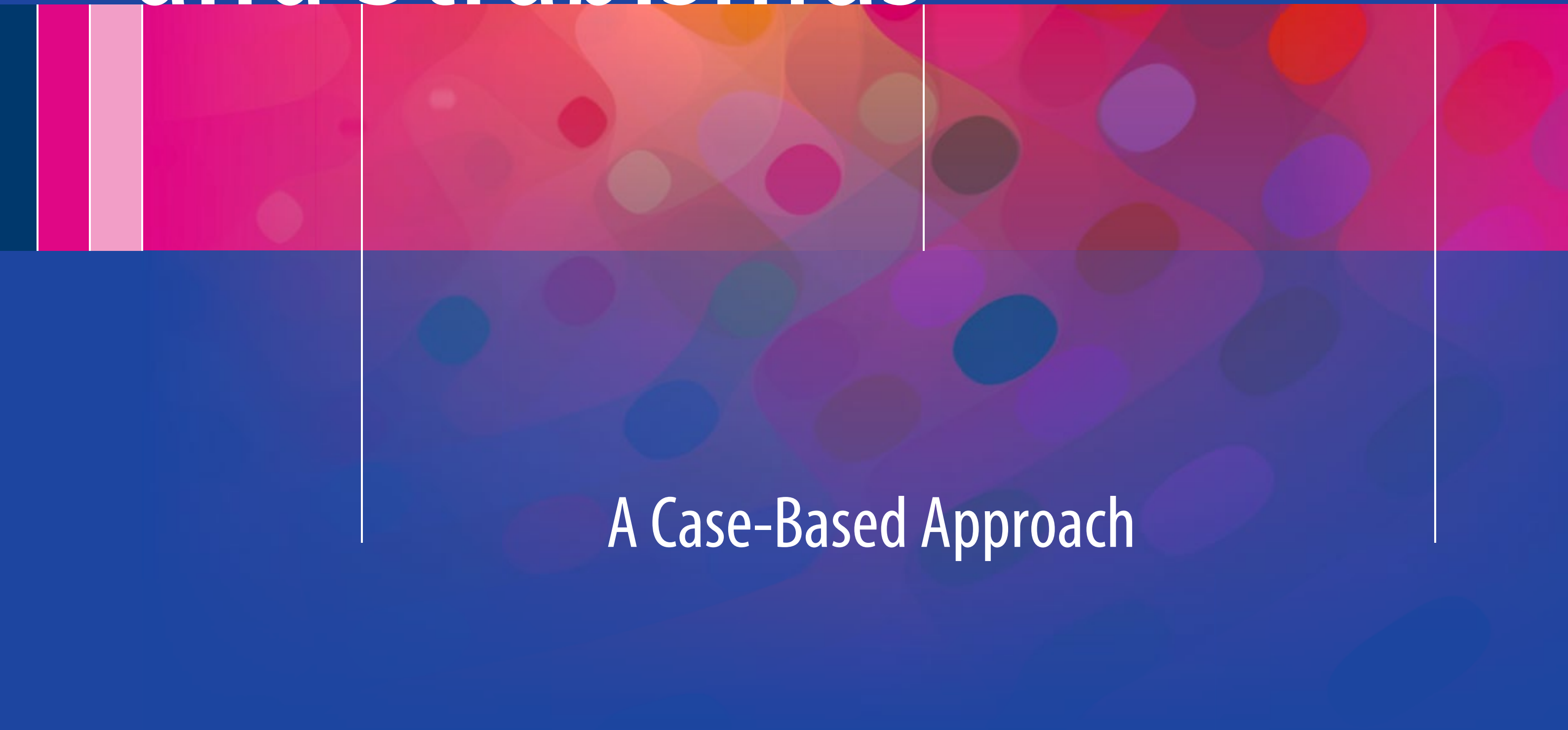
Elias I. Traboulsi
Virginia Miraldi Utz
Editors

Practical Management of Pediatric Ocular Disorders and Strabismus



A Case-Based Approach

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Editors
Elias I. Traboulsi, MD, M.Ed
Professor of Ophthalmology
Cleveland Clinic
Cole Eye Institute
Cleveland, Ohio, USA

Virginia Miraldi Utz, MD
Assistant Professor of Pediatric Ophthalmology
Cincinnati Children's Hospital Medical Center
Abrahamson Pediatric Eye Institute
Cincinnati, Ohio, USA

Assistant Editor
Michelle M. Ariss, MD
Assistant Professor of Ophthalmology
Children's Mercy Hospitals & Clinics
Department of Ophthalmology
Kansas City, MO, USA

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To Mayya, Alex, and Nadeem, loves of my life. To Marshall Parks, mentor and friend.

