Chapter 6
Preoperative Examination

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Proper preoperative counseling, planning, and management strategies require attention to a multitude of factors surrounding a pediatric patient presenting with a cataract. A thoughtful approach to these patients considers all of the possible medical and surgical needs of the patient. The timing of surgery, discussion of the surgical plan, preoperative measurements with biometry, and addressing postoperative expectations (aphakic contact lens versus spectacles) are important factors to consider. It all begins with the preoperative visit.

History

First and foremost, obtaining a thorough patient history is essential to understanding possible etiologies and the visual impact of the cataract. Pertinent information from the history includes prenatal and perinatal history, ethnicity, gender, history of maternal infection (TORCH infections), history of ocular trauma, and family history of childhood cataracts. History of juvenile idiopathic arthritis (JIA) can indicate that a cataract is secondary to uveitis and/or long-term corticosteroid use. In addition, the age of onset of visual symptoms and change from previous eye examinations should be ascertained. It is important to question parents about the child's perceived vision, as this can provide clues about the duration of the cataract and its visual significance. Nearly one-third of cataracts are inherited, so an examination of the eyes of the parents may be helpful and spare unnecessary testing associated with a workup [1].

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Examination

Immediately upon entering the room with a pediatric patient, one can begin gathering information about visual behavior and ocular alignment. These observations are easily made without even coming close to the patient.

One of the most useful skills includes the use of the direct ophthalmoscope to visualize the red reflex. It is a fairly quick test that does not require much cooperation from the patient but can provide a lot of information regarding the presence of a media opacity. The Bruckner test may be employed to compare one reflex to the other, allowing the provider to estimate the amount of visual significance produced by the cataract. Determining the location of the cataract and whether or not it is directly in the visual axis can also be noted while observing the red reflex.

Although red reflex testing can be a quick clue to the presence of a lenticular opacity, further testing is necessary to determine the visual significance of a cataract. Obtaining an accurate assessment of visual function in a young child can present a major challenge. Typically, the older and more cooperative the patient, the more likely the physician will succeed at obtaining an accurate visual acuity. Even though it may be difficult, it is not impossible to achieve a good examination in a young child or infant. Allow the child to participate in the exam. Use toys and other attention-grabbing props such as the "barking dog" to help assess the visual function. Encouragement from the physician and from family members can promote better cooperation and thus better exam results. Noting the patient's level of participation during an exam is also an important consideration when interpreting your exam findings.

To assess visual function in a preverbal child, check fix and follow behavior of each eye. Patch occlusion of a preferred eye during the exam may illicit a strong negative response from the child, signaling poor vision in that eye. This of course assumes minimal objection to occlusion of the non-preferred eye. The induced tropia test is also useful in a non-strabismic patient. This is done by placing a 20 prism-diopter base-down prism over an eye, one at a time, and allowing a couple of seconds to determine whether the child fixates on the second image. This prism amount is sufficient to provide two images for the child and two corneal light reflexes for the examiner to allow detection of which eye is being used for fixation [2]. In the strabismic patient, fixation preference can be determined by quantifying how long the non-preferred eye is able to maintain fixation. This may mean that the non-preferred eye immediately switches fixation back to the preferred eye when the cover is removed, indicating a very strong preference. Sometimes, the non-preferred eye can maintain fixation for several seconds after the cover is removed or will maintain fixation up to a blink, still indicating a preference for the other eye but less so than not maintaining fixation at all. Preferential looking techniques, such as Teller acuity cards, can also be used with good reliability in this age group.

In the verbal child, optotype visual acuity testing with HOTV matching, LEA symbols, or Snellen visual acuity testing can be employed. Each eye should be

occluded with a patch and separately tested for distance acuity with one of the aforementioned modalities. If it is determined that one eye is testing better than the other and you are unsure of the reliability of this result, the weaker eye should be tested first on a return office visit. Glare testing may also be useful in determining the visual significance of the cataract.

As mentioned previously, the assessment of the ocular alignment can be helpful in determining the amblyogenic effect of a unilateral cataract. The presence of strabismus typically indicates that the cataract is long-standing. The presence of nystagmus portends a poorer visual outcome, as it indicates visual deprivation from bilateral cataracts beginning in infancy. Sensory nystagmus usually develops around 3 months of age secondary to inhibited development of the fixation reflex that is normally formed by this age [3]. These eyes typically will not see better than 20/100 even after removal of the cataracts [1].

To test ocular alignment, the Hirschberg test is a good place to start and is done by assessing the position of the corneal light reflex. Cover-uncover and alternate cover testing at distance and near are superior for assessing ocular alignment and the degree of strabismic deviation. Overall, gathering information on the presence of strabismus or nystagmus may be useful for providing parents with predictive information regarding potential visual acuity [3]. It is important to counsel parents that the nystagmus is likely to persist after cataract surgery.

