. the upright Y is the front fetal nucleus suture while an inverted Y is the back suture. With the growth of the lens and

the elongation of the fibers, extra complicated sutures form, for instance stellate-shaped sutures. (W. Chen et al., 2017).

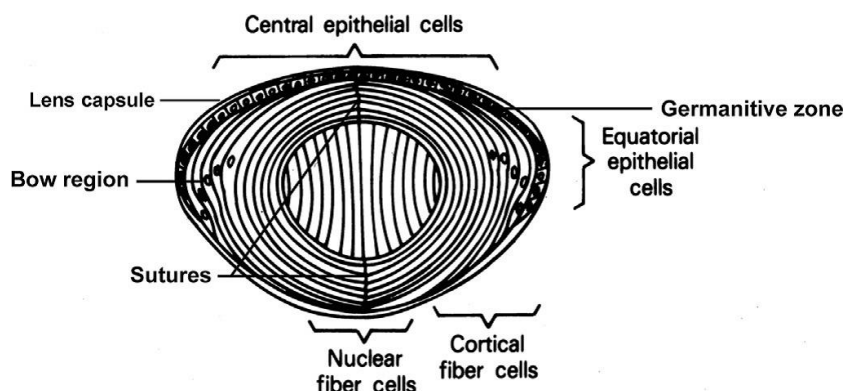


Image1: The human ocular lens composition (Hejtmancik & Shiels, 2015)

1.2. CONGENITAL AND DEVELOPMENT ABNORMALITIES OF THE LENS

1.2.1. ALTERATIONS IN EXTRACRYSTALLINE DEVELOPMENT

1.2.1.1. CONGENITAL APHAKIA

A condition known as non-existent of the crystalline lens, it has two types the initial one is occurred due to the lens placode may not have developed properly (primary congenital aphakia). The second, the lens developed before birth is reabsorbed during pregnancy (secondary congenital aphakia). Mutations in genes (FOXE3 mutation, PAX6 mutation) causing it genetically or by contagious disease including rubella. (Ionescu et al., 2016). These two types are frequently accompanied with other eye abnormalities. (American Academy of Ophthalmology, Jick, 2019)

1.2.1.2. CONGENITAL ANIRIDIA

Uncommon illness, (occurrence 1 of 64000 to 1 of 100000) that usually impacts bilateral and is linked to PAX6 genetic mutation. This gene is found on chromosome 11's short arm (11p13). Also, this gene is contributed to the eye's evolution, primarily the cornea, iris, lens, drain angle ocular, ciliary body. The hypoplasia of the iris is the main visible sign which varies from nearly complete to a minor form. Other eye signs include anomalies of the eye's drainage angle, glaucoma, the cornea with eye dryness and keratopathy, lens opacities, lens subluxation, and disorders of optic nerve, retina, and macula accompanied with nystagmus. The patients demonstrate decreased in visual acuity (VA), strabismus, as well as poor vision prediction that has been present from a young age. (Calvão-Pires et al., 2014). The anomalies in lens in the aniridia might be broadly divided into 3 categories clinically: changes in clarity (opacities or cataracts), in location (subluxation) that often reflect zonular weakening, and in shape and

size (microspherophakia), the opacifications of lens are quite common in aniridia eyes, with incidence rates ranging from 50 to 85 percentage, in the Korean population with up to 88 percentage, and the Norwegian population more than 90 percentage. Cataracts related to aniridia have a variety of morphologies, characterized as lamellar, subcapsular, cortical, polar, and seldom nuclear. If the vision in congenital aniridia with cataracts is slightly affected by the opacification, the surgery is not necessary. The gradual decline in vision usually happens during adulthood because of the evolution of cataract presenile and that requires surgical intervention. Treatment: Firstly, Continuous circular capsulorhexis technique (CCC) must be dealt with special attention. Histologic studies have displayed that the anterior capsules of people who have congenital aniridia are light and brittle, with deteriorating alterations in the epithelial cells. Secondly, utilizing the Capsular tension ring technique (CTR) may help to lower the danger of displacement of the intraocular lens and post-operative in posterior capsule fibrosis (PCO). Typically, congenital aniridia patients are juvenile, thus the occurrence of fibrosis in the capsule anterior and PCO are very common. Many procedures like Morcher intraocular lenses with a black diaphragm, prosthetic iris devices, or keratopigmentation, are available for rehabilitation of vision following crystalline lens surgery in these individuals, whether by cataract or displacement. (D'Oria et al., 2021)

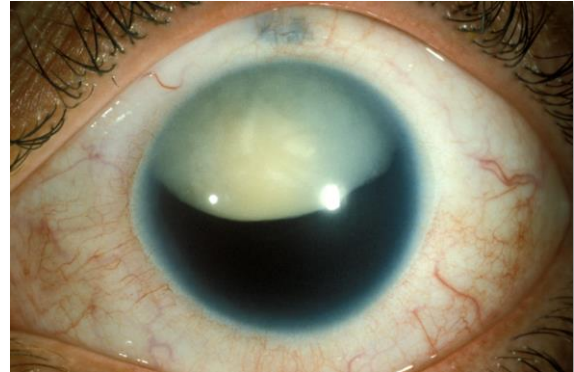
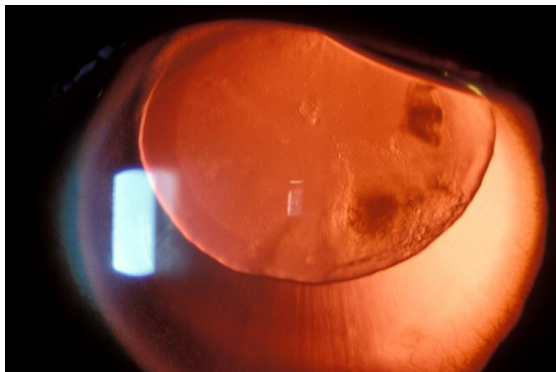


Image 2: Aniridia with microspherophakia

Image 3: Aniridia with subluxation

(Both extracted from Centro de Oftalmología Barraquer)

1.2.1.3. *PERSISTENCE TUNICA VASCULOSA LENTIS*

The structures of the systemic hyaloid vascular are artery of hyaloid, vasa hyaloide propria, tunica vasculosa lentis, in addition to membrane of pupillary. (McLeod et al., 2012)

The section of the ocular artery, that is a section of the carotid artery internal is called the Hyaloid artery. (Jeon et al., 2019)

VHP (vasa Hyaloidea propria) is comprised of sections of the hyaloid's arteries in the vitreous nearest to the retina; TVL (tunica vasculosa lentis) is constituted of sections of the hyaloid artery encircling the lens's posterior half-sphere, furthermore, the pupillary membrane is constituted of strands covering more the lens's anterior pole. (C. Chen et al., 2019)

Throughout the eye's initial fetal growth, the hyaloid artery is considered a transition vascular systemic, that guarantees nourishment for the eye's growing crystalline and posterior portion. Among third and fourth weeks of the gestation period, the HA joins the eye in the location of the optic nerve disc, develops distal till arrives at the eye lens among the fourth and fifth weeks, and forms the tunica vasculosa lentis in the ninth to eleventh weeks of intra - uterine growth. (Řeháková, Tereza and Stepanov, Alexandr and Jiraskova, 2017). The tunica vasculosa lentis's anterior section happens at the same time as the capsule begins to form, and it is directly linked to the evolution of the stroma of iris, then produces the "pupillary membrane." (Roberts et al., 2014)

Starting about fourteen weeks of pregnancy, this system is subject to natural retraction when a lens has completed and no more needs oxygen and nourishment. (McLeod et al., 2012).

The VHP is the first part of the hyaloid vasculature to regress, followed by the TVL, pupillary membrane, and then the hyaloid stalk. (C. Chen et al., 2019)

Any irregularities in the involution path might result in various clinical symptoms of PFV, which is caused by partial or full vascular regression failure.

The *PFV* is the abnormality of eye congenital, it has many different names which include persistent hyperplastic vitreous (PHPV), persistent tunica vasculosa lentis (TVL), persistent posterior fetal fibrovascular sheath of the lens, ablation falciform as well as congenital retinal septum. There are several pathological signs that related to it incorporate tiny congenital eye, cataracts, spontaneously retina bleeding, falciform folds of retina, detachment of retina as form stalk or funnel and glaucoma. (C. Chen et al., 2019)

Categorized as either anterior, posterior or both. The anterior PFV, vestiges of the tunica vasculosa lentis unable to retreat, resulting in a retrolental fibrovascular membrane which might obscure the vision axis and trigger early cataract creation. (<https://EyeRounds.org/cases/270-persistent-fetal-vasculature.htm>).

The anterior PFV include: Iridohyaloid blood vessels, Normally the vision is not impacted. These iridohyaloidal vessels result in existing of radial vessels extending on the iris's superior surface or appearing of the rings of hairpins on the collarette. In a few situations, a deformity of the limbic connector tissue might be found in the identical meridian.

Retrolental membrane: It's resulted from the posterior tunica vasculosa lentis failing to retreat. The PHPV is the classic name of it. It is distinguished by membranous fibers situated in the area of the retrolental. The retrolental membrane is usually pinkish or white in colour. The retrolental membrane's space differs extensively. In little circumstances, it could be as little as a spot, while in others, it may fill the whole of the lens posterior surface. The lens by itself fluctuates from fully transparent to extremely

opacified. It could appear the lengthened ciliary processes, and this is because of the spread and the concentric traction of the residual of the TVL posterior. (C. Chen et al., 2019).

And also, Persistence pupillary membranes (mentioned later in the point 1.2.1.7).

The posterior PFV: the hyaloid artery fails to degrade, resulting in a fibrovascular stalk connecting the retina and vitreous. (<https://EyeRounds.org/cases/270-persistent-fetal-vasculature.htm>). Other manifestations comprise hypoplasia of the optic nerve, detachment of the retina, proliferative of the retinal membrane, folding of retina.(C. Chen et al., 2019)

The most prevalent form is PFV which includes the anterior and posterior portions: it can lead to the opacity of the cornea, hemorrhage intra-ocular spontaneously, haze of cornea or secondary closed-angle glaucoma. If this situation isn't cured, it will ultimately lead the eyesight to no perception of light. (C. Chen et al., 2019)

1.2.1.4. PERSISTENCE OF THE HYALOID ARTERY

The Persistence Hyaloid Artery (PHA): is defined as the hyaloid artery's inability to regress, which can be partially or fully. It is an uncommon fetal residual that occurs during the 7th month of pregnancy due to developmental anomalies. The hyaloid artery's residual in the anterior part is termed Mittendorf's dot, the residual in the posterior part is termed Bergmeister's papilla (Jeon et al., 2019). It's a congenital condition, an infrequent with an unidentified etiology. The majority of them can be found in a microphthalmic eye. The Persistence hyaloid artery also named as (PHPV) persistence hyperplastic vitreous primary or the persistent fetal vasculature. It is not related to familial history. Typically, it can be detected within the 1st year of age. Usually, this condition is unilateral, however until to 11percent of situations, it can be bilateral, and it is accompanied with microphthalmos in two-thirds of patients. (Borbolla-Pertierra et al., 2014). Leukocoria is a common sign, (Borbolla-Pertierra et al., 2014) (Řeháková, Tereza and Stepanov, Alexandr and Jiraskova, 2017); so, it must be distinguished during the diagnosis such as (retinopathy of prematurity, retinoblastoma, cataract, (Borbolla-Pertierra et al., 2014). (Řeháková, Tereza and Stepanov, Alexandr and Jiraskova, 2017) and posterior uveitis, Coat's disease (Borbolla-Pertierra et al., 2014), and retinal dysplasia. (Řeháková, Tereza and Stepanov, Alexandr and Jiraskova, 2017). About 99% of babies born prematurely are affected by it and 3% of people are born on time. (Řeháková, Tereza and Stepanov, Alexandr and Jiraskova, 2017).

Complications: that are related to this defect such as amblyopia, nystagmus, strabismus (Řeháková, Tereza and Stepanov, Alexandr and Jiraskova, 2017)(Bjeloš et al., 2020), cataract (Jeon et al., 2019)(Bjeloš et al., 2020), Hemorrhage of vitreous (Jeon et al., 2019)(Bjeloš et al., 2020) and detachment retinal (Gandorfer et al., 2005)(Bjeloš et al., 2020).Traction vitreo-retinal (Řeháková, Tereza and Stepanov, Alexandr and Jiraskova, 2017).

