

Handbook of Pediatric Strabismus and Amblyopia

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Preface

The *Handbook of Pediatric Strabismus and Amblyopia* is a practical, easy-to-understand resource on the diagnosis and management of both common and the more esoteric forms of strabismus. Emphasis is placed on the understanding of the basis of the strabismus, not rote memorization of strabismus patterns. Concepts regarding sensory adaptations and sensory testing are described in a simple way to elucidate rather than confuse the reader. An in-depth chapter on visual development and the pathophysiology of amblyopia is included.

The goal of the *Handbook of Pediatric Strabismus and Amblyopia* is to make this often confusing subject simple and easy to understand. This book should make an excellent resource for board review. Chapters are reader friendly. They are organized with clear sub-headings that allow the readers to quickly find their area of interest. Diagrams and drawings are prevalent throughout the book to help illustrate otherwise difficult or complex concepts. Composite strabismus photographs are included to demonstrate the strabismus as it actually appears in the clinical setting and to help with pattern recognition. These composite strabismus photographs are very useful for board review. Each chapter is fully referenced to provide evidence-based practice guidelines and further in-depth reading.

An important use of the handbook is patient and family education. Families are rightfully concerned about the strabismus and they have often been told conflicting and confusing information about it. Information, including diagrams and photographs from the handbook, can be shared with the families to clarify their specific type of strabismus. This important information is often lacking in general texts on ophthalmology.

I hope you will find the *Handbook of Strabismus and Amblyopia* to be an invaluable adjunct to your practice and for board review.

Kenneth W. Wright, MD

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Pediatric Eye Examination

Ann U. Stout

THE HISTORY AND PHYSICAL EXAMINATION

In the nonpediatric eye clinic, the physician often views the presence of a small child in an examining lane with some anxiety, if not dread. Examination of a child is quite different from that of the adult. The history is largely from a source other than the patient, and the examination requires patience and talent. There are several tricks to make the visit go as smoothly and efficiently as possible (see the box on the following page).

HISTORY

Although ancillary personnel are often relied on to take the history, this is best obtained by the physician who knows how to direct the line of questioning to the most useful information. The old adage that "the patient is always right" is especially true in the case of parents' observations about their children. Most of the history is obtained from the parents or the referring physician, but any input from the child is equally important. Many children will not complain of blurry vision or diplopia, but should they describe these symptoms one must be very alert to an acute process. This is also an invaluable time to observe the child in an unobtrusive fashion and preliminarily assess head position, eye alignment, and overall appearance. Often this may be the extent of the physical examination that one can obtain; once children realize that attention is focused on them, they may become very uncooperative.

The problem precipitating the visit should be stated in the parents' or child's own words and then elaborated. Requisite

questioning for all pediatric eye problems should clarify whether the problem is congenital or acquired and should specify the age of onset in the latter case. If the chief complaint is a visual problem, it is helpful for the parents to specify what the child can or cannot see; that is, does the child respond to lights, faces, toys near or far, very small items? In cases of strabismus, the frequency and stability of the deviation and any associated head posture are important. Precipitating factors may include fatigue, illness, sunlight, and close or distance work. For nystagmus, medications and the past medical history may be pertinent. With cataracts, any history of trauma, medications, or associated medical conditions is important, as well as the family history. Tearing patients need to be questioned about any redness, photophobia, or crusting of the lashes. In ptosis, the stability or variability is important, as is any associated chin elevation or general neuromuscular problems. For difficulties in school, it is helpful to determine if the problem is only visual or is related to a particular subject area (reading, spelling, writing, or math) and if there are any stress factors in the child's extracurricular life.

Important aspects of past history include prenatal and perinatal problems, birth weight, gestational age, and mode of delivery. Any medical problems should be elicited, as well as current medication and allergies. Early development should be assessed by asking about specific developmental milestones, such as rolling over, sitting up, and walking. The Denver Developmental Scale is a good reference for developmental norms.¹⁰ Later development can be ascertained by asking about scholastic level and performance.

The family history is very important because often the young child does not have enough past history to be useful. The focus should be on the presence of strabismus, poor vision, and neurological problems. In the case of possible genetic disorders, the number and sex of siblings, possible consanguinity, and the number and gestational age of any miscarriages should be documented.

PHYSICAL EXAMINATION

Establishing Rapport

If you approach the examination with dread, the child will sense your personal tension and become uneasy. Children can be unpredictable, noncommunicative, and uncooperative, which

may make the examination both time consuming and frustrating for a busy practitioner. However, if extra time is taken initially to gain the trust of the child, the rest of the exam will go much more easily. This “friendship” is often first established in the waiting room, where toys, appropriate books, and even small furniture should be made available. In a general practice seeing children on a fairly regular basis, at least one exam room should be outfitted to make a child feel relaxed and make the exam go more smoothly. A 20-foot lane is best because of the frequent use of single Allen cards and the need for distance measurements in strabismus. Attention-getting distance targets may include a remote control cartoon movie or a motorized animal. Near targets should have variety and appeal, as one frequently finds that “one toy—one look” is the rule.

