Pediatric cataract

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List of Abbreviations

ACM- anterior chamber maintainer

GSH- glutathione

ECG- electrocardiography

WHO- World Health Organization

CCC- continuous curvilinear capsulorhexis

IOL- intraocular Lens

PCO- posterior capsular opacification

RGP- rigid gas permeable

Contents

1. Summary

Cataract is characterized by the clouding of the lens in the eye. In adults, it predominantly arises from age-related degenerative changes or as secondary effects from medications, ocular trauma, or metabolic disorders. In contrast, pediatric cataracts can be congenital, often resulting from TORCH infections, metabolic diseases, or genetic conditions. Newborns are routinely screened for cataract using the red reflex test, and any abnormalities warrant immediate referral to a pediatric ophthalmologist to prevent the risk of deprivation amblyopia. While cataracts in adults may develop insidiously and remain unnoticed until significant visual impairment occurs, the diagnosis is usually confirmed through a comprehensive patient history and visualization using slit-lamp microscopy. Treatment generally involves surgical removal of the cloudy lens and replacement with an artificial intraocular lens, especially when the visual function is severely compromised. If left untreated, cataracts can progress to complete blindness.

2. Sažetak

Katarakta karakterizira zamućenje leće u oku. Kod odraslih, ona uglavnom nastaje zbog degenerativnih promjena povezanih sa starošću ili kao sekundarna posljedica korištenja lijekova, traume oka ili metaboličkih poremećaja. Nasuprot tome, dječja katarakta može biti kongenitalna, često posljedica TORCH infekcija, metaboličkih bolesti ili genetskih stanja. Novorođenčad se rutinski pregledava za katarakту pomoću crvenog refleksnog testa, a bilo kakve abnormalnosti zahtijevaju hitno upućivanje pedijatrijskom oftalmologu kako bi se spriječio rizik od deprivacijske ambliopije. Dok se katarakta kod odraslih može podmuklo razviti i ostati nezapažena sve dok se ne dogodi značajno oštećenje vida, dijagnoza se obično potvrđuje kroz sveobuhvatnu povijest bolesnika i vizualizaciju pomoću mikroskopije s procjepnom svjetiljkom. Liječenje obično uključuje kirurško uklanjanje mutne leće i ugradnju umjetne intraokularne leće, osobito kada je vidna funkcija ozbiljno ugrožena. Ako se ne liječi, katarakta može napredovati do potpune sljepoće.

3. Introduction

Cataract, defined by the World Health Organization (WHO) as the clouding of the crystalline lens of the eye, significantly impair normal vision and represent a major global health challen ge. The term 'cataract' originates from the Latin word 'cataract' and the Greek 'catarrhines,' both resembling the white, rushing water of waterfalls, reflecting the eye's cloudy appearance in this condition. Historically, cataracts are depicted as early as in a 4500-year-old wooden statue found in Cairo, illustrating the long-standing human battle with this visual impairment.

Cataract formation involves multiple factors and can occur at any age, although it is predominantly observed in the elderly. Congenital cataract, present from birth or soon after, significantly impact visual development and can lead to amblyopia if untreated, as the immature visual system requires clear images for proper development. This condition can be unilateral or bilateral, with varying degrees of opacity and potential for preserving some visual function.

Globally, cataract is a leading cause of blindness, particularly in developing regions with limited surgical treatment access. In the Eastern Mediterranean region alone, cataract account for over 51% of blindness cases. Despite the lack of prevention methods for cataract development, advancements in surgical techniques have transformed treatment outcomes in developed countries, making cataract surgery one of the most effective interventions in modern medicine. However, challenges remain in the form of potential preoperative, intraoperative, and postoperative complications.

The significant burden of cataracts, especially congenital forms, extends beyond health to socioeconomic domains, influencing productivity and incurring rehabilitation costs. Early detection and treatment are crucial and are emphasized in the WHO’s Vision 2020 program, which aims to reduce the global prevalence of childhood blindness significantly.

This thesis will explore the pathogenesis, epidemiology, and advancements in treating cataracts, focusing on surgical innovations and public health strategies to reduce the incidence and impact of this prevalent condition.