Řeháková, Tereza and Stepanov, Alexandr and Jiraskova, reported two conditions of persistent hyaloid artery, they examined the fundus and discovered fibrosis pre-retinal, which caused local tractional retinal detachment (TRD) in one case and local retinoschisis in the other. (Řeháková, Tereza and Stepanov, Alexandr and Jiraskova, 2017)

Only two cases of vitreoretinal traction were found by Gandorfer et al., in a group of nearly 20 individuals with persistent hyaloid artery. A traction retinal detachment involving the macula was found in one patient near the optic nerve disc. The other patient exhibited retinal detachment but no traction maculopathy. (Gandorfer et al., 2005)

Persistent hyaloid artery could be diagnosed by Computed tomography, and magnetic resonance imaging Ultrasound A-B (Borbolla-Pertierra et al., 2014). According to prior publications, the Doppler ultrasound, fundus photography, fluorescein angiography (FAG), and optical coherence tomography (OCT) were utilized to assist in the evaluation bloodstream of this defect. (Jeon et al., 2019)

Jeon et al. described that OCT angiography will become a beneficial non - invasively method to verify the functional status of the persisting hyaloid artery. (Jeon et al., 2019)

Bjeloš et al. recommended that (FA) be conducted and that if contact with the retinal artery is found, fathers be told of the potentially grave consequences. (Bjeloš et al., 2020)

Treatment: for mild conditions, no is necessary surgery but in the severe conditions that lead to destroying the vision pars plana vitrectomy (Řeháková, Tereza and Stepanov, Alexandr and Jiraskova, 2017)

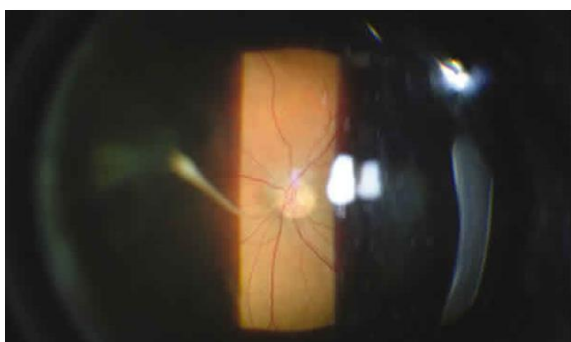


Image 4.a: Persistence of the hyaloid artery
(Both extracted from Centro de Oftalmología Barraquer)



Image 4.b: Persistence of the hyaloid artery

1.2.1.5. *BERGMEISTER'S PAPILLA*

It is a congenital rare alteration that happened to the optic nerve's disc (Rossi et al., 2020), constituted by the remnants of persistent hyaloid artery appearing as an epipapillary membrane which could cover a portion or the whole of optic nerve's disc, rendering it complicated for seeing and studying. Otto Bergmeister, an Austrian ophthalmologist, was who discovered it in the final quarter of the nineteenth

century. It is generally a harmless condition and does not have medical implications. However, it can generate diagnostic suspicions. It usually occurs unilaterally. (Santos-Bueso et al., 2015).

The vision is normal, but in serious situations, might be correlated to amblyopia, alterations pigmentary, tractional vitreomacular (Ray, Prajnya, Shruthy Vaishali Ramesh, 2021), cataract, microphthalmia, persistence of the primitive vitreous (Rossi et al., 2020) (Ray, Prajnya, Shruthy Vaishali Ramesh, 2021), in some cases tractional retinal detachment, hemorrhages in vitreous (Rossi et al., 2020). Vitreous thickening in the peripapillary area should be closely monitored since it can develop to vitreoretinal adhesive, which causes subsequent problems (vitreomacular traction, tractional retinal detachment). It is assessed by OCT will shed insight on the necessity for close surveillance and, if necessary, treatment. Further monitoring is not required in situations in which there is no traction; however, situations with vitreoretinal adhesion and traction require careful observation. (Ray, Prajnya, Shruthy Vaishali Ramesh, 2021).

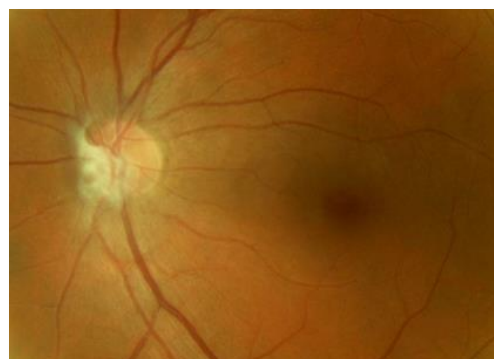
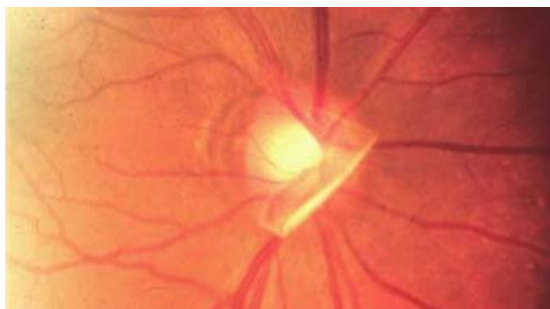


Image 5 a & b: Bergmeister's papilla, (Both extracted from Centro de Oftalmología Barraquer)

1.2.1.6. MITTENDORF DOT

Perhaps seen as a slight kind of (PHPV) persistent hyperplastic primary vitreous. It's positioned at the capsule posterior of the lens and manifests as a spot of tiny white opacity in the nasal region along the axis. It does not affect eyesight. (Amaya et al., 2003) (Czepita et al., 2018).

It is usually linked to the lenticonus posterior. It's sometimes related to a persistent hyaloid artery, which is similar to PHPV- feature. The surgical intervention is not necessary, but when it becomes advanced, which is uncommon, it must be noticed. it constitutes the remnants of the hyaloid artery's anterior end. frequently accompanied by a fine string. In addition to being referred to as a hyaloid body. (Amaya et al., 2003)



Image 6 a&b: Mittendorf Dot, (Both extracted from Centro de Oftalmología Barraquer)

1.2.1.7. PERSISTENCE PUPILLARY MEMBRANES

These membranes stand out as thin threads of iris around the pupil. They differ considerably in size, shape, composition, and thickness also might be unilateral or bilateral. They can become connected to the cornea's posterior surface, or the lens' anterior capsule, producing a tiny cataract. (Gavriş et al., 2015), usually seen in babies, but they are more frequent in premature neonates. They represent the vestiges of anterior tonica vasculosa lentis, which start to fade around the sixth month of pregnancy and vanish entirely by the eighth month. If they exist after delivery, they still retreat during the first year of life. Nevertheless, a thick membrane may exist in some situations, needing therapy. Ordinarily, they do not influence eyesight, but the vision is impaired in situations when the opening of the pupil is less than 1.5 mm, so that leads to reduced lighting of the retina and deviation. Depending on the size of the pupil's aperture and the membrane's size, treatment can be determined. Could be monitored with no treatment needed in conditions of tiny persistent pupillary membranes. Other situations might necessitate utilizing mydriasis, treating refractive error, and treating the lazy eye with patching. To disrupt membranes, the Nd: YAG laser could be performed, but it comes with the danger of causing hyphema and, consequently cataract, so it is because blood vessels are likely to be exist. Surgery removal of the membrane could be useful in some conditions. (Fard et al., 2016)

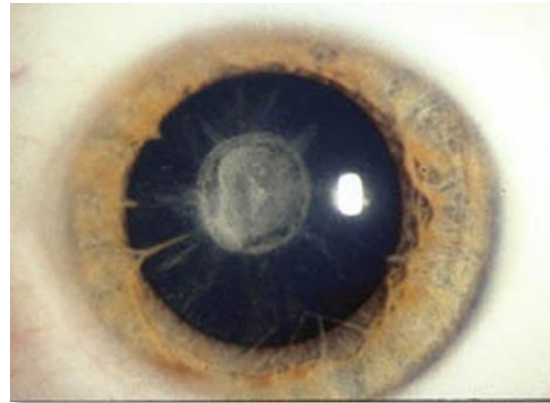
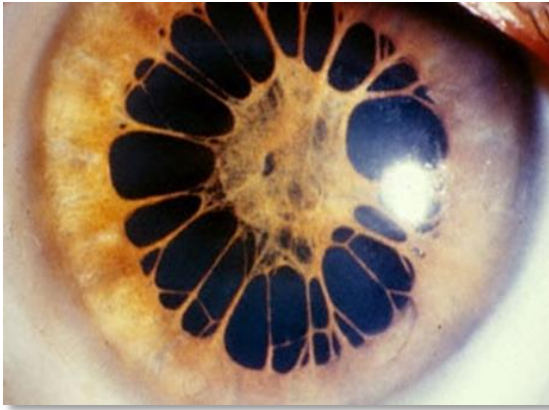


Image 7 a&b: Persistence pupillary membrane

(Both extracted from Centro de Oftalmología Barraquer)

1.2.1.8. *EPICAPSULAR STAR*

Could occur unilaterally or bilaterally and is a fairly popular condition of other residues of the tunica vasculosa lentis (Jick et al., 2019), consisting of brown or golden spots like stars, located on the anterior capsule of the lens. Usually referred to as chicken tracks. (Choudhry et al., 2001) (Deshmukh et al., 2018). The eyesight can be affected when the epicapsular stars become thick in the centre. Could be treated by a neodymium YAG laser (Nd: YAG) or when they're not attached to the anterior capsule of the eye lens, they can be surgically removed. (Choudhry et al., 2001)

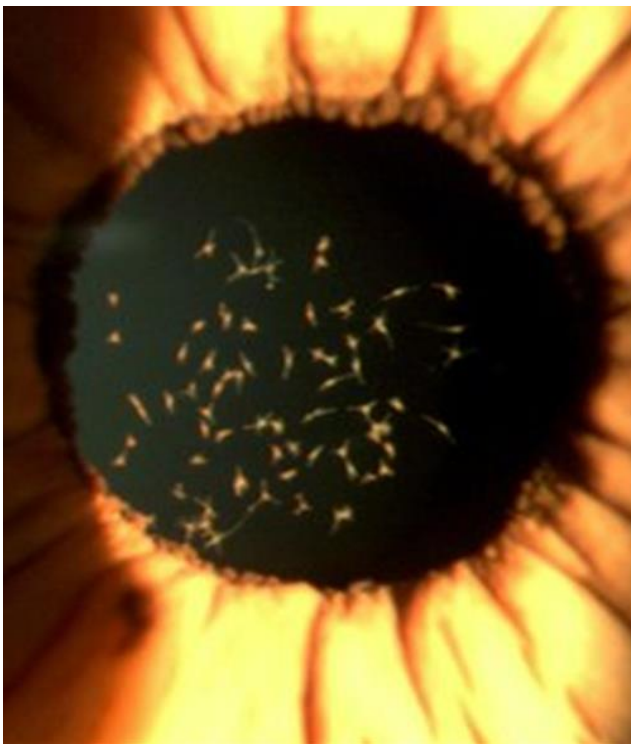


Image 8: Epicapsular star, (extracted from Centro de Oftalmología Barraquer)

1.2.2.ALTERATIONS IN SHAPE AND STRUCTURE

1.2.2.1. COLOBOMA

An uncommon neonatal illness. (Wang & Ma, 2015). It is a defect in the appearance of the lens. It has two types; Primary is a wedge-shaped disorder or notch of the lens outer edge that happens as a remoted irregularity. Secondary is a flatten or notch of the lens outer edge resulting from inadequate ciliary body or zonular growth. Generally placed inferonasal. Colobomas of lens generally related to an iris coloboma and with chorioretinal coloboma occasionally. The connections of zonules withinside the area of the coloboma is commonly weaker or missing. opacified lens cortical or capsule of the lens thickened might also additionally seem adjoining to the coloboma. (American Academy of Ophthalmology,Jick, 2019). Surgical remedy can acquire optical improvement. it could be handled with the aid of using elimination of the faulty lens. An intraocular lens might be positioned withinside the capsular bag at the side of a capsular tensor ring or placed withinside the ciliary sulcus. (Wang & Ma, 2015)

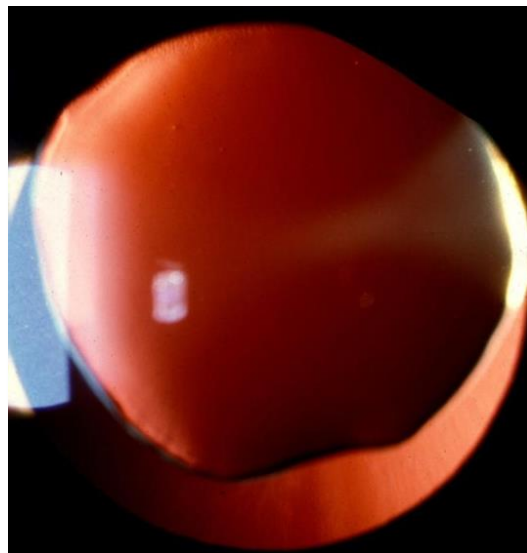


Image 9: a & b: Coloboma , (Both extracted from Centro de Oftalmología Barraquer)

1.2.2.2. LENTICONUS

A conical deformity located in the lens's anterior or posterior surface. Via retinoscopy the middle of the lens demonstrates a deformed and reflex of myopia in both anterior and posterior types. By red reflex both malformations could be viewed, however through retro -illumination they look like "oil droplet". (American Academy of Ophthalmology,Jick, 2019)

1.2.2.3. *ANTERIOR LENTICONUS*

75 percent of cases, the anterior lenticonus is bilaterally and occurs between the ages of 20 and 30, alongside indication of increasing myopic lens. Related to Alport syndrome. It is a hereditary disorder that comes in the form of X-linked, recessive autosomal or dominant autosomal. (Nihalani, 2014). It is present in approximately 10% of younger children impacted. Less frequent unlike the posterior type. Might be congenital. Could be a sign of an abnormality of the basement membrane. Lowe's and Waardenburg's syndromes have also been linked to anterior lenticonus. Generally, not related to cataracts. Despite the absence of lens opacities, severe astigmatism as a consequence of lenticonus can have a significant impact on vision, necessitating surgery. (Amaya et al., 2003)

