

Consensus Guidelines for Management of Congenital Cataract in Pakistan

Mian Muhammad Shafique, Muhammad Moin

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See end of article for authors affiliations

Correspondence to:
Mian Muhammad Shafique
Pediatric Ophthalmologists of
the Ophthalmological Society of
Pakistan
Email:
mianmshafiq@hotmail.com

Purpose: To formulate guidelines for management of congenital cataract in Pakistan through consensus of Pediatric Ophthalmologists.

Study Design: Mixed methods study using modified Delphi technique.

Place and Duration of Study: Among pediatric ophthalmologists of the Ophthalmological Society of Pakistan from June 2016 to February 2017.

Material and Methods: A survey questionnaire was constructed containing 40 questions after literature review, which covered almost all aspects of the management of congenital cataract. This survey was answered by 56 renowned pediatric ophthalmologists of Pakistan using the survey monkey online program. The analyzed data was presented in a planned face-to-face meeting of top 15 members of the Association of Pediatric Ophthalmology, Pakistan. The data was discussed point by point and consensus was built with some additions and modifications.

Results: 41 out of 56 pediatric ophthalmologists answered the survey. The most common presentation of a child with congenital cataract in our study was white pupil (72%) and 97.5% participants said that repeat eye examination should be done after dilation of pupil to confirm the diagnosis. 77% participants felt that the child should not be operated if cataract opacity is smaller than 3 mm. In our country, only one-third of the pediatric ophthalmologists were in favor of implanting intraocular lens (IOL) by the age of one year but there was 80% consensus to do so by the age of 2 years. Children with dense cataract should be operated by 2 months of age according to 80% participants. It is preferred to perform primary posterior capsulotomy with or without anterior vitrectomy in all children who undergo congenital cataract surgery in any age until 5 years (87%).

Conclusion: Management of congenital cataract by pediatric ophthalmologists in Pakistan is consistent with internationally available guidelines.

Key words: Congenital cataract, Consensus, Guidelines, Management.

Congenital cataract is considered to be one of the most important treatable causes of childhood blindness worldwide¹. Pediatric cataracts can have a profound impact on health and vision-related quality of life, and on functional visual ability². Its prevalence varies from country to country^{3,4}, in USA 3-4 per 10,000 live births⁵, and in UK 3.18 per 10,000 live births⁶. In developing countries like India, 7.4 – 15.3% of childhood blindness is due to

cataract⁷. The Pakistan National Blindness and visual impairment survey, published in 2007 indicates crude adult cataract prevalence as 1.75% but there is no mention of congenital cataract in this document⁸. According to our study survey, about 1% of children having eye diseases were found to have the congenital cataract.

In a population of 200 million in our country, the number and the percentage of children is rising day by

day and so is the constant rise in the number of children having eye diseases. Congenital and developmental cataract is a very common presentation in our outpatients and if not treated properly and in time may lead to lifelong disability. Unfortunately, there is a lot of confusion among our colleagues for its treatment plans and its execution. There is an urgent need to develop consensus guidelines for treatment of this disease in our children on priority basis so that we can save the vision of our future generations.

The main purpose of this effort is to establish consensus for treatment of childhood cataract in Pakistan and develop guidelines which could be followed by all pediatric ophthalmologists and also provide enough help to general ophthalmologists of our country. As there is no such document available in our country we took the responsibility to get a consensus among the renowned pediatric ophthalmologists of our country in the light of evidence of their experience of managing this disease in the previous 25 years.

MATERIAL AND METHODS

To start this project we did a literature search to see internationally available guidelines for treatment of congenital cataract. Based on this, a plan was made which comprised the following steps. A survey questionnaire was constructed on the basis of the knowledge acquired about the treatment of childhood cataract in excellent centers of the world. It contained 40 questions, which covered almost all aspects of management of congenital cataract. A survey was conducted with the help of ‘Survey Monkey’ and online opinion was sought from 56 renowned pediatric ophthalmologists of our country in all provinces and all big cities of Pakistan. Received data was compiled, analyzed and later presented in a face-to-face meeting of top fifteen pediatric ophthalmologists of our country on the platform of Association of Pediatric Ophthalmology Pakistan during OSP national conference in Peshawar in February 2017. The data was rigorously discussed in this meeting point by point and consensus was built with some additions and modifications. After the consensus, final guidelines for the management of congenital cataract in Pakistan were drafted.

RESULTS

Forty-one out of 56 pediatric ophthalmologists responded and gave their opinion. More than 90% of

them had been regularly managing congenital cataract for last 11 – 25 years. The results of the key questions in the survey are shown in Figures 1 – 19.