4. Lens

4.1 Anatomical and histological characteristics

The eye's lens is a vital optical element. Its anterior segment is transparent, flexible, and biconvex, positioned behind the iris and in front of the vitreous body. The lens is maintained in position by suspensory ligaments—radially arranged zonular fibers attached to the ciliary body (on the lateral side) and lens capsule (on the medial side)—allowing the accommodation process, which enables the eye to focus on objects at varying distances.

Histologically, the lens consists of three main components:

Lens capsule: The outermost layer is a strong basement membrane encapsulating the lens. It is the thickest basement membrane in the body, composed predominantly of type IV collagen and glycoproteins.

Lens epithelium: Situated beneath the anterior lens capsule, this single layer of cuboidal cells is crucial for the growth and maintenance of the lens fibers. These cells manage the lens's homeostasis and facilitate the transition of epithelial cells into lens fibers.

Lens fibers: These are elongated, transparent cells that evolve from the lens epithelium and make up the bulk of the lens. In their mature form, they lack organelles, contributing to the lens's transparency and organizing in a way that minimizes light scattering.

4.2 Embryonic development

The lens begins developing from the surface ectoderm, forming a lens placode that invaginates to create the vesicle. This process is closely regulated by signaling interactions between the optic vesicle and the overlying ectoderm. As the lens vesicle separates from the ectoderm, the posterior cells elongate to form the primary lens fibers. Lens development obtains nutrition via derivatives of the hyaloid artery. Later, after hyaloid artery regression, lens nourishment is supplied via the aqueous humor.

4.3 Lens metabolism

The metabolism of the lens is designed to maintain its transparency and protect against oxidative stress, largely through a unique biochemistry centered around glutathione (GSH). The lens contains an unusually high concentration of GSH, especially within the epithelium. In its predominantly reduced form, GSH plays a vital role in the lens's health and functionality.

GSH serves multiple critical functions in the lens:

4.3.1 Antioxidant protection

GSH is crucial in detoxifying hydrogen peroxide (H2O2), which is naturally present in the aqueous humor. This action helps prevent oxidative damage, which can contribute to cataract formation. The enzyme glutathione reductase helps maintain GSH in its reduced state, which is essential for its continued efficacy as an antioxidant.

4.3.2 Protein maintenance

GSH is vital in preserving the thiol groups of lens proteins in their reduced states, preventing the formation of high molecular weight (HMW) protein aggregates. This prevention is key in maintaining the clarity and function of the lens, as the aggregation can lead to opacities typical of cataracts. In conditions like X-ray-induced cataracts, the role of GSH in protecting these protein thiol groups from oxidation becomes particularly evident.

4.3.3 Ion regulation

The concentration of GSH influences the lens's permeability to cations and the activity of Na+, K+-ATPase, an enzyme critical for ion transport within lens cells. As observed in cataractous changes, decreased GSH levels can alter ion distribution, impacting protein synthesis and potentially halting lens growth.

Due to the lens's avascular nature, energy for these metabolic processes, including the synthesis and recycling of GSH, is largely derived from glycolysis. Additionally, carbohydrate metabolism supports the maintenance of GSH through the hexose monophosphate shunt, which generates NADPH, a reducing agent that assists in keeping GSH in its reduced (active) form.

Overall, the lens's metabolic activities are profoundly dependent on the efficient functioning of GSH-related mechanisms. Understanding these metabolic processes provides insights into how disruptions in these pathways could lead to lens pathology, such as cataract.

4.4 Lens function

The eye's lens plays a critical role in vision by focusing light rays onto the retina, a process that involves several precisely coordinated eye functions: accommodation, convergence, and the accommodation reflex.

4.4.1 Accommodation

This is the adjustment of the eye to different viewing distances, a key function for both near and far vision. The lens is crucial to this process, changing its convexity to adapt its refractive power for clear vision at various distances. This adjustment is achieved through the contraction of ciliary muscles, which are controlled bilaterally by the Edinger-Westphal nuclei. When the ciliary muscle relaxes, the ciliary processes become tense, leading to a decreased lens curvature for distant viewing. Conversely, when the ciliary muscle contracts, the ciliary processes relax, allowing the lens to increase curvature for near vision.

4.4.2 Convergence

Is the simultaneous inward movement of both eyes to maintain single binocular vision when focusing on close objects, such as when reading or looking at an object approaching the face. This movement is mediated by the contraction of the medial recti muscles, controlled by the oculomotor nerve (cranial nerve III).