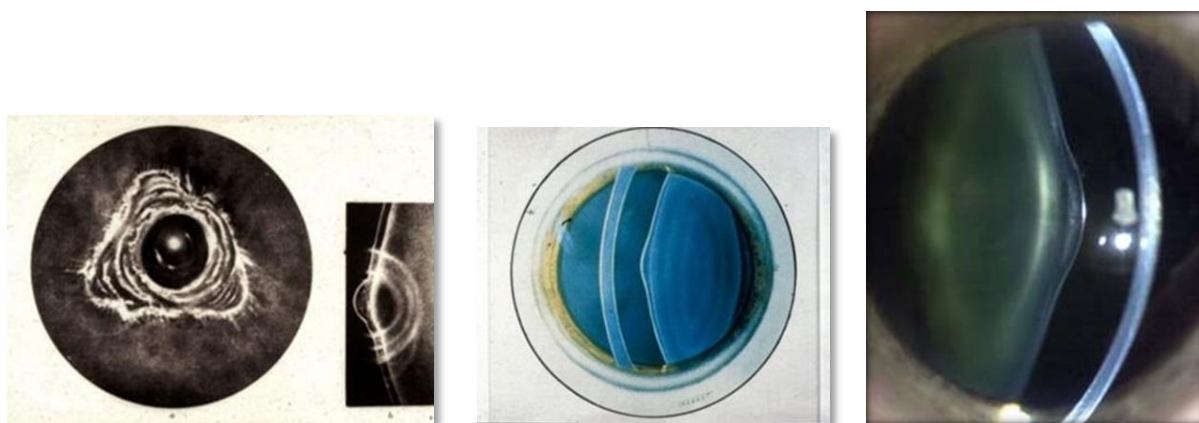


Image 10: a, b & c: Anterior lenticonus, (extracted from Centro de Oftalmología Barraquer)

1.2.2.4. *ALPORT SYNDROME*

An advanced inherited disorder of the glomerular basement membrane, that occurred due to the mutations in genes COL4A3,4,5, that encode the type IV collagen alpha 3 to 5 chains. It could be transferred as X-linked dominantly, autosomal dominant, or autosomal recessive. It could be transferred as dominant X-linked, autosomal dominant, or autosomal recessive. It is often related to anomalies in hearing and sight. Alport syndrome patients, 85% of them have X-linked type caused by a mutation in the gene COL4A5, which encodes the $\alpha 5$ chain of type IV collagen. Men with X-linked Alport syndrome are significantly more afflicted than women. Almost fifteen percent of patients with Autosomal recessive Alport syndrome (compound heterozygous or homozygous) are impacted by mutations of genes COL4A3 or COL4A4, that corresponded respectively to the collagen IV $\alpha 3$ and $\alpha 4$ chains. Alport syndrome autosomal dominant is extremely rare, resulting from the mutations in the genes COL4A3 or COL4A4.

The main symptoms include renal insufficiency, hematuria, and proteinuria. Extra-renal symptoms are deafness, visual problems, and spread leiomyomatosis.

The common eye defects involve degeneration of cornea, flecks retinopathy, and anterior lenticonus. The peripheral fleck retinopathy is the most usually observed ocular alteration.

Another ocular anomalies that could happen in Alport syndrome patients; comprise lamellar or partial-thickness macular holes and cataracts. Macular holes and cataracts can both cause loss of vision. In Alport syndrome, macular holes often lead to loss of vision permanently due to their nonwell response to surgical therapy. (Zhang & Ding, 2018)

1.2.2.5. *POSTERIOR LENTICONUS*

It's generally isolated and has no recognized cause. It is usually unilaterally; nevertheless, there were instances of inherited cases where it has been discovered at being bilaterally including an autosomal dominant inheritance or maybe X-linked. On the posterior side of capsule, it resembles a droplet of oil. there was a case in a kid has chromosomal translocation and revealed that has bilateral posterior lenticonus which caused by mutation of TDRD7 gene, that is correlated with posterior capsule. (Nihalani, 2014)



Image 11: Posterior lenticonus, (extracted from Centro de Oftalmología Barraquer)

1.2.2.6. *LOWE SYNDROME*

Lowe's oculocerebrorenal syndrome is related to X-chromosome multiple body system sickness described as a triple of congenital cataracts, acute mental impairment, and kidney tubes malfunction causing gradual progress of nephrotic failing. Further symptoms include postpartum developmental

delays unrelated to renal function, areflexia, nontender joint puffiness, subdermal nodulars, and joint pathology, which affects around half of all adult patients. The cause of this syndrome is variations inside the gene OCRL that is located on the Xq25-26 chromosome. It is encoding OCRL-1 (an inositol polyphosphate 5-phosphatase). It is unusual condition, so its incidence is about one in half a million in the public at large, according to the American and Italian Associations of Lowe's Syndrome (LSA) (AISLO) remarks. Their ages are seldom more than the age of forty.

Other ocular symptoms: approximately half of them have acute glaucoma accompanied by buphthalmos, that is needing urgent surgery; it is generally noticed at the age of one year; however, it may occur later in the second or third ten years. After they turn five, roughly twenty-five per cent of individuals evolve scar on the cornea and keloids, ordinarily with no antecedent injury. Because of the retina main malfunction, it is uncommon that their vision with a correction exceeds 0.2. In an embryonic stage, the cataract develops because the main posterior lens fibres are not formed correctly and then deterioration follows. (Bökenkamp & Ludwig, 2016)

1.2.2.7. MICROSPHEROPHAKIA

A state in which an uncommon inborn illness that frequently affects both eyes. (Yu et al., 2020). The lens size is fairly tiny; circular. (Wright, 2003) (Nihalani, 2014) (Yu et al., 2020). And thick. (Wright, 2003) (Yu et al., 2020). Once the pupil is completely expanded, the whole eye lens equatorial is seen. (Yu et al., 2020). its repercussions Myopia (Wright, 2003) (Nihalani, 2014) (Yu et al., 2020), lens displacement or subluxation, glaucoma, and impaired accommodation. (Nihalani, 2014) (Yu et al., 2020). Degeneration of the tapetoretinal. (Nihalani, 2014). It can exist by itself or in conjunction with the Weill Marchesani syndrome (most commonly). might be found in hereditary abnormalities that are autosomal dominant or autosomal recessive. (Nihalani, 2014) (Yu et al., 2020). Moreover, Marfan syndrome, Alport syndrome, Klinefelter syndrome. (Yu et al., 2020). to cure myopic (spectacles contact lenses),

Lens dislocation (surgery recommended in case of damaged zonules, other therapies such as dilator of pupil, hyperosmolarity therapy makes the vitreous shrunken, and supine position, so all of these can be used if the zonule is unharmed), Glaucoma (early laser peripheral iridotomy is frequently indicated, anti-glaucomatous drugs). (Yu et al., 2020)

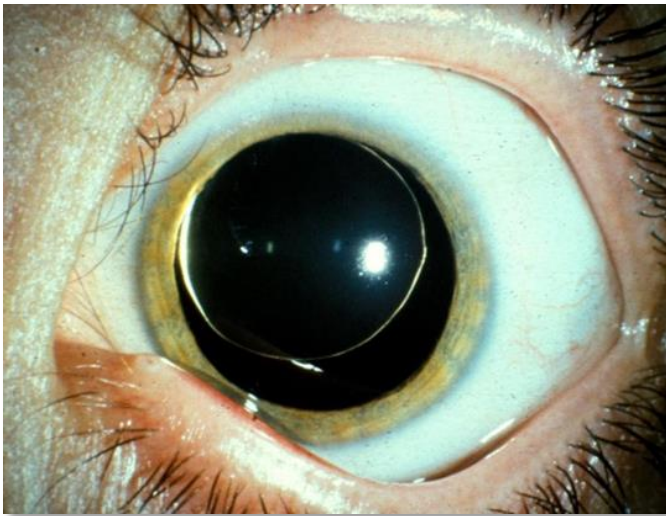


Image 12: Microspherophakia, (extracted from Centro de Oftalmología Barraquer)

1.2.3.ALTERATIONS TO THE POSITION

1.2.3.1. *SUBLUXATION, DISLOCATION, ECTOPIA LENTIS*

Uncommon disease (Simon et al., 2015), occurs when the lens is displaced from its typical location in the middle of the pupil. (Wright, 2003) (Piozzi & Del Longo, 2012), as a result of the zonular fibre distortion (Piozzi & Del Longo, 2012) (Simon et al., 2015). It could be partially (sub displacement) or total (luxation) (Wright, 2003) (Piozzi & Del Longo, 2012).

Subluxation: the lens is dislodged, nonetheless remains inside the pupil area, described too as subluxated. (Simon et al., 2015).

Luxation: The dislodged lens is separated totally from the ciliary body and can be found floating in the anterior chamber or vitreous cavity. (Simon et al., 2015).

Causes: Trauma is responsible for half of all lens dislocations; although can arise in the absent of damage in susceptible eyes since it happens regularly in the situation of disorder or metabolic dysfunction. The basic form of ectopia lentis, is a dominantly autosomic condition. it could be inborn or acquired, ordinarily dislocation of the lens is up and laterally. Ectopia lentis et pupillae is other disorder an autosomal recessive and usually affects both eyes, happens when the dislocation of lens and pupil together, habitually in different directions. Several system illnesses, including Marfan syndrome, Weill–Marchesani syndrome, and homocystinuria represent above 75 percent of documented non-traumatic dislocations of lens. (Simon et al., 2015)

Symptoms: Reduced eyesight, severe astigmatism, unilateral double vision, and iridodonesis. (American Academy of Ophthalmology, Jick, 2019)

Complications: alteration in refraction, lens opacification, pupil blockage, and closed angle glaucoma. (American Academy of Ophthalmology, Jick, 2019)

Management: for all of those patients, there is a range of surgeries available. However, before the surgery, amblyopic cases need to undergo vision therapy and best corrective refractive error must be done for phakic and non-phakic cases. (Simon et al., 2015).

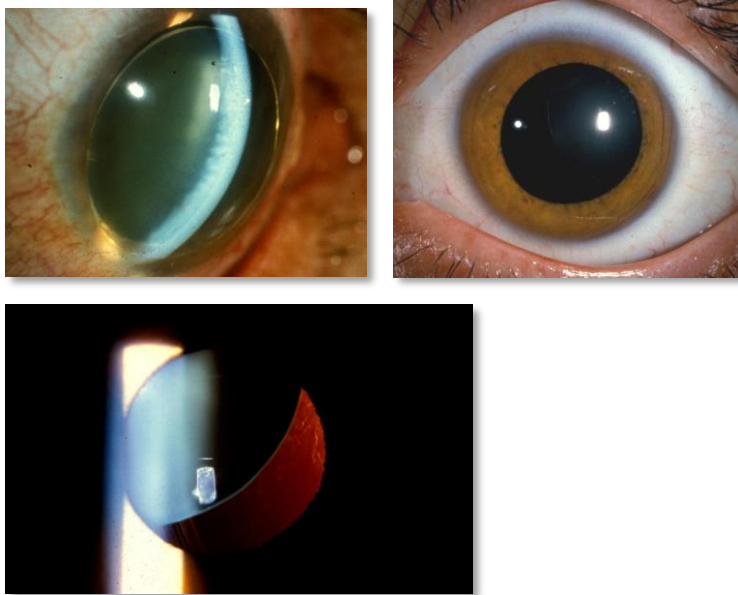


Image13: a,b & c: Subluxation, (extracted from Centro de Oftalmología Barraquer)

1.2.3.2. *MARFAN SYNDROME*

A hereditary condition affecting the bone, joint, cardiovascular also sight (Nihalani, 2014), Associated with autosomally dominance (Simon et al., 2015). The systemic signs of this illness are skinny extremities, long body shape, joints relaxation, and heart anomalies. Ophthalmic symptoms for example shortsightedness, periphery retina deterioration, detached retina, hypertension intraocular, lens opacity. Also, the displacement of the lens often happens bilaterally and symmetrically with a tendency to occur superotemporally in eighty per cent of Marfan syndrome sufferers; half of these instances arise before the child reaches the age of five. (Simon et al., 2015).



