

## Updates on the Surgical Management of Paediatric Cataract with Primary Intraocular Lens Implantation

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### Abstract

With the advent of modern surgical techniques, paediatric cataract has become much more manageable. Intraocular lens (IOL) implantation is the standard of care for patients over the age of 2 years. The use of IOL in young infants is still controversial. In addition, there are still unresolved issues, such as the minimum age at which IOL can be safely implanted, IOL power selection and IOL power calculation. The current trends in the management of the above challenges are discussed. Although numerous reports on the prevention and management of posterior capsule opacification have been published, there are ongoing intensive debates and research. Long-term postoperative complications like glaucoma and rhegmatogenous retinal detachment are problems that cannot be overemphasised and these issues are also reviewed.

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### Introduction

Paediatric cataract has a low incidence compared with adult cataract. The estimated incidence is about 1 to 6/10,000 births.<sup>1-3</sup> It is an important cause of childhood blindness in Hong Kong and in other countries.<sup>4</sup> Therefore the management of paediatric cataract remains a major challenge for ophthalmologists.

Identification of the aetiology is the first essential step in the management of paediatric cataracts. It can be an isolated occurrence, part of a multisystem or syndromal disorder, or an inherited disease, with autosomal dominance being the most common, usually of high penetrance and variable expression.<sup>5,6</sup> Therefore, ophthalmologists need to work with paediatricians to explore the family history and also the possibility of intrauterine infection, metabolic disorders or trauma as a cause of the condition.<sup>7</sup> Such causes can usually be found in approximately 50% of bilateral infantile cataracts, while it is unusual to find a cause of monocular cataracts.<sup>8</sup>

A comprehensive ophthalmic assessment is vital as it influences the management plan. Formal visual function tests may not be possible in very young infants and the decision to intervene will largely depend on the density of the cataract.<sup>9</sup> For older children, formal vision tests like

forced choice preferential looking or other vision tests appropriate to the age of the child may be possible.<sup>9</sup> A complete ocular examination including pupil reflex, optic disc, retina and macular status should also be assessed.<sup>9</sup> The presence of nystagmus suggests poor vision and a guarded visual prognosis.

Parental education and counselling is also an integral part of successful paediatric cataract management since patching, spectacles or contact lens wear may be needed before and after the operation. Realistic expectations of visual outcome should be given, especially for those with unilateral cataract, late presentation or poor initial vision.

In addition to all of the above, surgery remains an important part of the management of paediatric cataracts. The timing of an operation, surgical techniques, choice of aphakic correction and posterior capsule management are important factors influencing surgical outcomes.

### Timing of Surgery

The concept of a visually sensitive period has been well developed. It is known from animal studies that reduced visual input from birth to 3 months leads to reduced connections between the cortical cells and the affected eye and a reduced number of binocularly driven cells.<sup>10</sup> These changes can be reversed if there is early restoration of

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visual clarity. For humans, variations in visual outcomes are still seen despite significant improvement in treatment over the past 30 years. Patients with monocular cataracts have 2 predisposing factors for the development of amblyopia: binocular rivalry and visual deprivation.<sup>11</sup> To prevent otherwise irreversible amblyopia, patients with dense cataracts should have surgery and optical rehabilitation before the age of 17 weeks.<sup>10,12</sup> Superior visual acuity correlates with earlier surgery and vigorous amblyopia therapy.<sup>10</sup> Some authors believe that surgery should be done by the age of 8 weeks for a favourable outcome.<sup>13-15</sup> However, there are contradicting data concerning surgery within the first 4 weeks of life, with regard to a higher incidence of complications.<sup>16,17</sup> On the other hand, in patients with persistent hyperplastic primary vitreous (PHPV) the single most important indicator for future visual potential is the patient's age at the time of presentation and surgery, assuming no significant posterior-pole abnormalities.<sup>18</sup> In summary, the common and generally accepted practice is to perform surgery, for congenital visually significant cataract as early as possible to prevent irreversible amblyopia while timing of surgery needs balance between the effect on visual development and the risk of surgery. Type, density and laterality of the cataract are other important factors to consider.

### Surgical Technique

With the advancement of modern adult small incision cataract surgery, surgical techniques for paediatric cataract have also evolved significantly over the past 30 years. However, children's eyes are not miniature adult eyes, and multiple challenges distinguish paediatric cataract surgery from that performed on their adult counterparts.

