

Vision Assessment and its Implications for Children and the Pediatric Community



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OVERVIEW

PREVENTATIVE SERVICES AND ANTICIPATORY GUIDANCE PLAY AN IMPORTANT ROLE IN THE CARE OF CHILDREN. AMONG THESE SERVICES, WHICH ARE VITAL TO THE WELL BEING OF THE PEDIATRIC POPULATION, IS VISION SCREENING. THE FAILURE TO ACCOMPLISH VISION SCREENING CAN HAVE A SIGNIFICANT IMPACT ON A CHILD LATER IN LIFE. THIS WHITE PAPER ADDRESSES THE FUNDAMENTALS OF VISION ASSESSMENT AND ITS IMPLICATIONS FOR CHILDREN AND THE PEDIATRIC COMMUNITY.

The Committee on Practice and Ambulatory Medicine (COPAM) of the American Academy of Pediatrics has, as its primary objectives, the review of issues that affect the primary care pediatrician and pediatric practice, and the formulation of recommendations that will improve the efficiency and quality of the practice environment. One such tool created by COPAM is the Recommendations for Preventative Pediatric Health Care or the Periodicity Schedule (RE9535), a grid that recommends the timing, content, and procedures to be attempted or accomplished at each individual well-child or adolescent visit. The components of such a visit include comprehensive medical history, physical exam, vital signs, developmental and nutritional assessment, laboratory testing, anticipatory guidance counseling, dental evaluation, and assessment of hearing and vision.

Many organizations external to the AAP and constituencies within the AAP lobby forcefully for placement of individual priorities and initiatives within the confines of the periodicity schedule. However, only those interventions which can be performed in the context of the office setting, and which can be shown to have a significant impact on clinical outcome, are included in the schedule. As much as possible, the inclusion of parameters within the schedule is based on empirically based, research driven, and scientifically verifiable data. With this charge in mind, COPAM changed the vision assessment recommendation,

The Facts

- Most attempts to screen 3 year olds for vision problems yield interpretable results
- Only 25% of 3 year olds and 50% of 4 year olds can complete an eye chart exam successfully
- 50% of parents whose children had an abnormal vision screen didn't understand that there was a problem
- Only 21–27% of pediatric practices attempt to screen 3 year olds for visual dysfunction
- Amblyopia is the most common cause of preventable visual loss in children

OVERVIEW	1
BASIC SCIENCE	2
PATHOPHYSIOLOGY	2
VISION SCREENING AND TESTING	4
CONSIDERATIONS	5
TECHNOLOGICAL ADVANCES	6
REIMBURSEMENT	6
PROFESSIONAL LIABILITY	6
REFERENCES	8

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in 1995, from a subjective evaluation (historically-based data obtained from parents) to an objective evaluation (reproducible vision testing) for children at their three-year-old visit.

This decision was based on a number of studies performed by the AAP. Periodic survey #22 of the members of the Academy explored the vision screening practices of pediatricians. The study, completed in 1993, was a follow-up to periodic survey #3, completed in 1988. A total of 1137 responses, representing 71.1% of those surveyed, were received. Results of that study demonstrated that 91% of pediatricians performed visual acuity tests in both of the studied years. However, in 1993, only one third of pediatricians reported successful vision testing of children as young as age three.

Failure to diagnose a variety of pediatric conditions is now the #1 cause of allegations of medical negligence against clinicians who care for children. Within that category, failure to diagnose visual impairment is ranked #4, after failure to diagnose meningitis, appendicitis, and hip dysplasia

Studies have shown that only 25% of three year olds and 50% of four year olds can complete an eye chart exam successfully. Data obtained from the Pediatric Research in Office Settings Study Group (PROS) indicated that the majority of attempts to screen three year olds for vision problems led to interpretable results. Ironically, the study also pointed out that communications about abnormal results with the parents of children who failed vision screening was not uniformly completed or understood. Results of a questionnaire administered two months after the visit indicated that 50% of those parents whose children had an abnormal visual screen did not understand that their child had a problem. Some studies suggest that as few as 21–27% of pediatric practices even attempt to screen three year olds for visual dysfunction. This paper will focus on the importance of vision screening for children, especially younger patients, prior to school admission. Implications for children and the pediatric community regarding the failure to screen and implications for practitioners from an ethical and medical-legal perspective will be reviewed.