Approach young children as though you had come to play with and entertain them, and you will receive a lot of useful information in the process. Find out what they like to be called and use their name frequently, but speak softly and keep a respectful physical distance from them until they warm up to you. Also, find out from parents their favorite imaginary or cartoon characters and refer to these during your exam. Make a game of the exam; play peek-a-boo with cover testing, swoop near targets around like an airplane to evaluate the range of motility, refer to glasses and lenses used as “magic” or “funny

Useful Items for Pediatric Eye Exams

- Allen cards (single and linear)
- Wright figures (single and linear)
- Tumbling E (single and linear)
- Eye patches
- Interesting distance and near fixation targets
- Accommodative near targets (finger puppets, wiggle pictures)
- Portable slit lamp
- Papoose board
- Wire lid speculums (infant and child size)
- Loose retinoscopy lenses
- Loose prisms
- Fusional tests (Worth 4-Dot, Titmus or Randot, Bagolini lenses)
- 28-diopter lens
- Handheld tonometer (Perkins or Tonopen)
- Calipers

sunglasses," make funny sounds to get their attention. Do the noncontact things first: cover testing, fixation testing, pupillary and red reflex exam. Many small children object to physical contact by a stranger and once they are upset it is usually the end of the exam for the day. Sometimes they will more readily tolerate their parents placing glasses or a patch than a strange doctor. Remember, if you find yourself getting frustrated or impatient with a child in one area, stop and go on to some other aspect of the exam.

With older children, asking direct questions about their hobbies, school, and family shows an interest in them and often distracts them from the anxiety of the exam. They often appreciate a handshake or pat on the knee. Explain to them what you are doing before you do it and be honest; avoid talking down to them. If they ask, do not tell them the mydriatic drops will not hurt, but explain that they will only sting for a minute, like swimming in a pool with chlorine.

Examination of the Uncooperative Child

Sometimes, despite the best efforts, a child simply will not cooperate, and urgency of the problem or the need for further information may require physically restraining or sedating the child. A papoose board can be used to control a child up to around 5 years of age, depending on their size and strength. A lid speculum can then be used with a topical anesthetic to force the eyes open, although Bell's phenomenon of the eyes often makes a thorough examination difficult, and crying can affect intraocular pressure measurements.

SEDATION

For the child in whom relaxation is important, or when physical restraint seems too psychologically traumatic, as in older children, sedation should be used. The common modes of outpatient sedation include chloral hydrate (oral or suppository), Propofol infusion, or a combination injection of Demerol, Phenergan, and Thorazine (DPT). The first has the advantage of good sedation with a low level of respiratory depression and no effect on intraocular pressure (IOP). The latter two have analgesic as well as sedative properties, which may be useful in painful procedures, but there is slightly more respiratory depression and Propofol will lower IOP.^{24,37}

Although chloral hydrate is often not effective if the children are more than 2.5 years old, Propofol and DPT can be used in older children. Whenever sedatives are given, the child's vital signs including pulse oximetry must be monitored until awake, ventilatory equipment must be available, and appropriately trained personnel should be in attendance. Any sedative may have a greater effect in children with underlying neurological abnormalities.

CHLORAL HYDRATE

The minimal effective dose of chloral hydrate is 50mg/kg, but often 80 to 100mg/kg is needed if manipulation of the eyes is anticipated or if prolonged sedation is needed for electrophysiological testing.^{9,45,46} Half the initial dose can be repeated up to a maximum of 3g if the child is not sedated in 30 min.²² Any sedative is best given on an empty stomach (4h since eating) to increase absorption and decrease the risk of aspiration.

DPT

DPT is given in a dose of 2:1:1 mg/kg, not to exceed 50:25:25. Because of the better analgesic effects, this is probably a better choice for painful procedures (laceration repairs, chalazion excisions, cryotherapy). Potential complications include respiratory depression, apnea, dystonic reactions, hypotension, seizures, and cardiac arrest.³⁷ This mode of sedation should only be used when the child is under the supervision of a physician with appropriate training to manage complications, as in the emergency room.

