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A SYSTEMATIC REVIEW OF LITERATURE TO ASSESS THE USE OF INTRAOCULAR LENSES FOR TREATING APHAKIA IN ADULT AND PAEDIATRIC PATIENTS

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ABSTRACT

Background: Aphakia is a condition that involves the absence of the crystalline lens of the human eye. The leading cause of aphakia is cataract in the adult patients or the absorption of the lens after trauma in paediatic patients. The use of intraocular lenses, remain a major surgical treatment technique for the treatment of patients with aphakia.

Aim: The aim of this study is the systematic review of the literature in order to assess the use of intraocular lenses for the treatment of aphakia, in adult and paediatric patients.

Methods: A literature search was conducted in the databases Medline (Pubmed), Cinahl, Scopus and Cohraine using the following algorithm: “intraocular lenses” OR “Anterior Chamber Intraoccular lenses” OR “Posterior Chamber Intraocular Lenses” AND “Aphakia” OR “Cataract extraction” OR “Complications” OR “Visual rehabilitation”.

Conclusions: the use of intraocular lenses for the treatment of the aphakic eye, has generally been found to improve the most prominent outcome measure, the visual acuity of the patients in the post-operative period. However, the proposed surgical techniques have also been implicated for several complications including increase in intraocular pressure, papillary distortion, retinal detachement, corneal decompensation and cystoid macular oedema.

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1. Introduction

In ophthalmology, the term “aphakia” is used to describe any conditions that result in the absence of the eye’s crystalline lens. This pathological condition may be the result of post-surgical complications such as surgical intervention to correct visually significant cataract, the result of a trauma or a perforating wound, lens dislocation or sublaxation, or less frequently, the result of a congenital anomaly (Khurana et al., 2019). A serious complication that occurs with increased frequency in patients with aphakia is puppilary block, a complication that follows the extraction of round-pupil cataract without, however the sector iridectomy. There is also the distinct possibility that papillary block occurs during round-pupil extraction where irididectomy was performed but was placed in the extreme periphery and was small in extent (Barnir et al., 2018).

Aphakia causes high degree of hyperopia (farsightedness), a loss of accommodation and deep anterior chamber. Further complications may onlcude glaucoma or detachment of the retina or the vitreous (Plager et al., 2011). The occurrence of aphakia in infants is extremely rare, instead the pathology occurs as a post surgical complication to correct congential cataract. Congenital cataracts develop usually as a result of genetic modification in certain chromosomatic areas or may also be the result of an infection of the fetus. The exact causality of congenital cataracts is usually difficult to be clearly identified, especially in cases where only one eye is affected by the condition (Santana et al., 2011). A characteristic clinical manifestation of patients that suffer from aphakia is that their pupil are relatively small in comparison to pupils of healthy populations of the same age group and that the pupils tend to dilate at a smaller degree (Figure 1) (Scholtz et al., 2016).

The main causes of aphakia are the following:

- the surgical removal of the lens during cataract correction surgery
- the surgical removal of the clear lens to treat high myopic patients
- absorption of lens in children after trauma
- spontaneous absorption of the lens die to trauma (either dislocation of the lens into vitreous or traumatic exclusion of the lens from the eye)

- congenital absence of lens in congenital primary aphakia (rare genetic disorder)
- sublaxation or dislocation of the lens due to trauma or congenital anomalies (Anjum et al., 2010, Lindsay et al., 2010)

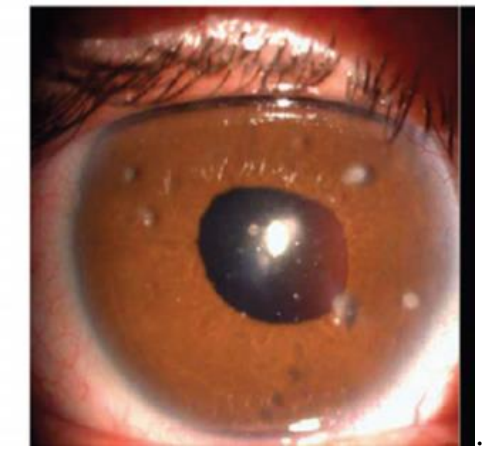


Figure 1. Aphakia after trauma, relatively small pupil

As mentioned above, the main cause for aphakia (aphakic glaucoma) is the application of early cataract surgery. More specifically, the underlying pathophysiology is based to the theory that early lensectomy is believed to conflict with maturation of the trabecular meshwork. More particularly, in healthy population the development of normal meshwork requires structural interactions between trabecular meshwork, ciliary body, zonules and the native lens (Chen et al., 2004). There is also the theory that aphakic glaucoma may be the result of chronic trabeculitis, Chronic trabeculitis is a condition that describes the inflammatory obstruction of the trabecula meshwork that causes severe angle closure and may be a secondary surgical complication that is caused by blockade of the trabecular network by retained material of the lens or by the development of postoperative inflammation (Figure 2) (Phelps et al., 1977).