4.4.3 Accommodation reflex

This reflex combines three synchronized eye movements: the synkinesis constriction of the pupil (miosis), the eyes' convergence, and the lens's accommodation. When an object moves closer to the eyes suddenly, preganglionic parasympathetic fibers from the Edinger-Westphal nucleus, traveling with the oculomotor nerve, prompt the ciliary muscle to contract. This increases the lens's convexity for near vision. At the same time, the iris sphincter muscle contracts to constrict the pupil (miosis), and the medial rectus muscles contract, causing the eyes to converge.

Together, these mechanisms ensure that the lens accurately focuses light on the retina, adjusting dynamically to different distances and environmental conditions. This is crucial for maintaining a clear and effective vision. This dynamic interaction underscores the lens's fundamental role in visual perception and ocular health.

5. Epidemiology of congenital cataract

Congenital cataracts are an important cause of visual impairment and blindness in children worldwide, representing a significant public health concern. The condition is characterized by an opacity of the lens present at birth or developing shortly thereafter, which can delay the passage of light to the retina, thereby disrupting vision.

5.1 Incidence and prevalence

The incidence of congenital cataracts varies significantly across different regions and populations. Studies indicate that congenital cataracts occur globally in about 1 to 15 per 10,000 live births. This variance can be attributed to differences in genetic factors, healthcare access, and diagnostic criteria among regions. The highest rates are often observed in areas where consanguineal marriages are more common and in populations with specific genetic predispositions.

5.2 Risk factors

The development of congenital cataracts is influenced by both genetic and environmental factors. Genetic mutations are the most significant cause, with many identified genes affecting lens development and clarity. Environmental factors, including maternal infections, such as rubella or chickenpox during pregnancy, can also lead to the condition. Moreover, metabolic disorders in the mother, like diabetes or her exposure to harmful substances, can increase the risk.

5.3 Challenges in public health

Effective management of the condition requires early detection and prompt surgical intervention. The availability of pediatric cataract surgery and postoperative care, which are crucial for optimal outcomes, varies widely, particularly in low-resource settings. These differences highlight the need for enhanced public health strategies, including better prenatal care and broader access to pediatric ophthalmological services.

5.4 Future directions

With ongoing research, there is potential to improve understanding and management of congenital cataracts. Advances in genetic technologies, like gene therapy, hold promise for future treatment options that could address the condition's underlying causes.

In conclusion, congenital cataracts continue to be a significant cause of childhood blindness that requires concerted efforts from healthcare systems worldwide to improve screening, treatment, and prevention strategies, especially in underserved areas.

6. Etiopathogenesis of congenital cataract

Congenital cataract can result from various etiological factors, including genetic mutations, intrauterine infections, and metabolic disorders. Understanding etiopathogenesis is crucial for diagnosing, managing, and potentially preventing these conditions.

6.1 Hereditary congenital cataract

Hereditary factors are the most common cause of congenital cataracts, often transmitted as Mendelian traits. These can be categorized based on their association with other ocular or systemic disorders or isolated cases.

6.1.1 Isolated cataract

Isolated congenital cataracts occur with no other associated ocular or systemic abnormalities. They may follow an autosomal dominant, autosomal recessive, or X-linked recessive inheritance pattern. Mutations in genes such as CRYAA, CRYBB1, and CRYGC are commonly implicated. These genes encode crystalline, which are crucial for maintaining the transparency and refractive index of the lens.

6.1.2 Cataracts associated with eye disorders

In some cases, congenital cataract occur as part of syndromes involving other eye abnormalities. For instance, cataracts associated with Aniridia involve mutations in the PAX6 gene, which plays a significant role in eye development. Other conditions may include anterior segment dysgenesis or retinal abnormalities, each requiring comprehensive ocular examination and management.

6.1.3 Cataract associated with systemic disorders

Congenital cataracts can also be a feature of systemic syndromes involving multiple organs or body systems. Examples include:

Galactosemia: An inborn error of galactose metabolism leads to the accumulation of galactitol within the lens, which promotes cataract formation.

Marfan Syndrome: A connective tissue disorder caused by mutations in the FBN1 gene, including ocular manifestations like lens dislocation and cataracts.