EXAMINATION UNDER ANESTHESIA

If it is impractical to sedate the child in the office, or if surgery is anticipated based on the exam findings, than examination under anesthesia should be arranged in the operating room. Modern anesthetic practices make general anesthesia a very safe procedure, even when done repeatedly. A disadvantage of general anesthesia is the purported intraocular pressure-lowering effects of inhalational anesthetics. Pressures taken under inhalational anesthetics have been lower than those measured in awake children, but this is probably a result of increased overall relaxation.¹³ Propofol has a similar effect on IOP.²⁵ The pressure may actually increase several points after intubation.⁴⁰ Use of laryngeal mask airways may eliminate this transient pressure rise.⁴³ It is probably best to record the pressure both before and after intubation.

External Examination

The child's overall appearance and level of alertness can be judged during the history taking. Head posture may also be noted then, as well as gross ocular alignment. The history or appearance may warrant more detailed examination of overall neuromuscular tone, cranial nerves, head circumference, extremities, or skin. The head may be assessed for symmetry, preauricular skin tags, ear position, and shape. The orbits should be appraised for ptosis, abnormalities in fissure size or shape, and orbital depth. Many of these findings do not require a systematic search but rather an overall heightened awareness of what is normal versus abnormal. The general assessment of a dysmorphic child should push the physician to more precisely define what abnormalities lead to that impression.

Visual Acuity Assessment: Preverbal

To judge vision in the preverbal child, one must rely on the smallest age-appropriate target that will hold attention and on the difference, if any, between the two eyes. An appropriate target for a 1-year-old child may be a small finger puppet; but a 1-month-old may fixate only on a human face and do that rather unsteadily. Infants are unable to pursue targets smoothly until 6 to 8 weeks of age but instead will track using hypometric saccades.⁵ Targets with fine detail that require accommodation and focused attention are best for children over age 1, for even though accommodation is appropriate to target distance by age 3 to 4 months, the macula is still immature even at the age of 15 months.⁶ Children who have developed a pincer grasp can be asked to pick up small particles from the palm of your hand. Often cake sprinkles are useful because they are edible and such a target often ends up in the mouth.

FIXATION

There are two types of fixation testing: monocular and binocular. In monocular fixation testing one assesses whether the patient fixes with the fovea (centrally) and the quality of fixation. Each eye should be occluded in turn and the smallest possible target that elicits a fixation response should be used. Monocular fixation should be assessed for three separate factors: *quality* and *accuracy* (good, fair, poor), *location* (central versus eccentric), and *duration* (maintained versus sporadic). Clinically, abbreviations

are often used to describe fixation including GCM for *good, central, and maintained*, CSM for *central, steady, and maintained*, or FF for *fix and follow*. Eccentric fixation is an important sign as this shows that the patient is not fixing with the fovea and vision is in the range of 20/200 or worse. It is important to remember, when testing monocular fixation, that the fixation target should be slowly moved through the visual field to assess the quality of fixation. The target size and distance should be estimated and documented in the chart. It is important to be aware of the normal timetable for visual maturation, although this may vary widely with individuals. The newborn has only sporadic saccadic eye movements with very poor fix and follow; by 6 weeks, most infants will show some smooth pursuit and central fixation, and by 8 weeks the vast majority of infants will have central fixation with accurate smooth pursuit and easily demonstrate optokinetic drum responses.⁶ Smooth pursuit is asymmetrical until age 6 months of age, with monocular temporal to nasal pursuit being better than nasal to temporal pursuit. Tables 1-1 and 1-2 outline normal visual development. One should remember that there is a subgroup of patients who are otherwise normal yet show delayed visual maturation. However, even this group of patients should show improvement of visual function and should have central fix and follow by at least 6 months, with occasional delays up to 12 months of age.

Binocular fixation preference compares the vision of one eye to the other. This test presumes that strong fixation preference in patients with strabismus indicates organic visual loss or

TABLE 1-1. Normal Visual Development.

Pupillary light reaction present: 30 weeks gestation
Blink response to visual threat: 2–5 months
Fixation well developed: 2 months
Smooth pursuit well developed: 6–8 weeks
Saccades well developed (not hypometric): 1–3 months
Optokinetic nystagmus (OKN)
1. Present at birth but with restricted slow-phase velocity.
2. Temporal to nasal monocular response better than nasal to temporal until 2–4 months.
Accommodation appropriate to target: 4 months
Stereopsis well developed: 3–7 months
Contrast sensitivity function well developed: 7 months
Ocular alignment stabilized: 1 month
Foveal maturation complete: 4 months
Optic nerve myelination complete: 7 months to 2 years

TABLE 1-2. Age-Related Visual Acuity Estimates by Test Method.