Image 14: Marfan syndrome, (extracted from Centro de Oftalmología Barraquer)

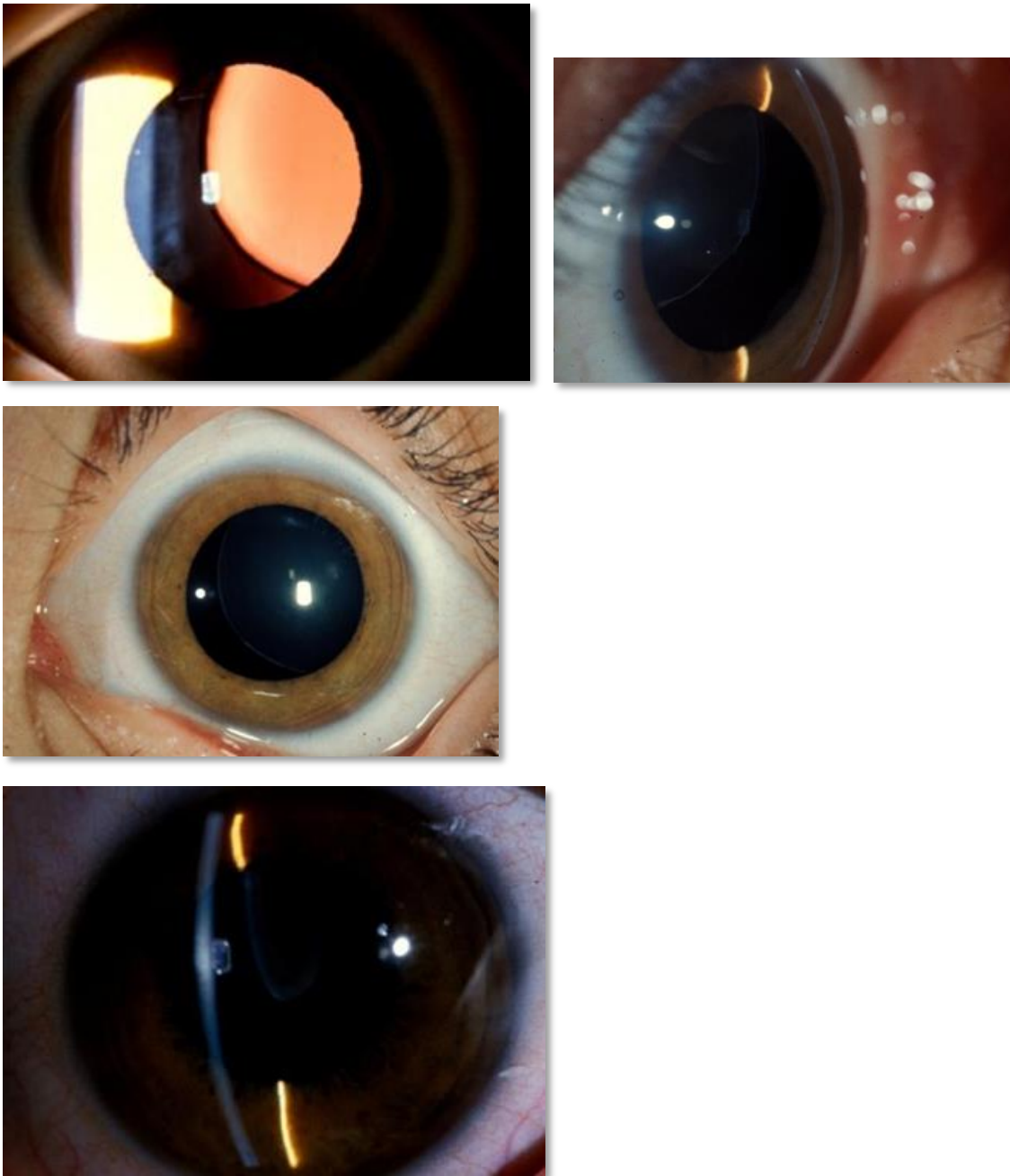


Image 15: a, b, c & d: Subluxation, (extracted from Centro de Oftalmología Barraquer)

1.2.3.3. *WEILL–MARCHESANI SYNDROME*

An autosomic recessive disease. (Simon et al., 2015).

The systemic signs: small size, tiny and thick digits, and a breast that is both little and big. (Nihalani, 2014), around half of individuals suffer a delayed in growth. (Simon et al., 2015). Ocular symptoms:

lens displacement inferiorly, (Simon et al., 2015). microspherophakia (Wright, 2003), short sightedness, glaucoma closed angle owing to anterophris synechia or pupil blockage. (Nihalani, 2014)



Image16: Weill–Marchesani syndrome,(extracted from Centro de Oftalmología Barraquer)

1.2.4.ALTERATIONS OF THE TRANSPARENCY

1.2.4.1. CONGENITAL CATARACT

The congenital cataract is a defect in the lens's transparency that appears in childbirth or a short time later. It's one of the most common reasons for curable blindness or impaired vision among kids. (Tătaru et al., 2020).



Image 17: Congenital cataract, (Extracted from Centro de Oftalmología Barraquer)

1.2.4.2. EPIDEMIOLOGY

The estimation of the WHO is that there are over 14 million children worldwide who have bilateral blindness due to cataracts, representing more than half of all blindness cases. (Tătaru et al., 2020) (Berry et al., 2020). In the United States and other developed nations, there are approximately six cases per

10,000. On the other hand, the incidence rate is predicted to be 15 per 10,000 in undeveloped regions. (Hernandez & Kumar, 2018)

1.2.4.3. THE ETIOLOGY

A large proportion of bilateral cataracts and the majority of unilateral cataracts are considered to be inherently unknown, and these situations are screened after other possibilities are ruled out. (SudarshanKumar Khokhar et al., 2017)

Etiological Classification:

Table 1: Bilateral and unilateral cataracts

Bilateral cataracts	Unilateral cataracts
Idiopathic	Idiopathic
Hereditary cataracts (autosomal dominant)	
Genetic and metabolic diseases	
Down syndrome	
Lowe syndrome	
Galactosemia	Ocular anomalies
Marfan's syndrome	PFV
Trisomy 13-15	Anterior segment dysgenesis
Hypoglycemia	Posterior lenticonus
Alport syndrome	Posterior pole tumours
Myotonic dystrophy	
Fabry disease	
Hypoparathyroidism	
Maternal infection	Traumatic (rule out child abuse)

Rubella	
Cytomegalovirus	
Varicella	
Syphilis	
Toxoplasmosis	
Ocular anomalies	
Aniridia	Rubella (rarely)
Anterior segment dysgenesis syndrome	
Corticosteroids	Asymmetric bilateral cataract

PFV: Persistent fetal vasculature, the table extracted from (Khokhar et al., 2017)

1.2.4.3.1. Hereditary

In congenital cataracts the heredity is noted 8.3–25% of conditions (Khokhar et al., 2017) (S. K. Khokhar & Dhull, 2019), The autosomal dominant heredity type is the most frequent modality of heredity. it's essential to register the history of the family, also for checking for congenital cataracts situations members of the family that are presented to the eye doctor. (S. K. Khokhar & Dhull, 2019). Mutations in genes encoding for proteins involved in lens shape or clarity cause around half of all juvenile cataracts. Most of the genes in this category are autosomal dominant; the remaining are autosomal recessive or Xlinked. Among isolated cataracts without systemic anomaly, the crystalline mutation represents 50% of cases, the connection mutation explains 25%, and the mutations in proteins such as Heat shock transcription factor-4, aquaporin-0, and beaded filament structural protein-2 account for 25%. (Wilson ME, 2015). The result of DNA sequencing tools has demonstrated the precision of genetic analysis in confirming Congenital Cataract in approximately 75% of patients, and 85% of patients with nonsyndromic Congenital Cataract have the likely pathogenic mutations. In addition, 63% of possible mutations linked to syndromic Congenital Cataracts were detected in these patients. (Gillespie et al., 2014). The pathologies that are related to the genetic cataract including: Norrie disease, Nance–Horan syndrome, Down's syndrome and Lowe syndrome (Khokhar et al., 2017), as well as, frequently related with myotonic dystrophy Fabry disease and Alport syndrome. (Nadeem et al., 2013).

The existence some of the interference between cataracts related systemic illnesses and those related-metabolic illnesses, for this they are included jointly in Table2. (Nihalani, 2014).

Table 2: Systemic disorders linked to cataracts. AD= Autosomal dominant; AR= Autosomal recessive; XD= X-linked dominant; XR= X-linked recessive, the table extracted from (Nihalani, 2014)

Systemic disorder	Chromosome (gene)	Type of cataract
Alpha-mannosidosis (AR)	9 cen (<i>MAN2B1</i>)	Punctate spoke like opacities
Alport (X-linked, AR, AD)	Xp22 (<i>COL4A5, COL4A6</i>)	Anterior lenticonus, spherophakia
Bardet-Beidel (AR, digenic recessive)	<i>BBS1-15</i>	Zonular
CHARGE association	<i>CHD7</i>	
Cerebrotendinous xanthomatosis (AR)	2q33 (<i>CYP27A1</i>)	
Chondrodysplasia punctata (AD, XR, XD)	6q22, Xp22, (<i>CPDX2, PEX7</i>)	Xp11
Cockayne (AR)	10q11, 5q12 (<i>ERCC</i>)	
Fabry (X-related)	Xq22 (<i>GLA</i>)	Anterior capsular or spoke-like posterior subcapsular
Galactosemia (AR)	9p13 (<i>GALT</i>)	Oil droplet/posterior subcapsular
Hallermann-Steriff (isolated)	6q21 (<i>GJAI</i>)	Spontaneous lens absorption
Hyperferritemia (AD)	19q13 (<i>FTL</i>)	Pulverulent
Hypoparathyroidism (AD)	Multiple (<i>GCM2</i>)	Multicolor flecks

Incontinentia pigmenti (XD)	(XD) Xq28 (<i>IKBK</i>)	Appear like PFV
Lactase deficiency (AR)	2q21 (<i>LCT</i>)	
Lowe's syndrome	oculocerebrorenal (XR) Xq26 (<i>OCRL</i>)	Male: thin discoid Female: fine cortical punctate
Myotonic dystrophy (AD)	9q13, 3q13 (<i>DMPK, ZNF9</i>)	Polychromatic, posterior, subcapsular
Nance-Horan (XR)	Xp22 (<i>NHS</i>)	Male: fetal nuclear and stellate Female: Y sutures
Neurofibromatosis II (AD)	22q12 (<i>NF2</i>)	Posterior subcapsular
Norrie (XR)	Xp11 (<i>NPD</i>)	
Smith-Lemli-Opitz (AR)	11q12 (<i>DHCR7</i>)	
Stickler syndrome (AD)	12q13, 1p21 (<i>COL2A1, COL11A</i>)	
Sturge-Weber síndrome	<i>GNAQ</i>	Nuclear
Trisomy 13 (Turner syndrome)	Trisomy 13	
Trisomy 21 (Down syndrome)	Trisomy 21	Cerulean
Usher (AR)	Multiple (<i>USH</i>)	
Waardenburg (AD)	Multiple	Anterior polar
WAGR (AR or somatic)	11p13 (del of <i>WT1</i> and <i>PAX6</i>)	Anterior pyramidal
Wilson disease (AR)	13q14 (<i>ATP7B</i>)	Sunflower
Zellweger (AR)	12 genes	Cortical

1.2.4.3.2. *Metabolic*

Galactosemia: it occurs when mutations occur in uridine diphosphate 1–4, galactokinase (GALK1), or galactose 1-phosphate uridylyltransferase, epimerase, which result in the galactose's level being unusually elevated. The lack of Galactokinase leads the cataract to be mild, moreover is linked to intellectual disabilities, growth failure, and icterus. When galactitol builds up; induces the lens osmotic damage, resulting in "oil droplet cataract", such these alterations can be reversed at the beginning through omitting the galactose from dietary. Also, can occur as nucleolar cataract as well as anterior or posterior subcapsular cataract. (Khokhar et al., 2017). Multicolour fleck cataracts are caused by hypoparathyroidism, vacuoles are linked to mannosidosis, and lamellar cataracts are a result from hypoglycemia. It has also been found that congenital cataracts are related to copper metabolism abnormalities. (Krishnamurthy & VanderVeen, 2008). Another diseases including diabetes, Wilson disease, and hypocalcaemia. (Hernandez & Kumar, 2018).

1.2.4.3.3. *Associated with infections*

Intrauterine inflammation is related to an elevated danger of developing cataract, such as Toxoplasmosis, Rubella, Cytomegalovirus, and Herpes. In addition, prenatal varicella is largely related to produce the cataract. (Morrison, 2017). Also, the possibility to occurring congenital cataracts increases 51% when the body's temperature increases by five degrees during the pregnancy phase. (Hernandez & Kumar, 2018).

1.2.4.3.4. *Corticosteroids*

As anti-inflammatory agents, corticosteroids are extremely useful for suppressing ocular inflammation. If they are advised, they should be given for the shortest time possible and side effects should be observed. (BECKER, 1964). Corticosteroid usage for a very long-time time can result in a posterior subcapsular cataract, the incidence of which varies depending on the intake and length of therapy. Various entry pathways of corticosteroids including systemic, topical, subconjunctival, and inhalation can lead to cataracts, and so if a patient receives systemic corticosteroid treatment superior than 15 mg/d of prednisone for over one year, more than 80% of patients will develop cataracts. When it comes to the topical method, it might occur after using corticosteroid dermatological formulations around the eyelids. (Gómez de Liaño et al., 2012)

1.2.4.3.5. *Traumatic cataract*

It is considered as the opacity of the lens permanently that manifests as a separate change after prolonged period of the eye injury (Zimmermann et al., 2019), where caused mostly by the injury of bow and arrow, for this, the trauma to the eye opened is more commonly three times than trauma to eye

closed. Also, among other causes of injury include metal, wood, stone, ball and firecrackers. (Khokhar et al., 2017). It is the main cause of blindness in children unilaterally; next to that, Its frequency accounts for 30% of childhood and adolescence in America and other developing nations, and it represents 41% of those aged 6 to 15.(Zimmermann et al., 2019).Accompanied commonly by scar or hole in the cornea, hyphema, shallow in the anterior chamber, deformation in the iris, bleeding in vitreous, tears in the capsular posterior, posterior synechiae, and vitreous in the chamber anterior. (Khokhar et al., 2017).

