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INVITED REVIEW

Contact lens management of infantile aphakia

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Richard G Lindsay BScOptom MBA FAAO (DipCL) FCLSA FVCO
Jessica T Chi BOptom
Melbourne, Victoria, Australia
E-mail: richard@lindsayassociates.com.au

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The visual outcomes for infants 18 months or younger with cataracts have improved dramatically over the past couple of decades. Earlier detection of infantile cataract and prompt surgical removal—with subsequent visual rehabilitation with contact lenses—mean that these patients now have a much better visual prognosis. Advances in contact lens technology have led to a significantly higher success rate with contact lenses and this has been a major factor in improving the visual outcomes for aphakic infants. This review outlines the contact lens management of infantile cataract, including a detailed analysis of the various contact lens options available and a discussion regarding the important factors that can cause issues with contact lens wear and affect the overall visual rehabilitation of the infant.

Key words: amblyopia, aphakia, cataract, contact lens, infant

Infantile cataract is one of the major causes of visual impairment in children. Historically, the visual prognosis for children with congenital and infantile cataracts was poor due to the effects of deprivation amblyopia.[1] Based on the work of Hubel and Wiesel[2] and the concept of the critical period of visual development,[2–4] we now know that vision is greatly improved if infantile cataracts are removed as early as possible and appropriate refractive correction provided for the resulting aphakia. Over the past decade the visual prognosis after surgical removal of infantile cataract has improved markedly,[5] although infantile cataract is still responsible for about 10 per cent of the world's childhood blindness.[6,7]

For this paper, we will adopt the definition of Lambert and Drack,[8] who class all cataracts developing during the first 18 months of life as infantile cataract. It is noted that many of these cataracts will be congenital, however, in some cases, it can be difficult to establish the age of onset with certainty. We believe it is simpler to use the term infantile cataract for all cataracts that develop in the first 18 months of life, rather than distinguishing between congenital cataracts—those that are present at birth—and those that develop after birth during infancy.

Studies have shown that the incidence of infantile cataract is about 1.5 in 10,000 in the USA,[8] 2.2 in 10,000 in Australia[9] and over three in 10,000 in the United Kingdom.[10] Approximately 65 per cent of infantile cataracts are bilateral.[11] Over 50 per cent of infantile cataracts, both bilateral and unilateral, will be idiopathic, with the percentage of idiopathic cataracts being higher in unilateral cases.[8] Bilateral infantile cataract is more commonly associated with systemic disorders and hereditary factors; unilateral presentations are more commonly associated with ocular anomalies including microphthalmia, aniridia and persistent hyperplastic primary vitreous.[11] In a small number (less than 10 per cent) of idiopathic cases, there will also be possible contributing perinatal or prenatal causative factors, such as prematurity and/or low birth weight.[11] The congenital rubella syndrome, caused by maternal rubella infection, was one of the most common causes of congenital cataract three to four decades ago. This condition is far less common now due to the widespread use of vaccination against rubella.[12]

Infantile cataracts can also be acquired. These often have a better visual prognosis than congenital cataracts due to the later onset of the amblyogenic factor (the cataract).[12] Trauma is the major cause of acquired cataracts in infants.[8] The trauma can be either blunt or penetrating and is more common in boys than girls.[13] Blunt trauma occurs more frequently than penetrating eye injury,[14] however, penetrating eye injury is more likely to be associated with the development of traumatic cataract.[13] While deprivation amblyopia is less of a factor with traumatic cataracts due to their later onset, the visual outcomes achieved following traumatic cataract surgery are variable, primarily as a result of damage to other parts of the eye including the cornea, retina and optic nerve.[13] Acquired cataracts that are associated with conditions such as long-standing uveitis and poorly-controlled diabetes are uncommon in infants; they will usually be diagnosed in children two years of age or older.[15]

TREATMENT OF INFANTILE CATARACT

Surgical removal of the cataract is now the management of choice for most visually significant infantile cataracts.[8] This should be done as soon as possible to reduce the amblyogenic effect,[16] although it has been shown that cataract extraction within the first month of life is associated with a marked increase in the risk of aphakic glaucoma.[17] Unfortunately, aphakic glaucoma is a common post-operative complication of infantile cataract surgery and this topic will be discussed in more detail later in this paper.

Once the cataract surgery has been performed and the eye is sufficiently healed, visual rehabilitation will be required. This consists of providing the infantile eye with a refractive correction for the aphakia and then initiating therapy to treat the amblyopia.[5,8]

The options for optical correction include spectacles, intraocular lenses, epikeratophakia and contact lenses. Refractive management of bilateral aphakia can be achieved with spectacles, however, the aphakic spectacles will generally be very heavy and difficult to fit to the infant.[18] The aphakic spectacles will also cause peripheral distortion and restrict the visual field due to the prismatic effect associated with the high plus lenses when the patient looks off-centre (the so-called 'jack in the box' effect). In some circumstances, it may not be possible to provide the infant with a full spectacle correction as the maximum power of spectacle lenses is restricted, even in lenticular form, usually to less than +30.00 D. Spectacle correction is not an option for unilateral aphakia due to the aniseikonia and induced prismatic imbalance caused by the marked anisometropia.[12]

While intraocular lenses (IOLs) are now the preferred option for correcting aphakia in adults and generally for older children, their use in babies and infants is still controversial.[8] IOLs have been implanted in babies as young as eight days[19] and they obviously have the advantage of providing a permanent correction to an aphakic eye with optics similar to those of the normal human lens.[8]