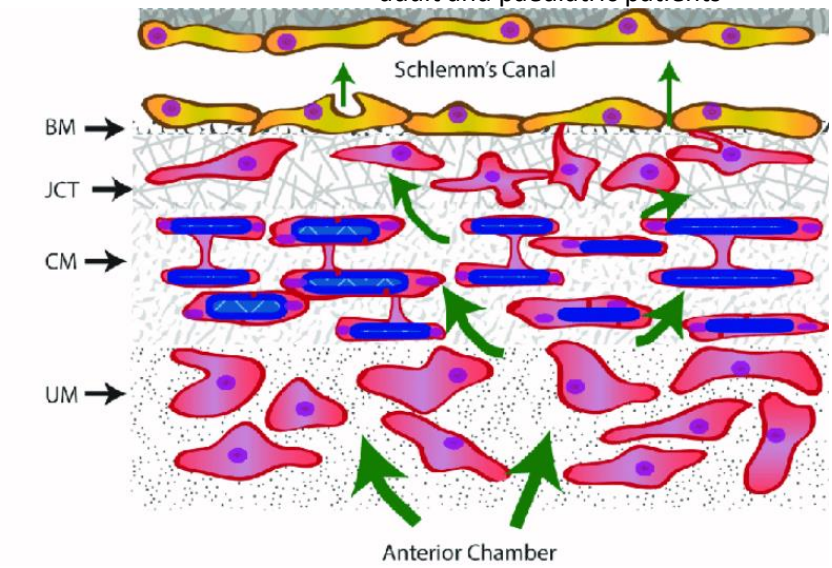


Figure 2. Schematic representation of the trabecular meshwork

Aphakic glaucoma is reported as the second most common form of open angle glaucomas and is also the second most common glaucoma in epidemiological studies that focus on paediatric patients (Olitsky et al, 2017). The most important risk factor that correlate with the manifestation of aphakic glaucoma include surgery within the first year of life, retained lens proteins, measurement of the corneal diameter less than 10mm, certain forms of cataract such as nuclear, complete or persistent hyperplastic vitreous), history of secondary surgeries or other ocular co morbidities (Baily et al., 2012). The most prominent of the above risk factors remains up to date the application of cataract surgery at young age. In a recent study by Olitsky et al., a sample of 137 patients was used that had surgery to correct congenital cataract. The same study reports that 12% of those patients developed aphakic glaucoma in a timeframe of mean 9,6 years after the operation. A meta-anaysis cited in the same study that used a large sample pool of 470 patients, reported that 17% of children that underwent surgery to correct infant cataract developed aphakic glaucoma. The risk was significantly higher in patients that underwent surgery at a an age less than 4 weeks and in infants that suffered from the condition known as micropthalmos (Olitsky et al., 2017).

Genetic anomalies have also been proposed as a risk factor for the condition. A large number of genes is reported as potential risk factors both for the development of glaucoma and cataracts. The most prominent of the reported genes is the PAX6 gene (Kirwan et al., 2006).

Furthermore, a higher concentration of lens proteins may remain after lens extraction and may further degenerate into lens byproducts that have a toxic effect to the trabecular meshwork. In a related literature review by Chen et al., a 15% of the patients that underwent early cataract surgery were found to retain lens material after the surgical procedure while the same study reported past researches with percentages up to 41,6-78% (Chen et al., 2004). The clinical presentation of aphakic glaucoma is typically an open-angle glaucoma, a fact that further supports the theory of the proposed interaction between lens epithelial cells and/or vitreous with the cells of the trabecula meshwork. This interaction is also believed to lead to the development of elevated intraocular pressure (Jaganathan et al., 2016).