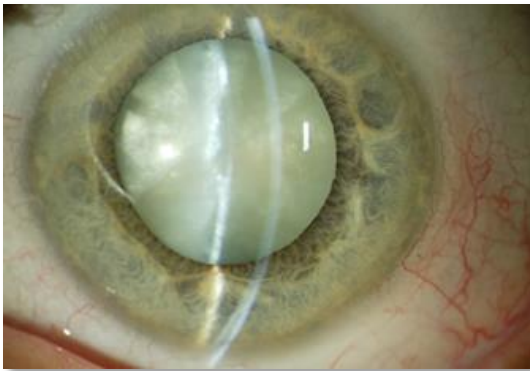


Image18: Traumatic cataract,(extracted from Centro de Oftalmología Barraquer)

1.2.4.3.6. *Uveitic cataract*

71 % of children with uveitis cataract suffer from juvenile idiopathic arthritis (JIA), which can contribute to severe intraocular inflammation which leads to uveitis cataracts in children. A posterior subcapsular cataract usually occurs in these patients as well as a posterior synechiae, iris bombe, in addition to peripheral anterior synechiae. However, bandshaped keratopathy and hypotony in cases of long time JIA. (Khokhar et al., 2017).



Image 19: Uveitic cataract, (extracted from Centro de Oftalmología Barraquer)

1.2.4.4. CLASSIFICATION MORPHOLOGICALLY

1.2.4.4.1. The Entire lens

1.2.4.4.1.1. Total Cataract

Opacity occurs in whole of the lens; most cases are bilateral and sporadic, though familial cases have also been reported. It might be caused by cataract advancement from zonular cataract, posterior lenticonus, or a preexisting posterior capsular defect. (S. K. Khokhar & Dhull, 2019) (Amaya et al., 2003). In addition to, the operation intervention is obligatory and directly because this type of cataract affects on the vision. (Amaya et al., 2003)

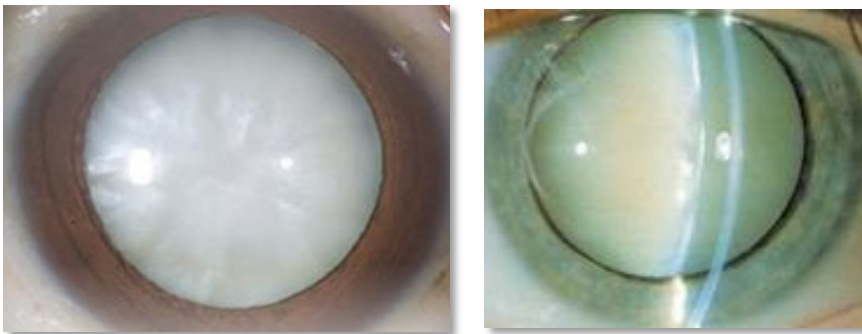


Image 20: a & b :Total cataract, (extracted from Centro de Oftalmología Barraquer)

1.2.4.4.1.2. Congenital Morgagnian Cataracts

In 1762, Giovanni Morgagni mentioned these rare, thick, completely opaque cataracts. (Amaya et al., 2003). It is when the nucleus remains undamaged; the lens's outer zones liquefy, permitting the nucleus to drop owing to the gravity in any direction, based on where the head is positioned.(Patil, 2015)(Amaya et al., 2003) , (the nucleus, which seems to be sunken)(S. K. Khokhar & Dhull, 2019).Eventually the fluid may be reabsorbed so anterior and posterior capsules adhere above the displaced nucleus or sometimes they can even completely reabsorb (Amaya et al., 2003)



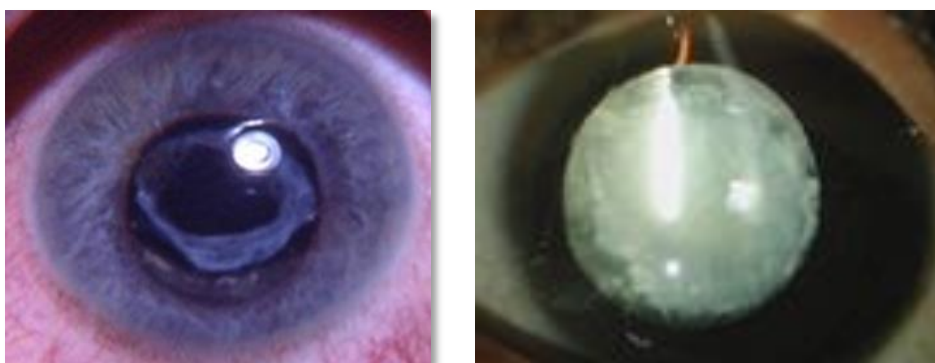
Image 21: Morgagnian Cataract (the image extracted from (S. K. Khokhar & Dhull, 2019))

1.2.4.4.1.3. Membranous Cataract

By permitting the merge of posterior and anterior capsules together and that is a result of re absorbance of the lens leaving either a disk of opaque material (or a thin disk of transparent material). (Amaya et al., 2003) (Patil, 2015) (American Academy of Ophthalmology, Jick, 2019).

Also, this form of opaqueness and deformation of the crystalline are responsible for vision impairment. (American Academy of Ophthalmology, Jick, 2019).

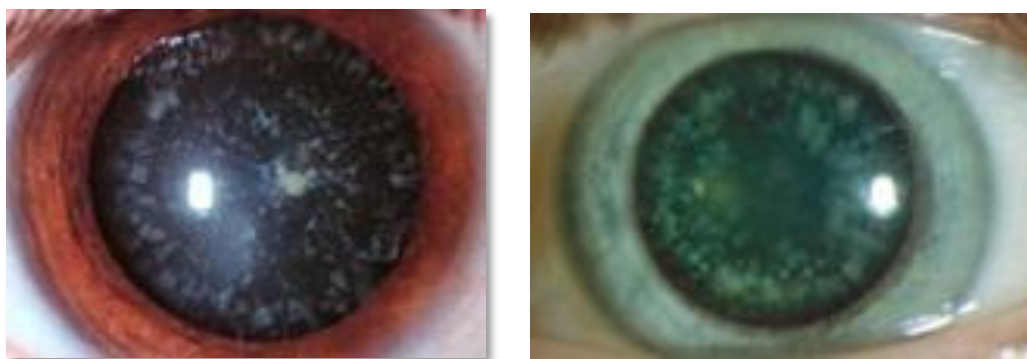
The membrane might be rather extremely dense in some cases. Ultrasound bio microscopy might be beneficial in determining the technique of surgery; because in these instances are more complicated, and intraocular lenses would not be simply introduced in the eye. (Amaya et al., 2003)



Images 22: a & b: Membranous Cataract, (extracted from Centro de Oftalmología Barraquer)

1.2.4.4.1.4. Cerúlean Cataracts

They appear as a tint of green or blue, bilateral (Amaya et al., 2003) (S. K. Khokhar & Dhull, 2019). on the microscopic slit-lamp, could be a lot of beauty. may also seen other hues such as brown and bright white resemble the crumbs of bread or red as is the case of hyperferritinemia. (Amaya et al., 2003). mostly the vision not damaged, termed as blue dot cataract, not advanced (S. K. Khokhar & Dhull, 2019). They are significantly steady, usually concentrated in the lens's equatorial area and have a shape dot-like with varying sizes, occasionally extended. The cataract visibly is inconspicuous, located in the lens's peripheral cortex. an autosomal dominant, early initiation, the genetic mapping of the cerulean cataract phenotype has been identified at 17q242 and 22q11.2-q13.1. (Amaya et al., 2003)



Images 23: a & b: Cerulean cataract ,(extracted from Centro de Oftalmología Barraquer)

1.2.4.4.2. Central Cataract

1.2.4.4.2.1. Nuclear

An opacification in the fetal or embryonal nucleus. (S. K. Khokhar & Dhull, 2019) (Ellis, 2002) (Amaya et al., 2003). Commonly, they are visually disturbing. (Ellis, 2002) (S. K. Khokhar & Dhull, 2019), typically bilateral (Amaya et al., 2003) (S. K. Khokhar & Dhull, 2019) and may be inherited autosomally dominantly (Amaya et al., 2003) (S. K. Khokhar & Dhull, 2019) which are the commonest form of bilateral nuclear cataract (Amaya et al., 2003).

Also, it is known that congenital rubella syndrome is related to nuclear cataracts, (S. K. Khokhar & Dhull, 2019) in which was described as dense nuclear opacities encircled by a comparatively pure cortex. (Amaya et al., 2003).

In developing countries, they are prevalent. Can occur insulated or in conjunction with cortical or zonular cataract. (S. K. Khokhar & Dhull, 2019).

They can be merged with suture opacifications. Mostly, they are merged with opacified cortical fibres that surround the nuclear opacity and that termed as riders. (Amaya et al., 2003).

Opacities of nuclear dominantly autosomal are connected with a greater danger of developing aphakic glaucoma post-surgery of cataract and could be linked to microphthalmos. (Amaya et al., 2003)

It is critical to identify these opacifications early life and operating during the initial few weeks to achieve the best potential visual result. (Ellis, 2002).

If the eyesight is intermediate, careful therapy is recommended to avert post - operative problems in some patients. Surgery, on the other hand, is required if vision is significantly damaged. (Amaya et al., 2003)

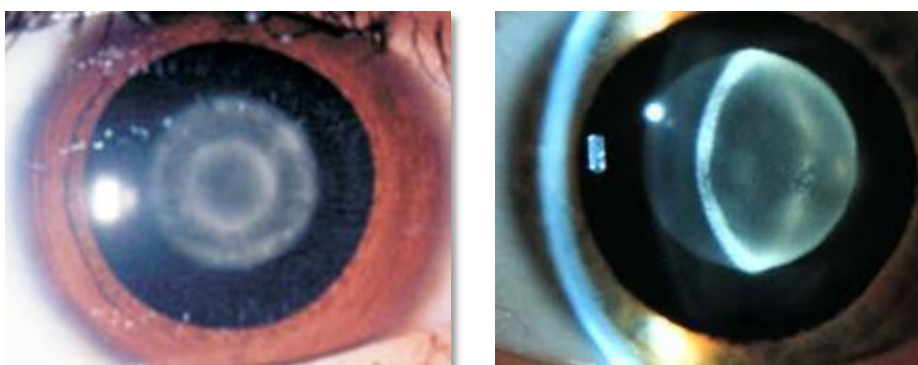


Image 24: a & b: Nuclear cataract, (extracted from Centro de Oftalmología Barraquer)

1.2.4.4.2.2. Sutural

These opacifications are existed in the sutures of the lens's fetal nucleus (Ellis, 2002) (Amaya et al., 2003) (S. K. Khokhar & Dhull, 2019); sutures like a Y-shape. (Ellis, 2002) (S. K. Khokhar & Dhull, 2019). The posterior more than the anterior. (Amaya et al., 2003). Usually are static, familial and are quite commonly. Mostly, during regular checks, they are noticed as an incidental outcome. (Amaya et al., 2003).

Typically, bilateral, nonsignificat visually, not advanced (Amaya et al., 2003) (S. K. Khokhar & Dhull, 2019); however, the intensity of sutural cataracts may differ (S. K. Khokhar & Dhull, 2019), that alters in their denseness as does their effect on eyesight and optical evolution. (Ellis, 2002)

They can be hereditary as x-linked recessive or an autosomal dominantly, and they were diagnosed in female with Nance-Horan syndrome carriers, in males impacted, having entire congenital cataracts. Sutural cataracts are occasionally the lone sign included in families without symptoms. Stellate cataracts are described as the opacity of the anterior and posterior sutures. Cataract triradiata is a condition in which all three sutures are evenly impacted. (Amaya et al., 2003).

They can vary in appearance from increasing in thickness of sutures to a diversity of cerulean or white spots grouped surrounding the sutures, nevertheless they can develop to produce central or nuclear cataracts. (Amaya et al., 2003). They can be linked to pulverulent, nuclear, or lamellar cataracts, that might necessitate surgery. (S. K. Khokhar & Dhull, 2019).

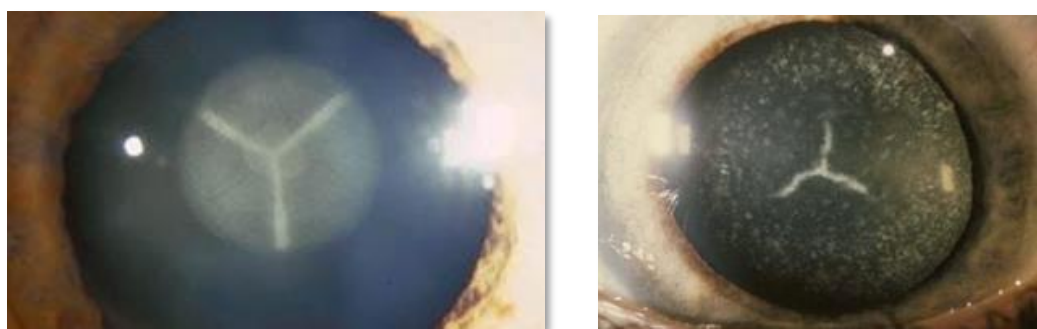


Image 25: a & b: Sutural cataract,(extracted from Centro de Oftalmología Barraquer)

1.2.4.4.2.3. Zonular

The commonest shape, which include one or more of lens's areas or layers. (Patil, 2015) (Amaya et al., 2003) (S. K. Khokhar & Dhull, 2019), which is located between the pure cortex and nucleus. (Amaya et al., 2003).