Unfortunately, there are many disadvantages associated with the use of IOLs in infantile eyes.[8] The infantile eye requires a smaller diameter IOL; this may lead to problems later in adult life, as there is a substantial risk of intraocular lens luxation into the vitreous cavity during subsequent growth of the eye. There is also a greater risk of complications such as posterior capsular opacity, corneal endothelial cell loss and uveitis.[5,19] The subsequent myopic shift that occurs in a pseudophakic infantile eye due to axial elongation means that when choosing the power of the IOL, the surgeon has to either leave the infant significantly undercorrected (hyperopic), based on the assumption that there will be movement towards emmetropia as the eye grows, or aim for emmetropia immediately after surgery and accept that the eye will become myopic as the axial length increases, based on the tenet that any optical defocus during infancy is more likely to lead to amblyopia.[5] Interestingly, for infants the incidence of glaucoma appears to be less with primary IOL implantation than in eyes left aphakic.[19,20]

Epikeratophakia (or epikeratoplasty) is a surgical procedure in which a lamellar graft is sutured onto the surface of the recipient cornea to reproduce the effect of a highly positive contact lens. This procedure has generally not been very successful for infantile aphakia due to clouding of the donor tissue for an extended period after surgery, therefore reducing visual acuity and increasing the risk of amblyopia. In addition, like the implantation of an IOL, this is a relatively permanent change, so continued axial growth after the procedure leads to a significant myopic shift in the patient's refractive error.[21] Epikeratophakia is rarely performed these days; it is occasionally used for patients who have a history of acquired cataract in one eye and therefore a reasonably good visual prognosis after cataract surgery but are contact lens intolerant and not a candidate for an IOL.[12]

Even though the use of primary IOLs for infantile cataract surgery is increasing,[19] contact lenses are usually the first option considered for the management of infantile aphakia. Most importantly, contact lenses are readily changed as infantile eyes grow and the refractive error (hyperopia) decreases.[1,8] With the developments in contact lens technology over the past 10 years, there are now many options available to contact lens practitioners, with which they can manage this special class of patients. As a result, contact lens complications such as microbial keratitis, corneal hypoxia, corneal vascularisation and giant papillary conjunctivitis are now less commonly observed.[22]

MENSURATION OF THE INFANT EYE

An understanding of the usual parameters—in particular, corneal curvature, corneal diameter and refractive error—of both normal and aphakic eyes during the neonatal period and infancy is essential for the contact lens management of infantile aphakia for three reasons.

First, when fitting infant eyes with contact lenses, it will not always be possible to obtain accurate measurements of these parameters. Therefore, the choice of the

Figure 1. Examination of an infant using a hand-held slitlamp

initial diagnostic lens for contact lens fitting is often based on the expected values for the patient's age. The average corneal curvature at birth has been found to be somewhere between 48.50 D (6.96 mm)[23] and 47.00 D (7.18 mm)[24] and the corneal diameter is approximately 10 mm at birth.[25] The refractive error at the corneal plane of an aphakic eye at age one month can be anywhere between +19 and +38 D, with an average of about +30.75 D, which is about 2.5 times greater than that of the average aphakic adult.[26]

Second, there is rapid growth of the eye during the first 18 months of life, leading to an increase in the corneal radius of curvature, an increase in corneal diameter and a reduction in hyperopia. Therefore, frequent changes in the contact lens prescription will be required during this period. The greatest increase in corneal radius of curvature occurs in the first three months[24]—anywhere between 0.2 and 0.8 mm—with the average corneal curvature approaching that of an adult (about 7.80 mm) by age three years.[27] By age two years the cornea has assumed its maximum diameter,[27] with an average value of 11.7 mm.[25] For aphakic eyes, the most rapid decrease in the large hyperopic refractive error occurs in the first year of life with an average reduction of approximately four dioptres during this period.[26] Moore[26] showed that for unilateral aphakes who had their congenital cataracts removed in the first six months, the rate of change per month decreased from 0.43 D between one and six months, 0.37 D between six and 12 months, 0.30 D between 12 and 18 months, 0.24 D between 18 and 24 months and less than 0.19 D thereafter.

Finally, the aphakic eye often demonstrates changes in ocular growth and refraction different from those of the normal eye. Corneal flattening has been observed to occur at a faster rate in the aphakic eye than the normal eye.[23] Congenital cataracts occur frequently in microphthalmic eyes with corneal diameters of less than eight millimetres.[16] Even for eyes that are not microphthalmic, most patients with unilateral congenital cataracts have smaller corneas in the affected eye and this difference in corneal diameter becomes more obvious during the first three years of life.[23] The reduction in hyperopia and the rate of reduction are markedly higher for an aphakic eye than a normal eye.[26] It is thought that one of the major factors contributing to this is the axial length. Eyes with unilateral congenital cataracts have a far more rapid growth in axial length following cataract removal than the fellow non-aphakic eye.[26,28]

PATIENT EXAMINATION AND CONTACT LENS FITTING

The following examination procedures need to be performed prior to fitting the infant with contact lenses.

- **Case history**

It is vitally important that the parents are asked some pertinent questions. Ascertaining whether a child was premature or full-term is important as premature babies have steeper corneal curvatures[24] and smaller corneal diameters.[23] Congenital cataracts can be associated with other systemic abnormalities,[8] which may preclude contact lens wear. Previous failed efforts to initiate contact lens wear may have traumatised the infant and this will decrease the chances of a successful contact lens fitting. If the child has had any previous problems with general anaesthesia, it would probably rule out the possibility of using anaesthesia for future examination and contact lens fitting.

- **Examination of the anterior segment**

This is best done with a hand-held slitlamp (Figure 1) and should include assessment of the eyelids, anterior chamber, cornea and conjunctiva, with fluorescein and a blue light filter to check for any corneal or conjunctival staining.

- **Keratometry to determine the corneal radius of curvature**

This is best done with a hand-held autokeratometer.

- **Refraction**

Determination of the refractive error of the patient can only be done using retinoscopy. This technique is also used for the contact lens over-refraction at the fitting appointment and subsequent after-care visits.