Congenital aphakia is a rare genetic disease, however despite the small percentage of its frequency in the general population it is further divided into two sub-groups:

- primary congenital aphakia and
- secondary congenital aphakia

In primary congenital aphakia there is no lens development while in the secondary congenital aphakia a lens is developed to a certain degree, however the lens is extruded or resorbed through corneal perforation during or before birth time. Primary congenital aphakia can be developed after the partial development and the invagination of the optic cup, as it has been reported by several histological studies that examine the human embryo at the 13 mm stage. The studied eyes demonstrated serous congenital deformities(Valleixetal.,2006).
The first signs of the development of lens in the embryo are the thickening of the surface ectoderm in the area where the ectoderm comes in contact with the primary optic vesicle. In this phase of embryonic development, the ectodermal cells have a cylindrical form their nuclei move to base of the cells. It is generally reported that in lens development an important part is being played by specific lens proteins that are detected during this placodal stage of lens development (Medina-Martinez et al., 2007).

Mesodermal cells move between the anterior wall of the lens vesicle and the surface ectoderm and form a directional membrane. It is clear that the formation of the directional membrane depends highly on the presence of the lens vesicle. For this reason it is not recommended for primary congenital aphasia to be diagnosed in eyes

that do not appear deformed in the anterior segment (Narroie et al., 2009). The above reasons result in the complete aplasia or the incomplete development of the anterior segment of the eye. So far, the underlying genetic causes of priamary and secondary congenital aphakia remain obscure, however, they are mutifactorial since the development of the lens is a complex process that involves interactions between tissues that are evolved from the neural crest mesechyme, the neural ectoderm and the the surface ectoderm (Graw et al., 2003).Many genes, including homebox genes, signaling molecules, and transcription factors are involved in the various stages of lens development. Furhtermore, mutations in the FOX3 gene, a transcription factor specific to the lens appear to play a critical role in defective formation of the lens (Valleix et al., 2006).

The main clinical manifestations of aphakia are briefly described in Table 1.

Table 1. main signs and symptoms of aphakia

Clinical sign	Symptoms
Hypermetropia	Farsighted eye that has lost its focus power
Loss of accommodation	Lens and zonules are responsible for adjusting the focus of vision to different lengths, so patients with aphakia have a total loss of accommodation
Defective vision	Loss of accommodation and hypermetropia cause defective vision
Cyanopsia	Absence of lens causes blue vision
Erythropsia	Reddish appearance of objects
Deep anterior chamber	Due to lens absence, deepening of the anterior chamber is evident
Iridodonesis	Agitated motion or vibration of the iris during eye movement
Iridectomy mark	Surgical aphakia
Astigmatism	Caused by corneal wound healing in surgical aphakia

A major participator of the refractive system of the human eye is the crystalline lense, a transparent lense that is in fact a bincovex, arched structure. In cases of metabolic diseases, diffuse opacification or localized opacification may occur in the lens, a pathology also known as cataract. One of the most prominent rehabilitation techniques of cataract is the use of intraocular lenses (IOL), that can be surgically placed either in the posterior or in the anterior chamber of the eye. The use of anterior chamber IOLs have been reported since the 1960s, however this particular technique was abandoned during the last two decades, due to the fact that the incidence of complications was exceptionally high and the progression of the cataract surgery was not favorable (Girard et al., 2003).

Since the development and application of anterior chamber IOLs, a great number of modifications has been made regarding their design. Initially the lenses' design was a biconcave form, which was later abandoned for a more convex-concave design. The modification to the lenses' design was conducted in order to ensure the protection of the corneal endothelium, a structure extremely susceptible to lens-induced damage. This clinical indications for the use of IOLs to treat cataract induce aphakia include intracapsular lens ectraction, intraoperative complications, penetratinc kerapoplasty and secondary implantation (Girarf et al., 2003).