Elias I. Traboulsi

To my parents, David and Leslee Miraldi, and my husband, Christopher, and son, Brendan David, for their unending love and encouragement.

To my mentor and friend, Elias Traboulsi, for his inspiration and continuing support throughout my career.

Virginia Miraldi Utz

Foreword

With the advent of the internet and effective search engines, there are few questions pertaining to pediatric ocular disorders, or most things for that matter, that cannot be answered by conducting a careful search online. This phenomenon is relatively new and expanding. The most widely used search engine, Google, started on September 4, 1998. As a generation, this makes Google a "millennial" and not so old—just entering adulthood.

A person wishing to learn more about a subject or condition as it relates to pediatric ophthalmology can find it online. Asking the right question while using the best search engine allows you to access information about diagnosis and treatment of fourth nerve palsy, management of cataract in a child, persistent fetal vasculature, and just about anything else you might want to find related to the subject in question. For example, more than 230 papers, parts of book chapters, and isolated comments pertaining to hyphema in a child can be accessed now. When you read this, there are likely to be few more or less, but there will be many. Some will offer gems and some will not. But all will have in common the need to validate the information you find and the time necessary to find the information you need to validate. Stated another way, "Am I asking the right question and is the information I am finding credible?" The need to even ask questions like these is eliminated by a book like *Practical Management of Pediatric Ocular Disorders and Strabismus.*

Pertinent material about disorders of the child's eye is covered in 15 sections containing 73 chapters and is followed by 13 appendices. In the case of this book, the editors asked the right questions for you. Then they chose qualified authors to sort through the material available, call on their own experience, and in many cases work with colleagues and mentors to put the best material available in a reader-friendly format. The authority commanded by this book relies on the careful editing of the senior editors, the conscientious efforts of more than two scores of junior authors and more than a dozen senior authors, and the unsung efforts of mentors known and unknown who have supported the work that went into comprehensive work.

In more than 50 years as a pediatric ophthalmologist, I have seen many changes. Knowledge has expanded greatly, diagnoses have become more accurate, and treatments have improved making better results possible. During this time, dozens of new books have been written dealing with pediatric ophthalmology, and that number includes a few that I bear responsibility for. I commend all who have worked diligently to make the latest information available to as many as they can reach. *Practical Management of Pediatric Ocular Disorders and Strabismus*, a comprehensive, authoritative, and updated book worthy of your attention, deserves a prominent place in the pantheon of literature on the subject of children's eye disease.

Eugene M. Helveston, MD
Department of Pediatric Ophthalmology, Indiana University
Medical Center
Indianapolis, IN, USA

Preface

This project stemmed from a desire to provide an educational resource for residents, fellows, and ophthalmologists that is practical and focused on management of the child with an eye problem or the adult with strabismus.

The book was not designed to be a comprehensive textbook on pediatric ophthalmology, but rather one that is replete with cases, diagrams, and tables that recapitulate the way we teach trainees or handle particular clinical situations. We tried to focus specifically on clinical pearls, an algorithmic approach to differential diagnoses, current evidence for therapy, or preferred practice patterns for diagnosis and treatment followed by case examples to reinforce the material. We gave the authors some discretion in terms of formatting, but we encouraged them to provide diagnostic/treatment algorithms and tables to supplement text content and to enhance its practical value. Every chapter was subjected to at least two and often several rounds of reviews and modifications before it reached its final and current state.

We believe that the reader will find the contents of help in their daily training and clinical practice. We are grateful to our families for their patience during the gestation of this book, to our patients for inspiring us to improve on our skills to take care of them, and to our residents and fellows for teaching us how to become better teachers.