- **Measurement of the horizontal visible iris diameter (HVID) as an approximation of the corneal diameter.**

Obviously, techniques such as biomicroscopy and keratometry are not performed easily on an infant in the consulting room. In addition, contact lens fitting and after-care can be a challenge, especially if fitting rigid or silicone elastomer lenses, where more precision is required compared to soft lens fitting. In regard to rigid lenses, for this paper the authors will adopt the convention proposed by Efron[29] that 'rigid lenses' will refer to all lenses made from rigid gas-permeable materials. The term 'rigid gas permeable' (RGP) will not be used because the adjective 'gas-permeable' is effectively redundant; virtually all rigid lenses today are made of gas-permeable materials.

There is the alternative of examining and fitting infants under general anaesthesia. The benefits of an accurate and efficient fitting in a still child must be balanced against the distress the family experiences by having to fast an infant and spend a morning in hospital, especially if repeated examinations are planned.

A recent report[30] states that although anaesthesia-related death 'is exceptionally rare, it is clear that the risk remains ever present, even in low-risk patients'. The report estimates the rate of death solely attributable to anaesthesia to be one in every 250,000 anaesthetics. The risk of mortality during a general anaesthetic is higher in infants than adults and even higher in infants less than one year of age, predominantly due to obstructed or shallow breathing.[31,32] Safety measures always need to be in place; the 1989 UK National Confidential Enquiry into Perioperative Deaths[33] recommended that anaesthetists should not undertake occasional paediatric practice, and facilities for the resuscitation of children should be mandatory at hospitals where paediatric anaesthesia is carried out.

The other concern is that it is generally accepted that general anaesthesia will bring about a reduction in intraocular pressure.[1] This decrease can affect the corneal topography and so the corneal curvature and refractive error determined under anaesthesia can be very different from those obtained when the infant is awake. The authors' clinical experience leads them to concur with the findings of Moore[1] that there are often significant differences in the fitting characteristics of contact lenses and the refractive correction of infants when examined under anaesthesia compared to when assessment is performed while they are awake.

Based on all these factors, the authors strongly endorse the recommendation of Moore[1] that examination and contact lens fitting under general anaesthesia should be performed only when absolutely necessary, that is, when it has been shown to be impossible in the consulting room. It is important to note that the use of mild sedating agents, such as promethazine hydrochloride (Phenergan, Aventis Pharma) and trimeprazine tartrate (Vallergan, Aventis Pharma), which are often used in young children, is not recommended with infants, as the safety of their use has not been established.[34]

While examination and contact lens fitting of the infant in the practice setting can be difficult, there are ways in which the contact lens practitioner can facilitate this process. If it is not possible to obtain an accurate measurement of ocular parameters such as corneal curvature and refractive error, then the selection of the initial diagnostic contact lens can be based on the normal values expected for age. This lens can be assessed on the eye and then appropriate changes made to the contact lens prescription.

Retinoscopy can also be a difficult technique to perform accurately. In particular, infants will not always respond well to having working distance lenses placed in front of their eyes. For determination of the aphakic refraction, it may be best to use a pair of +20.00 D flippers or a retinoscopy rack that incorporates a range of plus powers. Given that it is usual practice to over-correct aphakic infants in the first few years when prescribing contact lenses (this point will be discussed in more detail below), it should be possible to determine the over-refraction without the use of a working distance lens. For example, if performing retinoscopy over a contact lens, neutralisation of the retinoscopic reflex at a working distance of 50 cm without a working distance lens in place means that the over-refraction is -2.00 D.

CONTACT LENS OPTIONS FOR INFANTILE APHAKIA

There are several contact lens modalities that can be used for the contact lens correction of infantile aphakia. These include rigid lenses, soft (hydrogel and silicone hydrogel) lenses, silicone elastomer lenses and scleral lenses. While the soft lens is the most frequently used lens type in paediatric contact lens fitting,[18] the use of rigid contact lenses is increasing due to the many benefits associated with this lens modality. Silicone elastomer lenses are not as commonly used as either soft or rigid lenses, although there is still a significant number of aphakic infants fitted with this type of lens, especially in the USA. Scleral lenses are not commonly used for the contact lens management of infantile aphakia.

Soft contact lenses are relatively easy to fit to the infantile aphake and probably, this has been the major reason for their frequent use in paediatric contact lens fitting. They can be custom made in a wide range of parameters to fit any size of eye. The initial comfort with soft lenses is good, so infants generally adapt well to the wearing of these lenses. Soft lenses tend to be fairly stable on the eye, although they are more commonly dislodged and ejected from infant eyes compared to adult eyes due to factors such as excessive eye rubbing and lens dehydration.[18]

There are many disadvantages associated with the use of soft lenses for the contact lens management of infantile aphakia. The major problem is corneal hypoxia. Traditionally, hydrogel lens materials, which have a relatively low oxygen permeability (Dk),[35] have been used when prescribing soft lenses to aphakic infants. Obviously, high plus lenses are needed for these patients and it is not uncommon to require powers well above +30.00 D. Consequently, these soft lenses are very thick, with a centre thickness generally greater than one millimetre. The combination of the low Dk and the large centre thickness means that these lenses have a very low oxygen transmissibility (Dk/t)—even if higher water content materials are used—that is usually well below the level required to avoid hypoxic stress to the cornea during daily wear.[36] Long-term corneal hypoxia subsequently can lead to complications such as corneal vascularisation

Figure 2. Marked corneal vascularisation in an aphakic child who has been wearing a high plus (greater than +20 D) hydrogel soft contact lens for more than five years

(Figure 2), stromal oedema and chronic endothelial dysfunction.[37]

Lens dehydration is also a significant factor with soft lenses made of a hydrogel material with a high plus back vertex power (BVP). Fatt and Chaston[38] showed that lenses of the kind fitted to aphakic patients (greater than +15.00 D) may lose several dioptres of plus power when lens dehydration occurs on the eye. This necessitates that the aphakic lens needs to be made in an even higher plus power to allow for this power reduction on the eye. As a result, it is not uncommon to order soft contact lenses for infantile aphakes with back vertex powers of over +40.00 D. This even higher plus power further compounds the problem of the reduced Dk/t discussed above.