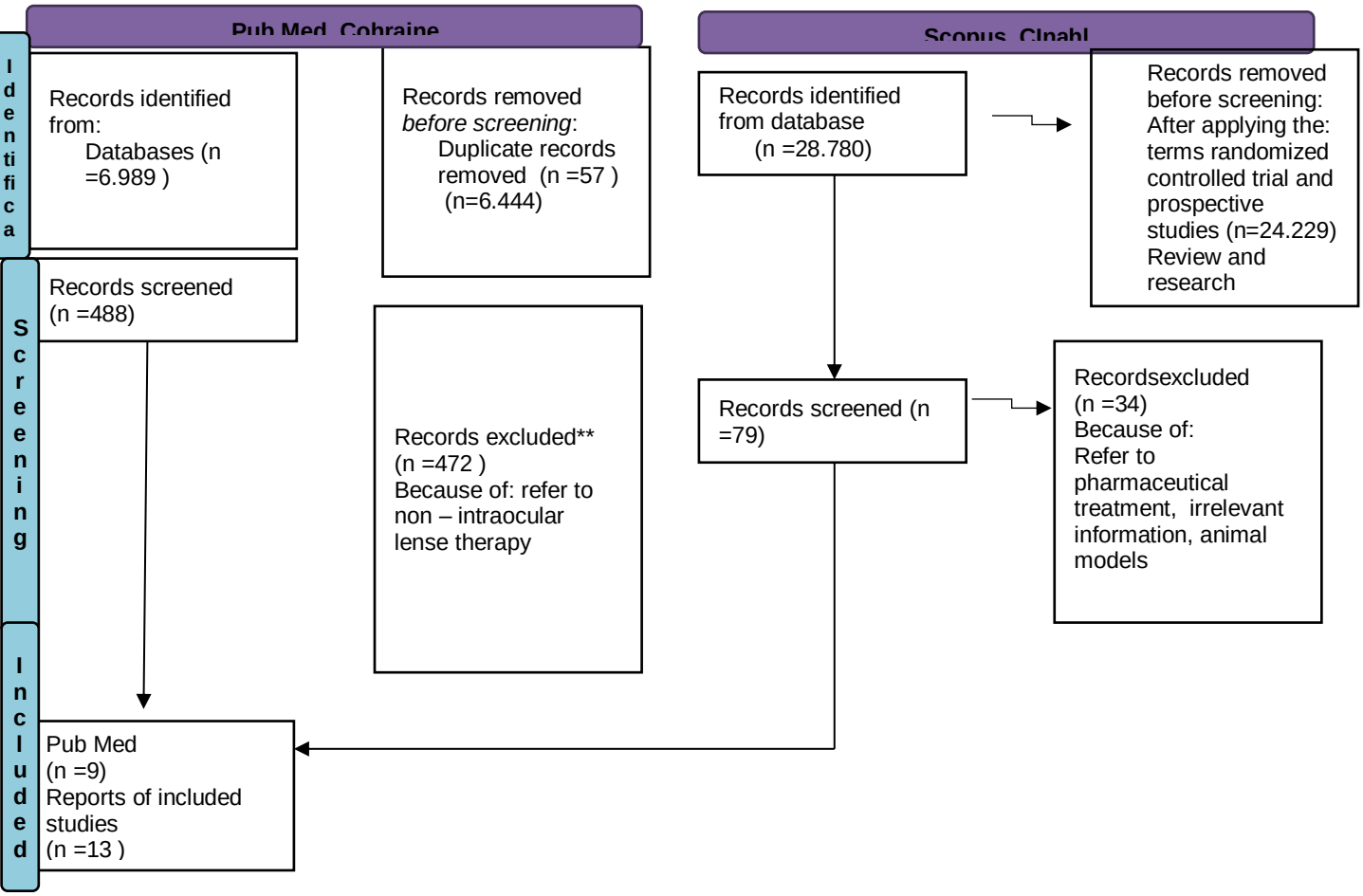
2. Methods

The aim of this systematic review is the assessment of the latest literature evidence regarding the use of intraocular lenses to treat the pathologies that are included under the term aphakia, as it is described in the introduction of the article. In order to obtain the latest published research, a search was conducted in the databases Medline (Pubmed), Cinahl, Scopus and Cohraine using the following algorithm: "intraocular lenses" OR "Anterior Chamber Intraoccular lenses" OR "Posterior Chamber Intraocular Lenses" AND "Aphakia" OR "Cataract extraction" OR "Complications" OR "Visual rehabilitation".

The inclusion criteria were the publication date (last decade), the language of publication (English), and studies that were focused in the rehabilitation or treatment of adult or paediatric patients that were diagnosed with aphakia. Studies that assessed

the other treatment approaches that did not include the use of intraocular lesnses were excluded. Furthermore, studies published in previews decades, as well as studies conducted in animal model were excluded. The initial search provided 79 studies that were related with the treatment of aphakia, from which the abstract was studied. From these 79 studies, 45 were selected and the full text was studies to produce a number of 13 studies that were included in the results of the current systematic review. This process is depicted in the PRISM flowchart of Figure 1.