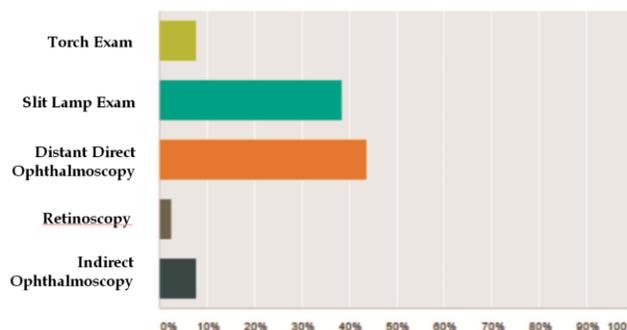


Fig. 1: Which of the following tests you think is most important in making a diagnosis of congenital cataract?

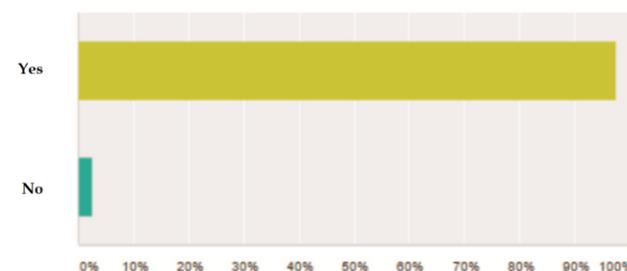


Fig. 2: Should the examination of eye be repeated after dilation of pupil to confirm the diagnosis of congenital cataract?

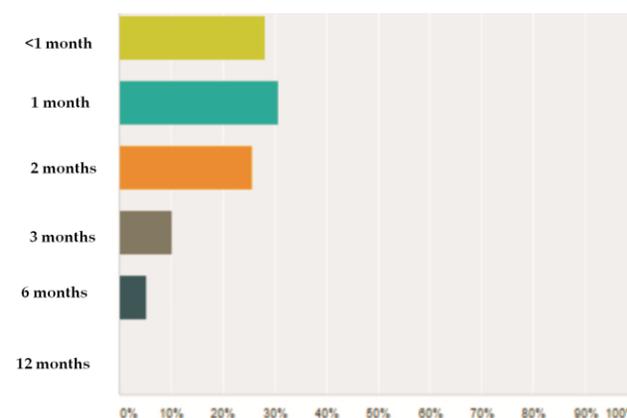


Fig. 3: At what minimal age of child with dense congenital cataract will you go for surgery?

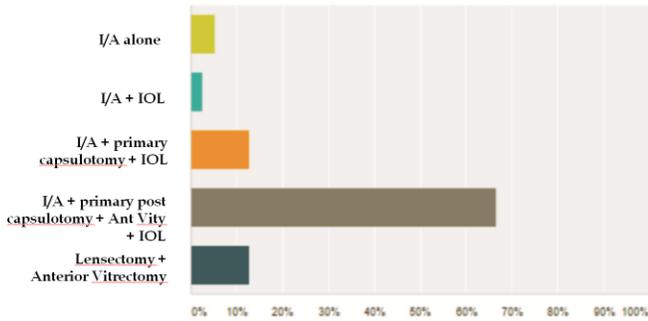


Fig. 4: Which procedure you prefer for treatment of congenital cataract?

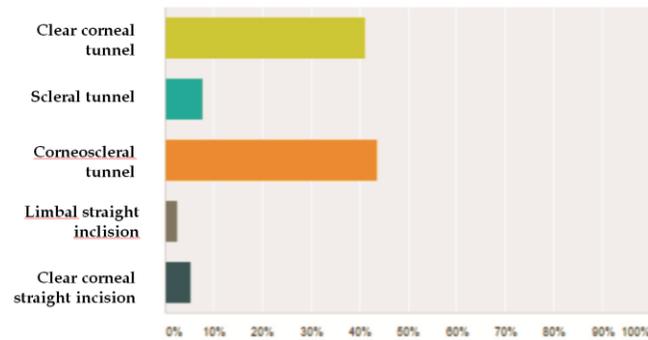


Fig. 5: What type of incision you prefer to make for removing congenital cataract and implantation of IOL?