The anterior segment examination follows the assessment of visual function and motility. Penlight or portable slit lamp examination of the anterior structures of the eye includes examination of the eyelids and eyelashes, conjunctiva, sclera, cornea, anterior chamber, and iris. In the event that abnormal findings such a blepharitis or nasolacrimal duct obstruction are found, treatment should be initiated prior to cataract surgery. When possible, corneal diameter should be assessed. The pupil examination is important both before and after dilation and can offer further prognostic indicators. Aniridia should be noted as there is a known correlation between this condition and infantile cataracts and glaucoma. Poorly dilating pupils may reflect immaturity of the anterior segment and place the eye at an increased risk of glaucoma after cataract surgery [1].

The slit lamp examination is important to help further classify the morphology, location, and possible etiology of the cataract. Anterior chamber depth should be assessed when possible. Signs of inflammation such as cell, flare, or posterior synechiae indicate an inflammatory etiology. The presence of lens subluxation, iridodonesis, or phacodonesis could indicate prior trauma or a genetic condition as a cause of the cataract [3].

Those children who are able to sit for a slit lamp examination may be able to cooperate for a neodymium-doped yttrium aluninum garnet (Nd:YAG) laser session postsurgically. This bit of information is crucial for surgical planning, as it can help the surgeon to decide whether or not to perform a primary posterior capsulotomy with anterior vitrectomy at the time of surgery.

If there is an adequate view through the cataract, a fundoscopic evaluation of the posterior pole and retinoscopy should be performed. The health of the optic nerve and fovea are important to note and assist in predicting visual outcome

postoperatively. Determining the refractive error of both eyes, if possible, and noting the presence of anisometropia are important in surgical planning. If there is no view through the lens, then B-scan ultrasonography can be done in the clinic or at the time of surgery to rule out posterior segment abnormalities such as a retinal detachment or persistent fetal vasculature.

Some of these exam components will not be possible in the clinic due to the patient's age and level of cooperation. These exam details can be obtained during an examination under anesthesia (EUA) before or at the time of surgery. Intraocular pressure and pachymetry measurements are both important data points to gather but will likely be more accurate in pediatric patients while under anesthesia. Note that it is best to check the intraocular pressure at the time of induction of anesthesia, as general anesthesia can artificially lower eye pressure.

Preoperative Testing

Biometry and Keratometry

Biometry and keratometry are both required for surgical planning regardless of a plan for IOL implantation or contact lens use. Biometry may be obtained successfully in the outpatient setting in children who are cooperative enough to sit for measurements. Infants and younger children who cannot sit for biometry typically have measurements conducted during an EUA preceding their cataract surgery [3]. In the clinical setting, choices for biometry include optical biometry with the IOLMaster (Carl Zeiss Meditec AG) or LenStar (Haag-Streit Diagnostics, USA) or via ultrasound biometry with immersion or contact techniques. In the operating room, measurements are obtained with ultrasound biometry. Keratometry can be performed manually in the cooperative patient; however, various handheld automated keratometers are available for use in clinic or the operating room. It should be noted that the cylinder axis measurement may not be as reliable with the handheld keratometer [3].

Laboratory Investigation

Studies show that nearly 86% of unilateral and 68% of bilateral cataracts have no apparent cause [4]. Therefore, most surgeons do not advocate laboratory workup for congenital cataracts unless the patient's history is concerning. The differential diagnosis of bilateral congenital cataracts includes genetic, metabolic, infectious, inflammatory, and idiopathic causes. The recommended workup is tailored based upon the clinical history and can include TORCH titers to rule out infectious causes such as toxoplasma, rubella, cytomegalovirus (CMV), herpes, syphilis, varicella-zoster virus, and parvovirus B19. Urine amino acid testing can be recommended to

rule out genetic causes such as Lowe or Alport syndrome. Blood glucose testing should be completed to rule out diabetes.

Case 1

A 4-day-old male infant in the neonatal intensive care unit (NICU) presented with a poor red reflex in both eyes. The baby was a term infant but had been sent to the NICU for persistent hypotonia, difficulty feeding, and hyperbilirubinemia.

On exam, he was noted to have blink to light vision in both eyes. His intraocular pressures were normal. Pupils were equal in size and slowly reactive to light. There was no evidence of a relative afferent pupillary defect. On slit lamp examination, the anterior segment was normal with the exception of dense nuclear and cortical lenticular opacities in both eyes (Fig. 6.1). The pupils dilated poorly after installation of mydriatic drops. There was no view of the posterior pole secondary the lenticular opacities. B-scan ultrasonography was conducted and found to be normal, without evidence of retinal detachment, mass, or persistent fetal vasculature.

Extensive laboratory workup was conducted, and the patient was found to have Trisomy 21 on genetic testing. Additional workup was performed by the NICU team to rule out other comorbidities commonly seen in patients with Trisomy 21, but the remainder of systemic workup was normal. There is a known correlation between Trisomy 21 and congenital cataracts; thus, this was the presumed etiology for this patient's cataracts.

