PRESCRIBING FOR PAEDIATRIC APHAKIA

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AIMS

This module aims to outline prescribing refractive correction and low vision devices for paediatric aphakia via developing a:

- Knowledge base of this condition
- Systematic approach to selecting the most appropriate refractive correction
- Protocol for implementing low vision services and prescribing magnification devices

LEARNING OUTCOMES

After completing this module, students should be able to:

- Recognise the importance of early detection of congenital cataract
- Describe the advantages and disadvantages of spectacles and contact lenses for aphakia
- Be aware of the ambylopia and strabismus risks with this condition
- Match the visual need with the most appropriate low vision device available
- Successfully involve parents and teachers in the visual rehabilitation of the child

INTRODUCTION

This chapter includes a review of:

- Paediatric aphakia (and its implications on visual function)
- Prescribing for patients with paediatric aphakia
- Optical and non-optical low vision interventions

PAEDIATRIC APHAKIA

Paediatric cataract is one of the leading causes of childhood blindness and accounts for 40% of blindness as shown in different studies of blind schools. The incidence of infantile cataract is about 1.5-2.2 per 10,000 births and in 65% of the cases the cataracts are bilateral.

The majority of cases are idiopathic and this percentage is higher in unilateral cases vs. bilateral cases. Bilateral cases are more commonly associated with systemic disorders and hereditary factors. Recent advancements in equipment and surgical techniques resulting in improved prognosis for surgery has led to more children with cataract being identified and operated on. Surgical outcomes need to be complimented by improvement in the refractive and low vision management of paediatric aphakia to minimize the negative impact of visual impairment.

Some of the challenges in management of these cases include:

- Inadequate expertise in refraction of children
- Unavailability of high power plus lenses required for the correction of paediatric aphakia
- High cost and poor compliance in aphakic contact lenses for children
- Psycho-social barriers to spectacle wear in children

Management of paediatric aphakia comprises of early detection and surgical intervention, refraction and prescription of spectacles or contact lenses as appropriate. Low Vision rehabiliation with devices and services are needed in cases where vision is impaired even with best correction. Addressing strabismus and amblyopia is critical early on. Patching therapy, especially in unilateral congenital cataracts, needs to be done during infancy and toddlerhood to maximize outcomes.

PRESCRIBING FOR PAEDIATRIC APHAKIA

Removal of a cataract will result in a clear visual axis, however, unless the resulting refractive error is corrected, the visual outcome will remain poor due to refractive amblyopia. Infantile corneal and lenticular powers are higher than in normal adult eyes. Surgical removal of the lens in infants leaves the eye highly hyperopic, and it is not unusual to find 30 D or more of refractive power, which is approximately 2.5 times higher than in adult eyes. Significant surgically induced astigmatism may also be present. As the axial length of the eye increases, the refractive power gradually decreases, and by three years of age the average aphakic power stabilizes around 14 D. A further reduction of 2-3 D may occur by the age of ten years.

REFRACTION

Because of the rapid changes in the refractive status in infants, patients with paediatric aphakia need to be refracted at regular intervals, which may not be feasible if under general anesthesia. Therefore, this needs to be conducted in a normal clinical setting. However, the initial retinoscopy may be carried out immediately after cataract removal while the child is still under general anesthesia. This could provide the baseline refraction and any significant difference on subsequent retinoscopy needs to be carefully evaluated. Aphakic correction is necessary and should be provided as soon as possible following surgery, usually within a week or so.

While carrying out the retinoscopy, it is possible with very young children to have the child lying in the mother's lap with an assistant sitting opposite holding the legs of the child. (Duke elder, 1962). Another option could be to wrap the child tightly in a sheet or a blanket. In both cases, care needs to be taken to complete the retinoscopy as quickly as possible to reduce the stress to the child. A good starting point for retinoscopy is 16 D and power is increased in 2 D increments up to 20 D sphere. If "with" movement on the retinoscopic reflex is still observed with a 20 D lens in place, the power is increased in 1 D increments by adding trial lenses over the 20 D lens until neutralization is achieved.

It is best to keep the retinoscope streak aperture at the widest setting to evenly illuminate the whole pupil. This helps in avoiding any peripheral artefacts. Another potential error is that of "off-axis" retinoscopy which can result in apparent astigmatism of oblique incidence. Therefore, it is important for the examiner to maintain the same eye level as the child during retinoscopy. The distance at which the trial lens is held from the eye is important and should be noted as it may have a significant impact on the final prescription.

Example: While performing retinoscopy the +20 D lens was held 15 mm from the eye. The back vertex distance in the prescription spectacle is 10 mm. What should be the power of the corrective lens?

