The ROYAL COLLEGE of OPHTHALMOLOGISTS

focus

An occasional update commissioned by the College.
The views expressed are those of the authors.

Paediatric cataracts

Meena Arunakirinathan, Fellow, Strabismus & Paediatric Ophthalmology, Moorfields Eye Hospital.

Lucy Barker, Consultant Ophthalmologist, Moorfields Eye Hospital.

Paediatric cataracts are relatively uncommon but can cause significant visual impairment and amblyopia. Prompt diagnosis, referral and treatment can lessen the burden of stimulation deprivation amblyopia, refractive error, and strabismus. In some cases, diagnosis can be lifesaving.

The challenges in managing this group of patients require a well-established pathway navigating a multi-disciplinary team of ophthalmic surgeons, orthoptists, optometrists, contact lens (CL) practitioners, paediatricians, paediatric anaesthetists, geneticists, genetic counsellors, and family support teams (**Figure 1**). As the child grows ongoing communication with their school and local paediatric visual support services facilitate their evolving visual rehabilitation as their visual demands change.

Globally the prevalence of paediatric cataracts is estimated to be between 2.2/10,000 and 13.6/10,000.[1] The incidence in the UK has been shown to be 2.49/10,000 by the age of 1 year, increasing to 3.46/10,000 by age 15.[2] Approximately two-thirds of children with congenital cataract have bilateral cataracts and one-third have unilateral cataract.

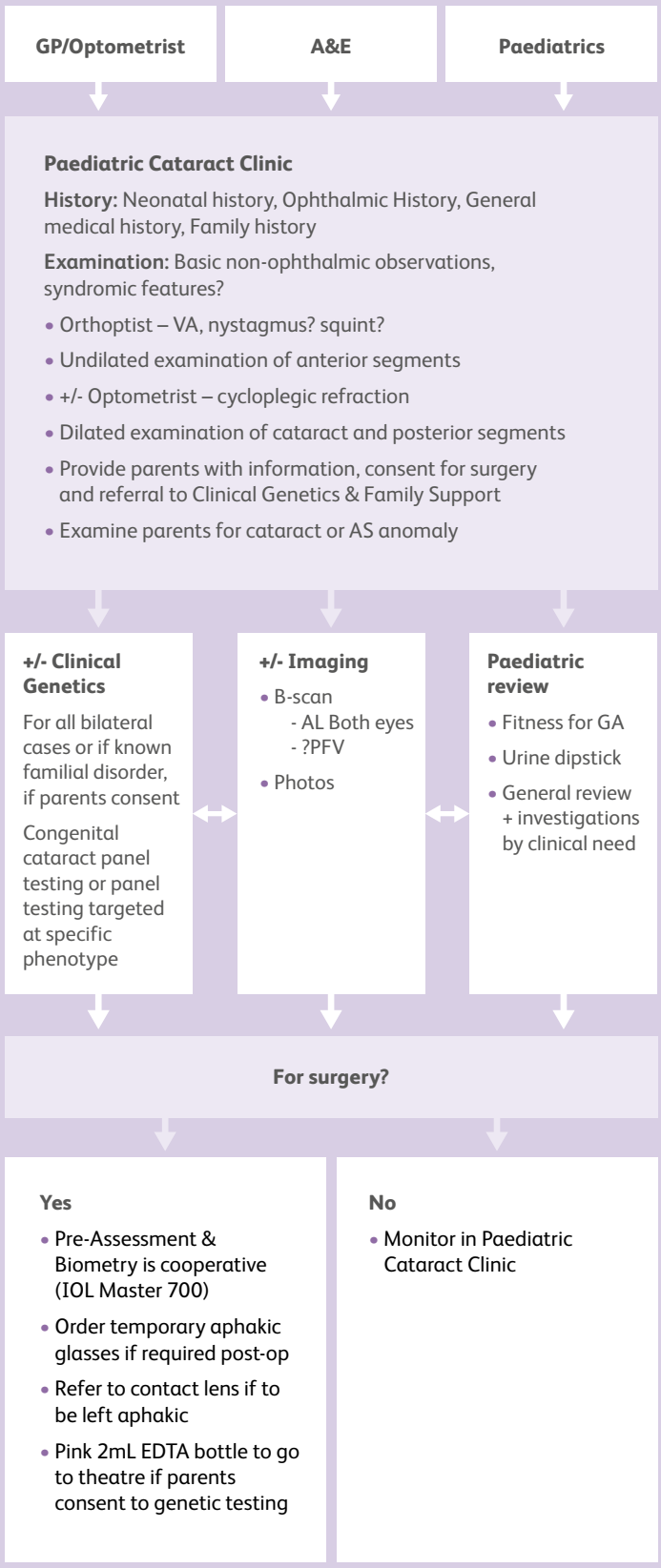
The UK National Screening Committee recommends all eligible neonates undergo the new-born and infant physical examination, with ocular examination within 72 hours of birth and at 6–8 weeks of age. Any children screening positive should then be streamlined to ophthalmology review by 2 weeks postnatally. This ensures prompt referral for treatment, thereby improving the chances of good visual outcomes.