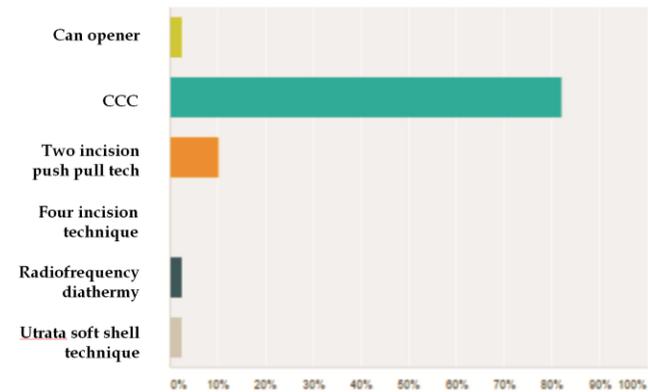


Fig. 6: What is your preferred technique for anterior capsulotomy?

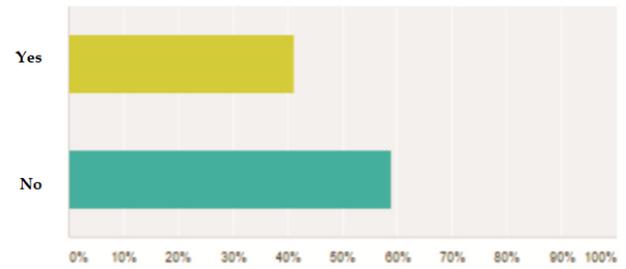


Fig. 7: Do you need to go for hydrodissection routinely in every case of congenital cataract to split nucleus for cataract?

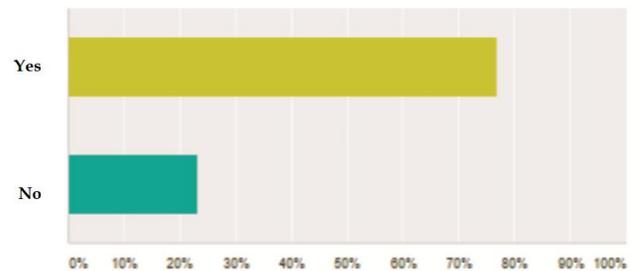


Fig. 8: Do you think that hydrodissection is contraindicated in congenital cataract of posterior polar type?

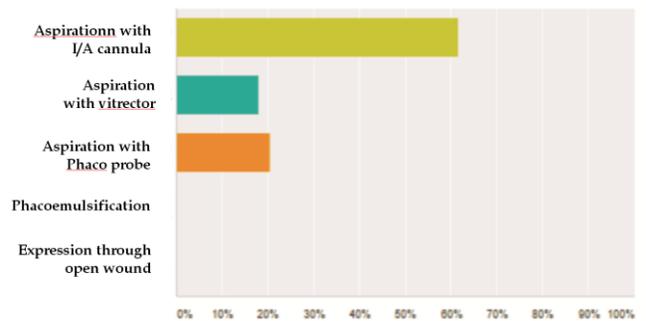


Fig. 9: What is your preferred method of removal of nucleus in congenital cataract?

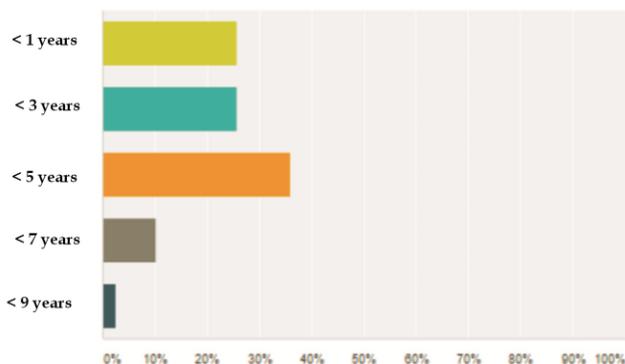


Fig. 10: At what age you will prefer to perform primary posterior capsulotomy with or without anterior vitrectomy while operating a child of congenital cataract?

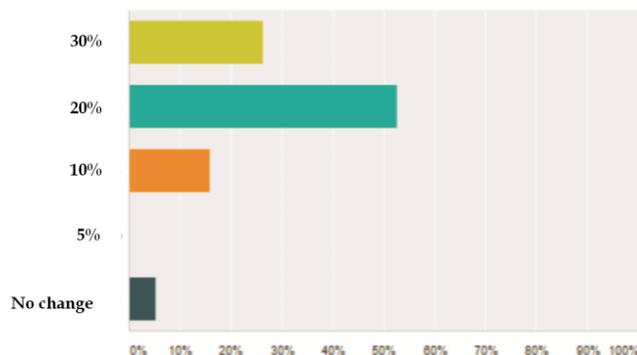


Fig. 13: What percentage of calculated power of IOL detected by biometry in a child of 2 years should be reduced to compensate for the change of refraction for adulthood?