2

BASIC SCIENCE

The eye, at its most basic, is the organ of sight and the most distal outpouching of the human brain. The spherical structures known as the eyeballs, or globes, are protected in the bony sockets of the skull, called the orbits, and, externally, by eyelids, lashes, eyebrows, and a film of tears. Six small muscles attached to the globe control the external motion of each eye. Light and visual stimuli enter the eye via the pupil and are focused by the cornea and lens to form an image on the retina. The retina contains two types of light sensitive cells – rods and cones. There are approximately six million cone cells in each eye responsible for sensing bright light and different light wavelengths, and 120 million rod cells which sense dim light, but not color. Rods and cones convert the received images into a pattern of nerve impulses that are transmitted along the optic nerve to the brain. Through a number of mechanisms, these binocular images are processed and incorporated into a single image via a number of integrated centers in the brain.

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and its Implications for Children
and the Pediatric Community**

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PATHOPHYSIOLOGY

Normal and equal visual inputs are necessary for proper growth and development and structure of the eye. If such inputs are not consistently received by the developing eye and visual cortex, the normal trophic response is blunted, and visual atrophy occurs. Visual system cell growth is initiated at birth and is finished by nine years of age. If during this interval visual inputs are not equal, cell growth is disturbed and vision becomes deficient.

From a clinical perspective, pediatricians observe several major types of visual loss, or amblyopia:

- Image–degradation amblyopia, associated with a structural problem involving the lids (ptosis, cyst, hemangioma, or tumor), or other periocular tissues, such as lens cataracts, corneal abnormalities, retinal edema, etc.
- Strabismic amblyopia, a condition in which the visual axis of the eyes are nonparallel so that they are not directed at the same object. Described as esotropia when the eyes are crossed inwardly, and exotropia, when the eyes are divergent. Less common is hyper– or hypotropia, when the eyes are directed up or down.
- Ametropic amblyopia results from abnormal cortical visual input due to uncorrected refractive errors, such as myopia (nearsightedness), hyperopia (farsightedness) or astigmatism, in which visual signals are inappropriately focused anterior or posterior to the retina.
- Anisometropic amblyopia refers to a visual condition where there is a significant imbalance in refractive error between an individual's eyes. This type of amblyopia is more common in patients in whom one eye is relatively hyperopic rather than myopic. Even small degrees of hyperopic anisometropia can induce clinically significant amblyopia.

Amblyopia can, potentially, develop from any of these conditions since the retina will transmit a blurred image to the brain. A potent stimulus with refractive error will cause suppression of the blurred image at a central level and ultimate loss of vision. Amblyopia is present in 2% of the population and is the most common cause of preventable visual loss in children. Strabismic amblyopia and ametropic amblyopia each occur in about 33% of patients, with a mix of the two conditions occurring in the remaining 33%. There is no genetic predisposition to amblyopia since it is an acquired illness. Some illnesses and conditions, such as syndromes of eye muscle abnormality or conditions associated with cataracts, would obviously predispose to amblyopia.

Once amblyopia is identified, treatment is directed at reversing or decreasing the stimulus to amblyopia. Image–degradation amblyopia may be treated medically, although surgical repair (ptosis repair or cataract extraction) may be definitive. Other types of amblyopia may be treated with glasses, lenses, or penalization, in which patching or occlusion of the non–involved eye for various periods of time during the day, may be employed. In the treatment of anisometropic amblyopia, glasses should, in most cases, precede patching therapy as a method of visual correction. Surgery is at times warranted to tighten or improve function of ocular muscles. Newer therapies involve selective injection of botulinum toxin into an ocular muscle to decrease its excursion.

If amblyopic stimuli are left undetected and untreated, the likelihood of normal visual function decreases. Congenital, or neonatal, amblyogenic stimuli, such as cataracts, should be detected in the first four months of life to ensure optimal

3

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visual function. Such evaluation in infants and in all children requires a visual physical exam by the clinician to assess appearance, movement, fundiscopic, and evidence of bilateral red reflexes for both eyes. Other important diagnostic signs may include head posturing, strabismus, nystagmus, abnormal pupil responses, lack of visual fixation and following, photophobia, and absent or unequal red reflexes. Parents are often excellent observers of their child's visual fixation, and their concerns should be carefully acknowledged and assessed.

