Chapter 2
Nonsurgical Management of Infantile and Juvenile Cataract

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Visually significant or total cataracts in infants and children, both unilateral and bilateral, typically involve straight-forward decision-making in terms of moving expeditiously to clearing the visual axis soon after discovery. However, partial opacities present more difficult management decisions, especially in young children, at the time of presentation. In addition, that decision-making process continues well into the future based on patient factors, cataract stability/progression, and visual needs. The decision to treat or monitor will also be influenced by laterality, type of lens opacity, age of patient, developmental outlook, visual prognosis, and family history. Bilateral partial cataracts allow more time to monitor the child's course than unilateral partial cataracts. Early treatment may prevent amblyopia but raises the risk of glaucoma [1, 2], retinal detachment, the need for additional surgery, and other less common complications. The outcomes for monitoring (and not operating) are not reported and likely quite heterogenous. The clinical tools to assist with informing those decisions are limited by age, measurement variability, and cooperation.

Crucial to decision-making are the ability of the clinician to examine the child, to be able to see the retina in spite of the partial cataract, measure the refractive error, and vision when it can be measured. The surgeon is trying to balance amblyopia development and visual impairment level with the issues associated with surgery, including the need for one or more future surgeries, as well as other complications.

Tools to Assess

The ophthalmologist needs access to a retinoscope, direct ophthalmoscope, indirect ophthalmoscope, and handheld slit lamp. All of the instruments are used to

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develop an overall judgment of the impact of the cataract on vision, hopefully as part of in office testing. On occasion, an exam under anesthesia may be needed for some infants and children, but contemporary pediatric care has moved to perform these exams as infrequently as possible. There has been recent concern about the impact of multiple anesthesia events on brain development, so the ophthalmologist needs to balance the needs of the eye exam with developmental concerns and neurological toxicity [3]. This area of concern needs to be monitored by the surgeon and discussed with parents if necessary.

Using each instrument, the surgeon assesses the quality of the red reflex and how much of the pupillary aperture is occluded. The most useful for monitoring the visual impact of the cataract is probably the retinoscope. Although a 3 mm size is often stated as a maximum guideline, it remains a consensus recommendation and the decision customized to the patient. The assessment of size is best done with a direct ophthalmoscope and distortion of the retinal reflex with the retinoscope. This size threshold for significance also assumes there is no disruption of retinoscopy beyond the opacity. If the retinoscopy uncovers significant distortions in the red reflex beyond the central opacity, the size including the area of distortion needs to be considered as the true size of the cataract. Try to be certain that the retinoscopic reflex distortion is from the lens and not the cornea.

The lens opacity for some children may be eccentric, often in the area of prior attachment of the hyaloid vessels (slightly nasal and inferior to the visual axis), and thus less damaging to the retinoscopy. This may require review at a subsequent visit to confirm if the central axis is indeed clear of the lens abnormality or not. The timing of that visit will depend on the overall level of concern. The use of a handlight to examine the eye can be misleading in some cases. Because the pupil will be constricted from the bright light, it might seem that the child cannot see around the opacity. For this evaluation, often the retinoscope on dim can be very useful.

The slit lamp is used primarily to determine the depth of the cataract. It too will constrict the undilated pupil. Posterior and posterior subcapsular lesions are generally more of a problem for vision development than anterior capsular and anterior polar lesions. In many cases, that is because they do not distort the retinoscopy and thus the image viewed as much as posterior lesions.

Visual acuity should be measured with optotypes whenever possible to guide therapy. Lea symbol charts and HOTV single surrounded optotypes can be introduced in the second or third year of life. Preferential looking techniques or fixation preference assessments are done in children unable to perform optotypes. These are sensitive only to large differences in acuity. Whenever possible, a child with a lens opacity should have their visual acuity measured a second time if a treatment decision is anticipated, to be certain they understood the test on the first occasion, performed their best, and it was not simply a poor test. This is usually simple to accomplish for those children with unilateral partial cataract as they will attempt a course of amblyopia therapy (e.g., occlusion) if there is an open optical pathway before moving on to surgery.