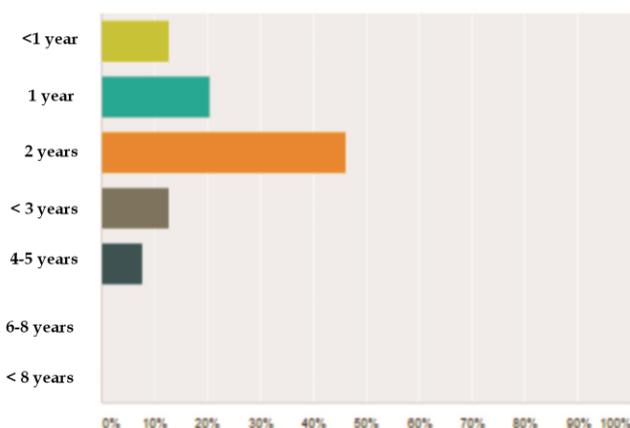


Fig. 11: At what minimal age you will prefer to implant an IOL in a child of congenital cataract?

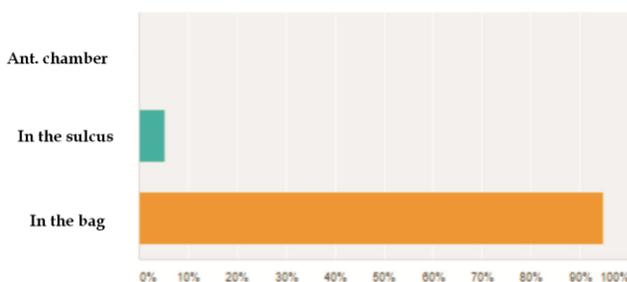


Fig. 14: What position of placement of an IOL is your target during surgery of congenital cataract?

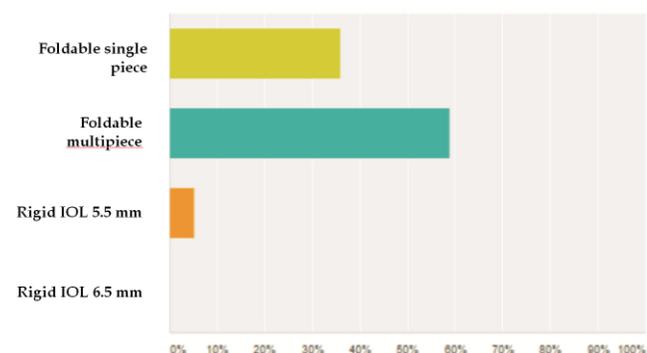


Fig. 12: What type of IOL you will choose for implantation in a child of congenital cataract?

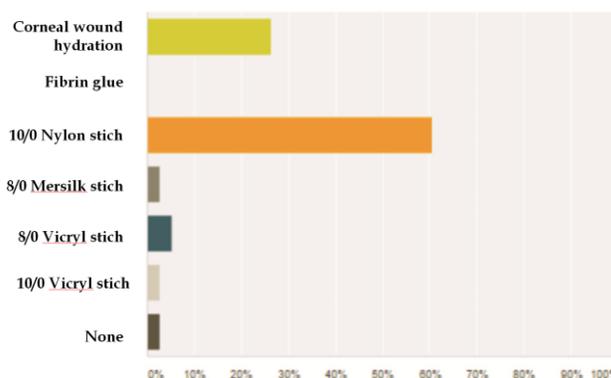


Fig. 15: What wound sealing measures do you prefer to take at the end of congenital cataract surgery?

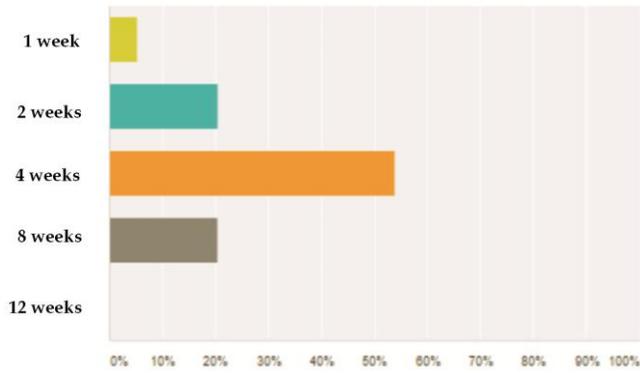


Fig. 16: How long do you advise to continue the use of post-operative topical antibiotics in a child undergoing congenital cataract surgery?

