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CONGENITAL CATARACT: SURGICAL APPROACHES, VISUAL OUTCOMES, AND POSTOPERATIVE CHALLENGES – A REVIEW

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1.1 Abstract

Congenital cataract forms a significant culprit of blindness in children especially those of the developing states. The early initiation of diagnosis and early intervention step are significant in success in vision process and reduction of long term visual disturbance. The following are the surgical interventions discussed in the paper that have been employed in treatment of congenital cataracts, evaluation of visual outcomes involving surgery and timings and postoperative complications which are among others comprising amblyopia, posterior capsular opacification (PCO) and secondary glaucoma. The research demonstrates the importance of the individualization of treatment planning, long-term monitoring, interdisciplinary treatment in the determination of the optimal visual development of children based on the comprehensive analysis of clinical material and the most current articles and publications.

Keywords: Congenital cataract, pediatric ophthalmology, surgery, visual outcome, postoperative complications, amblyopia, PCO

1.2 Introduction

One of the most important types of cataract is the congenital cataract which takes place during birth or earlier than infancy and it has to deal with an opacity in the lens of the eye. It remains one of the major causes of blindness among children that classifies up to nine per cent of the global numbers of the victims (Sheeladevi et al., 2016). Deprivation amblyopia is developed with interruption of visual axis at early age, and besides, not being treated, may also result in severe visual disability (Lambert et al., 2006). The etiology of congenital cataracts is multifactorial in which the mutation of the genes, other metabolic disorders such as

galactosemia, in utero infection of rubellosis or toxoplasmosis or even the cases could be idiopathic (Haargaard & Fledelius, 2006).

Timely surgical intervention is very essential particularly when the eyes are still at the delicate phase of the visual development in early childhood. The surgery is allowed at various times in the unilateral and bilateral cases, but recommended between 6 and 10 weeks of birth in order to obtain better success (Birch et al., 2009). Surgery is necessary but not sufficing, there should be the efficient process of visual recovery process entailing optical correction, amblyopia treatment and follow up. There is also need to state that the surgical procedure, the choice of the placement of intraocular lens (IOL) and management of contingencies of the post surgical treatment such as glaucoma and posterior capsular opacification (PCO) is crucial as far as visual prognosis is concerned (Wilson et al., 2003).

The present paper is going to discuss the modern way of surgical treatment of congenital cataract, the statement of long-term visual outcomes, and focus on the most relevant postoperative issues, the importance of interdisciplinary medical care, and long-lasting follow up is also of the utmost importance with the youngest patients to provide the needed effect in terms of visual development.

1.3 Surgical Approaches

Within the past few decades or even less than that, the area of surgical management of congenital cataract has improved and this has reached a level of ensuring that there is minimal risk of complication and maximising on vitreous rehabilitation. The decision of carrying out a surgery is subject to the age of the to the child, the morphology of the cataract, the length of the eye axis, the occurrence of any other eyes abnormality on the child, needs of the surgeon and the specialty practiced by him/her. The most common methods in the management of cataract in a child are the following.

1.3.1. Lens Aspiration with Posterior Capsulotomy and Anterior Vitrectomy

The Lens aspiration with primary posterior capsulotomy and anterior vitrectomy is the most common surgical procedure used among the infants and the younger children especially below twenty years. The pediatric eyes have high chances of becoming opaque when the lens is removed and affect the visual axis severely due to posterior capsule. Therefore, a posterior capsulotomy and a small anterior vitrectomy during the time of lenses aspiration is possible to prevent the later appearance of posterior capsular opacity (PCO) and hence give a clear line of sight (Vasavada et al., 2011).

According to the reports, the procedure has recorded a satisfactory mark concerning its security and effectiveness among children who have thick cataracts. Moreover, soft lenses cortical

cleanup may be implemented in a more optimal way using irrigation/aspiration bimanual procedure hence resulting in the reduced risk of inflammatory sequelae (Gogate et al., 2010). The process though is microsurgical and it required special equipment such as a pediatric vitrector.

1.3.2 Primary Intraocular Lens (IOL) Implantation

Intraocular lens implantation is progressively being applied in the children who are more than one year where the size of the eye is big and firm enough to accept an IOL. The insertion of IOL during infancy has caused controversy owing to the existence of the problem of biocompatibility, change in refraction during childhood and the risk of secondary glaucoma (Lambert et al., 2006). Nevertheless when it comes to children of 1-2 years of age, primary IOL implantation needs to be popularly carried out in both the bilateral cases in order to promote early visual rehabilitation and reduce the need to have spectacles as well as contact lenses.