<i>Technique</i>	<i>Birth</i>	<i>2 months</i>	<i>4 months</i>	<i>6 months</i>	<i>1 year</i>	<i>Age for 20/20 (months)</i>
OKN	20/400	20/400	20/200	20/100	20/60	20–30
FPL	20/400	20/400	20/200	20/150	20/50	18–24
VEP	20/800	20/150	20/600	20/400	20/20	6–12

OKN, optokinetic nystagmus; FPL, forced-choice preferential looking; VEP, visual evoked potential.

amblyopia in the nonpreferred eye.^{12,48} Binocular testing has an advantage over monocular testing, because vision can be very poor (20/100 to 20/200) and the patient will still show essentially normal monocular fixation. Binocular fixation preference testing, however, will identify even mild amblyopia (two lines of Snellen acuity difference).^{44,47} It is important to assess monocular fixation before fixation preference testing to rule out the possibility of bilateral symmetrical visual loss in preverbal children. Fixation preference testing is very accurate for diagnosing amblyopia in children with large-angle strabismus.⁴⁸

In patients with straight eyes or microtropias, binocular fixation preference testing can be done using the vertical prism test or induced tropia test.⁴⁷ In patients with straight eyes, one does not know which eye is fixing; therefore, it is impossible to determine fixation preference. The vertical prism test induces a vertical deviation and therefore allows assessment of fixation preference. In patients with small-angle strabismus (less than 10–15 prism diopters), the induced vertical tropia dissociates peripheral fusion and eliminates any facultative suppression scotoma associated with the patient's baseline ocular alignment. In turn, this eliminates the problem of overdiagnosis of amblyopia, previously described by Zipf.⁴⁸ Fixation preference testing is a quick and accurate way of diagnosing amblyopia in clinical conditions such as anisometropic amblyopia, unilateral ptosis, postoperative congenital esotropia, and other conditions that could cause unilateral amblyopia.

OPTOKINETIC NYSTAGMUS

Children with poor fixation to any targets as a result of either poor vision or central nervous system problems can be evaluated for the presence of *optokinetic nystagmus (OKN)* so long as they are able to generate saccades. Optokinetic nystagmus is an involuntary pursuit response to moving stripes filling up

most of the visual field, so a response may be seen in infants who are merely uninterested in other targets. Response to a standard OKN drum implies vision of finger counting at 3 to 5 feet.^{16,17} One can also assess the damping of the induced vestibulo-ocular reflex. By spinning the child around, either in your arms or on a swivel chair, a vestibular nystagmus will be induced, despite the level of vision. If there is visual input once the spinning is stopped, the nystagmus should damp in 30 to 60s due to the fixation reflex.

OTHER TESTS

Many ingenious tests have been devised to try to better correlate the vision to a linear acuity and detect amblyopia. The most popular are forced-choice preferential looking (FPL) and pattern visual evoked potentials (PVEP). The preferential looking test using Teller acuity cards assesses grating acuity by presenting the child with high-contrast gratings of various spatial frequencies along with a paired blank card. Infants will naturally prefer to look at patterns if they can be seen, and the examiner assesses whether the child fixates on the pattern or not.³⁶ This test reliably measures grating acuity, but it may take 20 to 30min to perform and may be difficult to perform monocularly in children younger than 2 years old³³ (Fig. 1-1). Although useful in the assessment of anisometric or deprivation amblyopia, this test



FIGURE 1-1. Teller acuity preferential looking apparatus.

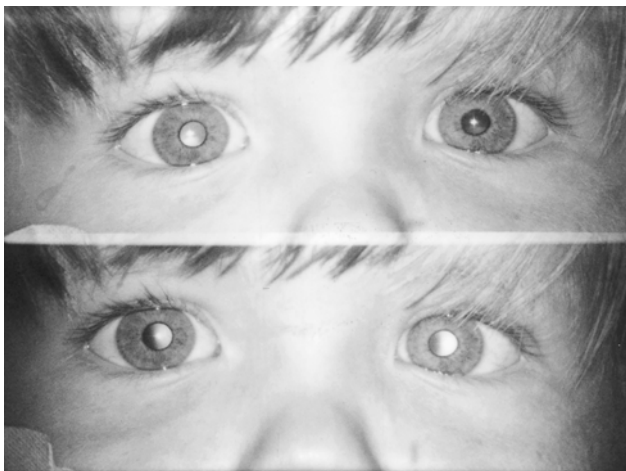


FIGURE 1-2. Two Polaroid photographs from MTI PhotoScreener, taken by a rotating flash. The *white crescent* in the pupils denotes a refractive error. The size and location of the crescent in each photo indicate the type and example of the problem. Findings: hyperopia, +2.00 D; astigmatism, >1.00 D. The displaced corneal light reflex demonstrates esotropia.

may underestimate strabismic amblyopia or decreased acuity secondary to macular disease, in which grating acuity may be much less affected than vernier and Snellen acuity. Therefore, although a detected difference confirms amblyopia, a normal test result does not rule out amblyopia.²⁶ Fixation preference testing is more reliable in detecting strabismic amblyopia.