The effective power of the final prescription can be calculated using formula:

$F_e = F / 1- dF$, where F_e is effective power, d = difference between the distance of the trial lens and the prescription frame in m, F = power of the trial lens

$F_e = +20 / 1 – (0.005) (+20)$

$F_e = +20 / 1 – 0.1$

$F_e = +22.22$ DS

This demonstrates the importance of knowing what distance a trial lens is held while performing retinoscopy.

CORRECTION OPTIONS

Careful choice of the appropriate technique for correcting the aphakic error is essential for a good visual prognosis. Available options include correction with spectacles, contact lenses and intraocular lens implants. The final choice of the prescription, however, may depend upon many factors including age of patient, technical expertise, access to surgical center, availability of materials, compliance, cost and social factors.

INTRAOCULAR LENS IMPLANTS

Paediatric cataract surgery and intraocular lens implantation is constantly evolving. Surgical removal of the cataract as soon as possible is important to limit the effect of occlusion on amblyogenesis. Proper correction of surgical aphakia is important for visual development. One option of correction is an intraocular lens implant. Based on the work of the Infant Aphakia Treatment Study, it is widely accepted that implantation of the intraocular lens is not recommended until a child is over 24 months old. The study group found that implanting before this age had no advantage over use of contact lenses/glasses for visual outcomes and had increased risk of complications. Adverse effects that are possible include: aphakic glaucoma, retinal detachment, corectopia, severe fibrosis of capsule, later lens subluxation, corneal endothelial cell loss and uveitis.

Another reason to wait is the change of prescription needs that happens as the eye develops and grows. The corneal curvature and diameter as well as axial length of the eye undergo rapid change in the first years of life. The axial length of the eye has its most rapid growth during the first 18 months of age. It has also been found that this will increase in the presence of ocular surgery. An infant's eye typically has an axial length of 16-17mm compared to 24.5

mm in adults. In the first 6 months of life, the eye grows 0.62mm/month, from 6-18 months of age, the eye grows 0.19mm/month and after 18 months of age up until 18 years old, the eye grows 0.01mm/month. This change in axial length causes significant changes of the aphakic correction. This contributes to a different prescription that would be in the implant done very early in life. By waiting until the child is older, adverse effects may be avoided and a more lifelong accurate presription may be achieved.

It is important to remember, even if a child has an implant, glasses are still necessary. Glasses can provide any cylinder correction and the necessary near addition. The glasses can also be polycarbonate and tinted as necessary. As technology and lens design continues to develop, multifocal and accommodating intraocular lenses will become more of a possiblity for this population group.

CONTACT LENSES

Contact lenses are the preferred choice to correct paediatric aphakia where possible. The advantages of contact lenses are include ensuring a clear retinal image at all times, an unrestricted field of view and cosmetic acceptability. Contact lenses are the only practical option for correction of unilateral aphakia.

Contact lenses are an invasive method for aphakic correction and there is a high risk of corneal oedema, eye infections and possible trauma during insertion and removal. They require frequent replacement due to lens deposition, loss of the lenses or rapid changes in refractive status during the first 18 months of life. There is also the question of cost and affordability when choosing contact lenses.

CONTACT LENS TYPES AND FITTING

When deciding what type of lens an infant should have, many factors need to be considered. In infants and toddlers, keratometry is difficult to perform. Therefore, the initial choice of lens parameters may be based on expected values for the patient's age.

High plus power, often in excess of +30 D, is required and therefore the high lens central thickness causes a reduction in oxygen permeability. The corneas are often very steep and the palpebral fissure narrow - requiring steep base curves and lenses in smaller diameters. Due to these factors, extra care needs to be taken in fitting contact lenses in infants.

Four types of lenses can be used for the correction of paediatric aphakia:

1. Soft (silicone hydrogel or hydrogel) lenses
2. Rigid gas permeable lenses
3. Silicone elastomer lenses
4. Scleral lenses

Silicone hydrogel lenses are the lens of choice as their Dk (oxygen permeability) is high and can therefore be worn for an extended period of time. These lenses are relatively cheaper and the fitting techniques are not significantly different from those for fitting adults with aphakia. Ideally, these lenses are prescribed based on the keratometric reading, horizontal visible iris diameter (HVID) and the palpaberal fissure height.

As extended wear of the lenses is required, complications such as microbial keratitis, lens deposits, mucin balls, superior epithelial arcuate lesions and contact lens induced papillary conjunctivitis must be considered. Another drawback with soft lenses is the inability of these lenses to mask significant astigmatism. Glasses with the cylinder correction will have to be worn over the contact lens. Lens loss can also happen due to excessive eye rubbing.