Cleveland, OH

Elias I. Traboulsi
Virginia Miraldi Utz

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Contributors

Ashley Abraham, MD Department of Ophthalmology, University of Missouri - Kansas City School of Medicine, Kansas City, MO, USA

Chrysavgi Adamopoulou, MD Department of Pediatric Ophthalmology, Metrohealth Medical Center, Cleveland, OH, USA

Fadia AlKhawaldeh, MBA, COT, ROUB Department of Refractive Surgery, Cleveland Clinic Cole Eye Institute, Cleveland, OH, USA

Luz Amaro-Quireza, OD Department of Ophthalmology, Columbia University Medical Center, New York, NY, USA

Michelle M. Ariss, MD Department of Ophthalmology, Children's Mercy Hospitals & Clinics, Kansas City, MO, USA

Hassan A. Aziz, MD Department of Ophthalmology, Cleveland Clinic, Cole Eye Institute, Cleveland, OH, USA

Allison E. Babiuch, MD Cleveland Clinic, Cole Eye Institute, Cleveland, OH, USA

Reecha S. Bahl, MD Department of Ophthalmology—Kresge Eye Institute, Wayne State University School of Medicine, Detroit, MI, USA

M. Tariq Bhatti, MD Department of Neuro-ophthalmology, Neurology, Duke Eye Center, Duke Medical Center, Durham, NC, USA

Charline S. Boente, MD, MS Department of Ophthalmology and Visual Sciences, University Hospitals Eye Institute, Cleveland, OH, USA

James D. Bowsher, MD Department of Ophthalmology, Indiana University School of Medicine, Eugene and Marilyn Glick Eye Institute, Indianapolis, IN, USA

Yasmin S. Bradfield, MD Department of Ophthalmology and Visual Sciences, University of Wisconsin Hospital and Clinics, Madison, WI, USA

Scott E. Brodie, MD, PhD Department of Ophthalmology, Mount Sinai Medical Center, Mount Sinai Hospital, New York, NY, USA

Emily Broxterman, MD Department of Ophthalmology, University of Kansas, Kansas City, MO, USA

Department of Pediatric Ophthalmology, Children's Mercy Hospital, Broadway, Kansas City, MO, USA

Paola Carrai, MD San Giuseppe Hospital, University Eye Clinic, Milan, Italy

Kara M. Cavuoto, MD Bascom Palmer Eye Institute, Miami, FL, USA

Maria Choudhary, MD Department of Internal Medicine, Cleveland Clinic, Cleveland, OH, USA

Rao Chundury, MD, MBA Department of Ophthalmology, Cleveland Clinic Cole Eye Institute, Cleveland, OH, USA

Heather L. Cimino, OD Department of Optometry, Cleveland Clinic Cole Eye Institute, Cleveland, OH, USA

Bryan R. Costin, MD Department of Ophthalmology, The Cole Eye Institute, Cleveland Clinic, Cleveland, OH, USA

Brita S. Deacon, MD Department of Ophthalmology, Arkansas Children's Hospital, Little Rock, AK, USA

Monte A. Del Monte, MD Department of Ophthalmology, Kellogg Eye Center, University of Michigan, Ann Arbor, MI, USA

Sonia B. Dhoot, MD Department of Ophthalmology, Sabates Eye Centers, Leawood, KS, USA

Eniolami O. Dosunmu, MD Department of Ophthalmology, University of Cincinnati College of Medicine, Cincinnati, OH, USA

Jared E. Duncan, MD Department of Ophthalmology, Duke Eye Center, Duke University Medical Center, Durham, NC, USA

William J. Dupps Jr. , MD, PhD Cleveland Clinic, Cole Eye Institute, Cleveland, OH, USA

Marina A. Eisenberg, MD Department of Ophthalmology, Kellogg Eye Center, University of Michigan, Ann Arbor, MI, USA

Mays Antoine El-Dairi, MD Department of Pediatric Ophthalmology and Strabismus, Neuro-ophthalmology, Duke Eye Center, Durham, NC, USA

Forrest J. Ellis, MD Inova Fairfax Hospital for Children, Northern Virginia Ophthalmology Associates, Falls Church, VA, USA

George S. Ellis Jr. , MD Tulane University Medical School, New Orleans, LA, USA

Louisiana State University Health Science Center, New Orleans, LA, USA

Sarah M. Escott, MD Department of Ophthalmology, Northwestern Memorial Hospital Feinberg School of Medicine, Chicago, IL, USA

Jill Foster, MD, FACS Department of Oculoplastics, The Ohio State University, Eye Center of Columbus, The Eye Center, Columbus, OH, USA

Sharon F. Freedman, MD Department of Ophthalmology, Duke Eye Center, Duke University Medical Center, Durham, NC, USA

Christopher Gappy, MD Department of Ophthalmology, Kellogg Eye Center, University of Michigan, Ann Arbor, MI, USA