Figure 1. PRISMA type flowchart of the systematic review of literature



3. Results

IOLs have two distinct categories and more specifically these categories are iris/iris-claw supported or angle-supported. The iris/iris-claw supported lenses are fixed in the anterior chamber in the anterior iris surface while the angle-supported lenses are attached in the anterior chamber of the eye by four haptic points (Kohnen et al., 2014). The most frequently used IOL is the Worst-Fechener lens also, referred to as the Artisan lens and was initially used in 2004, when the lens was approved by the FDA for the first time. Intraocular lenses also include other type of lenses that are used with smaller frequencies such as the open – loop intraocular lenses and the angle-supported Kelman Multiflex lenses (Kohen et al., 2014).

3.1 Artisan iris-claws IOLs

Artisan lens were first introduced for aphakia rehabilitation since the early 1970s. They have a lenth of 8.5 mm and for their fixation, two flexible appendages are used that are connected to the anterior chamber. The lens itself is made of PMMA (polymethylacrylate) and has a refractive power that extends from 2-30 D (increments of 1-D) or 14.5 -24.5 D (increments of 0.5-D). The fixation of the lens is centered over the pupil, in the periphery of the iris, in order to protect the mydriasis function and to preserve the finite angles. It is of great importance to mention that during the procedure to implant the lens, no sutures are required, but there is a possibility for pupil reformation through suture pupilloplasty. Pupilloplasty is sometimes necessary for the exact placement of the lens in the desired axis (Schallenberg et al., 2014).

In the study by Helvaci et al., a comparison was made between implantation of the artisan lens (iris claw) in the retropupillary area in patients diagnosed with aphakia. The study measured the visual acuity in two aphakic patient groups and revealed statistically

significant improvements. Other noteworthy study findings included similarities in the ireegularity of the pupil (3-5% prevalence) and a temporary but significant increase in the intraocular pressure. However, no significant differences were observed between the two groups that differed in the technique used to fix the lens Artisan lens (Helvaci et al., 2016).

Similar results are also observed in the study by Teng et al., where the main outcome measure was the loss of endothelial cells after the installment of Artisan lens in the posterior chamber of the eye. The study reported no statistically significant differences in the endothelial cell loss, and that the visual acuity was also reported in the iris claw lens group in comparison to the group treated with IOL in the posterior chamber. Furthermore, this particular research emphasizes in the postoperative complications that included spontaneous lens dislocation, papillary distortion and depigmentation in the iris claw lens group while the IOL posterior lens group experienced introaocular hemorrhage, retinal detachment and choroidal detachment (Teng et al., 2014).

In the research paper by Guel et al., the results show that the iris claw artisan technique is a promising rehabilitation methodology for aphakia. The research included a sample of 128 aphakic eyes that had no capsular support. The outcome measures included the correction of the visual acuity and showed that it was significantly improved 1 year after the procedure and this improvement was sustained for another 5 years, as was reported by a follow up research. The research also reported surgery complications that included penetrating keratoplasty, increased intraocular pressure and cystoids macular oedema (Guel et al., 2014).

Kherikhah et al., presented an approach that used Artisan IOLs to correct aphakia that is caused by the condition known as Fuch's heterocromic iridoclytis. The research used the case study methodology, where a patient that was treated with this approach had a 20/20 visual acuity, one month after the operation, and the main complications included few deposits in the lense's surface. The follow up study showed stability on the lens attachment and no sublaxation, iris atrophy, cystoid macula edema or vitreus inflammation (Kherikhah et al.,2014).
On the other hand, Chen et al., conducted a study to assess the safety and efficacy of the Artisan IOL to correct aphakia in patients that had no capsule support. This particular study was a cohort study and included 72 patients. The proposed curing

modality utilized lens fixation on the secondary anterior iris area. The main outcome measure was the improvement of the visual acuity and was measured periodically in a 3 year time frame. The results showed a significant improvement in the majority of the patients and only two patients reported complications: retinal detachment and ischaemic optic neuropathy. The authors did not observed any statistically significant changes in the postoperative intraocular pressure and the reported mean rate of endothelial loss was 9,78% (Chen et al., 2012).

In a recent study by De Silva et al., the intraocular implantation of IOLs when used to treat primary lens operations or when used at a secondary procedure dollowing a dislocated IOL. The sample size consisted of 116 patients and the study timeframe was approximately 22 months. Similarly to the above researches, the outcome measure was the visual acuity that was improved in the majority of the patients (69%), while the main postoperative complications involved IOL sublaxation (6,0% of the patients), increase in the intraocular pressure (9,5%), wound leak and consequent re-suture (2,6%) and cystoids macula oedema (7,7%) (De Silva et al., 2011).