Rigid gas permeable (RGP) lenses are another option for children with aphakia. Again, the fitting techniques of these lenses are similar to those with adults. Assessing the fit of these lenses is potentially easier as the flourescein pattern can be observed readily. Astigmatism can also be corrected with RGP lenses and complications observed

with soft lenses are rarely seen with hard lenses. RGP lenses are less comfortable on insertion and require a period of adaptation. It is easier to fit an infant than an older child with RGP lenses. Lens loss is less likely as eye rubbing causes a lot of discomfort.

Silicone elastomer lenses have higher oxygen permeability than silicone hydrogel lenses. They are also easier to handle than soft lenses as they tend to be less flexible. However, they are only available in limited parameters and are relatively expensive. Silicone elastomer lenses are also more susceptible to lens deposits and have to be coated with a hydrophilic coating as the material is hydrophobic. If the coating becomes damaged or is degraded, the lens becomes uncomfortable to wear. Fitting the lenses correctly is important but as they tend to flex on the cornea, a trial lens made of silicone elastomer should be used.

Scleral lenses manufactured from rigid gas permeable material offer another alternative for infantile aphakic fitting. These lenses offer better comfort than standard RGP lenses and are able to be modified as the eye grows. However, their use in infantile aphakia is not popular possibly due to perceived difficulty in performing the initial fitting and a higher cost.

Careful follow-up and serial over-refraction is important to make sure the child's eye is healthy and has an up-to-date refraction. Monthy follow-ups are recommended to carefully check the health of the cornea and the fit of the contact lenses. With all contact lens options, glasses will be needed for the near addition when children become more mobile, and for any residual cylinder that is not corrrected by the contact lenses.

SPECTACLES

Despite the many technical limitations, spectacles still remain the most commonly prescribed treatment for all forms of aphakia. They cannot harm the eye, and they are relatively inexpensive and simple to make. As the eye grows, its refractive state will change and glasses will need to be changed. It is easy to change the power of spectacles, either by changing the lenses, or providing a new pair. Unfortunately, there are many barriers to the provision of this simple solution. In many countries, spectacle frames in very small sizes, i.e. for infants and toddlers, are not available. There is a limited availability of high plus lenses required for the correction of aphakia and inadequate expertise for dispensing spectacles. The high plus lenses required for aphakic spectacle correction cause a functional ring scotoma, limit the field of view and cause spherical and chromatic aberrations. The weight of spectacle lenses and the additional issue of cosmetic appearance of the child also need to be considered. Spectacles with this type of prescription do cause magnification effect of up to about 25%. Glasses with high plus lenses will also gather light, so a hat with a brim or fit-over sunglasses are important.

Polycarbonate and CR39 (plastic or resin lenses) are the ideal lens materials for correction, typically in an aspheric or aspheric lenticular design with UV protection. UV protection is important as the natural lens has been removed from the eye and therefore cannot protect the inner ocular structures from UV harm. In developing countries these types of lenses may need to be imported, resulting in high cost. These lenses are also vulnerable to scratches and may need to be replaced frequently or have a hard coating for protection.

Glass lenses are relatively scratch resistant. They are thinner than plastic lenses for high prescriptions, however they are also considerably heavier and not safe for the paediatric population. These can be repolished and reground to incorporate small changes in power. However, glass lenses are not shatter-proof so glass is seldom used in the US and Australia for kids due to the weight as well as the small but possible risk of trauma due to lens breakage.

Keeping in mind the abovementioned solutions and their pros and cons, it is important to ensure their relevance to the circumstances. The major issues of cost and availability can be resolved by locally producing high plus lenses using glass or resin optical blanks and a lens generator. The procedure requires very basic training and is extremely affordable. As it is not always feasible to mass produce plastic frames locally, the most convenient solution is to maintain a reasonable inventory depending on the need.

As the distance of thebetween the lens and the eye has a significant impact on the effective power of the lens, it is important that the lenses sit as close to the eyes as possible without touching the eyelashes. An elastic band can be attached to both temples of the frame to ensure that the glasses do not slide down the nose and for better weight distribution. Another band can be attached to the nasal bridge and joined with the temple band to further reduce the

weight on the nose. A silicone or plastic pad, or surgical tape, can be applied to the nasal bridge to make the centration of the glasses better. The weight of the spectacles should be evenly distributed so that the glasses do not press heavily on the nose.

SPECTACLE LENS DESIGN

Ideally, the lenses used should have an aspherical design for better field of view, have a hard coating to reduce abrasions and provide UV protection. In case of limited availability or high cost of aspherical lenses, the lens design should be plano-convex, and the best form design should be used in higher powers (20 D and above). Biconvex lenses should be avoided if possible due to their poor optics.