Down Syndrome: Trisomy 21 is associated with a higher incidence of congenital cataracts alongside other systemic and developmental challenges.

Understanding the genetic and molecular basis of these disorders aids in accurately diagnosing and managing congenital cataracts, genetic counseling, and developing potential gene therapy strategies. The approach to managing these patients is multidisciplinary, often requiring collaboration between geneticists, ophthalmologists, and pediatricians to address the full spectrum of health needs.

7. Morphology of congenital cataract

The morphology of congenital cataracts varies widely, revealing the lens's diversity in underlying etiologies and developmental disturbances. The classification based on morphology helps us to understand the probable genesis and timing of the cataract during embryogenesis and can guide management strategies. Here are descriptions of common types:

7.1 Polar congenital cataract

Polar congenital cataracts are located at the anterior or posterior pole of the lens and are typically small and disc-shaped. They can be associated with remnants of the hyaloid system or a persistent fetal vasculature. Anterior polar cataracts are usually less visually disruptive than posterior polar cataracts, which are closer to the eye's nodal point and more likely to affect vision.

7.2 Lamellar or zonular cataract

Lamellar or zonular cataracts are characterized by an opacified lens layer packed between clear layers. They often appear as concentric rings. This type suggests a disturbance in lens fiber production during a specific period of embryonic development, sparing earlier and later fibers. They are often associated with systemic illnesses that the mother may have suffered during pregnancy or with inherited metabolic diseases.

7.3 Coronary congenital cataract

Coronary cataracts are named for their appearance, which resembles crowns or wreaths. They are often seen as radial, spoke-like opacities located in the peripheral cortex of the lens. They can be isolated or associated with other lens abnormalities and are less commonly linked to systemic conditions.

7.4 Punctiform congenital cataract

Punctiform congenital cataracts are tiny, dot-like opacities scattered throughout the lens. They are often too small to significantly affect vision unless they coalesce or occur in critical areas of the lens. This type of cataract may go unnoticed and be detected incidentally during an eye examination.

7.5 Nuclear congenital cataract

Nuclear cataracts affect the central nucleus of the lens and are often significant in terms of visual impairment. This type is commonly associated with a genetic origin and can be present in conditions such as Down syndrome or other chromosomal anomalies. Nuclear cataracts are dense and typically require surgical intervention early in life to prevent amblyopia.

8. Clinical presentation of congenital cataract

Congenital cataracts manifest in various clinical presentations, depending on the lens opacity's morphology, location, and extent. The clinical picture varies from subtle visual disturbances to profound visual impairment. Early detection and management are crucial to prevent amblyopia and ensure optimal visual development. Here is a detailed examination of the clinical presentations associated with congenital cataracts.

8.1 Visual symptoms

The primary symptom of congenital cataracts is blurred or cloudy vision. However, symptoms manifest in infancy or early childhood may not be immediately apparent until visual behaviors are observed. Parents or caregivers might notice an infant not responding to visual stimuli or tracking objects poorly. Older children may express difficulty seeing or describing what they see as "looking through a fog."

8.2 Physical signs

The most prominent sign of a congenital cataract is the leukocoria, or white pupillary reflex, which is often noticed in photographs where a flash is used or during routine pediatric examinations. This is contrasted with the normal red reflex indicative of clear optical media. Other signs might include abnormal eye movements such as nystagmus, which indicates a sensory deficit from birth, or strabismus, where misalignment of the eyes occurs due to unequal visual input.

8.3 Associated conditions

Congenital cataracts can be isolated or associated with other ocular or systemic abnormalities, requiring a comprehensive systemic evaluation. Conditions such as congenital rubella, galactosemia, and Down syndrome can present with cataracts as a component of broader systemic disease. A detailed family history and genetic testing are recommended to determine if there is a hereditary pattern or associated genetic disorder.

9. Diagnosis of congenital cataract

Congenital cataract are opacities in the eye's lens that are present at birth or develop shortly after birth. Prompt diagnosis is crucial as untreated congenital cataracts can lead to permanent vision impairment and amblyopia (lazy eye). The diagnostic process involves a comprehensive ophthalmologic examination. A comprehensive ophthalmologic exam allows for accurate diagnosis, classification of cataract type, and determination of appropriate treatment plan. Early detection is vital to prevent amblyopia or other complications of the disease.