VISION SCREENING AND TESTING

Formal testing of visual acuity should be possible for most children by the age of three, although some two year olds can take a test with Allen cards. Allen cards are pictographic representations of cakes, hands, birds, horses, and telephones of diminishing size linked to levels of visual acuity, and are usually the most appropriate test for younger children. Three to five-year-old children can use the Tumbling E Chart, which shows the letter E in different orientations (left, right, up, down). Children indicate which direction each E faces. For older children, the HOTV System (which requires that children recognize those four letters) or the standard Snellen chart may be employed. The difficulty of accurate visual assessment with these conventional methods has inspired new vision screening technologies, which will be discussed later in this paper.

The average visual acuity for a three to four year old is 20/40 in each eye, 20/30 for 5–6 year olds, and 20/20 for school age children. A significant difference between vision in each eye is a reason for ophthalmologic referral, even if one eye demonstrates vision of 20/20.

Although systems exist to perform vision screening in the pediatric office, the inherent chaos of the setting and the time the test takes (average: five minutes) may lead to failed or incomplete screening. The visual exam should be undertaken in an appropriately lighted room, or corridor, with the visual screening tool placed at the appropriate distance and height for the patient. Also, the environment should be relatively free from background noise and distraction. The examiner should be patient and experienced. Vision should be tested for both eyes, both simultaneously and individually.

The examiner should make a notation if a child squints, attempts to change position, or attempts to remove occluder paddles or a hand from the eye not being tested. A personal vignette: I have a patient in my practice who had an eye removed shortly after birth because of a retinoblastoma, and who has a prosthesis in one orbit. She managed to pass her exam for binocular vision in my office through some exceptional peering around her occluder. Any child who fails a vision-screening exam should be rescreened within six months. Any child who fails a rescreening should be referred to an ophthalmologist. Documentation of repeated attempts to screen a child with a notation that the child is “uncooperative” is clinically unacceptable. It should be assumed that the child who is labeled as repeatedly uncooperative cannot see.

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CONSIDERATIONS

Whenever a healthcare provider, facility, or system considers the introduction of a screening modality, the following issues should be considered:

- Determine if the condition being screened for is important to a patient's well being
- Consider whether the patient population considers the issue to be important
- Determine if adequate information exists to affect outcome if a patient is screened in a timely manner
- Determine if the condition has a latent phase that makes detection possible
- Consider whether or not we understand the natural history of the illness
- Consider whether or not there is an acceptable treatment for the illness
- Determine what a suitable test for the illness or condition might be
- Determine if the screening test is acceptable to the population to be screened
- Arrange for careful follow-ups of abnormal screens
- Establish policies about whom to treat
- Assure funding for the entire process

Although this list of considerations most clearly applies to a governmental agency or healthcare system initiating a health-care–screening program, individual providers can learn from it. Of particular interest is item #7. A “suitable test” should follow the following criteria: a test must have sufficient validity that it should be sensitive (i.e. that it correctly identifies those patients with the problem) and specific (i.e. that it successfully identifies those individuals without the condition). In fact, an eye chart is not sensitive to refractive amblyopic risk factors: it has only 27% sensitivity for three year olds and 45% sensitivity for five year olds.

5

With this in mind, one can create a grid with four possible outcomes: True Positives, True Negatives, False Positives, and False Negatives.

- True Positive. The patient is correctly identified with the illness being screened for; the patient is treated; and the family is grateful to the pediatrician for her diligence
- True Negative: The parent is relieved at the time of the encounter with the pediatrician that the child does not have the illness being screened for.
- False Positive: The child is referred for further evaluation. Although the family may be inconvenienced, they are reassured that their child is well – no foul, no penalty.
- False Negative: This one is a problem at several levels, and the Orinda study showed that the use of Snellen acuity tests in school age children resulted in 21% false negatives. A parent is reassured that their child does *not* have a condition when he actually does have that condition. Such false reassurance can lead to ignoring of subsequent symptoms (“the pediatrician said everything was fine”), delay in definitive diagnosis and treatment, and an impaired outcome. This scenario is a potential recipe for disaster for all of the interested parties.

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TECHNOLOGICAL ADVANCES

In response to the difficulty of visual assessment in children, technologies have evolved to augment the ability of the clinician to assess the visual axis. Because of cost and logistical issues, pediatricians have been reluctant to adopt photo screeners, which can involve the use of instant picture technology and off-site assessment. Newer technologies,

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however, represent an advance and opportunity not previously available to the pediatric clinician. Automated vision screening devices, such as the Welch Allyn SureSight™ Vision Screener, are an interesting addition to the diagnostic tool kit. Although such devices do not directly measure visual acuity, they assess the actual optics of the eye to demonstrate the existence of refractive error. The measurements include an assessment of the power, or sphere of the

eye, with a negative S calculation indicating myopia and a positive calculation hyperopia. The cylinder (C) parameter is a measure of astigmatism or irregularity of focus. The final measure known as the difference (D) is a calculation of the mean spherical power between the two eyes. We currently use visual acuity to evaluate visual dysfunction. Automated technology allows clinicians to strike at the root of the problem rather than its manifestations.