3.2 Open loop Intraoccular lenses for the treatment of aphakia

A surgical approach that included the attachment of open-looped IOLs in the iris was also proposed in order to improve instability of the IOLs but also to decrease the corneal damage that is progressively manifested after the aphakia correction with IOLs. The most recent study that proposed this particular modality was conducted by Dai et al., and involved 18 patients and a total of 20 aphakic eyes where open-lop secondary flexible IOLs were implanted. The lenses were maintained by the patients for a mean period of 6 years, and the main complications that were reported included lens dislocation, retinal detachment and secondary glaucoma. The main outcome measure was the assessment of visual deterioration, and the results concluded that none of the included patients reported any degree of visual deterioration in the 6 years timeframe (Dai et al., 2011).

3.3 The assessment of foldable acrylic IOLs for the surgical treatment of aphakia

Giles et al., proposed the use of angular supported foldable acrylic IOL in 21 patients that had unilateral aphakia. The study included 21 patients and the visual acuity of the patients was improved by 0,78 ± 0,57 in the postoperative period. Postoperative complications were reported in some cases (14,3% of the patients) and the most prominent ones were puppilar ovalization, retinal detachment and macular oedema. The study concluded that the use of foldable acrylic IOLs for the correction of aphakia is a promising technique that improves visual acuity without major complications (Giles et al., 2013).

These findings are in agreement with the finding of Omulecki et al., that used foldable acrylic IOLs. In this case, the proposed method attached the IOLs by a small incision and the visual acuity increased. However, the reported complications were more serious and were observed in a bog percentage of the patients. The most frequently reported post-operative complications included increased intraocular pressure, cystic macular oedema, reduction of the endothelial cell density, hyphaemia, distrorted pupil shape, blood in the vitreous, displacement of the IOLs and Descemet folds. However, it is concluded that these complications had no effect on the improvement of the visual acuity that is the main outcome measure (Omulecki et al., 2011).

3.4 Other studies

Sulves et al., proposed the use of the MTA3UO IOLs as well as the use of L122UV IOLS for patients with aphakia that had inadequate capsule support and in patients with a former history of inflammation in the intraocular area. These IOLs are constructed using a acrylic biomaterial that is derived from one particular type of polymer. The acrylic biomaterial has distinct chemical properties and more specifically in contains both hydrophilic and hydrophobic groups. The MTA3UO IOL is an open – loop convex-plan lens while the L122UV is a lens with a vaulted equiconvex design. The study supported the efficacy of the proposed methodology in the visual acuity of the patients. The procedure was also reported to be same and the reported complications were similar to the postoperative complications reported by most of the researches (Suleves et al., 2014).

The implantation of air-assisted IOLs is also proposed as a highly effective and safe technique for the treatment of aphakia, especially in patients that suffer from

progressive subluxation of the lens. More specifically, the mean improvement of the visual acuity was from 0,67 to 0,25 logMAR, and no decrease was reported to the intraocular pressure. Furthermore, the authors reported a decrease in the count of the corneal endothelium at 6 months in comparison to baseline measurements (Agarwal et al., 2015).

3.5 Surgical management of aphakia in paediatric patients using IOLs.

In the past years, transceral fixation of IOLs that consisted of one or three pieces was proposed for the treatment of aphakia in paediatric patients. Shuaib et al., reported satisfactory results regarding the improvement of visual acuity (0,69 ± 0,69 to 0.12 ± 0,13) in a follow up of 24 months. However, the authors report a high percentage of IOL dislocation caused by ruptures of the suture. This postoperative complication was reported in 6 of the 25 aphakic eyes of paediatric patients that used this particular modality (Shuaib et al., 2019). These findings are also supported by Sen et al., that also reported improvement of visual acuity and that a great percentage of the patients presented IOL dislocation with the same causality (Sen et al., 2018).