Silicone hydrogel lenses provide a marked increase in oxygen supply to the cornea compared to hydrogel lenses, such that it is now possible to make soft lenses, even in a high plus power, with a Dk/t that may satisfy the Holden-Mertz criterion for no corneal oedema during daily wear.[36,39] Initially, the silicone hydrogel soft lenses were not an option for the contact lens management of infantile aphakia, as they were available only in a disposable (frequent replacement) modality with a limited range of powers and fitting parameters. Custom-made lathe-cut silicone hydrogel lenses are available in a wide range of parameters, including high plus powers, so they are definitely the preferred option if prescribing soft lenses for infantile aphakia. Corneal hypoxia tends not to be a problem with silicone hydrogel lenses,[39] however, these lenses, especially if worn on an extended wear basis, can accumulate high levels of lipid deposition[40] and be associated with clinical complications such as superior epithelial arcuate lesions (SEALs),[41] mucin balls[42] and contact lens related papillary conjunctivitis (CLPC).[43]

The philosophy for fitting soft lenses to an aphakic infant is similar to that used when fitting soft lenses to an adult, with the optimal fitting lens showing good centration and corneal coverage in all directions of gaze and about 0.50 mm of movement on blinking. The fitting needs to be more precise if using silicone hydrogel lenses, as the increased modulus (stiffness) of the silicone hydrogel materials means that these lenses do not conform to the corneal shape as easily as hydrogel lenses.[44] It is important to note that this higher modulus of elasticity of silicone hydrogel lenses aids lens handling as the increased lens 'stiffness' means that lens insertion is easier.

Measurement of corneal curvature by either keratometry or videokeratoscopy may not always be possible, so the initial choice of back optic zone radius (BOZR) is often based on the age of the infant.[16] The BOZR will usually be about 0.5 mm flatter than the mean corneal curvature, so for a neonate, the first BOZR will generally be about 7.40 mm. This BOZR will have to be increased frequently, initially about every six months, as the eye grows and the cornea flattens. For the total diameter (TD), it is generally best to err on the large side, fitting about 2.5 to 3.0 mm larger than the horizontal visible iris diameter, especially as infants are more likely to rub the lenses off the eye. The choice of total diameter will depend on the infant's age. At birth, the HVID is normally about 10 mm, so a total diameter of either 12.5 or 13 mm is usually required. Just like the BOZR, the total diameter will also have to be increased regularly over the first 18 months, as the eye grows and the corneal diameter becomes larger. It is important to note that congenital cataract and posterior hyperplastic primary vitreous often occur in microphthalmic eyes with a corneal diameter of less than eight millimetres. For these patients, a total diameter of between 11 mm and 12 mm is usually required.[16]

There are other disadvantages associated with the prescribing of soft contact lenses for the infantile aphake. Soft lenses do not mask significant corneal astigmatism[18] and, due to the excessive lens thickness associated with the aphakic prescription, it is not feasible to fit these infants with anything other than spherical lenses.[16] The insertion of the soft lenses by the parents can by very difficult due to a combination of the very small palpebral aperture and the high plus power of the lens. Lens loss will be common as the lenses are frequently ejected from the eye due to excessive eye rubbing as well as lens dehydration.[18]

The use of rigid contact lenses in paediatric contact lens fitting is becoming more widespread, as these lenses can be manufactured from materials with Dk greater than 100 Barrer,[45] forgoing the hypoxic issues associated with soft hydrogel contact lenses that were discussed earlier. Rigid contact lenses made from high Dk materials such as Boston XO (Dk of 100 Barrer)[45] or Fluoroperm 151 (Dk of 151 Barrer)[45] easily satisfy the Holden-Mertz criterion for both daily and overnight wear, even at the very high plus powers required by aphakic infants.[45,46]

Rigid contact lenses can be manufactured readily and reliably in virtually any parameters. They are easily verified and evaluation of the fit is more accurate compared to soft lenses due to the ability to assess fluorescein fitting patterns. Rigid lenses are able to correct for any corneal astigmatism and offer far greater material stability than soft lenses as they do not

C5/6.60:6.40/6.90:7.60/7.30:8.80/8.00:9.80/9.60:10.80	+34.00
C5/6.70:6.40/7.00:7.60/7.40:8.80/8.10:9.80/9.70:10.80	+34.00
C5/6.80:6.40/7.10:7.60/7.50:8.80/8.30:10.00/9.80:11.00	+33.00
C5/6.90:6.40/7.20:7.60/7.60:8.80/8.40:10.00/9.90:11.00	+33.00
C5/7.00:6.40/7.30:7.60/7.70:8.80/8.50:10.00/10.20:11.00	+32.00
C5/7.10:6.40/7.40:7.60/7.80:8.80/8.60:10.00/10.30:11.00	+32.00
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C5/7.60:6.60/8.00:8.00/8.40:9.20/9.40:10.40/11.40:11.60	+29.00
C5/7.70:6.60/8.10:8.00/8.50:9.20/9.50:10.40/11.50:11.60	+29.00
C5/7.80:6.60/8.20:8.00/8.60:9.40/9.60:10.60/11.60:11.80	+28.00

Table 1. Rigid lens diagnostic fitting set for infantile aphakia

dehydrate on the eye. Contact lens complications such as microbial keratitis, corneal vascularisation and CLPC are less common with rigid lenses.[22,37,47] They also have the added benefit of ease of handling, as they do not fold over and may be inserted through a much smaller eyelid aperture than is required for soft lenses. Interestingly or even paradoxically, rigid lenses are less commonly rubbed out of the eye than soft lenses, as rubbing rigid lenses will usually cause the infant discomfort or pain.