The opaqueness might vary between minor opaque to high dense, so that including with partial layer to entire layer and linked to another morphology or zonular cataract isolated. (S. K. Khokhar & Dhull, 2019)

Usually, the fetal nucleus is preserved (Patil, 2015) (S. K. Khokhar & Dhull, 2019), however the fetal nucleus is occasionally involved, and from the outside the cortex is clear. (Patil, 2015)

Typically, they are heritable dominantly autosomal. (Amaya et al., 2003) (Patil, 2015) or idiopathic (Patil, 2015), and bilaterally but a little bit asymmetric, also they are made up of a thin layer whitish spots in a layer or many lens layers. There is frequently notable variation inter-ocular and intra-familial. (Amaya et al., 2003) (Patil, 2015).

It occurs as a result of damage to certain layers of lens fibres that were active in metabolism at the time of the damage. The zonular cataracts are visually insignificantly and might advance over time. (S. K. Khokhar & Dhull, 2019).

These Cataracts are frequently partial morphologically, and they might include protrusions from the outside borders called 'riders.'(Patil, 2015).

The aetiology of these riders is ambiguous. Depending on their appearance and progression pattern, (S. K. Khokhar and C. Dhull) defined the name "reverse cuneiform cataract" to describe riders with zonular cataract. (S. K. Khokhar & Dhull, 2019).

In the majority of instances, and in this morphology a well prognosis of vision seen. (S. K. Khokhar & Dhull, 2019).

Most instances could be treated cautiously, and surgical intervention in children is uncommon. (Amaya et al., 2003)

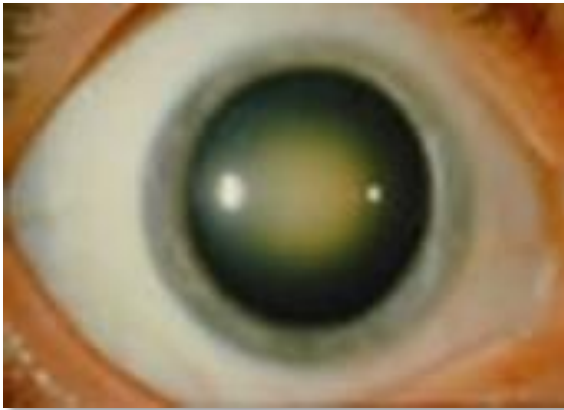


Image 26: Zonular cataract, (extracted from Centro de Oftalmología Barraquer)

1.2.4.4.3. Anterior Cataract

1.2.4.4.3.1. Anterior Polar

They situated at the center of the anterior pole of the lens capsule. (Ellis, 2002) (S. K. Khokhar & Dhull, 2019). On the opacification, might be tiny, inserted threads of iris. (Ellis, 2002)

They are white (Ellis, 2002), tiny (Ellis, 2002) (American Academy of Ophthalmology, Jick, 2019), not progressive which do not impact vision, symmetrical and bilateral (American Academy of Ophthalmology, Jick, 2019).

Generally, these little opacifications are not visually noticeable. (S. K. Khokhar & Dhull, 2019) and ordinarily not necessary to treat them (Ellis, 2002) (American Academy of Ophthalmology, Jick, 2019), yet they can result in amblyopia (Ellis, 2002) (S. K. Khokhar & Dhull, 2019) and anisometropia (Ellis, 2002) (S. K. Khokhar & Dhull, 2019) (American Academy of Ophthalmology, Jick, 2019) if not treated (S. K. Khokhar & Dhull, 2019).

In general, the forecast of eyesight is well. (Amaya et al., 2003).

They might advance in scarce situations. (Ellis, 2002).

These opacities vary from tiny opacity dot to a plaque-like or, denser pyramidal opacity which is less common. (S. K. Khokhar & Dhull, 2019).

The pathogen of this case is not fully intelligible. (S. K. Khokhar & Dhull, 2019).

The ocular anomalies such as anterior lenticonus, persistent pupillary membrane and microphthalmos, occasionally are seen in conjunction with anterior polar cataracts. (American Academy of Ophthalmology, Jick, 2019).

They sometimes may be associated with persistent fetal vasculature that could play a pathogenic role. (S. K. Khokhar & Dhull, 2019).

Most often are inherited (Amaya et al., 2003) (American Academy of Ophthalmology, Jick, 2019) may be in a dominantly autosomal trait (American Academy of Ophthalmology, Jick, 2019), and non-hereditary could be novel mutations that pose a danger of recurrence in the patients' kids. (Amaya et al., 2003).

The type of dot-like is an opacification on the center of the capsule of the lens that is not visually noticeable. The plaque-like, an opacification that is linked with anterior capsule which is centrally flat and fibrous. It can occur separately or in conjunction with membraned or entire cataract. (S. K. Khokhar & Dhull, 2019).

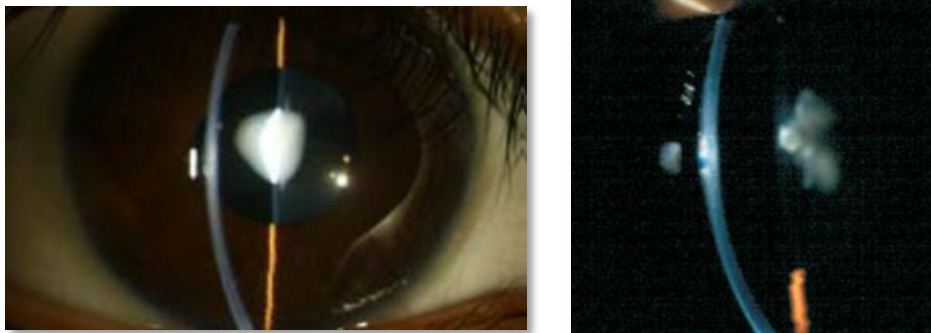


Image 27: a & b: Anterior polar, (extracted from Centro de Oftalmología Barraquer)

1.2.4.4.3.2. Anterior Pyramidal

Compared to anterior polar cataracts, they are more probably of being visible significantly and advance. They might separate and constitute a foreign body in the anterior chamber. (Patil, 2015). An opacification is observed centrally in the anterior capsule with a rising that resembles the pyramid shape. In general, are observable and could be linked to cortical or anterior subcapsular cataracts. (S. K. Khokhar & Dhull, 2019).

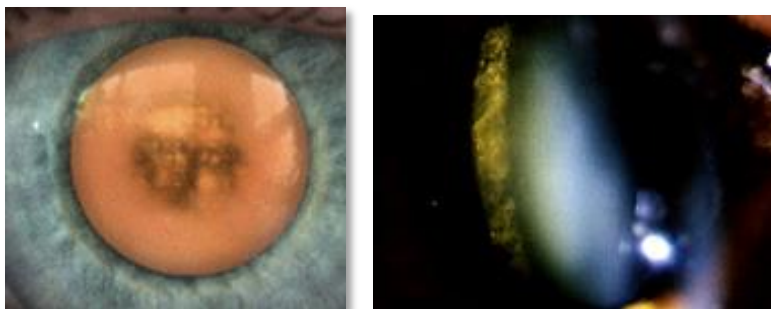


Image 28: a & b: Anterior pyramidal, (extracted from Centro de Oftalmología Barraquer)

1.2.4.4.3.3. Anterior Subcapsular

In situations of congenital, it can be observed in conjunction with an anterior polar cataract. (S. K. Khokhar & Dhull, 2019)

The opaqueness exists underneath the lens's anterior capsule. Usually, the cause is unknown. When the manifestations are bilateral, Alport syndrome must be considered. (Ellis, 2002)

In the congenital isolated form, the cataract could perhaps be visually invisible, and the vacuoles may be seen. (S. K. Khokhar & Dhull, 2019)

Despite opacities may be minor and have no effect on vision, nevertheless, in certain cases, vision is impaired; therefore, surgical intervention must be taken into account. (Amaya et al., 2003)

The acquired illness is the commonest linked to this cataract which appeared in trauma, uveitis (Amaya et al., 2003) (S. K. Khokhar & Dhull, 2019), instances of caused by using medicines (S. K. Khokhar & Dhull, 2019), radiation exposure, atopic dermatitis (in which the opacity of lens resembles a shield) (Amaya et al., 2003), which can be of visual significance. (S. K. Khokhar & Dhull, 2019). Nevertheless, they might be the portion of a most prevalent cataract; they are seldom linked to pulverulent cataracts or anterior lenticonus, such as in Alport's syndrome. It is unknown the precise role of growth factor of connective tissue in the pathogenesis of anterior subcapsular cataracts even though indications of its presence in these lens opacities. (Amaya et al., 2003).



Image 29: a & b: Anterior subcapsular, (extracted from Centro de Oftalmología Barraquer)

1.2.4.4.4. Posterior Cataract

1.2.4.4.4.1. Posterior Polar

They have opacities in the lens's posterior pole, as in the anterior polar cataracts. They are strange owing to the susceptibility to posterior capsular deformity throughout the operation. Minor blunt trauma might potentially cause the posterior capsule to rip. A preexisting posterior capsular deformity may be present in some situations. (S. K. Khokhar & Dhull, 2019)

The fragility of the capsule has been notified in conjunction with these cataracts and which are typically steady but can develop. Since they are bigger and nearer to the eye's nodal point, these cataracts are related to a greater substantial reduction of sight compared to the anterior polar cataracts. (American Academy of Ophthalmology, Jick, 2019)

They may occur sporadic or familial. The sporadic are usually unilaterally and can be linked to a posterior lens surface anomaly or residues of the tunica vasculosa lentis for example, lentiglobus or lenticonus. The familial are often bilaterally and autosomally dominantly inherited. (American Academy of Ophthalmology, Jick, 2019)

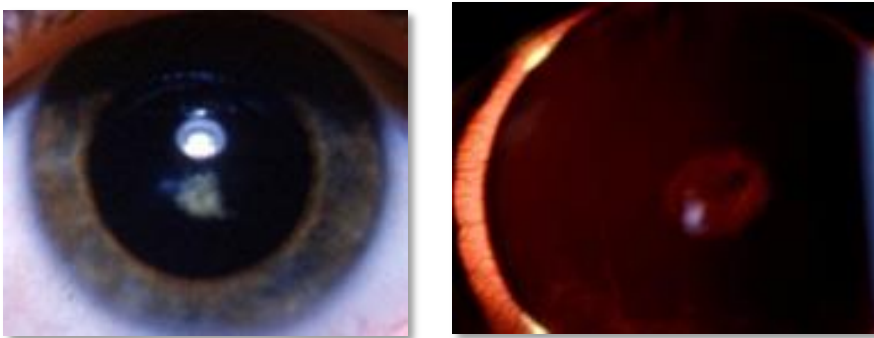


Image 30: a & b: Posterior polar, (extracted from Centro de Oftalmología Barraquer)

1.2.4.4.4.2. Mittendorf's Dot

Mentioned in the point (1.2.1.6).

1.2.4.4.4.3. Posterior Subcapsular

It could appear as a plaque or vacuolar. (Amaya et al., 2003) (S. K. Khokhar & Dhull, 2019). In the pediatric age group, it can occur congenitally or acquired. (S. K. Khokhar & Dhull, 2019). This may occur after severe trauma. (Ellis, 2002) (S. K. Khokhar & Dhull, 2019), after therapeutic corticosteroids topically or systemically. (Ellis, 2002) (S. K. Khokhar & Dhull, 2019) or uveitis. (S. K. Khokhar & Dhull, 2019).

Posterior subcapsular opacities that adhere to suture lines can be seen in Fabry's disease. These are typically found following trauma which commonly results in the lens swelling and extremely opaque, subsequently a membrane after cataract. The cortical and posterior subcapsular cataracts happen before senility in type 2 neurofibromatosis. The vacuolar type is nearer to the lens's posterior capsule while the plaque is more cortical. The plaque form can be present in myotonic dystrophies, the syndrome of Turner, as well as congenital cataract. This kind of cataract is significant because of its location in the centre or axial and the posterior, it reduces vision early. In the plaque kind, the main histopathologic

alteration of lens fibers is breaking the typical orderly parallel rows into round pellets. (Amaya et al., 2003). Frequently requires surgical treatment. (Ellis, 2002)

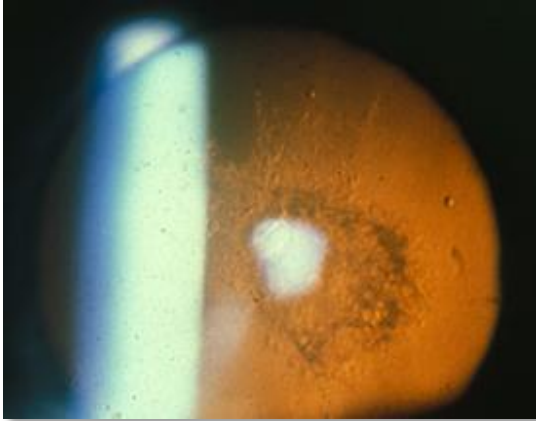


Image31: Posterior subcapsular cataract, (extracted from Centro de Oftalmología Barraquer)

1.2.4.4.4. Oil droplet

Despite the morphology is rare.(S. K. Khokhar & Dhull, 2019), however it is usually observed in children who have galactosemia.(Ellis, 2002)(Amaya et al., 2003)(S. K. Khokhar & Dhull, 2019).The opacity is dim in the posterior lens cortex's central portion.(Ellis, 2002).Inside the lens's center portion, a distinct alteration happened in the index of refractive which resulted in appearing as an oil droplet.(Amaya et al., 2003)(S. K. Khokhar & Dhull, 2019). When galactosemia is detected, a proper work and therapy are required. the Mild galactosemia-related cataracts could well be cleared with suitable nutritional limitations. (Ellis, 2002). The Lamellar cataract could evolve and grow if a dieting regimen is not strictly observed or if the treatment is delayed. (Amaya et al., 2003).