Modifying Factors

Bilateral cataracts are much less amblyogenic than unilateral cataract and, generally, the surgeon can wait longer to pursue surgery. Bilateral cataracts are not always symmetric, so the surgeon should carefully consider if both eyes are affected sufficiently and do both require surgery. Guidelines for visual acuity thresholds should be considered consensus-based and subject to individualized decision-making. For unilateral surgery, the outcomes for younger children are much poorer than for older children and poorer for those with bilateral cataracts. Thus, for younger children with measurable accurate visual acuity of better than 20/80 with a unilateral cataract, it is probably best to monitor. For older children, the threshold to perform surgery may be 20/50 or worse or if the opacity is judged to cause at least three lines of vision impairment. For bilateral cataracts, vision of 20/60 or better is typically monitored until the children are older or begin to have functional deficits. As children age, needs will likely become more visually demanding, and surgery may become necessary. This is a very common situation with central and lamellar cataracts.

It is much harder to decide on treatment timing for children unable to perform optotype acuities. For those children, the decision is largely driven by how poor vision appears to be while wearing an occlusive patch over an unaffected eye and clinical judgment about the degree of severity of the opacity.

Anterior polar cataracts are typically much less of a problem than central or posterior polar cataracts. Lamellar cataracts are also usually associated with much better vision and frequently bilateral, allowing surgery to be delayed until significant decline in ability to function. Lamellar cataracts look far worse than their impact on visual acuity.

Family history is increasingly important. In the developed world, cataracts are increasingly familial or inherited (with reduction in the number related to infectious disease). Thus, the family will often have specific concerns about parental and sibling experiences, which will affect the discussion about when to do surgery.

Expected development of the child also plays a role. If there is substantial developmental delay, yet the cataract is significant, then surgery should be done sooner just as in children with no developmental delay. However, if the cataract is mild and the child is able to achieve daily goals, then a delay in treatment would be reasonable.

Management of Cataract

If the patient can be adequately refracted and the decision is to initially manage without surgery, correct the refractive error and have the child get used to wearing glasses full time. This is often the most important part of the treatment. The child should return in a few weeks for reassessment of the vision. If a deficit remains and there is a reasonable visual axis, then an attempt with occlusion therapy for as many hours per day is possible for the parents. Half of waking hours is a useful guideline

to maximize the chance for improvement. In some cases, atropine penalization of the fellow eye is offered with unilateral cataract, but there are no data to affirm its efficacy.

Pupillary dilation may also be used for very central opacities with good peripheral clarity and normal retinoscopy. Topical phenylephrine 2.5% eye drops are often a first choice with partial opacities, but the surgeon needs to verify the adequacy of the dilation to accomplish the goal of providing an optical pathway. In some cases, topical atropine 1.0% to the affected eye can be used for its more potent mydriasis, but the patient will then need to wear appropriate glasses to correct for the cycloplegia. For a child under 30 months, single vision at an intermediate distance works well along with occlusion therapy, whereas for older children bifocals would be needed.

For some cataracts, a delay in surgery will be for a lifetime because of relative advantages and disadvantages of surgery for that child. However, in many instances, surgery is simply deferred until the child is older. The advantages of older age will include a better measured IOL power, easier posterior capsule management, possibly less risk for glaucoma, and perhaps better technology will be available for surgery, the implant, and subsequent care.

Case 1

A 32-month-old boy was found by his pediatrician to have central crystalline lens opacities. His vision was 20/40 in each eye with symbols with +2.50 D refractive error and no heterotropia. No prescription and 6-month follow-up exams planned; this continued for the next 13 years. Over that time, the nuclear cataracts remained unchanged, retinoscopy was crisp, and the hyperopia decreased. At the most recent exam, he noted classroom seating accommodation in the front half of the room worked, although there was some glare when he was outdoors. Visual acuity was 20/40–2 in each eye, normal stereo, and low hyperopic refractive error. There was some parental concern about driving and glare, but the decision to follow was reaffirmed (Fig. 2.1).

Comment In this case, the child's ability to perform a visual acuity at an early age along with the crisp and symmetrical retinoscopy helped drive the decision to delay surgery. In addition, classroom accommodations and his excellent grades in school may have allayed any parental concerns. The issue of driving is important as this issue, especially with nuclear and lamellar cataracts, serves as the impetus for teenagers to undergo surgery.