Rigid contact lenses do have disadvantages. They may require a period of adaptation as initially, they are generally less comfortable than soft contact lenses. Adapting to the rigid lenses tends to be more of a problem as the child becomes older. Clinical experience shows that fitting a child with rigid contact lenses prior to the age of 18 months tends to be more successful than initiating treatment with rigid contact lenses as the child progresses to being a toddler at two years and older. As they do not mould completely to the shape of the cornea, rigid lenses are more prone to foreign bodies being trapped underneath compared to soft lenses. Mechanical irritation and subsequent corneal abrasions can be a problem if infants rub their eyes.[16] Rigid lenses are more easily ejected from the eyes than soft lenses.

As with soft lenses, fitting rigid lenses to an aphakic infant is similar to the technique used when fitting high plus rigid lenses to an adult. The optimal fitting lens should display a well-centred alignment fit, with adequate edge clearance evidenced by approximately a one millimetre band of fluorescein around the edge of the lens.[48] The fitting needs to be more precise than with soft contact lenses, as the rigid lenses do not conform to the corneal shape as easily as soft lenses. If the BOZR of the rigid lens is too flat (that is, greater than required), excessive edge lift, lens discomfort and instability may ensue. If the BOZR is too steep, that is, less than required, there will be central fluorescein pooling with restricted tear exchange and lens movement. This can cause the lens to tighten and become uncomfortable after prolonged wear.[48] Very steep-fitting rigid lenses may cause bubble formation, which can reduce vision and cause hypoxia.

The main difference in fitting rigid lenses to infants compared to adults is the choice of total diameter. For infants, it is generally best to err on the large side with respect to the TD. The aim is to fit a lens with complete corneal coverage, that is, slightly larger than the horizontal visible iris diameter. The choice of total diameter will depend on the infant's age; at birth, the HVID is normally about 10 mm,[25] so a total diameter of 10.8 to 11.2 mm is usually required. Just like the BOZR, the total diameter will have to be increased regularly over the first 18 months as the eye grows and the corneal diameter becomes larger.[23] Larger lenses provide better centration and stability, and they are less likely to mislocate on the eye or be rubbed out of the eye. Caution must be taken not to prescribe lenses that are too large, as lens insertion and removal may become too difficult for the parents.

For the fitting, it is imperative to use a diagnostic rigid lens set that has been specifically designed for aphakic infants with appropriate BOZR, TD and BVP. An example of such a fitting set is shown in Table 1. Note that the front optic zone diameter (FOZD) should be smaller than would usually be used with a lower refractive correction to reduce the overall lens thickness and weight. As an approximation, the FOZD should be about the same as the back optic zone diameter (BOZD), which is generally less than seven millimetres. If the rigid lens fitting is being performed in the practice setting, and therefore not with the infant under general anaesthesia, it is recommended that a drop of topical anaesthetic be instilled into the eye prior to inserting the rigid lens.[16]

Ideally, as with other contact lens modalities, a measurement of corneal curvature by either keratometry or videokeratoscopy will give a good starting point for fitting the infantile aphake, with the aim of fitting a lens on the mean corneal curvature. As discussed previously, it is not always possible to obtain an accurate measurement of corneal curvature, so in these circumstances, the initial choice of back optic zone radius for fitting will be based on the age of the infant.[16] For a neonate, the first BOZR will generally be about 7.00 mm.[23,24] The initial contact lens BVP can range from between +18.75 to +40.00 D with the average being 31 D.[49] Due to the greater precision required with rigid lens fitting, these lenses will need to be modified more often than soft lenses. In particular, the BOZR will have to be increased more frequently, about every three months for the infant's first 12 months, as the eye grows and the

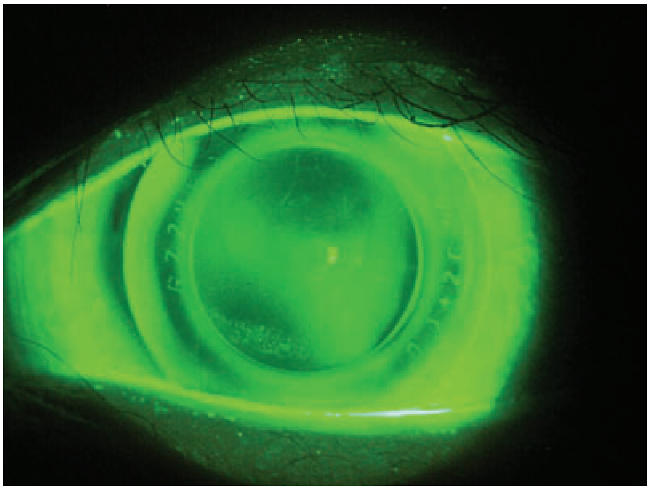

Figure 3. Fluorescein fitting pattern of a Silsoft lens showing good central alignment and appropriate edge clearance

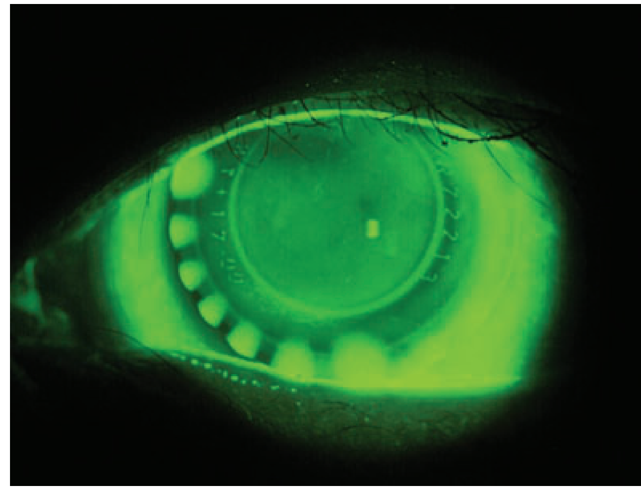

Figure 4. Fluorescein fitting pattern of a Silsoft lens showing lens fluting as a result of excessive edge clearance

cornea flattens.[49] This is in addition to the usual changes in BVP (reduction) and TD (increase) that are required during the first 18 months of life as a result of the relatively rapid growth of the eye.[23,26]