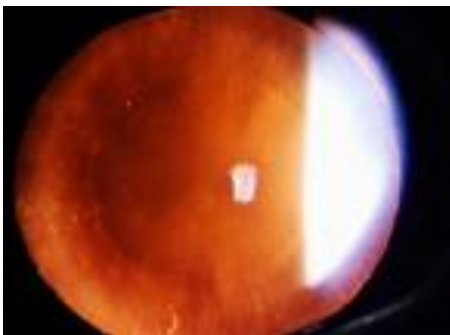


Image 32: Oil droplet,(extracted from Centro de Oftalmología Barraquer)

1.2.5. DIAGNOSIS AND EVALUATIONS

1.2.5.1. TAKING HISTORY

A thorough a complete history can provide an aetiological hint. Good predictions are linked to the patient's complain and the presentation of symptoms. For example, patients with leucocoria (white pupil) indicate a better diagnosis than patients with strabismus. Likewise, long-term opacity can contribute to amblyopia caused by image deprivation, makes prognosis more difficult. It's also crucial to record the entire family history, including intranatal, postnatal, and any related diseases, as well as the trauma history in detail. (Patil, 2015). The most important thing to document is if the child has any delay in development milestones or is not growing well. Moreover, pediatricians should be consulted to aid the general evaluation of the child for possible systemic disease. (Aryee & Dumont Jones, 2020).

1.2.5.2. DETECTING FOR CATARACT

When a mother is concerned about her infant's eyesight, pupil shape, or if the baby squints or has nystagmus, she usually visits a child's doctor for an examination. (Taylor & Rice, 1982). A red reflex examination for all newly born is crucial, which should be tracked with the same test at 6-8 weeks. And so, to perform the test red reflex, direct ophthalmoscope is used. A pediatric ophthalmologist should be consulted if any of these factors appear during the examination, such as opacities, a missing red reflex, or leukocoria. (Wilson ME, 2015).

In an older child, parents may bring their child to a doctor due to the child's failure in a vision assessment at school or they are concerned about their child colliding with objects. (Aryee & Dumont Jones, 2020).

1.2.5.3. VISUAL ACUITY

There are many types for testing the acuity of sight depending on the age of child:

To perform the acuity of sight in babies, lateness to grow or a child who is unwilling to cooperate by evaluating Fixation (central, steady, maintain), following (Dhull et al., 2019), (Aryee & Dumont Jones, 2020), with this method it can be strong evidence of an eye fixation preference, suggesting impaired vision in the other eye. In situations of unilateral or asymmetric cataracts, it's effective too. (Kraus et al., 2016). Also Evoked visual potential (VEP) (American Optometric Association, 2020)

Before one year: visual-evoked response (VER) (American Optometric Association, 2020), Catford drum, optokinetic nystagmus (Dhull et al., 2019), Teller's acuity cards, Cardiff acuity test too (American Optometric Association, 2020) (Dhull et al., 2019), (Aryee & Dumont Jones, 2020)

Between one and two years: Worth's ivory ball test, Boeck's candy test, the Screening Test, Retards (STYCAR) (Dhull et al., 2019), Cardiff's acuity test and Evoked visual potential (VEP) (American Optometric Association, 2020) (Dhull et al., 2019), (Aryee & Dumont Jones, 2020).

Between two and three years: miniature toy test, coin test, and LEA symbols. (Dhull et al., 2019).

Between three and five years: Allen's picture card, Lippman's HOTV test, letter test.(Dhull et al., 2019), and LogMAR chart. (Dhull et al., 2019), (Aryee & Dumont Jones, 2020)

More than five years: Tumbling E, Landolt's broken ring, Snellen's chart, ETDRS chart (Bruyn & Parbhoo, 2013), (Dhull et al., 2019), and LogMAR chart. (Dhull et al., 2019), (Aryee & Dumont Jones, 2020)



Image33: cards for vision acuity called "Cardiff", extracted from (Dhull et al., 2019)

1.2.5.4. *SLITLAMP BIOMICROSCOPY*

Through using this equipment as standard or movable, it aids the physician to assess any abnormalities in the anterior segment (irregularity in growth, synaechiae, microphthalmous), determining the degree, location and form of the opacification of lens (In the peripheral, central, anterior, or posterior). In addition to the posterior capsule should be evaluated to look if there are any co-existing abnormalities linked to congenital cataract (posterior lentiglobus or persistent foetal vasculature). (Aryee & Dumont Jones, 2020)



Image34: slit-lamp movable, extracted from (Hoonpongsimanont et al., 2015)

1.2.5.5. *ULTRASOUND (BIOMIROSCOPY /B-SCAN)*

1.2.5.5.1. *Biomicroscopy (UBM)*

Is a helpful instrument for a thorough evaluation of pediatric cataracts and accompanying eye abnormalities. An accurate evaluation of angle and the ability for quantitative analysis of the anterior chamber can be made with UBM using high-resolution. When the cornea is completely opacified, it gives information about the anterior segment morphologically and topographically, which allows the surgeon to build a customized surgical plan. (El Shakankiri et al., 2009)

1.2.5.5.2. *B-scan Ultrasound*

Provides a wealth of information, aids in evaluation during the operation, prognosis of a case, particularly in cases of unilateral cataract. (Khokhar et al., 2018) It's also useful when the posterior portion isn't seen. (Dhull et al., 2019)

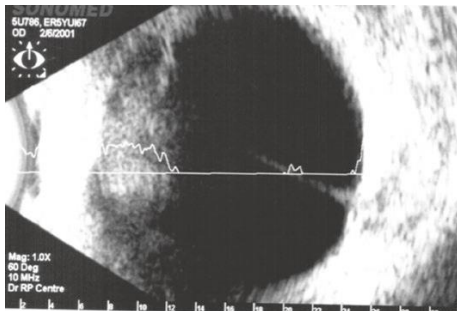


Image35: B-scan for diagnosing posterior portion diseases, extracted from (Dhull et al., 2019)

2. OBJECTIVES AND METHODS:

2.1.OBJECTIVE

The congenital cataract is very crucial issue which causing the impairment of the vision in children and amblyopia in the eye,therefore, there are some questions about treating cataract in children, including:

It can be treated or no?, when the surgery is necessary?

What is the most adequate solution for managing cataract in infants and managing the amblyopia resulted from cataract?

2.2. METHODS

According to these questions, we have searched about the best techniques for the management of congenital cataract

3. RESULTS

3.1. NON-INDICATIVE AND INDICATIVE SURGICAL

In congenital cataracts, not all of them are considered for surgery. The anterior polar cataracts are never surgical, they never advance or impact the maturity of vision, however, they should be monitored because frequently they are related to anisometropia and refractive error. As with pyramidal cataracts, are also not surgical at first but are more likely to progress. (Morales, 2007).

The surgery is indicated when a cataract is optically noticeable more than 3 mm, cataracts that prevent retinal examination, cataracts accompanied with strabismus or nystagmus, and intensive posterior or nuclear cataracts. (S. K. Khokhar & Dhull, 2019). To avert irreparable deprived amblyopia, significant visual cataracts in a neonate must be eliminated during the initial few weeks after birth. The more serious cataracts definitely benefit from surgery in older kids who have measurable eyesight. (Ellis, 2002)

3.2. KERATOMETRY

The data of Keratometry is often taken using a portable auto-keratometer underneath general anesthesia. In older kids, whereas, it can be evaluated by utilizing the keratometer manually. Also, Keratometry measurements must be done with no speculum this is due to the speculum's deformed impact on the eyeball in that could cause inaccurate findings. (S. K. Khokhar & Dhull, 2019). In the initial six months of age, keratometry decreases sharply (-0.40 D/month), thereafter (-0.14 D/month) with in the following six months, then (-0.08 D/month) in the age of two, attaining the level of adults at around three years old. (Khokhar et al., 2017). The keratometry in eyes with cataracts is steeper than in non-cataractous eyes; also, in eyes with unilateral cataracts keratometry has been noted to be steeper than in eyes with cataracts bilaterally. The median pre-surgical keratometry for congenital cataracts (47.78 D) is greater than that of developing cataracts (44.35 D). (S. K. Khokhar & Dhull, 2019).



Image36: Automated Keratometer Hand-held extracted from (Dhull et al., 2019)

3.3. BIOMETRY AND INTRA-OCULAR LENS POWER

In the majority of the reports, correcting below the IOL power for paediatric cataract is indicated, predicting myopia shifting after an intraocular lens implant. (Ram & Sukhija, 2010). Frequently, young children's axial length (AL) metrics are performed in the operation theatre beneath general anesthetic. (Patil, 2015). The Ultrasonic biometric is utilized with the indentation of the cornea or technique of immersion and that is the favored method. (Joseph & Meena, 2018). In the first six months, the (AL) gets to increase swiftly (0.46 mm/month), thereafter in infantile stage the growth is slower (0.15 mm/month) until 1.5 years, subsequently in youthful stage slowly (0.10 mm/month). (Khokhar et al., 2017). The optical biometry which is dependent on fractional coherence interferometry (Patil, 2015), is widely utilized in cooperating kids. (Joseph & Meena, 2018). Children have a greater absolute error than adults. (Khokhar et al., 2017). In adults, a 1 mm error in measuring the axial length can result in error in IOL power a 2.5 diopter; nonetheless, in short eyes, the error increases to 3.75 D. Relying upon the eye length, the fault of 0.1mm in measuring AL can lead to diopter variation between 0.25 to 0.75 in the calculation of the intraocular lens and this may be in certain persons medically important. (Joseph & Meena, 2018). The newborn's eye has a median axial length of 17.0mm, comparable to 23 - 24mm for adult people. (Ram & Sukhija, 2010). There was not disclosed formula for calculating power IOL to be superior to another, also no exists general agreement on which formula to use. The Physicians either could opt hypermetropia, with a projected of shifting of myopia by time, or emmetropia, aid in treatment of amblyopia. (Patil, 2015).

The IOL could be placed for eyes with axial length more than 17mm as well as the diameter of cornea more than 10 mm; and the power of it is relies upon a variety of aspects, including the patient's age, cataract morphology, cataract development period (if congenital or developmental), presentation of acuity of eyesight and biometry, refraction condition of the other eye and if the cataract is in one eye or two eyes. (Khokhar et al., 2017). In children instances, modified theoretical and regression formulae utilized in adults, such as Holladay, Hoffer Q, SRK T, and SRK 2, are applied. (Joseph & Meena, 2018). In kids below the age of two, the IOL's power must be beneath corrective by 20%, while it must be less corrective by 10% among the ages of two and eight. (Kumar et al., 2018). Enyedi et al. proposed a post- operative refractive target of:

+6 D for a one year old , +5 D for a two years old, +4 D for age- three years, +3 D for age 4 years , +2 D for age 5years , +1 D for age 6 years, Plano for age 7 years , and -1 D to -2 D for age 8 years and old people.(Enyedi et al., 1998).In children, the perfect IOLs have to be between 10.5-12mm.(Kumar et al., 2018).



Image37: beneath general anesthetic, axial length is measured by immersion extracted from(Wilson & Trivedi, 2012)

3.4.IOL MATERIALS

The implant of intra-ocular lens has become a popular manner of visual rehabilitative following removal of cataract.(Ellis, 2002).There are methods for inserting the IOL including, in the ciliary sulcus, optic of IOL in back of the bag's capsular, or inside the bag.(Kumar et al., 2018).The preferred technique of IOL is within the bag, but when circumstances require, such as secondary IOLs when opening the bag is complicated, as well as when a substantial posterior capsular removal is present in posterior polar cataracts, then the insertion of Sulcus is preferable.(Joseph & Meena, 2018). It has been shown that acrylic lenses have reducing ratios of posterior capsule opacification (PCO) in comparison with polymethylmethacrylate (PMMA) or silicone IOLs in pediatrics, Indicating the best biocompatibility of substances for acrylic lenses, in addition to; there were informed of well results with acrylic hydrophobic and hydrophilic acrylic IOLs in kids, despite of no exact experiment was conducted. Regarding shape, the acrylic lenses with one piece appear significantly better than 3-pieces in the ratios of issues intraoperatively and postoperatively. (Zhao & Zhao, 2021).