PVEP measures the summed occipital cortical response to a pattern stimulus (Fig. 1-2). This method reflects the activity from the central retina and is therefore a good assessment of macular function.³² The resulting cortical potential can be evaluated by a trained electrophysiologist and Snellen acuity can be roughly estimated. The important parameters of the PVEP waveform are the amplitude and the latency of the spikes. The first major large positive deflection is the P1 spike, with a normal latency of 100ms. Amblyopia diminishes the amplitude, as do uncorrected refractive error or organic problems. The difficulty in the test lies in the need for specially trained personnel to administer and interpret the test and the frequent need for seda-

tion to get good results. Although the test can be administered while the child is awake, poor fixation may give artificially low results. In the sedated child (chloral hydrate), cycloplegia and appropriate refractive correction are necessary because retinal blur also affects test results.^{45,46} Despite these drawbacks, the test is often useful to monitor the progress of amblyopia therapy and to diagnose amblyopia in preverbal children.

Visual Acuity Assessment: Verbal

OPTOTYPE

Older children who are able to identify character shapes can be assessed with Allen cards, Wright figures, or Lea symbols.¹⁸ Often, a child who is not quite verbal, or is too shy to talk, can be asked to match a sheet of pictured figures to the displayed cards. The single cards are most useful in the beginning, because often the child does not attend well to distance targets such as projected figures. The child can also take a photocopy of the cards home to practice as a game with parents. It is best to start close to the child and work backward. In young children, the endpoint is more often demonstrated by a loss of attention than by an incorrect response. The disadvantage of single cards is that they cannot detect the crowding response that is so often seen in amblyopia. A child may test equally on single optotypes but show a marked discrepancy with linear optotypes.³⁴ Once equal acuity is obtained on single cards, it is important to move to linear testing to confirm the findings. Both single and linear figures test only to the level of 20/30, which is adequate for children under the age of 3 years.

The next step in preliterate testing is use of the tumbling E, Wright figures, or HOTV.¹⁸ With the "E game," the child is asked to point his fingers in the direction of the "legs on the table." Often, vertical orientations are more readily confused by children than horizontal directions, and this should be taken into account when determining the endpoint.³⁹ The HOTV letters can be easily identified even if the full alphabet is not known. Again, single cards or linear projection can be used, the latter being best for amblyopia. These tests go to the 20/20 level and are slightly more challenging than picture cards.

Once the child is literate, traditional Snellen letters can be used. For most children, this occurs around age 5 to 6, but do not confuse knowing the alphabet with being able to distinguish

random letters. If in doubt about the reliability of letter testing, return to an easier test.

Visual Fields

As soon as a child is able to fix steadily on a target, a rough estimate of visual fields can be obtained. Even if the child will not tolerate a patch, binocular fields can be checked for homonymous or bitemporal defects. Infants with good fixation will usually move to an interesting peripheral target once it comes into view as a result of the fixation reflex. The examiner captures the attention with a central target and then slowly brings in a peripheral target, watching for the first jump to the peripheral target. In this manner all four quadrants of the peripheral field can be tested. Patients with posterior optic pathway lesions that respect the vertical meridian will often ignore a peripherally advancing target until it crosses the midline and then suddenly move and pursue it. A slightly older child of 3 or 4 years may be able to respond accurately to finger counting by making a game of copying the examiner's actions.

Formal visual field testing such as Goldmann perimetry can sometimes be performed in preschool children. It is important to have a good idea of the suspected field loss and concentrate on these areas first. Usually the largest brightest target (V4e) is best, but smaller targets should be used if the child is capable of cooperating. Automated fields require prolonged concentration and steady fixation and are usually not reliable in children less than 9 to 10 years old. Some newer user-friendly programs are being developed that shorten testing time enough to make them more applicable for children (Welch Allyn, frequency doubling technology or FDT). The tangent screen is also not useful until reliable verbal responses can be made because it is difficult to monitor fixation.