Silicone elastomer lenses were first approved for use with aphakic patients in the mid-1980s. Presently, the Silsoft (Bausch & Lomb, Rochester, NY) lens is the only silicone elastomer lens on the market and this lens has been approved by the Food and Drug Administration in the USA for use as a 30-day extended wear lens for aphakia.[50] The main feature of the silicone elastomer material is its very high oxygen permeability,[51] much higher than the currently available silicone hydrogel materials, so that the silicone elastomer lenses have an oxygen transmissibility high enough to meet the oxygen requirement for extended wear according to the Holden-Mertz criterion.[36]

Silicone elastomer lenses do have other advantages. Due to their increased modulus they are easier to insert than soft lenses.[18] Their rigidity means that they will usually give better visual acuity than soft lenses.[52] Lens dehydration on the eye is generally not a problem because of the hydrophobic lens material.[52] Silicone elastomer lenses are more stable on the eye and not as easily rubbed out or mislocated.[16]

Unfortunately, there are many problems associated with the use of silicone elastomer lenses. They come only in a limited range of parameters; the steepest available BOZR (7.50 mm) is often too flat for infant aphakes[16] and the highest available plus power (+32.00 D) can be below the required aphakic prescription,[53] especially in the first few months of life. Due to the hydrophobic nature of the lens material, these lenses incorporate a hydrophilic surface coating, which can become scratched or damaged, leading to the lens having a very uncomfortable hydrophobic surface.[18] Alternatively, the coating can become heavily deposited with lipid[54] or mucus,[55] especially if the lenses are worn on an extended wear basis,[53] resulting in a degradation of vision and the need for frequent lens replacement. The fitting process is more complex; the lenses have to be fitted very carefully and they can bind to the cornea if the fitting is too tight.[56] The lenses are very expensive, which is a significant factor given that they have to be replaced frequently, probably about every three months, due to a combination of lens spoliation and required changes in the lens parameters as the eye grows.

The method of fitting the silicone elastomer lenses is similar to that for rigid lenses. As for rigid lenses, the fit is assessed by use of fluorescein and an ultraviolet light source, however, using rigid trial lenses to determine the best fitting silicone elastomer lens is not a very successful technique, so silicone elastomer diagnostic lenses should always be used to determine the optimal fit.[53] For the initial trial lens, fit on flattest K for both toric and spherical corneas. If in doubt or if the keratometric readings are not available, the results of one study suggested that it is best to use the 7.50 mm BOZR if the infant is under seven months and a BOZR of 7.70 mm if the child is between the ages of seven and 17 months.[53] When specifying the BOZR, it is best to err on the flatter (greater) side, as the lenses will tighten on the eye and a steeper fitting lens may bind and be difficult to remove.[16] The optimal fitting lens will show good central alignment with no tear pooling; the edge clearance should be plentiful as denoted by a ring of fluorescein around the edge of the lens of at least about one millimetre (Figure 3). Excessive edge clearance can lead to lens fluting as shown in Figure 4.

Scleral lenses, made from a gas permeable material with a high Dk, are rarely used these days for the contact lens management of infantile aphakia, which is perhaps a little surprising given that they offer many advantages including superior initial comfort compared to rigid lenses, good stability on the eye, excellent vision and relatively easy handling.[57] Scleral lenses are also a good option where there is an irregular corneal surface as a result of trauma to the eye.[16] The complication rate with these lenses is also very low, partly because the lenses are generally fitted with corneal clearance.[57] Scleral lenses can also be modified in office as the eye grows and the prescription changes, obviating the need to order replacement lenses.[16,57] The reason for scleral lenses being used sparingly in paediatric contact lens practice is probably two-fold. First, they are perceived as being difficult to fit; second, the initial cost of these lenses to the patient is usually very high, about 10 times the cost of a rigid contact lens.

IMPORTANT CONSIDERATIONS IN THE CONTACT LENS MANAGEMENT OF INFANTILE APHAKIA

Lens handling

In most cases, the infant will need to be restrained for both contact lens insertion and removal. This will often require two adults, as the child's arms, legs and head need to be immobilised. For babies, restraint can sometimes be achieved by swaddling them in a sheet or blanket and placing them on the floor or on a change table. As children get older, they get stronger, thus firm restraint with rapid lens insertion/removal is important. If restraining the child proves to be very difficult, regardless of the age, lens insertion and removal can be performed on the child about one hour after they have gone to sleep, with the assistance of a dull torch light. If the contact lens can be tinted this is definitely recommended, as this will make it easier for the parents to confirm that the lens is on the eye and find it if lens mislocation occurs.

For contact lens insertion, it is usually not possible to pull the infant's eyelids wide enough apart to allow the lens to be placed directly onto the eye, regardless of the lens modality. For all types of contact lenses, it is best to insert the contact lens under the upper lid and then place it under the lower lid while ensuring at all times that the contact lens is kept in the same plane as the cornea. As previously noted, soft lenses are far more difficult to insert than rigid lenses due to their larger diameter and more flexible nature.

For contact lens removal, soft lenses are removed, as they are for adults, by simply pinching and lifting them off the eye. In a small number of cases, where the soft lenses are extremely thick due to a very high plus power, they may need to be removed by the technique usually associated with rigid lenses, whereby the eyelids are used to lift the lens off the eye. Rigid and silicone elastomer lens removal is best done using a moistened rubber suction holder. This should have the end moistened with saline or the lens storage solution. Lens centration must be confirmed prior to attempting lens removal.[16] With the child's eyelids held apart, the suction holder is pressed gently against the lens and the lens removed directly. For some infants, rigid lens removal can also be done by the same method used for adults, whereby the lens is removed by index finger manipulation of the upper and lower eyelids.