There are a variety of surgical options, depending on the age of the patient and the personal experience of the surgeon. The standard surgery for infants less than 6 months old is lens aspiration with primary posterior capsulectomy and anterior vitrectomy.<sup>19</sup> Whether IOL should be implanted in infants less than 6 months old is still controversial, although there are increasing numbers of surgeons opting for intraocular lens (IOL) implantation for patients less than 6 months old.<sup>20</sup> The ongoing USA multicentre trial – Infant Aphakia Treatment Trial – may provide an answer to this issue. There is increasing evidence that IOL implantation is safe for patients under 2 years old.<sup>21,22</sup> It has been suggested that this should be the standard of care for patients over the age of 2 years.<sup>23</sup>

In wound construction, scleral-tunnel, limbal and clear corneal approaches have their own advantages and disadvantages, and studies have shown there is not much difference in long-term postoperative astigmatism.<sup>24-26</sup> Performing anterior continuous curvilinear capsulorhexis (CCC) is always a surgical challenge to paediatric cataract

surgeons as the capsule is much more elastic and is notoriously difficult to control. High-viscosity viscoelastic is preferred in manual anterior CCC, which is still the most tear-resistant procedure.<sup>27</sup> A cystotome is usually used to create a central small flap and then anterior capsulorhexis forceps are usually used for the rest of anterior CCC as it provides better control throughout the whole process. With the introduction of trypan-blue or indocyanine green (ICG) staining, anterior CCC has become more feasible even in white cataract.<sup>28-30</sup> Anterior vitrectorhexis, by means of mechanised vitrector and radiofrequency diathermy anterior capsulectomy, have also been advocated.<sup>31-33</sup> Anterior chamber maintainer through a corneal paracentesis is gaining popularity for better control of anterior chamber depth. The cataract is usually soft and can often be removed with aspiration alone through a phaco probe or with a vitrector. Good hydrodissection can also help in cortical clean-up as the cortex is sticky. Acrylic IOL seems to induce less PCO than polymethyl methacrylate (PMMA) IOL and is currently the preferred choice of IOL although there has not been any controlled trial on this issue. In-the-bag IOL implantation could provide a good centration and reduce the chance of PCO.<sup>34</sup>

The management of posterior capsule and anterior vitreous is another challenging and controversial area. A primary posterior CCC is recommended by some surgeons for IOL implantation for patients younger than 6 years old. However, achieving an optimal size of posterior CCC is technically challenging as it is thinner and less visible.<sup>35</sup> In addition, a reopacification of the anterior hyaloid face after primary posterior capsulotomy that requires a second operation is well reported especially in young infants. Plager et al<sup>36</sup> found that 80% of children aged 6 months or below receiving primary CCC developed secondary opacification requiring a second pars plana vitrectomy. PCCC is performed in the following way. The posterior capsule is punctured with a cystotome, and viscoelastic substance is then injected through the opening to push the vitreous backward. The aim of the PCCC is to try to achieve an opening about 1 mm smaller than the optic of the IOL used. It has also been suggested that posterior capsulorhexis may be performed more easily with posterior capsule staining.<sup>28,37</sup> The posterior capsulotomy and anterior vitrectomy can also be performed with a pars plana approach after IOL implantation, depending on the surgeon's preference and experience. Whether or not an anterior vitrectomy or optic capture should be performed is still an area of debate.<sup>38-41</sup> To capture the IOL, the optic is slipped inferiorly and superiorly through the PCCC using a spatula or cannula. However, there is increasing evidence that posterior CCC with anterior vitrectomy may decrease the risk of posterior capsule opacification.<sup>35</sup> The management of established posterior capsule opacification will be discussed later.

## Refraction and IOL

It has been well documented that infants, on average, are hyperopic and that the hyperopia gradually decreases during infancy and early childhood. These natural changes in refractive error are presumed to reflect finely regulated eye growth.<sup>42</sup> The process is known as emmetropisation. The rate of change appears to be most rapid in the first 12 months. Emmetropia has yet to be reached at 4 years of age.<sup>43</sup>

Primary IOL implantation is becoming more popular in the management of paediatric cataract. In addition, the minimum age of IOL implantation has been lowered. When determining the power of IOL to be used, one should take into account the changing axial length and refractive power of the children's eyes and also the effect of IOL on ocular growth. Eyes with lens extraction during infancy may grow less than normal.<sup>44</sup> On the other hand, it has been found that paediatric pseudophakic eyes have a slightly lesser rate of refractive growth than aphakic eyes while unilateral pseudophakia has an accelerated growth compared with bilateral pseudophakia.<sup>45</sup> This implies that the refractive growth of the eye is affected by the presence of an IOL. Griener et al,<sup>46</sup> however, found that there might be an actual reduction in the axial length in infantile eyes following cataract extraction and IOL implantation. Other reports indicate that the pattern of axial elongation and corneal flattening was similar in eyes with congenital cataract compared with that observed in normal eyes.<sup>47</sup> In summary, the effect of IOL on ocular growth remains controversial.