What power to prescribe?

In infants, the aphakic correction prescribed is for close viewing distance. An infant's visual world is mostly restricted to their parent or caregiver's face and they are also not mobile at this time. Taking these factors into consideration, it is standard practice to overplus the infant by +3.00 D up to the age of one year. As the infant/toddler starts to visually explore their world and becomes mobile, the extra plus should be reduced to +1.50 to +2.00 D. Ideally, children over 2 and a half years of age should be prescribed bifocals with an addition of +2.50 to +3.00 D. Higher adds can also be precribed to help children with visual impairment see smaller print. Please refer to the next section (Low Vision Considerations) for more information.

LOW VISION CONSIDERATIONS

MAGNIFICATION DEVICES

Early detection and treatment is critical to maximize visual outcomes. Better visual outcomes require less magnification devices and services for the child in their lifetime. Prior to prescribing any low vision magnification devices it is always important to make sure the proper refractive correction is in place. This allows the low vision device to provide the prescribed magnification and not have to use some of its power to compensate for uncorrected refractive error.

There are several options of magnification for near: spectacle addition, hand-held or stand magnifiers and electronic magnification. The spectacle addition is important in this population as these infants have no ability to focus. The near addition may have to be prescribed to allow more functionality, rather than to allow for print reading. A functional near addition allows a child to see things like food on their plate, toothpaste on their toothbrush or playing with toys. This power is typically set at their arm's length, resulting in an addition between +2.50 to +4.00 D. This addition may not enable them to see smaller print depending on their level of visual impairment; in these cases magnifiers will also be used. As a child gets older, the prescription of the spectacle addition can be done to meet the child's requirements. If a child has contact lenses or an intraocular lens, they can use either a bifocal or single vision near glasses. They can have a power that is more functional in nature for activities of daily living and a second higher powered pair to see smaller print sizes/fine details. Children do very well with stronger additions, it is natural for them to pull things closer to see them better. In doing so, they also benefit from relative distance magnification, and the job of the optometrist is to ensure that the correct bifocalnear addition power is prescribed to provide a clear retinal image. For example, if a child is holding things at 10 cm, a +10.00 D addition is appropriate to provide a clear in-focus retinal image. Other benefits of bifocal or reading glasses include a wide field of view due to its proximity to the eye and leaving the hand-free.

When using magnifiers, remember that parallel light leaves a hand-held magnifier and divergent light leaves a stand magnifier. Therefore when a child uses a hand-held magnifier they should use the distance portion of their glasses or contact lenses. If they use their near addition with the hand-held magnifier it will reduce the magnification power. For stand magnifiers, it is important to use their near addition. The addition will neutralize the divergent light leaving the magnifier and put the magnified image clearly in-focus on the retina, allowing the child to benefit from the full power of the magnifier. Younger children tend to be more steady with stand magnifiers over hand-held magnifiers. The stand

magnifier can rest on the page and slide back and forth, whereas the hand-held magnifier one has to be held parallel and at a certain distance from the page to provide a clear magnified image. Stand magnfiers may also lend themselves better to more extended reading tasks. Both hand-held and stand magnifiers are good options and illuminated models should always be considered.

When using electronic magnification or adaptive techology, it is important to prescribe reading glasses or an addition that corresponds to the eye-to-screen distance. For example, if a child had a portable hand-held electronic magnifier and uses it at 33 centimeters, a +3.00 D addition should be used to keep the screen in focus for the child. The device will provide the magnification, but the addition will provide a clear image. (The addition also contributes slightly to the overall magnification in this system.) However, if an addition is not prescribed, the child will have to increase the magnification on the screen of the electronic device to compensate for the image being a few diopters out of focus. The same applies to desktop electronic magnifiers (CCTVs) or computers with software enlargement programs. With these devices it is important to consider a pair of full-field reading glasses dedicated to using these devices. These near glasses with only have a +2 to +4 D addition, but will bring the screen into focus and limit the enlargement needed. The benefit of this is using less magnification which allows for more information to be displayed on the screen, leading to faster reading rates. There will also be less eye strain and neck strain as the patient does not have to tilt their head backwards to view through the near segments of bifocal glasses.

There are a few things to consider when prescribing and training with telescopes. Due to the high amount of plus in aphakic prescriptions, it is best to use a telescope over the patient's correction. If the patient has contact lenses or IOLs, this is easy, but glasses pose more of a challenge. If the telescope is used with glasses, it is important to teach the child to fold the rubber eye cup of the telescope down, this allows the back of the telescope to be closer to the eye. By doing this, the exit pupil of the telescope will be closer to the entrance pupil of the eye and the child will have a better field of view.