Assessment of Color Vision in Children

Although color vision testing is not often done in children, it helps in the diagnosis of decreased acuity of uncertain etiology and monitors progression in cases of macular degenerations or progressive optic neuropathies. Children on retinal toxic drugs also need to be evaluated. Congenital red-green color defects are often detected first by the pediatric ophthalmologist as an incidental finding. Screening questions are often useful in detecting

these cases, which occur in 8% to 10% of the male population. The child may confuse green with brown crayons and purple with blue crayons; he may confuse yellow and red traffic lights or green and red lines on a paper. Often he will have no trouble accurately naming colors of large objects but will mistake smaller colored objects subtending less than 2° .⁵

The easiest way to screen for color vision defects is with color plates. There are two popular types of plates, which are each useful in specific situations. The *Ishihara pseudoisochromatic color plates* work on the principle of color confusion, which is common with dichromats and anomalous trichromats. These plates are extremely sensitive for red-green defects, which are usually congenital. Most acquired color defects show some loss in the blue-yellow range, and the Ishihara plates will miss these patients unless the loss has extended into the red-green range. The advantage of these plates is that they come in an illiterate form with geometric shapes that can be traced with a finger. This design is useful for children who do not know numbers but still requires the comprehension and fine motor skills of a 3- to 4-year-old. The *Richmond pseudoisochromatic plates*, formerly called *American Optical Hardy-Rand-Rittler (AO-HRR) plates*, work on color saturation and can detect both red-green and blue-yellow defects, making them more useful in acquired defects; unfortunately, these do not come in an illiterate format. Both tests use many two-digit numbers, which can intimidate young children, and often their responses are better if they are asked to name each digit separately. Another good test for children is the *City University Color Vision Test (TCU test)*; this uses the colors in the Farnsworth D-15 in a book format, so that manipulation of the color discs is not necessary. Unfortunately, it is not the best test for screening, as 20% of color defectives will pass the test. In general, optic nerve disease is more likely to affect red-green perceptions, whereas retinal disease affects blue-yellow discrimination, although there are many exceptions to this rule.^{16,17}

Assessment of Contrast Sensitivity

Contrast sensitivity represents the minimal amount of contrast required to resolve various-sized objects from the background. As such, it is perhaps a more sensitive test of visual function than Snellen acuity, which only assesses high-contrast resolution. The contrast sensitivity threshold is the minimal amount

of contrast required to detect sinusoidal gratings of different spatial frequencies. The contrast sensitivity function (CSF) is the curve obtained by plotting contrast sensitivity against spatial frequency. The peak of the curve is usually at three to four cycles per degree, although the maximal contrast sensitivity for each spatial frequency increases with age to stabilize in adolescence. Contrast sensitivity may show decrements in many disease processes despite a normal Snellen acuity, including cerebral lesions and multiple sclerosis.⁴ Amblyopes demonstrate a reduction in the CSF curve that may occur only at the peak (high-frequency loss) or throughout the curve (high- and low-frequency loss).²⁰ This difference persists after Snellen acuity returns to normal and may be useful in detecting a continued need for amblyopia therapy.^{31A} Occlusion therapy for amblyopia has also been shown to reduce contrast sensitivity in the dominant eye, even without decreased acuity, a type of mild occlusion amblyopia.²⁴ Although testing previously required fairly complex equipment and was not suitable for young children, contrast sensitivity function can now be reliably measured on children over 4 years using the Vistech wall chart. The chart presents eight levels of contrast sensitivity (horizontal axis) for each of five levels of spatial frequency (vertical axis). The gratings are oriented in one of three directions, +15, 0, or -15, and the child imitates grating orientation with his hand as in the "E game." The minimum contrast detectable at each spatial frequency is recorded and used to plot a contrast sensitivity function curve.

Red Reflex

Evaluation of the red reflex is often forgone in adults because of the better sensitivity of other available tests; that is, visual acuity and high-power biomicroscopy of the anterior segment, lens and vitreous. In children, these tests may not be applicable for reasons of youth or lack of cooperation. Evaluation of and especially binocular comparison of the red reflex are invaluable in assessing media opacities or refractive aberrancies. The red reflex is best tested by staying far enough away from the child to illuminate both pupils with the same direct ophthalmoscope beam and comparing the quality and intensity of the reflexes between the two eyes. The exam should be done in dim illumination, to encourage pupil dilatation, and with the child's attention focused in the distance, to avoid a near response. If the direct ophthalmoscope beam is too strong, the pupils will con-

strict and the child will react to the brightness by blinking or turning away. It is important to assess the reflex both before and after dilation, especially if there is a visually significant opacity, to see how much of the undilated pupillary space is obscured. Dimming of the red reflex is also an important sign in early endophthalmitis after cataract or strabismus surgery.

Bruckner described a useful test for strabismus using the red reflex from the direct ophthalmoscope. In the presence of strabismus, the red reflex will be brighter and the pupil will appear slightly larger in the deviated eye as the patient fixates on the light. This test can detect deviations as small as three prism diopters and is especially useful in evaluating postoperative alignment.³⁸ The test can be carried a step further to evaluate amblyopia by narrowing the light beam and illuminating one eye at a time. Fixation with each eye should be steady on the light; if amblyopia is present, fixation may waver or the eye may remain deviated in the presence of strabismus.