Children normally have a small amount of myopic shift as they grow, despite the large increase in axial length (from 16.8 mm at birth to 23.6 mm in adult life). This is because of the correspondingly large decrease in lens power from +34.4 D to +18.8 D.<sup>44</sup> Eye growth and refractive change are most rapid during the first year of life. O'Keefe et al<sup>21</sup> reported that the mean myopic shift was 6.0 D after a mean follow-up of 41 months in children receiving cataract extraction and IOL implantation during the first year of life. Much of the myopic shift occurred in the first 24 months. This is in keeping with the fact that most eye growth takes place in the first 2 years of life. Therefore, because of the constant power of an IOL, a child with pseudophakia might be expected to experience a large myopic shift as the eye grows. This is particularly significant in very young infants. The difficulty in predicting refraction is one of the main disadvantages of IOL implantation. If an IOL calculated for emmetropia is implanted in the first few years of life, myopia of the eye will result after only a few months. However, the use of hyperopic target refraction for an IOL may be associated with amblyopia postoperatively, if refractive correction is not initiated immediately.

To compensate for the myopic shift, several authors have

recommended IOL powers that yield an initial postoperative hyperopia that varies based on the child's age.<sup>48,49</sup> In the first year of life, 6.0 D were subtracted from the calculated IOL power.<sup>21</sup> Hutchinson et al<sup>50</sup> recommended under-correcting most children 3 to 9 years of age by 1 D from the IOL power predicted to achieve emmetropia. Dahan and Drusedau<sup>49</sup> suggested that an infant should receive 80% of the IOL power needed for emmetropia, while in a toddler or a younger child the IOL power should correct 90% of the aphakia. Gimbel et al<sup>51</sup> recommended the implantation of IOLs with a power closer to that predicted for immediate emmetropia. Crouch et al<sup>52</sup> preferred to target a postoperative refraction of 1.0 to 2.0 D hyperopia in children under 6 years old, emmetropia for those over the age of 6 years, avoiding anisometropia greater than 2.5 D. Awner et al<sup>53</sup> and Buckley et al<sup>54</sup> advocated a postoperative refraction of +4.0 D for children under 2 years, +3.0 D for those 2 to 4 years old, +2.0 D for children 4 to 6 years old, +1.0 D for those 6 to 8 years old, and emmetropia in children over 8 years old, adjusting for the fellow eye to avoid anisometropia greater than 3.00 D.

The younger the child is at the time of implantation, the greater will be the myopic shift. To reduce the necessity of IOL exchange, these eyes should be under-corrected. This leads to initial hypermetropia that gradually moves to emmetropia or moderate myopia in adulthood. However, the immediate target refraction should be carefully selected. A balance between risk of amblyopia, future myopic shift and refractive error in the fellow eye should be considered. It is important that the residual refractive error be corrected by spectacles and/or contact lenses that are adjusted throughout life according to refractive development.

IOL calculation formula is also an unresolved issue in paediatric IOL implantation. In adults, older formulas such as the SRK and SRK II have been demonstrated to be less predictable than the newer "theoretical" formulae such as the SRK-T, Holladay, and Hoffer Q, especially in shorter eyes.<sup>55</sup> Andreo et al<sup>55</sup> analysed the predictive value of regression and theoretical IOL formulas in paediatric IOL implantation. Theoretical formulas did not outperform the regression formula. Further studies on special IOL design and IOL formulas for paediatric cataract are warranted.

## Complications

### *Posterior Capsule Opacification (PCO)*

PCO is a frequent complication after cataract surgery in children. The reported risk of PCO in children can be as high as 95%, and its occurrence prevents visual rehabilitation.<sup>56-58</sup> The incidence of capsular opacification is related to the age of the patient and degree of postoperative intraocular inflammation.<sup>59</sup> Hopefully, improving surgical techniques and IOL design will decrease the incidence of

PCO to spare children further operations and lead to prompt and better visual rehabilitation.