9.1 Ophthalmologic examination

The ophthalmologic examination for suspected congenital cataract typically includes the following:

9.1.1 Patient history

- Prenatal history (infections, metabolic disorders, etc.)
- Family history of congenital cataracts or other eye disorders
- History of vision problems or leukocoria (white pupil)

9.1.2 Visual acuity testing

- Age-appropriate vision testing (e.g., preferential looking, Lea symbols)
- Assessment of fixation behavior
- Red Reflex Test: A crucial initial screening that helps detect abnormalities in the refractive media of the eye

9.1.3 External eye examination

- Inspection of the eye and adnexa with good illumination
- Evaluation of pupillary light reflexes

9.1.4 Slit lamp biomicroscopy

- Detailed examination of the anterior segment
- This is vital for visualizing the lens opacity and assessing its morphology
- Important considerations include the location, density, and extent of the cataract

9.1.5 Fundus examination

- Evaluation of the posterior segment after pupillary dilation

- Assessment of the optic nerve and retina as retinal disorders can also lead to visual impairment

9.1.6 Transillumination

- Used to define the edges of the cataract and evaluate the involvement of the posterior lens capsule, which is particularly important in posterior polar cataracts where there is a higher risk of capsule rupture during surgery due to defective capsules

9.1.7 Additional testing

- Intraocular pressure measurement
- Imaging techniques (ultrasound, CT, MRI) if indicated
- Electrodiagnostic tests (ERG, VEP) in some cases
- Ultrasound B-Scan: performed when dense cataracts prevent visualization of the posterior segment from checking

for structural integrity and ruling out conditions like persistent fetal vasculature.

9.2 Additional research

For a comprehensive diagnosis, additional tests may be required:

- Photo documentation: useful for documenting the progression of lens opacities over time and evaluating the effectiveness of any interventions
- Keratometry and axial length measurement are critical for planning cataract surgery, especially in determining the power of the intraocular lens to be implanted. In children, this is often done under general anesthesia
- Genetic testing is recommended in cases of suspected hereditary patterns or associated systemic diseases, as it can provide insights into the etiology and guide management strategies

Congenital cataracts can also be associated with systemic diseases such as galactosemia, where dietary management can reverse lens changes, or chromosomal anomalies like Trisomy 21, emphasizing the need for systemic evaluation.

The comprehensive approach to diagnosing congenital cataracts ensures that all potential factors contributing to lens opacity are considered, allowing for tailored management plans that address visual and systemic health needs.

10. Treatment of congenital cataract

10.1 Indications for operation

Congenital cataracts are primarily indicated for surgical intervention when they are visually significant and obstruct the normal visual development of the child. The primary indications include:

- Visual axis obstruction: cataracts obstruct the central visual axis and impair the child’s ability to fixate and follow objects

- Bilateral dense cataracts: especially when presenting in infancy, which can lead to severe visual impairment and amblyopia if not treated promptly
- Unilateral cataracts: presenting before the child reaches 4-6 weeks old to prevent the onset of irreversible amblyopia.
- Cataract associated with other ocular anomalies, such as nystagmus or strabismus, where early intervention may improve the visual prognosis

10.2 Ideal age for operation

The timing of the surgery is crucial for optimal visual outcomes:

- Bilateral congenital cataracts: surgery is typically recommended between 6-8 weeks of age to avoid amblyopia and allow for better visual development
- Unilateral cataracts should ideally be operated on before the infant reaches 4-6 weeks of age due to the higher risk of amblyopia associated with these cases
- older children: with visually significant cataracts, surgery should be planned as soon as possible once the diagnosis is made and the child is fit for general anesthesia

10.3 Classifications of congenital cataracts

Congenital cataracts can be classified based on their morphology, location, and etiology:

10.3.1 Morphological classification

Total cataract: complete opacification of the lens.