Photoscreening is a sophisticated application of the red reflex test. The MTI photoscreener creates a Polaroid picture of the red reflex that allows assessment of media opacities and refractive errors (see Fig. 1-2). Although not as sensitive as a traditional eye examination, it is a useful screening tool for general practitioners.^{29,30}

Pupillary Examination

Normal pupil size varies with age. The newborn has small, miotic pupils that increase to an average diameter of 7 mm by age 12 to 13 and then gradually decrease again throughout life. Reaction to light in infants is often difficult to assess due to the natural miosis and uncontrolled near response to the examination light. With careful observation, a small light response can be seen in addition to the near response, but care must be taken to not mistake one for the other. Older children should have the near response controlled as much as possible by a distance fixation target. If the examination light is too bright, the child will close his eyes; when necessary, they must be held open to get an adequate exam. It is important to rule out an afferent pupillary defect, especially with unilateral visual loss or strabismus. The swinging flashlight test is routinely used, as in adults, but care must be taken to aim the light directly into the pupil, especially in strabismus, or an artificial afferent defect may be produced because of incomplete retinal stimulation. It is important

to remember that only dense amblyopia can produce an afferent defect and that even then it is barely detectable.³¹ Any afferent defect of significance in a patient with presumed amblyopia must be investigated further.

Slit Lamp Examination

Although most young children who cannot cooperate with a slit lamp examination can be adequately evaluated with a muscle light, at times more detailed examination is mandatory. There are several handheld slit lamps, which have the advantage of being portable and useful in examining supine patients. The disadvantages are their cost and somewhat limited resolution. They are not very good for assessing mild intraocular inflammation or subtle corneal abnormalities, but they are the best alternative. There are several other more readily available magnifiers such as the direct and indirect ophthalmoscope, both of which can be focused on the anterior segment. Whichever source is used, the exam is still limited by the child's resistance and movement, even while restrained. At times, sedation is required to obtain the necessary information. Often a child of 1 or 2 years will allow a quick look at the standard slit lamp; the key is to keep the light as dim as possible, have in mind what you most want to see, and look at that first.

Intraocular Pressure Measurements

Certain children are more prone to develop elevated intraocular pressure, and these patients must have accurate measurements. Such patients include aphakes, those with any anterior segment anomaly, those with orbital vascular lesions, or those on steroids. Most children under 3 years will not cooperate with routine applanation tonometry and they must be supine (sedated or restrained). There are several useful handheld tonometers. The original Schiøtz tonometer is easy to use and read but often is too large for the infant eye. It has the advantage of being less sensitive to pressures induced by lids or extraocular muscles but is more affected by ocular rigidity, which tends to be lower in infants. The Perkins tonometer is an applanation device using fluorescein and a split prism to assess the pressure. It is highly accurate at all ranges of pressure but requires some experience on the part of the examiner to read and is unreliable with an abnormal corneal surface. Contact must also be made directly

on the cornea, which is often difficult in a struggling child with a good Bell's phenomenon. The Tonopen is easier to use and can obtain approximate readings off the sclera or an irregular cornea, but it is not accurate at high or low pressures, and it is difficult to hear the tones that signal endpoint when the child is crying.²³

The pneumotonometer is easy to use and does not require corneal contact, but the equipment is less portable than the other two. Struggling and crying both cause swings in the intraocular pressure, which must be taken into account, and pressures taken with a calm child are more accurate. Small infants who are drowsy will sometimes allow tonometry, especially if they are held in their mother's arms and given a bottle or pacifier. Sedation may be necessary for truly accurate readings. Chloral hydrate has the advantage of not lowering the pressure, whereas inhalation anesthetics and Propofol can.^{3,25} If an exam under anesthesia is done, the pressure should be measured as soon as safely possible after induction because it will become artificially lower with time.¹³

Keratometry

Assessment of corneal shape can be done qualitatively or quantitatively. Several conditions predispose to corneal astigmatism, which may be amblyogenic or require contact lens correction. Children with limbal dermoids or corneal scars may have poor retinoscopy reflexes, which make accurate assessment of astigmatism difficult. Placido's disc is a keratoscope that images a series of concentric light rings on the cornea. The reflected image can be used to assess the axis of astigmatism and corneal regularity. It is handheld and nonthreatening to most children but only gives a rough qualitative assessment. More accurate measurements must be obtained with a keratometer; this may require a sleeping or sedated infant to get accurate readings, and the standard keratometer can be mounted on a special bar to use with supine infants.³⁵ Such measurements are helpful for contact lens fitting. Alcon Corporation has produced a handheld keratometer that has been very helpful in the pediatric age group.