Fatema F. Ghasia, MD Department of Ophthalmology, Cleveland Clinic Lerner College of Medicine of Case Western University, Cleveland, OH, USA

Debra A. Goldstein, MD Department of Ophthalmology, Northwestern Memorial Hospital Feinberg School of Medicine, Chicago, IL, USA

Michael Eric Gray, MD Abrahamson Pediatric Eye Institute, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA

Department of Ophthalmology, University of Cincinnati College of Medicine, Cincinnati, OH, USA

Laura L. Hanson, MD Department of Ophthalmology, University of Cincinnati College of Medicine, Cincinnati, OH, USA

Gena Heidary, MD, PhD Department of Ophthalmology, Boston Children's Hospital, Boston, MA, USA

Christopher T. Hood, MD W.K. Kellogg Eye Center, University of Michigan, Ann Arbor, MI, USA

Timothy E. Hug, OD Department of Ophthalmology, Children's Mercy Hospital, Kansas City, MO, USA

Mohamad S. Jaafar, MD Department of Ophthalmology, Children's National Health System, Washington, DC, USA

Viral Juthani, MD Department of Ophthalmology, Cole Eye Institute, Cleveland Clinic, Cleveland, OH, USA

Aaron R. Kaufman, BA School of Medicine, Boston University, Boston, MA, USA

Adam H. Kaufman, MD, FACS Cincinnati Eye Institute, Department of Ophthalmology, University of Cincinnati, Cincinnati, OH, USA

Natalie C. Kerr, MD Department of Ophthalmology, LeBonheur Children's Hospital, Hamilton Eye Institute, Memphis, TN, USA

Terry Kim, MD Department of Ophthalmology, Duke University, Durham, NC, USA

Greg Kosmorsky, DO Department of Ophthalmology, Cleveland Clinic, Cole Eye Institute, Cleveland, OH, USA

Jessica J. Kovarik, MD Department of Ophthalmology, Indiana University School of Medicine, Eugene and Marilyn Glick Eye Institute, Indianapolis, IN, USA

Courtney L. Kraus, MD Johns Hopkins Hospital Wilmer Eye Institute, Baltimore, MD, USA

Priyanka Kumar, MD Cleveland Clinic Foundation, Cole Eye Institute, Cleveland, OH, USA

G. Robert LaRoche, MD, FRCSC Department of Ophthalmology and Visual Sciences, Dalhousie University, Halifax, NS, Canada

Jason Lee, OD Cincinnati Children's Hospital Medical Center, Abrahamson Pediatric Eye Institute, Cincinnati, OH, USA

James Lin, MD Department of Ophthalmology, New York Presbyterian-Columbia, New York, NY, USA

Timothy P. Lindquist, MD Durie Vision, Overland Park, KS, USA

Grant T. Liu, MD Department of Ophthalmology, Children's Hospital of Philadelphia, Philadelphia, PA, USA

Sarah L. Lopper, OD Department of Ophthalmology, University of Cincinnati College of Medicine, Cincinnati, OH, USA

Careen Lowder, MD, PhD Cole Eye Institute, Cleveland Clinic, Cleveland, OH, USA

Kelly Lusk, PhD, CLVT Department of Ophthalmology, Cincinnati Children's Hospital Medical Center, University of Cincinnati School of Medicine, Cincinnati, OH, USA

Julia F. Malalis, MD Department of Ophthalmology, Northwestern Memorial Hospital Feinberg School of Medicine, Chicago, IL, USA

Andreas Marcotty, MD Cleveland Clinic, Cole Eye Institute, Beachwood, OH, USA

Meghan J. Marino, MS, LGC Department of Ophthalmic Research, Cleveland Clinic, Cleveland, OH, USA

Shana E. McCormack, MD Department of Pediatrics, Division of Endocrinology and Diabetes, Children's Hospital of Philadelphia, Philadelphia, PA, USA

Mary Lou McGregor, MD Department of Ophthalmology, The Ohio State University Wexner Medical Center, Columbus, OH, USA

Department of Ophthalmology, Nationwide Children's Hospital, Columbus, OH, USA

Craig A. McKeown, MD Bascom Palmer Eye Institute, Miami, FL, USA

Carlos A. Medina, MD Bascom Palmer Eye Institute, Miami, FL, USA

Mary E. Moffatt, MD Department of Pediatrics, Children's Mercy Hospital, Kansas City, MO, USA