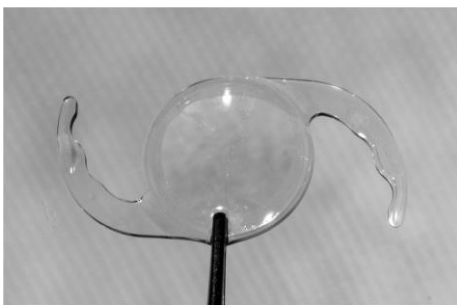


Image38: Intraocular lens made of one piece of acrylic,extracted from(Bar-Sela & Fleissig, 2013)

3.5. SURGERY

A general anesthetic is necessary for pediatric surgical cataract. (Self et al., 2020). In pediatrics, the surgery differentiates from adult people, due to the reduced rigidity of their cornea and sclera, lens is smooth, the capsular anterior is extremely flexible and vitreous is good-shaped. (Khokhar et al., 2017). The surgical technique's purpose is to afford a pure axis for long-extent through prevention of the formation of secondary membrane or PCO. (Ram & Sukhija, 2010); and there are many techniques that existed include such as lensectomy, anterior vitrectomy, and/or combination with primary posterior capsulotomy. The cataract in children can be extracted by two primary methods: the methods of pars-plana and the limbal, with pars-plana as most diverse. (Kumar et al., 2018). The technique superior incisions are the commonest employed, which is the most favored modality; since there is a smaller danger of injury and endophthalmitis after surgery compared to the alternate. An anterior capsulotomy is conducted to assure secure lens implant with manual continuous curvilinear capsulorhexis (CCC) being the “standard golden technology”, that has a minimum danger of tearing or stretching. The combination of primary posterior capsulotomy with anterior vitrectomy is necessary for newborns and young kids, so as to evade multipliers like posterior capsule opacification (PCO). (Hernandez & Kumar, 2018).



Image 39: Capsulorhexis,(extracted from Centro de Oftalmología Barraquer)

3.6. COMPLICATIONS

Direct consequences are including problems of the wound like prolapse of iris, hole wound and suture break. (Joseph & Meena, 2018). The most commonly post-surgical complication extensively mentioned in several research; is the Visual axis opacification (VAO) (Zhao, 2021); and when this happens, as earliest as feasible the ophthalmologist must contemplate surgery of membranectomy or Nd: YAG laser capsulotomy underneath general anesthetic. Uveitis or a serious fibrinous reactive is one of the gravest complications. There is also, a most dangerous and dreaded issue that might happen in either early or late stages is endophthalmitis postoperatively. It can be seen the strabismus and amblyopia before and

after surgery. Kids are more likely to long-term danger of retinal detachment. (Mohammadpour et al., 2019). Glaucoma, decentralization of IOL, the membrane of pupillary, and dispersion of iris pigmentation, are comparatively popular post-surgical complications. (Zhao, 2021).

3.7. VISUAL REHABILITATION

After removing paediatric cataract, there are approaches for vision rehab that incorporate the implant of IOL, contact lenses, and aphakic spectacles, thus; monitoring is required regardless of the rehabilitative approach chosen. It is essential to educate parents regarding the necessity of amblyopia treatment following surgery. Ultimately, following the success of cataract operation in kids, the result of vision is relied on the treatment of amblyopia at the appropriate time, particularly following unilaterally cataract excision. (Medsinge & Nischal, 2015). Also, to promote the growth of the vision and to give a concentrated image of the retina, its essential rectifying any error in refraction as quickly as feasible post-surgical. (Self et al., 2020).

3.7.1. GLASSES

As the child's eye suffers from substantial hyperopia post -surgical cataract, which could be in eyes with aphakia reach +30 diopter sphere, while it is lower obvious in eyes with pseudophakia, though based on the child's age and the refractive desired. The weak optical characteristics of glasses are related to elevated hypermetropia in aphakic children; so, the spectacles are weighty and certain kids do not endure them well; Besides this, they cause a discrepancy in the size of the image among the eyes of pseudophakia or aphakia and phakia in kids that have previously undergone a unilateral operation, and so contact lenses reduce this. (Self et al., 2020). In the case of children accompanied with strabismus, the complete prescription of spectacles is given; that is done at a 1-meter distance and beneath atropine 1%. For kids over the age of three and who are attending school, following distance correction, the spectacles with the addition of near are given. (Khokhar et al., 2018).



Image40: aphakic spectacles with designing of biconvex, extracted from(Repka, 2016)

3.7.2. CONTACT LENSES

The contact lenses are regarded to be visually preferable to glasses, however, it is not possible to wear in the lengthy-term.(Mohammadpour et al., 2019), they are the best choice in many situations because the lens's refractive power stays concentrated above the optical axis regardless of where the kid is

seeing.(Self et al., 2020);and have many features; the most crucial is diminishing variability in retinal image size in aphakia unilaterally;another additional advantages comprising reducing in dazzle and glare, increased optical areas, the disappearance of ring scotomas,simple adjustment with variable refractive error, and lower occurrence of nystagmus.(Mohammadpour et al., 2019).

Each four to six weeks, lenses are replaced, based on how they good properly cared for them, and usually, three or four pairs are supplied at once. Typically, the preferred method is the soft silicone hydrogel lenses for daily use, as they are fairly in price, steady in the eye, and well-tolerated. (Self et al., 2020);some other types such as; Mini-scleral and scleral lenses, despite their very difficult in fitting, however, provide a high quality of sight, especially when there is an abnormal cornea or corneal astigmatism; Gas-permeable lenses, their oxygen permeability is significantly higher, possible to add the UV filter, able to correct irregular corneal astigmatism and this is especially useful in a traumatic cataract that injures cornea. The incidence of neovascularization of cornea and infections is substantially lower than with soft lenses. (Corbeil et al., 2014).



Image41: A child that is bilaterally aphakic and wears contact lenses, in both eyes the eyesight is good

Extracted from(Repka, 2016)

3.7.3.SECONDARY IOL IMPLANTATION

In aphakia children, when the cornea gets to the admissible diameter, then consideration must be seen to implant secondary IOL, which is normally performed during 1.5 years old. After the planting of IOL, they must undergo refraction assessment regularly, as well as prescribing for suitable power depending on eye development, are also provided with add +2.50 to 3.00 of progressive or bi-focal for the near sight. (Joseph & Meena, 2018). So, the implant of IOL, affords the best vision rehab and reconstruction of binocular vision, with the exclusion of a few side effects. Furthermore, it is effective in terms of selection and cost; by contrast, the contact lens in the developing countries is difficult to admit broadly by majority families of congenital cataract, that's because of its elevated load financially, complicated care actions, and reduced popularity. In spite of this, there is no proof convinced that IOL is superior to contact lens. (Zhao, 2021).

3.7.4.PATCHING

After the operation, the occlusion treatment is initiated as quickly as feasible. So, the occlusion therapy and also well commitment provide excellent outcomes in situations of cataract were operated unilaterally. To prohibit the amblyopia from occurring in the better eye due to occlusion, the weak eye is covered. Under the age of 8 months in kids, occlusion is administered in a ratio 1:1, since relying on the age, both of eyes are covered; occlusion for child two months old is prescribed for two hours of waking in alternating eyes every day. And the occlusion in the better eye is more than in the weaker eye for a kid above one year old; in which; occlusion for three year-old kid is used for eight hours of waking daily in the better eye for three days and in the weaker eye one day. (Khokhar et al., 2018). On the other hand, in the bilateral cataract postoperatively, the treatment of occlusion is just needed if there is strabismus that appeared and is favored one eye. Also, throughout occlusion, the Bi-focals are especially necessary. (Lagrèze, 2021).



Image42: treatment using patch extracted from(Narendran, 2015)

3.7.5.PENALIZATION

An efficient therapeutic option for kids that do not improve with just glasses. And it's the use of atropine as a penalty causes paralysis in the good eye. This Atropine drops (1%), causes blurring optical and losing of accommodation in the dominant eye, thus the weak eye is used by force. Also, its efficacy is larger if the better eye is long-sighted. For curing the amblyopia, in the beginning is taken every day. In curing moderate amblyopic kids, it seems that its usage at the weekend as efficacious as everyday usage. (Park, 2019).



Image43: Atropine 1%, extracted from(<https://www.akorn.com/prod-details-list.php?group=Atropine+Sulfate+Ophthalmic+Solution%2C+USP&ndc=3810>)

4. DISCUSSION

The congenital cataract is known as any opacity of lens evident at born or early infancy (Mohammadpour et al., 2019), and it is a leading cause of vision impairment in children globally. It can also influence substantially on a child's development neurobiologically (Nischal & Medsinge, 2015), and consequent amblyopic deprivative (Mohammadpour et al., 2019). Therefore, the earlier screening, diagnosing, as well as suitable medical attention, are crucial for attaining the best results. (Self et al., 2020). A trial study was performed for comparing the precision of reflex of infrared evaluation utilising a model photographic apparatus to routine non-mydriasis red reflex test by a direct ophthalmoscope in diagnosing the cataract in newborn and pediatrics, which the result was shown that fetal remains in the eye's anterior portion were discovered in 5% in eyes of neonatal by photography infrared reflex, that was not observed using the red reflex test. Also, In detecting pediatric cataract, Infrared evaluation demonstrated much higher sensibility and precision; and When comparing "non-Caucasian eyes" to "Caucasian eyes", the accuracy of red reflex was especially low, so this study summarized that the imaging by infrared reflex has the capability to enhance the accuracy of diagnosis of cataract screening by untrained medical personnel, especially in pediatric "non-Caucasian". (Duret et al., 2019)

In pediatric cataract, it must be considered while treating it, which is based on the age of its appearance, the form, the laterality, and any related systemic and ophthalmic diseases. (Nischal & Medsinge, 2015). So, the ophthalmic staff confronts particular challenges such as the age of manifestation and examining these patients. (Aryee & Dumont Jones, 2020). Most the Cataracts in children spite of that are unknown causes, However, depending on the history of family and antenatal, as well as the cataract's kind, for this, it is useful to perform the initial lab work also, is indicated for lab examinations which incorporate; assessments of lowering amino acids and compounds in urine, CBC, BUN, Disease of Venereal Research lab test, tests of red cell calcium, phosphorus and galactokinase, in addition to TORCH tests.(Mohammadpour et al., 2019).

Qi-Hui Zhao and Yun-E Zhao suggested for IOL transplantation for congenital cataract newborns, particularly for these unilateral cataracts and having good grown eyes, is indicated at the age of 6 months or more. In contrast, the implant of secondary IOL for unilateral cases at the age of 1.5 years and for bilateral cases at the age of 2 years. For the power of IOL, they favoured leaving a specific grade of hypermetropia, whilst they pointed to the refraction of the corresponding unaffected eye in the case of children with unilateral, in addition to, recommendation for two- open haptics of acrylic IOL when selecting an IOL design.(Zhao, 2021). A meta-analysis was conducted by J. Chen et al. on children on children with congenital cataract under two years to compare acuity of vision and complicates between implant of a primary intraocular lens and utilizing a contact lens, therefore, statistical there was no substantial change in the incidence of strabismus and glaucoma among both two groups, then they

concluded that inserting a primary IOL through surgery is superior than using a contact lens for children under the age of two.(J. Chen et al., 2020).

There are some “clinical tips” by (Self et al., 2020):

1- In the case of unilateral dense cataract in childhood, the majority of surgeons schedule to operate between the ages of six and eight weeks and between the ages of six and ten weeks in a bilateral cataract.

2-The corrective of refraction for pediatric with pseudophakia and aphakia is as crucial as surgical in obtaining good vision.

3- 3-In spite of technical challenges, nowadays a variety of choices of the insertion of secondary IOL, that allow for effective long rehabilitation of vision and optic successfully. The IOL insertion can be considered for aphakic kids during whatever age post early childhood, especially those who are uncomfortable with using contact lenses or spectacles.

5. FOR CONSIDERATION

In situations of strabismus or nystagmus should be treated immediately. It seems to raise the secondary glaucoma danger if the cataract is removed prior to four weeks. (Joseph & Meena, 2018). Also, because of the danger of the possibility occurring opacification of the posterior capsular, hence usually, children are very small to undergo YAG laser capsulotomy, for this, it is suggested to a primary posterior capsulorhexis. (Aryee & Dumont Jones, 2020).

The treatment of amblyopia following pediatric cataract surgery is crucial for the successful, especially in the cases of unilateral, which must commence soon post lens removal (Kletke et al., 2018); therefore, the parents need to be informed of the necessity of treating amblyopia post-surgical. (Saha et al., 2017)

The advance of the equipment and technology of surgery have improved the results of vision following cataract surgery of children to be better than ever. nevertheless, the collaboration work of the team of multi-disciplinary and well examination of eye with optometrists, orthoptics, specialists, as well as medical professionals, stays critical to attaining the best results of vision. (Self et al., 2020)

The splendid techniques for investigating hereditary cataracts are "next-generation sequencing " and "future ultra-high-throughput sequencing". A better understanding of the functional outcomes of mutations of cataract pathology may lead for the future to new therapy choices. (Bremond-Gignac et al., 2020)

6. CONCLUSION

-The congenital cataract is known as the lens opacification which happens when the child gets birth or later in early childhood and is considered one of the most critical issues and threatens children's visual blindness that leading to amblyopia if not treated directly

-Among the main challenges faced in the treatment of congenital cataracts include age, and eye size when taking measurements during the investigations, time of surgery, selection of the appropriate intraocular lens, postoperative complications, as well as when rehabilitating vision postoperatively; therefore, these all represent great challenges, especially for ophthalmologists, optometrists as well as the other medical professionals, and they still exist.

-In cases of mild cataracts that are not considered to be in need of surgical intervention, they should be followed up through periodic visits to an ophthalmologist and optometrist.

-There are many surgical techniques has been developed in surgical pediatric cataract available for minimizing the consequence post-surgical.

-After surgery, the most important thing is the visual rehabilitation for treating the aphakia, refractive errors, and amblyopia, which can be done by optometrists that prescribe aphakic spectacles, fitting contact lenses, occlusion therapy, as well as cooperating with ophthalmologists for reaching the ideal visual results.

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