Common IOL ingredients Polymethyl methacrylate (PMMA), hydrophobic acrylic and silicone are some of the common IOL ingredients. The Acrysof is hydrophobic and it is used because it is foldable, less inflammation and PCO (Kugelberg et al., 2008). The artificial lens is typically placed in the capsular bag but in some conditions of insufficient capsular support a insertion into the sulcus can be performed.

The safety of primary IOL insertion along with the posterior capsulotomy and vitrectomy is also proven by the latest research and reported outcomes are stable regarding vision in children aged 12-24 months (Cortina et al., 2018).

1.3.3 Secondary IOL Implantation

The implantation of secondary IOL may be performed during the process of stabilization in the eyes in children undergoing an operation when they are too young (less than 6 months) or those that have some problems in the anatomy of the eye such as microphthalmia and/or persistent fetal vasculature (PFV). This phased procedure can be used to avoid false biometry, and refractive surprises. To correct aphakic children, the use of contact lens or aphakic glasses is placed on child first and the second implantation of IOL is performed after the child grows to attain the age between 2-4 years (Plager et al., 2014).

The insertion of secondary IOL is dependent on the preserved capsular support that requires surgery that depends on how the IOL is inserted. In the case of acquisition of capsular residues, IOL may be put in the sulcus although none of them considers capsular residue, presence of residual capsule considers scleral-fixated lens and iris-fixated lens. Newer modalities of the sutureless intrascleral fixation are resulting in improved outcomes of the secondary IOL, which includes Yamane technique (Yamane et al., 2014).

1.4 Visual Outcomes

The postoperative effect after cataract surgery of the newborn baby is influenced by the multiplicity of concomitant factors, among other things, the age in which the operation should have been made, the lateness of cataract (unilateral or bilateral), the treatment of the amblyopia that should have happened along with the operation and the time of administration and repetition, the postoperative peripheral complications, such as opacification of the posterior capsule or secondary glaucoma.

1.4.1 Timing of Surgery and Visual Development

In the infants, there is a significant role that early visual experience has to play so that it can develop the visual pathway healthily. The deprivation of vision in this critical period could result in deprivation amblyopia that becomes irreversible once the cataract could not be timely extracted. When the cataract is not bilateral, surgery will begin at the earliest moment and when it is bilateral, it should be done with all possible speed. The dense cataracts that are responsible in causing more effect on the visual development and are unilateral should ideally be removed either in the first 4 or 6 weeks at birth (Birch & Stager, 2006). Quite the contrary, bilateral cataracts get treated mainly by means of surgery before 8 or 10 weeks of age so as to avoid binocular amblyopia development (Lambert et al., 2006).

The studies of researchers such as Infant Aphakia Treatment Study (IATS) have revealed that surgery at an early stage produces better long-term outcomes of vision especially in bilateral cases. However, the excessive number of surgery (prior to 4 weeks) is associated with the high postoperative glaucoma; hence, visual rehabilitation and risk care are to be balanced (Plager et al., 2014).

Most children who have bilateral congenital cataracts have relatively improved visual outcomes than the unilateral ones. That is mainly because greater symmetry of visual input and relative facility of optical correction applies to bilateral ones. In unilateral instances, the visual system will be inclined to block the input of the damaged eye culminating in considerable amblyopia. In such incidence, the use of sustained occlusion therapy (patching of the unaffected eye) is essential to enhance stimulation to the weaker eye to improve the visual acuity (Wright et al., 2012).

1.4.2. Impact of Amblyopia Therapy and Optical Correction

The results of postoperative visualization also largely depend on the observation of the rules of treatment of amblyopia. These comprise patching treatment, wear of contact lenses or spectacles and frequent assessment of the visual acuity. Wilson et al. (2016) conducted a meta-analysis and found that almost 60 percent of bilateral cases had better than 6/18 visual acuity

compared to only about 30 percent when the surgery was done early and aggressive amblyopia treatment applied.

In unilateral situations, there is more heterogeneity of outcome, with just 25-30% of children being able to see better than 6/18 as a result of any complexities in treating monocular deprivation (Birch et al., 2009). Also the residual refractive errors should be corrected in a timely manner as it is necessary at this critical phase of visual development to have appropriate focus at the retina.

1.4.3 Long-Term Follow-Up

Long follow up studies are required to screen delayed complications which include secondary glaucoma, anisometropia and alteration of axial length. Regular change in refractive correction, along with constant visual rehabilitation, serves as the important factor to achieve best possible results with time (Dahan et al., 2004).