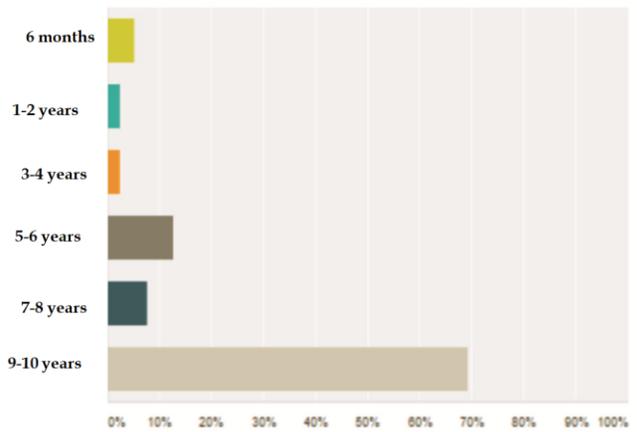


Fig. 19: How long do you prefer to follow up the child of congenital cataract postoperatively?

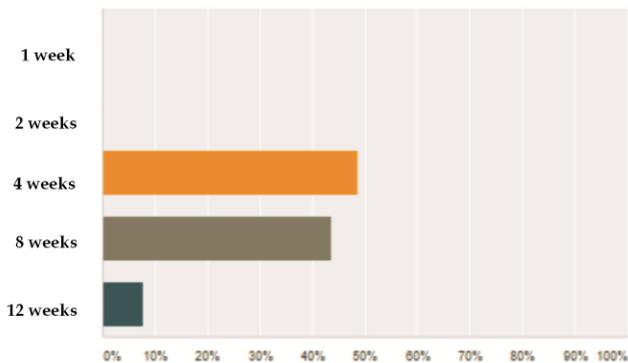


Fig. 17: How long do you advise to continue the use of post-operative topical steroids in a child undergoing congenital cataract surgery?

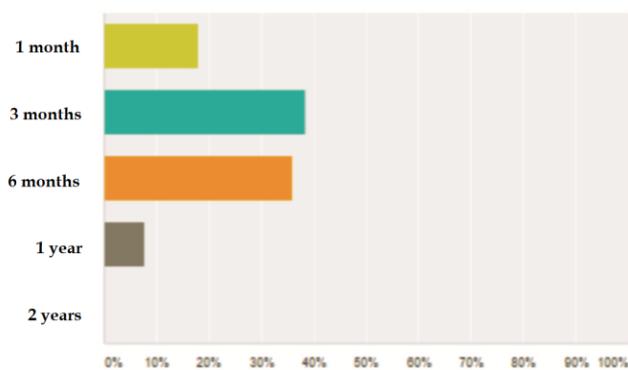


Fig. 18: If the posterior capsule becomes thick after congenital cataract surgery, how long after the surgery will you prefer to go for Nd-YAG laser capsulotomy?

DISCUSSION

The most common presentation of a child with congenital cataract in our study was white pupil (72%) followed by ‘abnormal visual behavior’ (23%). Only these two symptoms helped the ophthalmologist to think of congenital cataract in 95% of cases. The presentation of congenital cataract with a white pupil in our study was far more common as compared to findings in a neighboring country (72% vs. 24%)⁹. Among the uncommon presentations of the condition were strabismus and nystagmus. These two are considered as important risk factors for poor visual outcomes¹⁰.

Although the torch examination is a very common practice, the use of the slit lamp for anterior segment examination followed by distant direct ophthalmoscopy are the two pivotal tests for making the diagnosis of congenital cataract (82%). If possible, indirect ophthalmoscopy and retinoscopy may also be performed to rule out other causes of abnormal visual behavior. For uncooperative children, a special method of slit lamp-adapted anterior segment photography with assistance technique and sleep aid administration has been used in China successfully for monitoring and classifying pediatric cataracts¹¹.

There is a strong consensus (97.5%) among ophthalmologists of our study on repeat examination of the child's eye after dilatation of the pupil before confirming the diagnosis of congenital cataract. The dilating drops if having the character of cycloplegia will also help to perform cycloplegic retinoscopy at the same time. If the intention is to perform the later test also then use Atropine eye ointment for children

younger than 2 years and cyclopentolate eye drops for children older than this age.