If the infant has been fitted with a rigid lens, it may be preferable for the parents to instil a drop of unpreserved (unit dose) topical anaesthetic into the child's eye prior to inserting the contact lens. This is to facilitate the infant's adaptation to contact lens wear and is usually required only for the first month or two of wear. Topical anaesthetics that come in a bottle should not be used for this purpose, as these will usually incorporate preservatives such as benzalkonium chloride, which can have a toxic effect on the cornea if the cornea is exposed to them for a prolonged period.[58–60]

Care must be taken if advising the use of anaesthetic eye drops to aid in lens adaptation, due to the potential for misuse by parents and the increased risk of epithelial damage, especially through overuse. Parents should be counselled that the topical anaesthetic is to be used only prior to lens insertion and for a short period of time, to aid with adaptation; it should not be used on an ongoing basis to provide comfort during contact lens wear. It is the authors' clinical experience that the risks of using topical anaesthetic, albeit in specific cases and only for a short period, are outweighed by the marked benefit of assisting with lens adaptation. In several cases, this has been the critical factor in achieving successful contact lens wear.

Daily wear versus extended wear

Given the dependency that infantile aphakes have on contact lenses for both their visual correction and visual development, it is essential that complications associated with contact lens wear are minimised. Consequently, daily wear is the preferred option for these patients. It is true that there are contact lenses now available that meet the Holden-Mertz criterion for Dk/t of 87 Barrer/cm to avoid excess oedema during overnight wear,[36] namely some silicone hydrogel lenses, all silicone elastomer lenses and rigid lenses made from materials with very high Dk.[46] Even with this higher oxygen transmissibility, the complication rate and the risks associated with extended wear are higher than for daily wear.[41–43,46,47,52,61–63] It is important to note that hydrogel lenses incorporating an aphakic prescription will not meet the Holden-Mertz criterion for daily wear, let alone overnight wear.[35,36]

Daily wear may be too difficult for the parents to manage due to problems with lens insertion and removal. In this case, the infant is more likely to be successful with the contact lenses if the child is permitted to wear the lenses overnight so that lens handling is avoided. We recommend that only extended wear—lenses worn for up to one week—is acceptable; continuous wear—lenses worn for up to one month—is undesirable due to the risks involved. Again, we stress that daily wear should be used as the first option, with extended wear reserved for those patients for whom lens insertion and removal is too difficult to perform on a daily basis. Inserting a contact lens is much easier on very young babies and neonates than on children a few years older, who are stronger and more fearful of being restrained. Establishing a pattern of daily wear from the beginning means that the child will be far more accepting of the lens handling procedures as they get older.[1]

Contact lens back vertex power

Aphakic infants are unable to accommodate. They are, akin to other infants, more likely to be interested in objects that are close to them such as faces and toys.[16] Consequently, they should be overcorrected by about +2.50 to 3.00 D to focus them at this near viewing point.[1] This overcorrection should be reduced to about +1.00 to 1.50 D at 18 to 24 months of age, when the young child starts to become more aware of distant objects. At about three to four years of age, the contact lens should have the appropriate distance correction, with the reading correction then incorporated into a pair of spectacles, usually bifocals or multifocals, to be worn over the contact lenses.[18]

Glaucoma

Unfortunately, aphakic or secondary glaucoma is a common post-operative complication of paediatric cataract surgery. Depending on the length of post-operative follow-up, the incidence can vary between 15 and 32 per cent.[17,64–66] A recent study[64] showed that glaucoma developed in 15.4 per cent of children who underwent surgery for congenital cataract, where the mean follow-up time was 6.3 years and the mean time to diagnosis of glaucoma after surgery was 4.9 years. Obviously, a longer follow-up is associated with an increased prevalence of aphakic glaucoma. There have been reports[67] of aphakic glaucoma being diagnosed over 10 years after the original cataract surgery and this is consistent with the authors' clinical experience.

As noted earlier in the paper, cataract extraction within the first month of life is associated with a marked increase in the risk of aphakic glaucoma.[17] Microcornea, persistent hyperplastic primary vitreous and posterior capsulotomy have also all been confirmed as significant risk factors for the development of this condition.[64,65] The actual mechanism by which the secondary glaucoma occurs is not well understood, although the two most popular theories are that it is due to post-operative inflammation of the filtration angle or it is related to the mechanical absence of the lens.[64]

Consequently, intraocular pressures (IOP) must be closely monitored in infants who have undergone cataract surgery. Initially, children with aphakic glaucoma can be asymptomatic, particularly if the IOP is only slightly elevated.[64] With marked increases in IOP, ocular changes will usually become apparent, most noticeably corneal haze caused by epithelial oedema and corneal clouding. Lacrimation, photophobia and blepharospasm, all of which are associated with the corneal haze, may also be observed.[64,68] Buphthalmos, which is defined as an enlargement of the eye due to raised IOP prior to the age of three years, can happen along with Haab's striae (these represent healed breaks in Desçemet's membrane). Optic disc cupping is not always seen in the early stages of aphakic glaucoma.[68]

An important point to consider is that the fit of the contact lens may change dramatically if buphthalmos occurs in the affected eye. Enlargement of the eye will cause an increase in corneal diameter, a significant flattening of the cornea and axial myopia leading to a marked reduction in the hyperopic refractive error. Clinicians should be suspicious of any sudden change in the fit of a contact lens on the eye of an aphakic infant. For example, if fitting a rigid contact lens, there would be cause for concern if the lens went from being a good alignment fit to one that now demonstrated marked apical clearance (central pooling) or if the lens now looked relatively smaller on the eye or if the refraction over the lens showed the infant to be overcorrected by an amount much greater than the +2.00 or +3.00 D that is usually prescribed.