Aetiology

Paediatric cataracts are highly heterogenous and a paediatric review is particularly crucial for conditions with systemic associations. Important environmental factors to consider include congenital infections such as toxoplasma, syphilis, varicella zoster, rubella, cytomegalovirus, and herpes simplex virus I and II (TORCH). Trauma and iatrogenic causes such as medications and radiation exposure are also relevant but rare in this age group. Inherited congenital cataract may occur in isolation (70%), with other ocular abnormalities (complex) (15%) or form part of a syndrome (15%).[3] (**Table 1**).

Traditionally comprehensive work-up for congenital cataract include biochemical, genetic, clinical, and imaging modalities. However, these require frequent clinic visits at great burden to health services and significant engagement from patients' parents.[2, 3] Musleh et al demonstrated, in a work-up of 27 consecutive bilateral cases, that only 3.4% diagnostic yield was found with no positive TORCH results and no unexpected

Figure 1 - Cataract Pathway



medical findings.[4] Major advancements in genetic testing now include a targeted panel of cataract-related genes using next-generation sequencing technology with over 90% coverage.[3, 5] The addition of whole genome sequencing is thought to increase diagnostic pick-up by a further 40%.[3]

Timing of surgery

The early neonatal period, from birth until any visual deprivation begins to interfere with the maturation of the visual system, is deemed the latent period. During this time the immature visual system relies on sub-cortical pathways.[6] This period appears to be approximately 6-8 weeks for babies with unilateral visual deprivation. Unilateral cataract surgery performed during this critical window has been demonstrated to produce good outcomes. However, operating after this time, even with good surgical results and excellent compliance, can result in a much poorer outcome.[7] An equivalent latent period for bilateral cataract surgery is within 8-12 weeks.[8] During the sensitive period of 7-8 years, the developing visual pathway can be subject to amblyogenic effect and treatment. Therefore, parents are counselled about the importance of long-term post-operative visual rehabilitation with aggressive patching. Occlusion therapy commences with 1 hour per day for each month of life until 6 months old and thereafter 6 hours per day of patching is recommended until age 7-8 years or until the post-operative visual acuity stabilises. Development of binocularity and fixation stability also have a 'latent period'; therefore, the timing of surgery and length of visual deprivation are important factors in the development of nystagmus. Even with early surgery some children go on to have nystagmus which in most cases is manifest latent nystagmus.[9]

A Summary of the Surgical Approach to Paediatric Cataracts

Infant eyes pose considerable challenges beyond just being smaller in axial length. They often have increased corneal thickness and a shallower anterior chamber with a highly elastic capsule. They may be significantly microphthalmic (axial length < 16mm) with microcoria exacerbated by pupillary membrane. The iris is commonly hypoplastic and more vascular. The sclera lacks rigidity and trabecular meshwork is more rudimentary. Examination under anaesthesia of both eyes with measurement of corneal diameter, pachymetry and keratometry should be undertaken before cataract surgery in infants. For older children planned for primary intraocular lens implantation, a scan biometry may be required under anaesthesia if non-contact biometry is not possible pre-operatively.