In our study, 77% pediatric ophthalmologists felt that one should not be in hurry to operate if the opacity in congenital cataract is small (< 3 mm) and the vision improves with dilatation of the pupil. Tropicamide dilating eye drops were generally preferred (60%) as these are short-acting mydriatic and weak cycloplegic. These drops should be used twice a day. Through the dilated pupil the child will be able to focus both for distance and near as still, we have saved the accommodation in this crystalline natural lens. Other drugs like cyclopentolate and atropine are strong cycloplegic and are less commonly used for this therapeutic purpose. Such children who are under treatment with dilating drops should be kept under monthly follow up to observe the progress of this small opacity and save the child from the risk of sensory deprivation amblyopia.

Once decision on surgery for dense cataract has been made and conveyed to the parents, they may ask about the visual prognosis. Always be ready to answer rightly, which can only be done if macular function tests have been performed. The easiest and reliable test for younger children is brisk pupillary light reflex (70%). Other tests like visual acuity, color vision, and if possible, the two-point discrimination test should be performed.

Motivation and education of the family members especially the parents before and after the surgery of a child of congenital cataract is of utmost importance (100%) and should not be ignored. If surgery is not yet indicated then the parents must be made to realize the importance of regular and frequent follow-ups. On the other hand, if the child has undergone surgery then the reasons for regular follow up and meticulous application of post-operative measures is even more important. All this can be achieved only if good rapport with parents is developed by giving them the knowledge of the disease and motivating them for cooperation to achieve the targeted outcome.

In our survey, 100% of the participants were in favor to address the following three major aims of treatment of congenital cataract. The first one was to remove the cataract by the procedure, which suits the type of cataract. Second was to correct the high hypermetropia related with lens removal either by intraocular lens implantation per-operatively or by glasses postoperatively. The third aim was to treat any

associated amblyopia, which should never be forgotten.

All the following conditions related with congenital cataract should be considered an indication of surgery: lens opacity 3 mm or larger, posterior sub-capsular lenticular opacity, lens opacity with no fundal view, lens opacity with nystagmus and lens opacity with strabismus.

The participants felt that if the child was having dense congenital cataract then his surgery should not be delayed much. Operating before the age of one month was not considered safe, as there are more chances to develop aphakic glaucoma.¹² 85% of pediatric ophthalmologists of our country were of the opinion that the child should be operated by the age of two months, which is consistent with international standards¹³. If there was any reason for avoiding general anesthesia like some serious cardiac congenital abnormalities, cataract surgery can be undertaken with necessary precautions like intensive monitoring, keeping a standby cardiac setup¹⁴. In these circumstances, the operation can be delayed until 6 months.

General anesthesia was the main choice for surgery of congenital cataract (100%). Endotracheal intubation was preferred over laryngeal mask (LMA) for maintenance (69% vs 39%). For some reasons, if these two were not possible to use, then anesthesia with ketamine injection may be a safe and potent option¹⁵. Everybody felt that there was no place for relying solely on local anesthesia in this surgery.

The most favored surgical technique for congenital cataract was a combination of several procedures. Irrigation and aspiration of lens matter alone was not liked as almost 100% of these operations were complicated by posterior capsular thickening. The participants felt that if the child was younger than 2 years of age it was preferable to combine irrigation, aspiration and IOL implantation with primary posterior capsulotomy with or without anterior vitrectomy (67%). Primary IOL implantation should also be done in the same session if the age of the child is more than one year.

Tunnel incision was more popular (92.3%) for reason of more stable anterior chamber during surgery and sealing of wound without any stitch in the majority of cases. A keratome was preferred to make the tunnel incision of 2.7 mm or 3.2 mm. Both corneo-scleral tunnel and clear corneal tunnel were equally

liked as compared to the scleral tunnel. Straight limbal or straight corneal incisions are now less used as these need stitching to close the wound and the post-op astigmatism is quite high.

Continuous curvilinear capsulorhexis (CCC) was the most popular technique (82%) as it was easy to master with cystotome or capsular forceps. Due to the higher elasticity of capsule in this age group, there are more chances of extending the rhexis out as compared to adults. In case of failure to make a complete CCC the anterior capsulotomy can be safely converted to can opening capsulotomy. Other innovative techniques like two incisions push-pull technique, radiofrequency diathermy and plasma blade (Fugo blade) although applied uncommonly in our country are becoming popular in some places around the world¹⁶. In white congenital cataract during anterior capsulotomy, the capsular edge is seen with great difficulty. In such cases, use of Trypan Blue dye is a good adjunct for handling a smooth anterior capsulotomy and was liked by most of the pediatric ophthalmic surgeons (90%).