3.6 Intrascleral fixation without sutures

The postoperative complications reported in the use of sutures for the attachment of the IOLs led to the development of novel surgical approached regarding the fixation of the IOLs in the sclera area, in paediatric patients. Kumar et al., proposed an approach where single or three piece IOLs are glued without sutures intrasclerally. The improvement in the visual acuity were similar to the trasccrelar fixation methods, however , the dislocation of the IOLs due to suture rupture was not reported. In this case the most frequently reported post-surgical complication included intraocular haemorrhage, one case of hypotony, and two cases of IOL decentration. The reported complications have relative low frequency, if the number of the sample size is taken into account (41 aphakic eyes) (Kumar et al., 2012).

In a case series study that was presented recently by Sternfield et al., the Yamane technique was adapted for use in paediatric patients. This technique involved the use of

30G thin needle to externalize the haptics. Then the haptic is bradened into a flange and is depressed into the intracleral area. The case series used a sample size of 12 eyes of 10 paediatric patients. The findings concluded statistically significant improvement of visual acuity, insignificant fluctuation of intraocular pressure and rarely reported postoperative complications such as IOL sublaxation and mil hypotony (one patient) and choroidal effusion that did not present a clinical leakage (one patient) (Sterbfeld et al., 2020).

3.7 Iris – claw IOLs fixated in the anterior area for the correction of paediatric aphakia

An alternative surgical approach for the correction of aphakia in children with inadequate capsular support is the installment of the Artisan IOLs, in a procedure that is similar to the procedure proposed to the adult patients. This modality has the advantage of the preservation of the entothelial cell count. In the recent study by Gawdat et al., encouraging outcomes are reported regarding the visual outcomes and more specifically the mean visual acuity differed from 0,36 ± 0,26 to 0,04 ± 0,09 logMAR during a 12 months follow up. The study compared two groups of paediatric patients with aphakic eyes that underwent Artisan IOL installment surgery for the treatment of traumatic cataract. The authors reported better results regarding the surgical repair using iris-claw IOLs, however the loss of endothelial cells was not mitigated. For this reason, longer follow-up studies are proposed and deemed as necessary (Gawdat et al., 2015).

3.8 Retropupillary – attached Iris Claw IOLs for the treatment of aphakia in paediatric patients.

The retropupillary fixation method was also proposed in order to avoid the postoperative complications regarding the deterioration of the endothelial cell count. Gonnerman et al., reported their findings using this particular surgical application in seven aphakic eyes of paediatric patients that had no capsular support as well as lens sublaxation. The mean decrease of the endothelial cell count was 6,4% after one year of post-operative follow up. Furthermore, no serious complications are reported such as IOL dislocation while there was significant improvement of the visual acuity. More specifically, the reported post-operative visual acuity was 0,13 ± 0,17, 21 12 monts post-operatively (Gonnerman et al, 2013).

4 Conclusions

The aim of the current study was to review the most recently published literature regarding the use of IOLs to treat aphakic eyes, both in adult and paediatric patient populations. In adult populations, even though there is a significant improvement in the main outcome measure of visual acuity, the installment of IOLs is also causality for several post-surgical complications. The complications that are reported with greater frequency in the studies included in this review are increased intraocular pressure, corneal decompression, papillary distortion, retinal detachment and cystoids macular oedema.

The surgical interventions that include the use of IOLs to treat aphakic eyes, either in paediatric or in adult patients represent a significant degree of challenge to the ophthalmic surgical team, especially if the patient has inadequate capsular support or/and lens sublaxation. In the past decade a number of surgical techniques and different lens designs have been proposed in order to improve the visual acuity of the patients but also to decrease the possible post-surgical complications. The studies included in this review propose that the artisan iris-claw IOLs as well as the scleral fixation of the IOLs provide adequate visual improvements. However, these methods also demonstrate higher rate of complications that involve the rupture of sutures. These complications include the dislocation of the IOLs, suture erosion, endophtalmitis and ocular inflammation. In the intrascleral IOL fixation surgical modality (Yamane technique), the studies report fewer post-surgical complications that are related to the deterioration of the sutures. The main reason is the smaller degree of complications and the faster healing time.

This systematic review has several limitations the most prominent of which is the small number of studies that has been recently published for each of the proposed surgical techniques that use IOLs to correct aphakia. Furthermore, the majority of the studies are retrospective and lack comparative data. For this reason it is difficult to safely generalize the optimal surgical approach to correct aphakia, using IOLs, in the two major patient categories that are presented: adult and paediatric. Therefore, it is recommended that more research is conducted in the field in the form of randomized

controlled trials, which can provide data that will better clarify the advantage of each of the proposed surgical techniques as well as the percentages of the presented post-operative complications.

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