Case 2

A 33-month-old girl was seen with a history of bilateral asymmetric cataracts. She had a lensectomy performed in the left eye as an infant, which was subsequently treated with an extended wear contact lens. The right eye's mild anterior polar

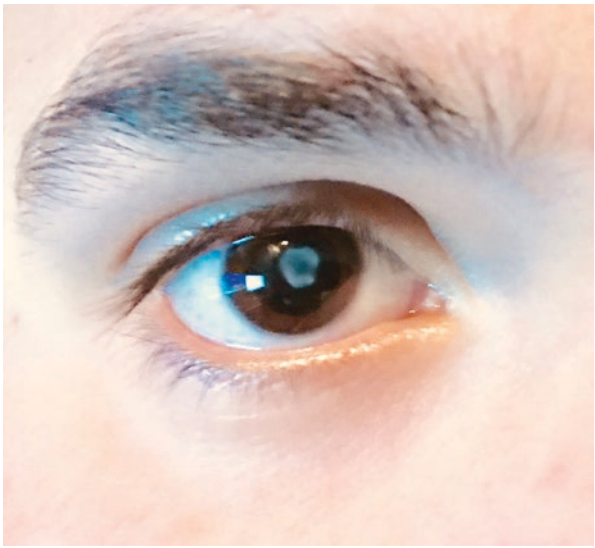

Fig. 2.1 Prominent central nuclear cataract. Central opacity was present and unchanged for many years. The retinoscopy through the lens around the opacity remains normal, and the retinoscopy reflex was crisp

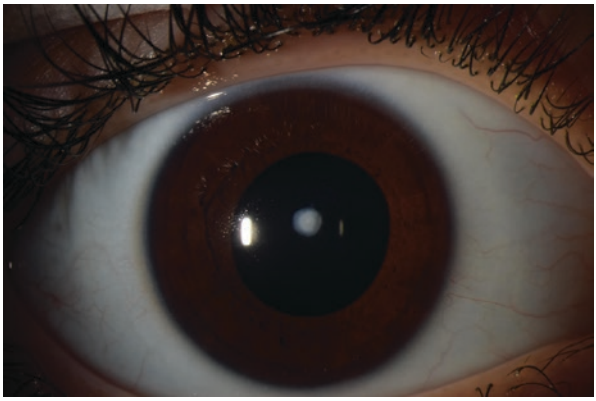

Fig. 2.2 Anterior polar cataract about 1.5 mm in diameter with good vision was not disruptive of the visual axis or the retinoscopy

cataract was monitored (Fig. 2.2). She had a secondary intraocular lens placed at 16 months of age in the left eye with a resultant refractive error of +1.50 D. Amblyopia therapy was performed by underplussing the more hyperopic, phakic right eye (+4.50 D) along with administration of atropine penalization. At 33 months of age, she was 20/25 in the right and 20/20 in the left. The cataract in the right eye was a 2 mm anterior polar opacity with no disruption of the retinoscopy. The optical axis of the left eye was clear. There was no strabismus with positive response to the Fly stereo test. She was to return in 4 months and would have follow-up visits about twice per year.

At 10 years of age, visual acuity was 20/25 OD and 20/20 OS with normal stereo and no strabismus. The refractive error remained anisometropic with +4.50 OD and − 0.50 OS (pseudophakic), but the intraocular pressure was in the mid-20s to upper 20s in the left eye, while the phakic eye was normal.

Comment It is common for children who have frequent eyecare visits to be able to perform optotype visual acuity at a young age easing some decision-making. The localization of the polar cataracts to anterior or posterior lens surface can be difficult with the retinoscope and direct ophthalmoscope in children, but the absence of retinoscopy distortion suggests anterior capsule. This unoperated anterior polar cataract has had a very good outcome, but that can be a tough decision early on in a patient with bilateral cataracts in whom one eye was operated. Early surgery is known to be associated with sustained increased intraocular pressure in the mid-20s and is of increasing concern in this child. Lastly, bilateral cataracts do not always require surgery in both eyes. That decision has to be made considering the nature of the cataract in each eye and the impact of each on vision.

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