There was split opinion among the pediatric ophthalmologists regarding the need to go for hydrodissection routinely in every case of congenital cataract to separate the nucleus from the cortex (59% vs. 41%) However, it is very important not to be aggressive to perform hydrodissection in posterior polar and posterior subcapsular cataract where it is rather contraindicated. In such type of cataracts, it was preferred (77%) to avoid the hydrodissection but if performed the pressure of injecting fluid should be very low and gentle otherwise there can be a risk of posterior capsular rupture even before irrigation and aspiration.

The nucleus in congenital cataract is very soft and hardly needs removal by expression through an open wound. This has given place to small tunnel incision in surgery for congenital cataract. The majority of surgeons (62%) removed the nucleus of congenital cataract easily by I/A cannula, which normally is used to aspirate cortex. If it fails then phaco-probe or vitrector can be applied to aspirate the nucleus.

I/A cannula was the preferred instrument to aspirate cortical matter in congenital cataract surgery (82%). Hardly ever, there was any need to use phaco-probe or vitrector for this purpose.

Almost all children operated for congenital cataract before the age of 4-5 years develop posterior capsular (PC) thickening postoperatively if the

posterior capsule is left intact¹⁷. It is preferred to perform primary posterior capsulotomy with or without anterior vitrectomy in all children who undergo congenital cataract surgery in any age until 5 years (87%). The chances of development of PC thickening are reduced after this age and if it occurs, can be treated with YAG laser in children older than this age under topical anesthesia, so the primary posterior capsulotomy and the anterior vitrectomy should be avoided after this age.

77% pediatric ophthalmic surgeons liked to use preservative-free triamcinolone acetonide during congenital cataract surgery after posterior capsulotomy for better visualization of prolapsed vitreous and as a safe adjunct to post-op steroid drops.

There has been great controversy and debate among the pediatric ophthalmologists regarding the minimum age to implant an IOL in a child of congenital cataract. In a survey, approximately 70% of the American Association of Pediatric Ophthalmology and Strabismus members worldwide preferred to implant an IOL in children^{18,19}. In our country, only one-third of the pediatric ophthalmologists were in favor of implanting IOL by the age of one year but there was 80% consensus to do so by the age of 2 years. There is no evidence in favor of delaying it beyond the age of 4 years.

The best choice is the foldable IOLs (95%) among which multi-piece foldable lens was preferred over single piece foldable lens. Both these can easily be injected through the same 2.75 mm/3.2 mm sized tunnel incision. The Rigid IOL 5.5mm may be used if the foldable is not available or if the given incision is straight, otherwise the tunnel will have to be extended to more than 5.5 mm with larger keratome.

The eye of an infant at birth has high hypermetropia, which continuously decreases until the age of 10 - 13 years¹⁸. This decrease is rapid in first five years. As the implanted IOL power remains static, the choice of IOL power should be adjusted according to the expected adult power of lens for this child. The majority of participants of our survey were in the favor to reduce the biometric power of IOL by 30% until the age of one year, 20% up to the age of 2 years, 10% between 2 - 5 years and no reduction at the age of 10 years.

There was no controversy regarding the choice to place the IOL during congenital cataract surgery. The best choice was 'in the bag' (95%). If for some reason bag is not intact and there is a risk of IOL sinking to

vitreous then the second choice is 'in the sulcus, and the placement in the anterior chamber or posterior fixation are only done if above two choices are not available. Some studies have advocated the use of capsular tension ring (CTR) in children with subluxated lenses²⁰.

When a primary IOL is not implanted, residual aphakia should be treated using either aphakic glasses or contact lenses (CL). Aphakic glasses can be used for the correction of bilateral aphakia but these are not suitable for eyes with unilateral aphakia. Many physicians insert a silicone CL immediately at the end of cataract surgery under general anesthesia but others delay it until 1 to 2 weeks after surgery. To calculate the power of CL preoperatively, use the formula for IOL power calculation with an A constant of 112.176, which provides a good CL estimation²¹.

Although the opinion was divided for use of intracameral steroids (46% vs. 54%) at the end of surgery of congenital cataract, the majority was against the use of intracameral antibiotics (60.5%). However, these two can be injected intracamerally when manipulation has been done for a longer period or there is any doubt about the strict application of sterilization rules.

The wound sealing measures at the end of congenital cataract surgery depend on the type of given incision. The tunnel incision whether clear corneal or corneoscleral may close itself or may require just corneal hydration of the wound. However, majority liked to apply one stitch (71%) to secure the wound and sleep with comfort as the tensile strength of the wound lips in this age group is less as compared to adults.