- Zonular or lamellar cataract: opacities that are concentric around the nucleus.
- Nuclear cataract: opacification of the central nucleus of the lens.
- Posterior polar cataract: located at the back of the lens, often associated with posterior capsular involvement
- Anterior polar cataract: opacities on the front surface of the lens
- Sutural cataract: opacities along the Y-sutures of the lens

- Posterior subcapsular cataract: located just in front of the posterior capsule of the lens

10.3.2 Etiological classification

Genetic: hereditary cataracts linked to specific gene mutations

- Infectious: resulting from maternal infections during pregnancy (e.g., rubella, toxoplasmosis)
- Metabolic: associated with metabolic disorders (e.g., galactosemia)
- Traumatic: resulting from ocular trauma during or after birth

10.4 Preoperative processing

Preoperative preparation involves several steps to ensure the safety and effectiveness of the surgical procedure:

- Mydriasis and cycloplegia: achieved using 2% homatropine eye drop a day before surgery or 1% atropine eye ointment twice daily for three days preoperatively. This helps in sustaining the mydriasis
- Antibiotic prophylaxis: topical antibiotics such as 0.3% tobramycin or 0.5% moxifloxacin are administered four times daily for three days preoperatively
- Antiseptic preparation: one drop of 5% betadine solution is applied immediately before surgery
- Systemic prophylaxis: intravenous mannitol may be administered to reduce vitreous volume, especially in cases requiring posterior capsulotomy
- Fasting: ensuring the child is nil orally as per anesthesia guidelines

10.5 Anesthesia

General anesthesia (GA) is the standard for pediatric cataract surgery due to the need for complete immobility and the challenges posed by pediatric anatomy:

Preoperative assessment: this includes evaluating any associated systemic conditions that may affect anesthesia, such as congenital heart defects in children with maternal rubella or thromboembolism risk in homocystinuria.

Induction and maintenance: careful induction and maintenance of anesthesia are recommended to prevent rises in intraocular pressure. Laryngeal masks are also recommended for secure airway management.

Pain management: postoperative pain is usually managed with non-steroidal analgestics, avoiding opioids due to their side effects. subtenon blocks and topical anesthetics can also be used to manage perioperative pain.

10.6 Operation

The surgical procedure for congenital cataracts involves several key steps:

Capsulorhexis: creating a continuous curvilinear capsulorhexis (CCC) is crucial for long-term intraocular lens (IOL) centration. Due to the elasticity of the pediatric capsule, this step requires precision and experience.

Lens Aspiration: hydrodissection mobilizes the lens material, followed by bimanual irrigation and aspiration to remove the lens cortex and nucleus.

Posterior capsulotomy and anterior vitrectomy: often necessary to prevent posterior capsular opacification (PCO). This step is typically performed using a vitrector through the pars plana or limbus.

IOL Implantation: primary IOL implantation is generally recommended for children over six months of age. Secondary IOL implantation is considered around 3-5 years of age if initial surgery was performed without IOL placement.

Wound closure: ensuring all incisions are securely closed to prevent postoperative complications such as wound leakage and iris prolapse.

10.7 Postoperative treatment

Postoperative care for children following cataract surgery is critical to ensure optimal recovery and to prevent complications. Key aspects include:

Medication: postoperatively, children have typically prescribed a regimen that includes 0.3% gatifloxacin (or 0.5% moxifloxacin) and 1% prednisolone acetate eye drops six times a day for four weeks, along with 1% cyclopentolate eye drops twice daily. Alternative medications include difluprednate 0.05% instead of prednisolone acetate and homatropine 2% instead of cyclopentolate.

Bandaging and monitoring: the operated eye is bandaged for 24 hours. Close monitoring is essential, with daily examinations for the first three days, followed by weekly check-ups for a month, and then at three to six-month intervals.

Inflammation control: in cases where significant postoperative inflammation is expected, such as with associated uveitis, a subconjunctival or subtend on triamcinolone acetonide injection may be administered.

Follow-up: children are examined frequently postoperatively to check for complications like posterior capsular opacification (PCO), glaucoma, and retinal issues. The schedule typically includes visits on postoperative days 1 and 2, then weekly for the first month, and subsequently at increasing intervals depending on the child's recovery.

10.8 Corrections of sight

Following cataract surgery, children require appropriate correction to ensure proper visual development. The main options are optical lenses, such as contact lenses and intraocular lenses (IOLs).

10.8.1 Spectacles

Types: spectacles are a common method of optical correction for both aphakia and pseudophakia. Children under 2 years old are typically prescribed near correction lenses to aid with near activities. As they grow older, bifocals or progressive lenses may be used.