The PCO could either be managed surgically with pars plana anterior vitrectomy and capsulectomy or with laser.<sup>60</sup> If the child is unable to cooperate in the clinic, a specially mounted YAG laser machine may be required (Micrupter III, A.S. Meridian, Bern, Switzerland) to perform laser under general anaesthesia. Paediatric membranes can sometimes be quite dense and cannot be safely treated with laser. Reopacification of the anterior hyaloid face by migration of lens epithelial cells across the vitreous face requiring repeated laser treatments is not unusual.<sup>61</sup>

The thickened membrane can be approached anteriorly through the limbus or posteriorly through the pars plana. A limbal approach can render the IOL unstable.<sup>62</sup> On the other hand, a posterior approach through the pars plana was preferred by some surgeons. In addition, sutureless pars plana vitrectomy through self-sealing sclerotomies has been successfully performed in children with PCO.<sup>60</sup> Sudden changes in intraocular pressure during operation are reduced, minimising complications like intraoperative haemorrhage, vitreous herniation and others. Suture-related problems, such as loosening, exposure, and infection, are also avoided. These would have been more difficult to manage in children as extra sessions under general anaesthesia may become necessary. The exposure for surgery in younger children could be suboptimal; use of the 25-gauge transconjunctival sutureless vitrectomy system (TSV) would obviate the need for dissecting scleral tunnels, and may also increase the ease of instrument manipulation in children with small eyes and palpebral fissures.<sup>63</sup> Because the pars plana is not well developed in young children, the site of sclerotomies differs according to the age of the patient, as shown in Table 1 which is derived from the literature, the authors' experience and on an anatomical basis.<sup>63-65</sup>

One main problem of studies on PCO prevalence is the various methods of defining PCO. This renders the comparison of PCO rate with different surgical techniques less reliable. An established method of documentation of PCO is yet to be available although several measurement systems have been suggested.<sup>66-68</sup>

In-the-bag placement of the IOL is preferred because it diminishes the risk of lens dislocation or decentration, iris capture and uveal inflammation. It is also required to consistently reduce the incidence of PCO. Thorough removal of lens substance, including hydrodissection-assisted cortical cleanup, and in-the-bag posterior chamber IOL fixation seem to be the most important factors in reducing PCO.<sup>34</sup> Implantation of posterior chamber IOLs may delay PCO because of a barrier effect created by the IOL optic. This barrier reduces the migration of lens epithelial cells (LECs) from the equatorial zone onto the

Table 1. Sites of Sclerotomies According to Age of Patients<sup>63-65</sup>

Age of patients	Site of sclerotomies (posterior to limbus)
≤3 months	1.5 mm
4 to 6 months	2.0 mm
7 to 12 months	2.5 mm
13 to 36 months	3.0 mm
>36 months	3.5 mm

central posterior capsule.<sup>69</sup>

Acrylic IOL has been related to a significantly lower PCO rate than older IOL types, especially PMMA lenses. Following the “no-space-no-cells” concept, an IOL with a high adhesion property to the posterior capsule should be equated with a lower incidence of central PCO. Moreover, proliferation of LECs is inhibited by the sharp optic edge of the IOL.<sup>70</sup>

The use of a 3-mm to 4-mm primary posterior capsule opening and anterior vitrectomy has been advocated to prevent posterior capsule opacification in children 5 years old or younger.<sup>71</sup> A retrospective review by Jensen et al<sup>72</sup> found that primary posterior capsulorhexis seems to be advisable for children less than 6 years old when cataract extraction with PCIOL implantation is performed. In one study involving children who had received cataract extraction and IOL implantation during the first year of life, 15 eyes had a primary posterior capsulorhexis and only one eye needed further intervention while 10 out of the 11 eyes that did not have primary capsulorhexis developed posterior capsule opacification that required one or more further operations. On the other hand, the disadvantage of this technique is that secondary membranes covering the initial posterior capsulorhexis may develop.<sup>73,74</sup> In addition, Plager et al<sup>36</sup> also found that 80% of children aged 6 months or below with primary IOL implantation and primary PCCC required second operations for recurrence of posterior capsule opacification involving the visual axis. Posterior capture of the IOL optic through the PCCC may prevent secondary membrane formation.<sup>72</sup> As acrylic IOLs were well tolerated in paediatric eyes, Mullner-Eidenbock et al<sup>75</sup> suggested that optic capture was not necessary to ensure a clear visual axis in older children. However, primary posterior capsulorhexis should be performed in preschool and uncooperative children and in eyes expected to have relatively high postoperative inflammation. Anterior vitrectomy is now advocated more often even after primary posterior capsulotomy in order to prevent PCO. This is believed to reduce the scaffolding for the LEC to migrate to the anterior hyaloid face resulting in secondary membrane formation.