REIMBURSEMENT

The American Academy of Pediatrics Committee on Coding and Reimbursement finally obtained approval, in 2000, for the use of a vision screening code as part of the preventative services exam. This code, 99173 (screening test of visual acuity, quantitative, bilateral), should not be used if the patient's chief complaint at the time of a visit is related to a visual problem. In that case, the appropriate E&M code is utilized to describe the service, and the visual assessment is bundled into the office visit code. If the patient has no visual complaint (i.e. screening is done as part of a well child exam), however, 99173 is appropriate. Reimbursement for this code has been spotty, but current data suggests that it is improving. The ophthalmology section of CPT 2002 Manual contains the code for determination of a refractive state, 92015. There is no specific prohibition in CPT against the use of this code by a pediatric generalist when using a device such as the SureSight™ device. Again, negotiation with regional HMOs to obtain their acquiescence to provide coverage for a previously uncovered service or as an acknowledgement of an improvement and advancement in the profession is an important tactical step.

PROFESSIONAL LIABILITY

Although healthcare professionals should always be motivated by desire to care for their patients based on sound scientific principles and evidence-based practice parameters, we need also to acknowledge the risks inherent in failure to perform at levels expected by our patients or professional organizations. The Periodicity Schedule of the AAP is held out as a minimum frequency visit guideline, with an acknowledgement that some patients will require more frequent

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and the Pediatric Community**

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assessments as part of their preventative services. Within the context of each visit, insurers and patients expect certain historical, physical, and assessment interventions to be successfully completed. Some assessments of the time necessary to accomplish all of the necessary interventions at each well encounter suggest a minimum time of approximately 45 minutes. Since the average well encounter lasts for 15–20 minutes, office systems that operate efficiently and in a reproducible fashion are critical to ensure the health and safety of our patients.

With the overall health of America’s children improving, and with the expected continued disappearance of traditional health scourges, the focus of pediatric practice is shifting to concerns related to developmental practice and expectations. Failure to diagnose a variety of pediatric conditions is now the #1 cause of allegations of medical negligence against clinicians who care for children. Within that category, failure to diagnose visual impairment is ranked #4, after failure to diagnose meningitis, appendicitis, and hip dysplasia. Because clinicians have multiple, potential opportunities to diagnose visual dysfunction in children, failure to do so may particularly anger families. When patients appear for what is euphemistically described as “well baby care,” they often hear: “Your baby is doing great.”

It is hard for families to accept that a child’s potential is limited even when there has been advanced warning or when the change is abrupt or linked to a specific event. When parents think the clinical situation was preventable and merely went unnoticed or misdiagnosed, the problems for the clinician are significantly multiplied. Although visual screening and assessment are expected as part of the well child experience, they are not universally documented if performed and, in fact, current data suggests that such evaluations are not being universally performed.

From 1990 through 1998, I was a member of the AAP’s COPAM and chaired that committee during the period when the vision screening recommendation was changed to an objective assessment for children beginning at age three years. The committee’s deliberations were, and are, based on the belief that the pediatric community was ready, willing, and able to evaluate, assess, and refer children appropriately with visual needs. I believe that that statement is true but, more importantly, it is also the belief of our Academy of Pediatrics, the American Academy of Ophthalmology, the American Academy of Family Practice, and a variety of governmental and advocacy organizations committed to the prevention of blindness.

If you are successfully and appropriately screening your patients for visual problems, I salute your commitment to the health and well being of patients in your care. If you are not, I would urge you to do so for the benefit not only of your patients, but also of you and your practice.

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Dr. Peter Rappo is a practicing pediatrician who is an assistant clinical professor of pediatrics at Harvard University School of Medicine. He has a particular interest in the care of children with special health care needs and serves as the head of the Beansprout pediatrics team. The research firm Woodward/White recently named Dr. Rappo one of the top doctors in America.