After-care

Given the complexity of fitting contact lenses to infants and the fact that there is rapid growth of the eye during the first 18 months of life, it goes without saying that frequent after-care is essential for these patients. During the fitting process, the infant should be reviewed at least weekly. Once contact lenses have been fitted successfully, after-care visits every four to six weeks are recommended. If the infant is wearing the contact lenses on an extended wear basis, more frequent review is recommended.

The following examination procedures need to be performed at the after-care appointments.

- **Case history**

Is the child rubbing the eye? Are the contact lenses causing any distress to the child? Has any redness, watering or discharge been noted? Are the contact lenses stable on the eye? Does the child appear to be fixating well with the lenses? Are the appropriate lens maintenance procedures being performed? Have there been any problems with lens handling?

- **Assessment of the contact lens fit**

Lens centration and movement should be evaluated. The fluorescein fitting pattern should be assessed for rigid, silicone elastomer and scleral lenses.

- **Slitlamp examination**

This should include checking for any corneal or conjunctival staining; noting any limbal vascular changes or any corneal vascularisation; eversion of the upper eyelids to check for any signs of CLPC; noting if there are significant deposits on the contact lens.

- **Retinoscopy to determine the contact lens over-refraction.**

After-care visits can generally be performed in the practice setting, with the option of examining the infant under general anaesthesia rarely needed. Intraocular pressures need to be closely monitored at these after-care appointments given the risk of developing glaucoma. Frequent changes to the contact lens prescription will be required during the first 18 months due to the growth of the eye and the practitioner must not hesitate to make these modifications, even if it

Figure 5. Marked left esotropia in an infant with bilateral aphakia

means ordering new contact lenses every two to three months.

Amblyopia

The visual outcome for eyes with infantile onset cataracts depends on factors such as the age at diagnosis, the density of the cataract, the age at which surgical treatment and visual rehabilitation is initiated and whether the cataracts are unilateral or bilateral.[8] This last point is very important as the visual prognosis for bilateral infantile cataracts is much better than for unilateral infantile cataract. Amblyopia is inevitable with unilateral cataract, whereas bilateral aphakic infants do not always have significant amblyopia.[1] The age at which treatment is initiated is also especially critical for unilateral infantile cataract, with studies showing that earlier treatment of unilateral cataract is associated with a better visual outcome.[69] Bilateral nystagmus may also occur with both unilateral and bilateral cataracts[8] and its presence is usually an indicator that vision is affected significantly.[12]

Treatment of the amblyopia is by penalisation of the non-amblyopic eye with either patching or atropine therapy. These have been shown to be equally successful for the treatment of amblyopia,[70,71] although achieving compliance with patching will be more difficult as the child gets older and will also depend on the degree of visual improvement in the amblyopic eye. Care should be taken with atropine penalisation, as this can result in amblyopia in eyes with normal acuity.[72] Most aphakic children will require some patching until about eight years of age.[12] Atropine penalisation is not an option if the infant is bilaterally aphakic due to the absence of accommodation in both eyes.

For bilateral infantile aphakia, there is also the option of refractive penalisation by wearing a contact lens only in the amblyopic eye. Leaving the contact lens out of the non-amblyopic eye will result in significant hyperopic blur, however, it is the authors' experience that patching is more effective for treating the amblyopia, as the child will often still try and sight with this better eye even though it may be uncorrected by an amount greater than 30 D.

While better visual outcomes are obtained in bilateral cases, there have been cases of unilateral infantile cataract documented where visual acuity of better than 6/12 has been achieved by appropriate surgical treatment and contact lens management.[73,74] Unfortunately, there is a wide variation in visual outcomes with some cases of unilateral infantile cataract ending up with visual acuity of less than 6/60, despite having the proper management.[73] As a result, a significant percentage of children with a history of unilateral infantile cataract, probably about 30 per cent based on the authors' experience and other studies,[75] will discontinue contact lens wear and this will be more likely to be due to poor visual acuity than problems related to the fitting and wearing of contact lenses.[75]

Strabismus

Strabismus occurs in a large number of infants with either unilateral or bilateral cataract, although it is more common with the former.[8] There is also a higher incidence post-operatively compared to pre-operatively.[76] Congenital cataracts are more likely to lead to an esotropia, whereas acquired cataracts are associated more often with an exotropia.[76] Strabismus will generally follow deprivation amblyopia, as the visual system's stimulus to fuse is diminished as a result of the degraded retinal image in the amblyopic eye.

In strabismic cases of infantile aphakia, amblyopia therapy is initiated first to increase the quality of vision in the amblyopic eye, thus giving more opportunity for the retinal images to be fused so that there is some degree of binocularity. Generally, surgery for strabismus is delayed until the misalignment between the visual axes is stable and the visual acuity of the amblyopic eye has been optimised,[77] however, strabismic surgery may be performed earlier to facilitate contact lens wear. This is especially the case if the infant has a relatively large esotropia (Figure 5), such that the cornea is close to the nasal canthus. In this situation, the proximity of the cornea to the inner canthus means that the contact lens is more likely to be dislodged from the eye, particularly when the direction of gaze is nasal, and lens irritation may be greater due to the lens rubbing against the nasal canthus.

CONCLUSION

The management of infantile cataract has improved dramatically over recent years. Early cataract detection and prompt surgical management at the appropriate time, along with advances in contact lens technology, have greatly improved the visual prognosis for these children. While the contact lens management of infantile aphakia is often very challenging, it is extremely rewarding as the benefits to the patient are immeasurable. Regular review of these patients is essential. Contact lens complications must be avoided given the dependency on this modality for refractive correction. Therapy for amblyopia may be required and its importance should be stressed to the parents so that they and their child are more likely to comply with this treatment. The infant's ocular health should be monitored closely; in particular, IOP needs to be measured regularly given the increased risk of glaucoma.

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Corresponding author:
Richard Lindsay
Level 5, 376 Albert Street
East Melbourne VIC 3002
AUSTRALIA
E-mail: richard@lindsayassociates.com.au