Dilatation and Cycloplegia

Cycloplegia is essential to eliminate uncontrolled accommodation and adequately assess the refractive error in children. Several agents are available, but the adequacy of cycloplegia, not

the maximal pupil dilatation, is most important. Tropicamide is not a strong enough cycloplegic for young children; instead, cyclopentolate, homatropine, or atropine should be used. Cyclopentolate has the most rapid onset and shortest duration and thus lends itself to clinic use. For most children, one drop each of cyclopentolate 1% in combination with phenylephrine 2.5% is adequate.² Lighter pigmented eyes may only require one or two applications, whereas darkly pigmented eyes may require more than three. For children less than 6 months old, it is safer to use diluted drops such as Cyclomidril (cyclopentolate 0.2% and phenylephrine 1%). Homatropine 5% is another choice used for clinic dilation, especially in darkly pigmented patients, but this drop lasts up to 3 days. Both cyclopentolate and homatropine produce maximal cycloplegia within 30 min to 1 h, but the former recovers within 1 day.

Table 1-3 lists the various agents available and their effects. If cycloplegia seems inadequate, based on either pupil size or changing retinoscopy streak, it is best to use atropine; this is usually given to the parents to take home and administer. To avoid toxicity of frequently administered atropine, the drops are given twice a day for 3 days prior to the visit.¹⁵ For infants and very young children, the drops should be given only once a day to each eye, with one eye receiving one drop in the morning and the other eye receiving one drop at night. As an alternative, atropine 0.5% could be used. Atropine should not be given to children with possible heart defects or reactive airways. Punctal occlusion can be performed for 1 min after the drops to decrease systemic absorption. Parents should be alerted to discontinue the drops if signs of toxicity or allergy develop (flushing, tachycardia, fever, delirium, lid edema, redness of the eyes). Most cases of toxicity respond to discontinuation of the drops, but more severe cases can require treatment with subcutaneous physostigmine (Eserine), 0.25 mg every 15 min, until improvement occurs. This treatment is useful for toxicity with any of the antimuscarinic agents. The phenylephrine drops will occasionally cause blanching of the periocular skin, especially where the drop contacts the skin either by tears or a tissue; this is seen most often in infants and does not require treatment or discontinuation of the drops. For examination of premature babies, dilate with cyclomidril (combination of cyclopentolate and phenylephrine) and tropicamide 0.5%.

Refraction is always an art, especially in a preverbal child. Care must be taken to control the working distance and the

TABLE 1-3. Cycloplegic/Mydriatic Agents in Children.

Agent	Strength (%)	Mydriasis		Cycloplegia		Side effects
		Maximum (min)	Recovery time	Maximum (min)	Recovery time	
Phenylephrine	2.5	20	2-3 h	None		Tachycardia, hypertension
Tropicamide	0.5, 1	20-40	2-6 h	30	2-6 h	
Cyclopentolate	0.5, 1, 2	30-60	6-24 h	25-75	6-24 h	Psychosis, seizure
Homatropine	2, 5	40-90	1-3 days	30-60	1-3 days	Ataxia
Atropine	0.5, 1	30-60	7-14 days	60-180	3-12 days	Flushing tachycardia, fever, delirium

visual axis, or inaccurate readings will be taken. Expertise with loose trial lenses or a skiascopy rack is important as the phoropter is useless in small children. If the endpoint is unclear, it is best to get a second or even third reading, either by repeat refraction on another visit or by a second refractionist. There are several handheld *autorefractors* on the market now that show promise in pediatric application (Nikon Retinomax, Welch Allyn Suresight).^{1,8,42} Cycloplegia may still be necessary to optimize results.

Fundus Examination

An adequate fundus examination is imperative for all children who present to the ophthalmologist. The extent of the fundus exam necessary will vary widely depending on the patient. For most patients visualization of the posterior pole (optic nerve and macula) is adequate; this is done quickly and easily in most children by keeping the indirect light low and not touching the child. A brief look may be all that is obtainable by this technique, but this is often adequate. The optic nerve can be examined in more detail with the direct ophthalmoscope if the examiner is unhurried and stays several inches away from the child. By staying focused on the retinal vessels, the observer can see the nerve as it wanders into view while the child is busy watching a distant target (moving targets, especially videos, work best for this). For more detailed fundus examination or examination of the periphery, sedation or restraints are usually needed because most children will not tolerate the examining light for extended periods of time. As fundus examination comes at the end of the clinic visit, the child may be sleeping, especially if they have taken a bottle after the eyedrops, and this makes the examination much easier. Children less than 2 years old can usually be restrained adequately to allow a thorough fundus examination, even to the periphery, whereas older children require examination under anesthesia if uncooperative.

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