1.5. Postoperative Challenges

Although surgery and early intervention have improved cataract surgery especially in cases of congenital cataract, the procedure is usually followed by various postoperative complications that sometimes jeopardize the results of the treatment. Such problems are to be monitored and thoroughly managed in the long-term.

1.5.1 Amblyopia

Amblyopia is the major cause of poor vision following surgery to remove congenital cataracts especially when it is unilateral. This is the case because; despite a successful extraction of the lens, there is even a possibility of the visual cortex suppressing the picture of the affected eye in case of a lack of proper visual stimuli. Patching therapy, occlusion therapy it is done by patching the better seeing eye so that the child is forced to use the amblyopic one, is necessary but hard to keep going on a regular basis, especially in the younger children (Lambert et al., 2006). Lack of good compliance during patching routine is a massive roadblock towards achieving the goal in developing world where parental enlightenment might not be high (Rahi et al., 2002).

1.5.2. Posterior Capsular Opacification (PCO)

PCO is highly prevalent among the pediatric patients as compared to the adult population because of increased capability of lens epithelial cells to regenerate in children. The reports indicate that between 50-90 percent of children who have undergone cataract extraction run the risk of developing PCO provided that post-extraction posterior capsulotomy is not made (Kugelberg et al., 2008). Older kids in whom a posterior capsulotomy was not performed in the first operation may need capsulotomy performed by a neodymium-doped yttrium aluminum

garnet (Nd:YAG) laser but this may require the use of general anesthesia to sedate uncooperative patients.

1.5.3. Glaucoma

One of the more dangerous and potentially vision- threatening complications is secondary glaucoma that may occur months, and even years after the surgery. It also has an increased incidence in aphakic eyes as well as in subjects who receive surgery during the initial weeks of life. According to the Infant Aphakia Treatment Study, glaucoma was observed in 17-20% of aphakic infants after five years the surgery had been implemented (Plager et al., 2014). Hence, a regular check of intraocular pressure is a compulsory necessity including treatments that can be topical or requiring surgery like trabeculotomy.

1.5.4. Refractive Errors

This is because the eyes of a child keep developing and refractive results can hardly be obtained reliably after the surgery. After surgery, children are commonly prescribed to be left hyperopic to address the natural myopic drift when the eye develops. Nevertheless, refractive changes cannot be predicted with certainty and regular changes in the spectacle or the contact lens prescription have to be made (Dahan & Drusedau, 2004). Secondary intraocular lens (IOL) implantation (particularly of partially sighted individuals who had not been implanted earlier, when they were already older) is done to alleviate the spectacle grind, later in life.

1.6 Discussion

Successful early treatment of congenital cataract leads to prevention of accidental blindness which is achieved by early surgery, although this treatment creates complexity in management. One of the biggest dilemmas is the issue of the benefits of early surgery compared to the risk of secondary glaucoma and PCO that are more likely with surgery. It requires remote, multidisciplinary follow-up to observe the development of vision constantly, increase ocular pressure, and the state of visual axis maintenance.

The involvement of parents is very crucial in the postoperator care success. Amblyopia treatment education, refractive correction education and education on follow-up timeliness is crucial. Access to reliable care can be impeded by social economic issues, especially in venues of low resources, increasing the necessity of the implementation of public health initiatives and local support programs (Gogate et al., 2010).

New hope is emerging with the innovations. Adaptable IOLs to fit the ocular development, better biometric devices in children and digital devices that can aid in treatment of amblyopia (e.g. visual stimulation through games) are promising. Such developments, along with effective

screening and parent education, may greatly improve visual results in the children affected with congenital cataract.

1.7. Conclusion

Management of congenital cataract is one of the greatest issues in the field of pediatric ophthalmology. It requires not only timely surgical treatment but also a multidisciplinary approach when all conditions should be combined in terms of ocular surgery, corrective visualization, and active amblyopia and a long-term follow-up assessment. Surgery at an early age and preferably during the sensitive period of visual development gives a good opportunity of restoring functional vision. Nonetheless, postoperative regimes of rehabilitation should be applied to ensure success which involves patching, optical correction and checkups.

Although improvements in surgical procedure and intraocular lenses have made surgery safer, posterior capsular opacification, secondary glaucoma and refractive unpredictability still remain problematic. These issues make the significance of a personalized therapy plan and engagement of a caregiver to make sure the therapy would be adhered.

Moreover, unequal access to care, particularly the resource-poor environments, indicates that there is a necessity of the public health efforts being introduced to aid early detection, educating parents, and follow-up structures. The current innovations, including pediatric-adjustable IOL and digital amblyopia therapy devices, provide a hopeful future in terms of treating patients.

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