Prevention is the first step in management. Besides the prevention of PCO with PCCC and anterior vitrectomy to decrease the migration of LEC across the anterior hyaloid

face, a variety of pharmacologic strategies like mitomycin, anti-inflammatory agents, gene therapy or virus have also been tested for prevention of PCO.<sup>76-85</sup> Though some of these studies are preliminary or were carried out on animals, they may provide useful information for further studies with longer follow-up to reveal the ideal method preventing PCO.

#### *Inflammation*

Since postoperative intraocular inflammation is much more severe in children after cataract surgery and the formation of fibrinous membrane is not unusual, subconjunctival steroids and antibiotics are frequently used at the end of surgery, together with intensive topical steroid therapy postoperatively. Topical prednisolone acetate 1% every hour to 4 hourly is commonly used, occasionally in conjunction with oral steroids. Mydriatic eye drops are important in preventing posterior synechiae and reducing the chance of pupillary block.<sup>9</sup>

#### *Glaucoma*

Aphakic glaucoma has emerged as a serious long-term problem after lensectomy in infancy. The mechanism of aphakic glaucoma is not exactly understood and is most often of an open angle type.<sup>86,87</sup> Johnson and Keech<sup>88</sup> reported a 32% incidence with a mean interval of 47.5 months. Most glaucoma occur only after about 7 years after surgery,<sup>89</sup> but it can also occur months to decades after surgery. Medications and multiple glaucoma surgeries may be needed.<sup>90,91</sup> A more recent study reported a decreased incidence of open-angle glaucoma in pseudophakic eyes compared to aphakic eyes after cataract surgery.<sup>92</sup> The authors proposed that the IOL acts as a barrier between the vitreous and trabeculum, preventing a vitreous chemical component from acting on the trabeculum. Microcornea is another risk factor for the development of glaucoma. There may be an optimal time for cataract surgery to reduce the risk of developing glaucoma.<sup>16,17</sup> All patients will probably need lifelong surveillance.

#### *Rhegmatogenous Retinal Detachment*

The occurrence of rhegmatogenous retinal detachment after paediatric cataract surgery is one of the late complications. There is scant literature regarding this issue. The reported average interval between lens operation and retinal detachment is 23 to 34 years.<sup>93-95</sup> Seventy-two per cent of the detachments occurred after more than 10 years.<sup>95</sup> An internal approach for detachment repair is often needed after congenital cataract surgery and high reattachment rates are achieved.<sup>95</sup> It has been suggested that the detachment rate in children after cataract surgery is similar to that in adults, but the incidence of this complication in childhood is likely to be underestimated, given the relatively short follow-up periods of most studies. However,

the incidence of postoperative retinal detachment after modern surgical techniques can be appraised only after follow-up observation for about 30 years.

#### **Conclusion**

Paediatric cataract surgery remains a challenge to ophthalmologists. Surgical management is however evolving with the advance of microsurgical techniques in adult cataract surgery and these will no doubt be modified and translated to paediatric cataract management.

There are increasing numbers of paediatric cataract surgeons who opt for IOL implantation rather than leaving the infant aphakic.<sup>20</sup> The generally accepted minimum age of IOL implantation is 1 year to 2 years and more surgeons implanting IOL for infants between 6 months and 1 year old. For those under 6 months old, IOL implantation is still highly controversial and is currently being studied in the Infant Aphakia Treatment Trial.

While IOL implantation is becoming more and more common in paediatric cataract patients, there is still no general agreement on IOL power calculation and selection. The basic concept is to leave them more hyperopia the younger they are, but whether this should be on a fixed scale, a percentage scale or some other forms remains unresolved. Although there is evidence of better visual outcomes in infants managed with IOL implantation rather than with contact lens as in aphakia, this is also accompanied with increased number of subsequent operations.<sup>96</sup> Future studies will be needed to reduce the risk of glaucoma and PCO. Refined surgical techniques with posterior CCC, anterior vitrectomy, in-the-bag IOL implantation with or without IOL capture help to reduce PCO. Improved IOL material like acrylic and square edge design further decrease the rate of PCO. Pharmacological agents like cytostatic, anti-inflammatory drugs, gene therapy and even viruses have been studied to maintain a clear visual axis. Established PCO is usually managed with pars plana anterior vitrectomy and capsulectomy in infants and young children or with YAG laser in older or cooperative children and good results can usually be achieved.

Surgery is only one part of the entire management of the paediatric cataract patient. Participation in the visual rehabilitation of the child involving parents, ophthalmologists, paediatricians and optometrists cannot be overemphasised.

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