Material: polycarbonate lenses are preferred due to their shatter-resistant properties and inherent UV protection. Flexible plastic frames are recommended to minimize breakage and ensure proper fit.

Compliance: it is crucial to ensure children wear their spectacles consistently. Strategies to improve compliance include using headbands to secure the spectacles and choosing larger frames to prevent peeking over the top.

10.8.2 Contact lenses

Advantages: contact lenses provide superior optical resolution, especially for unilateral aphakia. They can be customized and changed as the child's eye grows, making them suitable for young children.

Types: the main types of contact lenses used in children are rigid gas permeable (RGP), hydrogel, and silicone elastomer lenses. Each type has advantages and disadvantages regarding safety, ease of use, and comfort.

Challenges: contact lens use in infants can be challenging due to difficulties with insertion and removal, maintenance of hygiene, and compliance. Parental education and motivation are critical for successful contact lens use.

10.8.3 Intraocular lenses (IOLs)

Primary IOL implantation: this method is preferred for children over 6 months without other ocular comorbidities. IOLs provide a stable and continuous correction, eliminating the need for glasses or contact lenses.

Considerations: the timing of implantation and accurate power calculation are crucial due to the dynamic nature of the growing eye. Over time, the eye's axial length changes, necessitating adjustments in refractive correction.

Secondary IOL implantation: secondary IOL implantation is typically performed after 2 years of age for children initially left aphakic. This approach accounts for most eye growth and stabilization of the corneal curvature, making it easier to achieve accurate biometry and optimal IOL placement.

10.9 Occlusions

Occlusions are an essential part of the postoperative management in pediatric cataract surgery, particularly for preventing and treating amblyopia. Amblyopia, also known as "lazy eye," occurs when one eye develops poor vision due to abnormal visual development early in life. This section discusses the types and protocols of occlusion therapy used in managing pediatric cataract patients.

10.9.1 Types of occlusions

Total Occlusion: this involves completely covering the better-seeing eye with a patch to force the use of the amblyopic eye. Depending on the severity of amblyopia, it is often recommended for several hours a day.

Partial occlusion: this method uses occlusive contact lenses or filters over glasses that partially obscure vision in the better-seeing eye. It can be more comfortable and cosmetically acceptable, improving compliance.

Penalization: involves using atropine drops in the better-seeing eye to blur vision, encouraging the use of the weaker eye without needing a physical patch. This is especially useful in children who resist wearing patches.

10.9.2 Protocols and monitoring

Duration and frequency: occlusion therapy typically starts with a few hours per day, increasing based on the child's response and age. The duration is adjusted according to the improvement in visual acuity.

Monitoring: regular follow-up visits are essential to assess the effectiveness of the occlusion therapy and adjust as necessary.

This includes measuring visual acuity and ensuring no reversal of amblyopia to the previously better-seeing eye.

10.10 Postoperative complications

Postoperative complications in pediatric cataract surgery can significantly impact the visual outcome and overall prognosis. Prompt identification and management of these complications are crucial.

10.10.1 Early complications (within 4 weeks of surgery)

Increased intraocular pressure (IOP): commonly managed with topical medications. If persistent, surgical intervention may be required.

Inflammation: more intense in children than adults, it can lead to fibrin formation and secondary membranes. It is managed with aggressive anti-inflammatory therapy.

Endophthalmitis: a rare but severe infection requiring immediate intervention with antibiotics and possibly vitrectomy.

Wound leakage: can cause a shallow anterior chamber and hypotony, requiring prompt repair.

10.10.2 Late complications (after 4 weeks of surgery)

Visual axis opacification (VAO): common in children due to the proliferation of lens epithelial cells. Prevented by primary posterior capsulotomy and anterior vitrectomy during initial surgery.

Secondary Glaucoma: requires lifelong monitoring as it may develop years after the initial surgery.

Retinal detachment: though rare, it necessitates immediate surgical intervention to prevent permanent vision loss.

IOL-related complications: includes decentration, dislocation, and pupillary capture, which may require surgical correction.

10.11 Prognosis

The prognosis for children undergoing cataract surgery is influenced by several factors, including the timing of surgery, the presence of additional ocular or systemic abnormalities, and adherence to postoperative management protocols.