If a child takes their aphakic glasses off to use a telescope, clear image will have to be obtained by focusing the telescope, a child will increase or decrease the power of the telescope depending on the telescope design. As a review, the magnification of the telescope is calculated by dividing the focal length of the telescope by the focal length of the eyepiece. Keplerian telescopes have plus objective and eyepiece lenses whereas Galilean telescopes have plus objective and minus eyepiece lenses. If a child with aphakia is uncorrected and focuses the telescope to see clearly, they will be either adding to or taking away from the value of the eyepiece lens. For Keplerian telescopes, the uncorrected aphake will 'steal' or 'borrow' plus power from the eyepiece lens, resulting in a lower telescope power. For Galilean telescopes, the uncorrected aphake will add to or increase the power of the eyepiece lens resulting in a higher telescope power. This needs to be considered when prescribing telescopes for this population of patients.

Table 1: Example of using Keplerian and Galilean telescopes with and without a +20 D aphakic correction

	Objective lens power	Eyepiece lens power	Magnification of telescope
Keplerian telescope	+20	+60	60/20=3x
used with glasses	+20	+60	60/20=3x
used without glasses	+20	+40	40/20=2x
Galilean telescope	+20	+60	60/20=3x
used with glasses	+20	+60	60/20=3x
used without glasses	+20	+80	80/20=4x

Finally, hand-held telescopes are recommended for younger children. Mounted telescopes are a good option when they get older as mounted systems are more expensive and require more care in handling.

For more information on this topic, see chapter 5 in the *Low Vision 1* module, O*ptical low vision devices*.

NON-OPTICAL AIDS

In addition to magnification devices, non -optical devices are important and need to be prescribed, depending. These aids depend on the age of the infant/toddler/child. Glare control is very important and can be achieved with tinted

lenses, fit-over sunglasses and/or a hat with brim. All children with aphakia may be photophobic, but there are several situations where the photophobia will be more extreme. These include cases where there is a surgically enlarged pupil, use of aphakic glasses (light gathering properties of plus lenses) and cases of contact lens use where the anterior segment of the eye may be slightly uncomfortable or irriated.

Other non-optical devices that are important include: reading stands, high contrast printed material, bold lined paper with a thicker writing device (black crayon better than pencil) and audio products (audio books).

ADDITIONAL SERVICES

Depending on the age of the infant/toddler/child and the level of the vision loss, other service providers and interventions are important to maximise visual and developmental outcomes.

This population can benefit from:

- Early intervention: services and therapists during infancy and toddlerhood. These include professionals that can help families understand the needs of a child with visual impairment and initate therapies and/or environmental modifications to help with the child's overall development.
- Teacher of the visually impaired: once a child enters the school system, a teacher of the visually impaired can help the child with using prescribed low vision devices, test taking accommodations (additional time, large print), advocate for the child and family, make sure the child is seated in the front of the classroom. The teacher can also provide things such as non-optical devices (reading stand, etc.) and audio products.
- Orientation and Mobility specialist: to train child to safely navigate their environment. Can introduce mobility cane, GPS system, etc.
- Adaptive technology specialist: to evaluate the needs of the child, recommend and train with adaptive technology. This can include computers with software enlargment and speech output.
- Braille Instructor: teaching Braille to children with no functional vision. Exposing child to technology such as a refreshable Braille display.

For more information on this topic, see chapter 6 in the *Low Vision 1* module, *Non-optical low vision devices*.

OTHER CONSIDERATIONS

Amblyopia and strabismus need to be screened for and managed. Typically, esotropia occurs in this patient population and surgical correction is often necessary. Sometimes the surgey needs to be done very quickly if contact lenses are going to be fitted. In a large angle esotropia, a contact lens will often dislodge in the eye that is esotropic.

Occlusional amblyopia can be unilateral or bilateral. In bilateral cases, management typically consists of best correction and vision stimulation. Patching may be required if there is a difference difference between the two eyes and/or strabismus.In cases of unilateral cataract, more patching will have to be done. If a child has unilateral cataract with surgical removal, that eye is at high risk for amblyopia. If glasses correct for the aphakia, the brain will suppress the blurrier magnified image. However, patching needs to be done with the correction in place. It is better to correct with contact lenses or intraocular implants in unilateral cases. When this is done, differences in image size due to anisometropia are eliminated and patching will be more successful.

For more details, please refer to the *Binocular Vision and Therapy 2* module.

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