Lensectomy requires a surgical technique that can adapt to the age of the child and co-existing ocular pathology such as persistent foetal vasculature (PFV) and/or anterior segment dysgenesis. A superior approach with temporal and nasal 20G or 23G incisions can enable a more stable AC which is prone to collapsing due to the lack of scleral rigidity. Adjuncts such as Trypan blue to stain and stiffen the capsule and viscoelastic with greater cohesive properties such as Healon 5 or Healon GV are preferred. Pupillary dilatation can be enhanced with diluted intracameral phenylephrine, but this requires communication with the anaesthetist to monitor for cardio-respiratory changes. Further iris manipulation can be facilitated with hooks, blunt and visco-synechiolysis and surgical pupilloplasty. Anterior capsulotomy can be performed either manually with forceps and capsule scissors or by automated vitrectorrhexis and is followed by lens aspiration, posterior capsulotomy and anterior vitrectomy. Posterior capsulorrhexis either manually or with a vitrector is recommended in young children (up to age of 5 years) due to the risk of re-opacification and subsequent need for Nd:YAG laser requiring further general anaesthesia. Special consideration dependent on the intra-operative findings can determine whether to implant an IOL or leave aphakic or perform a surgical iridotomy. Optic capture of the IOL is preferred for stability and reduced risk of posterior capsular opacification (PCO). Finally, all wounds are sutures ideally with an absorbable suture (10/0 vicryl) to prevent leakage.

Intraocular lenses – to implant or not to implant?

The IOLunder2 and Infant Aphakia Treatment Studies (IATS) are two large cohort studies with 5 year follow-up conducted in the UK and USA, respectively, which provide evidence that post-operative vision following concurrent intraocular lens (IOL) implantation is similar when compared to aphakia plus CLs.[10, 11] Furthermore, IOL implantation does not confer protection against post-operative glaucoma, and visual axis re-opacification (VAO) occurs more frequently and earlier with use of IOLs in children under 2 years versus aphakia.[10, 12] Therefore the evidence suggests that implantation of IOLs should be reserved for children over 2 years and when used, aim to leave them hypermetropic.

Complications of Surgery

The post-operative inflammatory response in children is more exuberant. Topical corticosteroids are used to reduce pain and posterior synechiae or pupillary membrane formation which can lead to IOL decentration and pupil-block glaucoma. The detection of glaucoma requires long-term surveillance, although the IOLunder2 study showed median time to development was 6 weeks following unilateral lensectomy and 4.5 months in bilateral lensectomy. Though the pathophysiology is unclear, it is evident that each additional week surgery is delayed slightly reduces the post-operative glaucoma risk but slightly increases the risk of deprivational amblyopia.[10, 13] Development of PCO or recurrence of VAO from capsular phimosis, Soemmering ring or vitreous and pupillary membranes is amblyogenic and therefore the most common reason for re-operation. Retinal detachment following lensectomy is thankfully rare and the risk is not increased by anterior vitrectomy or posterior capsulotomy. However, incidence may be increased by the presence of co-existing PFV.[9]

Options for Refractive Correction

Following lensectomy, children can have a refractive error up to +30 dioptres if left aphakic and significantly less if pseudophakic. Therein begins intensive efforts for visual rehabilitation. Options for refractive correction include aphakic (high power) contact lenses with bifocal glasses, aphakic (high power) glasses, and intraocular lenses with bifocal glasses. The choice of IOL power is made by considering how the eye will focus clearly when fully-grown because a marked myopic shift is expected. Therefore, an individual approach to each child should be made, dependent on their age, when choosing

an IOL power to produce undercorrection. Younger children, irrespective of whether they are pseudophakic or aphakic, will require significant hypermetropic correction as leaving them with some near vision is important to for visual development. High plus spectacles can induce optical aberrations such as restricted visual field, pincushion distortion, magnification and in the unilateral cases, aniseikonia between the phakic and operated eye making glasses difficult to tolerate. The latter can be overcome with the use of silicon hydrogel daily CLs which are often very well tolerated but require a high degree of engagement from parents. As children become more mobile and learn to read the use of bifocals and progressive lenses for older children over their aphakic contact lenses or with pseudophakia are highly effective.