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Academy Lauds Identification of High-Performing Screeners In Vision in Preschoolers (VIP) Study

April 1, 2004

SAN FRANCISCO—The American Academy of Ophthalmology praised the first phase findings of a study on preschool vision screening for identifying four screeners that significantly outperform other options in accurately detecting amblyopia, refractive error and other targeted eye disorders.

The study, published in the *Ophthalmology Journal* today, noted that “a striking finding of this initial phase...is the accuracy of NCR (noncycloplegic retinoscopy), the Retinomax Autorefractor, and SureSight Vision Screener in detecting children who have more than one targeted condition, as well as the most severe of these conditions...in the hierarchy of targeted disorders.” Although Lea Symbols did not perform as well on targeted disorders, it was ranked with the other three in terms of overall superiority.

The Vision in Preschoolers (VIP) Study Group assessed four conditions: amblyopia, strabismus, significant refractive error and unexplained reduced visual acuity. It was conducted with 2,588 children in Head Start programs in Berkeley, CA, Boston, MA, Columbus, OH, Philadelphia, PA and Tahlequah, OK. The 3- to 5-year-old children were selected to over-represent children with vision problems.

A spokesperson and immediate past president of the American Academy of Ophthalmology found the information on screening efficacy especially useful for screening practitioners.

“It’s very helpful to know which screening techniques performed well and we applaud the National Eye Institute (NEI) for its role in evaluating the different screening processes,” said Michael Redmond, M.D.

“This is the first study to compare traditional screening approaches with more highly advanced technology. It’s clear that ongoing research is needed to identify the strengths of different approaches for different audiences and to evaluate the cost to benefit ratio, but this illustrates the folly of mandating one technique of identifying children with eye problems over all others,” said Redmond.

“If we straitjacket professionals with government mandates on screenings and exams, we’ll hamper the development and use of new techniques that could improve efficacy.”

The study evaluated three criteria for success with preschool children: testability, or the ability to administer the tests to high numbers of three to five-year olds; sensitivity, or the ability to correctly identify a high proportion of children with vision disorder; and specificity, or the ability to correctly identify as normal a high proportion of children with no eye disorder.

When the standard for specificity was set at 90 percent, there were four techniques with similar sensitivity: the noncycloplegic retinoscopy (NCR), the Retinomax Autorefractor, SureSight Vision Screener and Lea Symbols. Sensitivities of the

Random Dot E stereoacuity and Stereo Smile II tests were similar to each other and lower than the sensitivities of the first four, while the cover-uncover test had very low sensitivity (16 percent) but very high specificity (98 percent).

Pediatric ophthalmologist Mary Louise Collins, M.D. felt that the study bodes well for advances in eye screening. "This study shows that with the appropriate commitment to research and development, we are headed toward producing a variety of highly accurate techniques for vision screening."

The VIP Study design incorporates a phased approach, with Phase I evaluating screening efficacy when administered to a select population by eye care professionals. Phase II will evaluate test results when administered to a select population by pediatric nurses and lay people, and Phase III will monitor screening results for the general population in a realistic screening setting.

Dr. Redmond cautioned policy makers to consider cost as they review the study findings. "This study is unparalleled in its efficacy value and studies like it are an important step in identifying the best screening methods to accomplish our goal of screening all children before kindergarten, but it didn't address cost. And you simply cannot influence policy without that critical element."

The Academy position is that regular pediatric care should detect significant eye disease well before pre-school. In fact, most ophthalmic and pediatric professionals recommend primary care screening from birth to three years of age. The Policy Statement on Vision Screening by the Academy, the American Association for Pediatric Ophthalmology and Strabismus, and the American Academy of Pediatrics recommends rigorous vision screening during the preschool years because early detection of treatable eye disease can have far reaching implications for vision and, in some cases, general health care.

"Screenings aren't perfect and we want to improve what we're doing," said Redmond. "But even full comprehensive eye exams can miss some disorders. Society's goal is to find a balance of efficacy and cost-efficiency that leaves some funding for care after a problem is diagnosed."

"With some follow-up cost analysis, this study should direct eye care professionals to screening techniques with an efficacy for cost ratio that leaves adequate funding for treatment of the disorders themselves—many of which are detected well before the pre-K screenings."

Dr. Redmond is a pediatric ophthalmologist and currently practices in a multi-specialty group practice in Pensacola, Fla. Dr. Collins practices at the Greater Baltimore Medical Center and is active in both the Academy and the American Association for Pediatric Ophthalmology and Strabismus.