Nylon 10/0 was the most used suture (60.5%) for closing wound for congenital cataract. Its disadvantage of being non-absorbable mounts an additional session of general anesthesia for removal of stitches after 8 weeks. This problem can be overcome by use of absorbable suture 8/0 Vicryl or 10/0 Vicryl out of which the latter is becoming more popular⁹. If available, fibrin glue is another good option to seal the wound without a stitch.

During the wound healing phase after congenital cataract surgery, it is advised to continue the use of topical antibiotics and steroids for some time. The majority of pediatric ophthalmologists liked to use postoperative topical antibiotics for 2 - 4 weeks. However, the topical steroids were used for a

relatively long period ranging from 4 - 8 weeks depending upon the condition of the operated eye.

The frequency of post-surgery refractions in an operated child of congenital cataract may vary with advancing age. In first 5 years of life, the six monthly visits were ideal and liked by the majority of pediatric ophthalmologists (82%). After 5 years of age, annual refractions may be sufficient to cater for expected changes in glasses.

If primary posterior capsulotomy has not been done during the congenital cataract surgery, the chances of posterior capsular thickening are quite high. Younger the child more are the chances. If the capsule becomes thick then it cannot be left as it is. We must perform YAG Laser capsulotomy or surgical capsulotomy to avoid development of sensory deprivation amblyopia. It should not be performed until 3 months after the operation but should not be delayed more than six months after surgery.

To perform Nd-YAG laser capsulotomy under topical anesthesia in children less than 5 yrs of age is not an easy task. However, a very good percentage of pediatric ophthalmologists (49%) feel confident in successfully performing this procedure after the age of 5 years while the high majority (92%) can do so at the age of 9 years. This needs to prepare the child and the parents in several frequent visits before embarking upon this procedure.

Congenital cataract and amblyopia go side by side for the array of reasons i.e. cataract develops in amblyogenic age, under correction by reducing the biometric power of implanted IOL during surgery, posterior capsular thickening, wrong refractions and ignorance about the importance of the regular use of glasses.

The amblyopia should be diagnosed vigilantly and treatment started immediately. The cheapest and most popular method (97%) is occlusion therapy. The normal eye is patched and the amblyopic eye is kept open and is forced to focus the colorful objects. The prescribed glasses must be worn during the session of patching. There are different patching regimens but 'one hour daily for every year of age' is the simplest and the successful rule for treatment of amblyopia related to congenital cataract. Other methods like the use of Atropine eye drops to blur the normal eye can be applied in highly uncooperative children in place of patching.

Although the healing process after cataract surgery takes only a few months to complete, the child

cannot be discharged from care so early due to continuous growth and changes occurring in refraction of the eye. The post-operative management and follow up is the cornerstone of successful pediatric cataract surgery.²² In our study, there was a strong opinion (69%) to keep the child under constant follow up during the whole time of growth or at least until the age of 10 years. However, the literature review shows evidence of poor follow-up in these children. All efforts should be made to improve this important part of management.²³ The loss to follow up is much more commonly seen in free eye camps of rural areas as compared to surgery done in tertiary care centers.²⁴

The limitation of the study is that it includes only the members of OSP (Ophthalmological Society of Pakistan) and APOP (Association of Pediatric Ophthalmology of Pakistan). There are other pediatric ophthalmologists who are working quite successfully all around the country but they were not included in the study. In future, the study can be expanded to a broader base by increasing the number of its participants as well as adding up the individual interviews to collect additional data from pediatric ophthalmologists.

CONCLUSION

Data received by a national survey on "Evidence-based management of congenital cataract" from renowned pediatric ophthalmologists of our country has helped us formulate "consensus guidelines for management of congenital cataract in Pakistan". These are very much consistent with internationally available guidelines for management of this disease. All aspects of management right from symptoms, signs, diagnosis, surgery and postoperative follow-up has been covered. An attempt has been made to address all controversial issues related with management of congenital cataract in light of survey results and international practices.

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Nil.

Conflicts of Interest

There are no conflicts of interest.

Author's Affiliation

Prof. Mian Muhammad Shafique
Head of the Ophthalmology Department
Lahore Medical and Dental College,
Lahore.

Prof. Muhammad Moin
Head of the Department of Ophthalmology,
Postgraduate Medical Institute,
Ameer-ud-Din Medical College,
Lahore.

Role of Authors

Prof. Mian Muhammad Shafique
Study Design, Manuscript drafting, Data Collection.

Prof. Muhammad Moin
Study Design, Critical review. Data analysis.

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