Subsequent amblyopia treatment is key to overcoming the high anisometropia also complicated by loss of accommodation. Regardless of this intensive rehabilitation, strabismus may still occur.[13] IATS provided evidence that occlusion therapy of at least 4 hours per day resulted in better visual outcomes than occlusion of less than 2 hours. Close monitoring is required to avoid disruption of developing binocular function, reverse amblyopia and to promote compliance. Children with surgery for bilateral cataracts will require occlusion if visual acuity difference or fixation preference manifests.

As in adults, implantation of a secondary IOL may be indicated if there is intolerance to CLs or glasses, CL-related infections or patient and parent aversion to their use. Careful selection of cases for secondary IOL implant is required and ascertaining the extent of capsular and iris support and structural abnormalities. Contraindications are relative microphthalmia, poorly controlled uveitis and glaucoma and a poor visual prognosis. Though no IOL formula specifically calculated for paediatric patients exists, SRK/T is the formula of choice but prediction errors can be highly variable in paediatric populations, especially in infants under 36 months.[9] Emerging evidence for Barrett Universal II shows comparable accuracy than previous generation formulae in children with posterior chamber IOL implantation.[14, 15]

Options for IOL implantation include in-the-bag or sulcus fixation of a foldable 3-piece or PMMA IOL, ideally with optic capture. Pre- or retro-pupillary iris claw IOL, sutured IOL and intrascleral haptic fixation provide further options for secondary pseudophakia depending on the degree of structural support from the capsule and iris. Long-term endothelial cell loss associated with anterior chamber IOLs remain a concern therefore these are not recommended. Professor Tassignon developed the 'bag-in-the-lens' technique for primary and secondary IOL implantation. Two identical anterior and poster curvilinear capsulorrhexes are created and both margins are inserted in a flange of the IOL which result in very low VAO rates.[16]

Summary

- Paediatric cataracts are an important cause of lifelong visual impairment requiring prompt diagnosis and surgical treatment within the critical windows
- Next-generation sequencing technology can now yield more precise molecular diagnosis
- Timing of surgery is key to optimising visual outcomes by balancing the risk of amblyopia (if surgery done too late) and risk of glaucoma (if surgery done too early)
- Following an evidence-based approach to refine surgical techniques can improve visual outcomes and lower the risk of complications associated with this
- Ultimately, a specialist network of expert optometrists, orthoptists, geneticists, and clinicians following these patients closely throughout their childhood is necessary to support the child and their family through their long journey of visual rehabilitation

Table 1. Examples of associated risk factors for development of Paediatric Cataracts

Cause	Example
Isolated	No other ocular or systemic disease
Associated Ocular disease	PFV Microphthalmia Aniridia Retinitis Pigmentosa ROP Endophthalmitis
Inherited	Autosomal Dominant Autosomal Recessive X-linked Recessive
Inherited with systemic abnormalities	Marfans Weill-Marchesani Stickler syndrome Bardet-Biedl syndrome Alport syndrome Lowe's syndrome Hallermann-Streiff-François syndrome Cockayne syndrome Incontinentia pigmentii Myotonic dystrophy Dermatological and Atopic dermatitis
Chromosomal	Trisomy 21 Trisomy 18 Trisomy 13 Cri du chat syndrome
Infection	Toxoplasmosis Syphilis Rubella Herpes group (CMV, HSV, VZV)
Metabolic	Galactosaemia Galactokinase deficiency Diabetes Mellitus Hypoglycaemia Hypocalcaemia
Other	Trauma Laser photocoagulation Radiation JIA/Uveitis Drug-induced e.g. Corticosteroids, Chlorpromazine

Andrew Tatham
Editor, Focus

Acknowledgements: Professor Christopher Lloyd & Dr Joyce Chan for images