Timing of surgery: early intervention, ideally within the first few months of life, is crucial for preventing amblyopia and ensuring optimal visual development. Delays in surgery can lead to irreversible vision loss.

Associated conditions: the presence of other ocular conditions, such as persistent fetal vasculature or systemic conditions like metabolic disorders, can adversely affect the visual outcome.

Postoperative care: rigorous postoperative care, including timely correction of refractive errors, consistent use of occlusion therapy, and management of complications, plays a critical role in determining the long-term visual prognosis.

Parental involvement: active participation by parents in following the prescribed postoperative regimen is essential for achieving the best outcomes.

11. Conclusions

The treatment of congenital cataracts in children is a complex and multifaceted process that demands a comprehensive approach involving early diagnosis, timely surgical intervention, meticulous postoperative care, and continuous visual rehabilitation. This thesis has explored the various dimensions of managing pediatric cataracts, emphasizing the importance of each stage in ensuring optimal visual outcomes.

Early diagnosis and intervention

Early diagnosis of congenital cataracts is crucial to prevent amblyopia and other complications that can result in permanent visual impairment. Routine neonatal screenings, such as the red reflex test, are essential for early detection. Upon identifying abnormalities, immediate referral to a pediatric ophthalmologist can significantly enhance the prognosis by facilitating timely intervention.

Surgical considerations

The ideal timing for cataract surgery is critical, particularly for congenital cataracts. For bilateral cataracts, surgery is typically recommended between 6-8 weeks of age, while unilateral cataracts should be operated on before the infant reaches 4-6 weeks old to prevent amblyopia. The surgical technique must be tailored to the specific type and severity of the cataract, with considerations for primary or secondary intraocular lens (IOL) implantation depending on the child's age and ocular conditions.

Postoperative management

Postoperative care is vital for ensuring successful visual outcomes. This involves a regimen of anti-inflammatory and antibiotic eye drops, and regular monitoring for complications such as increased intraocular pressure, endophthalmitis, and visual axis opacification (VAO). The role of occlusion therapy in managing amblyopia post-surgery is highlighted, with strategies tailored to individual patient needs.

Visual rehabilitation

Post-surgical visual rehabilitation includes using spectacles, contact lenses, or IOLs to correct refractive errors. The choice of corrective measures depends on factors such as the child's age, compliance, and specific visual needs. Regular follow-ups and adjustments are necessary to adapt to the changing visual requirements as the child grows. The involvement of low-vision rehabilitation teams can significantly enhance the quality of life for children with suboptimal visual outcomes despite surgery.

Long-term prognosis

The long-term prognosis for children with congenital cataracts depends on several factors, including the timing of surgery, the presence of additional ocular or systemic abnormalities, and adherence to postoperative care protocols. Early intervention and consistent management can significantly improve visual acuity and overall visual development, enabling children to integrate into mainstream education and daily activities more effectively.

In conclusion, managing congenital cataracts in children requires a multidisciplinary approach that combines surgical precision, diligent postoperative care, and robust visual rehabilitation strategies. By addressing each aspect comprehensively, healthcare providers can significantly improve affected children's visual and developmental outcomes, thereby enhancing their quality of life and future prospects.

12. Literature

This literature review provides a comprehensive overview of the current knowledge regarding congenital cataracts in children, focusing on epidemiology, etiology, clinical presentation, diagnosis, treatment, and prognosis.

The scope of this thesis is focused on the most recent books, articles, and case studies published in the last three years.

Books/Textbooks:

- Pediatric Ophthalmology and Strabismus by Drs. David Taylor and Creig S. Hoyt (Popular pediatric ophthalmology textbook)
- Ophthalmology by Drs. Myron Yanoff and Jay S. Duker (Comprehensive ophthalmology reference)

Review Articles:

- Childhood cataracts: Etiology, clinical features, and management (Lim et al., Nature Reviews Disease Primers, 2021)
- Pediatric cataract: Challenges and future directions (Vasavada et al., Eye, 2012)

Professional Guidelines:

- Pediatric Eye Evaluations: I. Vision Screening in the Healthy Child (American Academy of Ophthalmology, 2022)
- Childhood Cataract (American Association for Pediatric Ophthalmology and Strabismus, Guidelines).

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