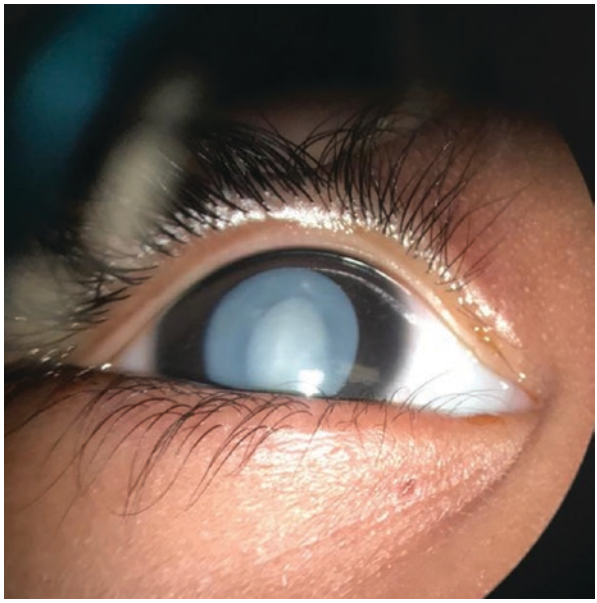

Fig. 6.1 Total lenticular opacity involving the nucleus and surrounding cortex of the lens. Also note the incompletely dilated pupil despite multiple rounds of mydriatic drops. A similar cataract was present in the fellow eye

The patient underwent immediate sequential bilateral cataract surgery at 6 weeks of age. The decision for immediate sequential surgery was made given the patient's age and risk of anesthesia-related adverse events. A complete examination under anesthesia and contact ultrasound biometry and automated keratometry measurements were done in the operating room. He was left aphakic, and refractive correction was planned with contact lenses.

Comment This case illustrates the importance of an infectious, genetic, and metabolic workup in infants with bilateral congenital cataracts and other concerning findings in their history and/or physical exam. As mentioned previously, in an otherwise healthy patient, a cause is not often found; thus, workup is not recommended [4]. It is imperative that any systemic condition that could lead to additional morbidity or mortality for the patient be detected and treated appropriately. It also illustrates the importance of discussing the timing of surgery, as well as the decision for immediate sequential cataract extraction in infants. The risk of anesthesia was felt to be greater due to this patient's Trisomy 21, and therefore, immediate sequential cataract extraction was performed. In an otherwise healthy baby, this same decision may not be pursued.

Case 2

A healthy 4-year-old girl presented to clinic as a referral for cataract evaluation. She did not have any medical conditions with known cataract associations nor did she have a history of trauma to either eye. She did not take any medications. Her first eye exam occurred approximately 1 month prior to this clinic visit. Her uncorrected Snellen visual acuity was 20/250 in the right eye and 20/30 in the left eye. Her pupillary examination was normal. Intraocular pressures, motility, and muscle balance testing were all normal, with the exception of reduced stereo acuity. Her slit lamp examination revealed a posterior subcapsular cataract in the superior temporal quadrant of the right eye. The cataract was mostly outside the visual axis when looking through the un-dilated pupil. After dilation, the opacity was noted to be largely occupying the superior temporal quadrant of the lens but again was mostly outside of the visual axis. The remainder of the slit lamp and fundoscopic exam was normal in both eyes. Cycloplegic retinoscopy revealed a refraction of +5.00 sphere in the right eye and +2.00 + 0.75 × 180 in the left eye. Because of significant anisometropia, the patient was prescribed glasses for full-time wear with the following prescription: +3.00 sphere in the right eye and plano+0.75 × 180 in the left eye. She was also instructed to start patching the left eye 3 h daily until her follow-up visit.

Three months later, the patient returned to clinic for follow-up. Her vision with correction was 20/150 and 20/30 in the right and left eyes, respectively. Mom reports she patched well for the first 2 months but had not been as compliant in the past few weeks. The remainder of her exam appeared stable; thus, the patient was instructed to continue full-time glasses wear, and the importance of patching was reinforced.

She returned several months later, and her vision with correction was 20/200 in the right eye and 20/30 in the left eye. Her mother admits that they have not been compliant with the glasses or patching. On slit lamp examination, the posterior subcapsular cataract appeared to be occupying more of the visual axis compared to her two previous examinations. The decision was made to proceed with cataract extraction with intraocular lens implantation. This patient underwent keratometry and contact ultrasound biometry in the operating room. Postoperative refraction of +2.00 was chosen in order to match her fellow eye and to allow for emmetropization with axial elongation.

Comment This case illustrates the importance of considering the etiology of the amblyopia. Just because a cataract is present, it is not always surgical. The leading reasons for amblyopia in this case include deprivation amblyopia from the cataract and refractive amblyopia from her uncorrected anisometropia. In this case, reinforcing the importance of a thoughtful and attentive preoperative exam, the cataract was not fully obstructing the visual axis, and there was a significant amount of anisometropia present. Treating her with refractive correction and patching first allowed the surgeon to be certain that deprivation amblyopia was causing a significant amount of her amblyopia. It is also important to note that the posterior subcapsular cataract progressed during this time. Sometimes, it is necessary to examine the patient several times in the clinic or perform an EUA prior to making a decision regarding surgery. This was evident not only on slit lamp exam, but the patient's vision also worsened compared to the previous visit.

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