Lileth Mondok, MD Department of Pediatric Neurology, Cleveland Clinic Foundation, Cleveland, OH, USA

W. Walker Motley, III, MS, MD Abrahamson Pediatric Eye Institute, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA

Department of Ophthalmology, University of Cincinnati College of Medicine, Cincinnati, OH, USA

Cameron Nabavi, MD The Eye Center of Columbus, The Ohio State University, Nationwide Children's Hospital, Columbus, OH, USA

Paolo Nucci, MD San Giuseppe Hospital, University Eye Clinic, Milan, Italy

Scott E. Olitsky, MD Department of Ophthalmology, University of Missouri—Kansas City School of Medicine, Children's Mercy Hospital, Kansas City, MO, USA

E. Mitchel Opremcak, MD Department of Ophthalmology, The Ohio State University, Columbus, OH, USA

Faruk H. Örge, MD Department of Ophthalmology and Visual Sciences, Case Western Reserve University School of Medicine, Rainbow Babies and Children's Hospital, University Hospitals Eye Institute, Cleveland, OH, USA

Grace L. Paley, PhD Department of Ophthalmology, Children's Hospital of Philadelphia, Hospital of the University of Pennsylvania, Philadelphia, PA, USA

Cajal K. Patel, MD Department of Ophthalmology and Health Sciences, University of Wisconsin–Madison, Madison, WI, USA

Julian D. Perry, MD Department of Ophthalmology, Cleveland Clinic Cole Eye Institute, Cleveland, OH, USA

Francesco Pichi, MD San Giuseppe Hospital, University Eye Clinic, Milan, Italy

Jennifer B. Pierce, MD Department of Pediatrics, University of Missouri-Kansas City, Children's Mercy Hospital, Kansas City, MO, USA

Patricia Ann Ple-plakon, MD Department of Ophthalmology and Visual Sciences, University of Michigan Kellogg Eye Center, Ann Arbor, MI, USA

Cindy Pritchard, CO Department of Ophthalmology, Children's Hospital of New Orleans, New Orleans, LA, USA

Rachel E. Reem, MD Department of Ophthalmology, Nationwide Children's Hospital, Columbus, OH, USA

Department of Ophthalmology, The Ohio State University Wexner Medical Center, Columbus, OH, USA

Johane Robitaille, MDCM IWK Health Centre, Ophthalmology and Visual Sciences, Halifax, NS, Canada

Daniele P. Saltarelli, OD Abrahamson Pediatric Eye Institute, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA

Department of Ophthalmology, University of Cincinnati College of Medicine, Cincinnati, OH, USA

Melanie A. Schmitt, MD Pediatric Ophthalmology, Cleveland Clinic Foundation/Cole Eye Institute, Cleveland, OH, USA

Terry L. Schwartz, MD Abrahamson Pediatric Eye Institute, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA

Department of Ophthalmology, University of Cincinnati College of Medicine, Cincinnati, OH,USA

Jonathan Eliot Sears, MD Cleveland Clinic, Cole Eye Institute, Cleveland, OH, USA

Nathanial Clarks Sears, MD Cleveland Clinic, Cole Eye Institute, Cleveland, OH, USA

Hande Taylan Şekeroğlu, MD Department of Ophthalmology, Hacettepe Universitesi Tip Fakultesi Goz Hastaliklari Anabilim Dali B Kati, Sihhiye, Ankara, Turkey

Emin Cumhur Sener, MD Private Practice, Ankara, Turkey

Claire A. Sheldon, MD, PhD Department of Ophthalmology, Children's Hospital of Philadelphia, Philadelphia, PA, USA

Christine Shieh, MD Department of Ophthalmology, Duke University, Durham, NC, USA

Arun D. Singh, MD Cleveland Clinic, Cole Eye Institute, Cleveland, OH, USA

Robert Andrew Sisk, MD Department of Vitreoretinal Surgery, University of Cincinnati, Cincinnati Eye Institute, Cincinnati Children's Hospital Medical Center, Cincinnati Eye Institute, Cincinnati, OH, USA

Alison E. Smith, MD Greenville Hospital System: University of South Carolina School of Medicine-Greenville, Greenville, SC, USA

Oriel Spierer, MD Bascom Palmer Eye Institute, Miami, FL, USA

Derek T. Sprunger, MD Department of Ophthalmology, Eugene and Marilyn Glick Eye Institute, Indiana University School of Medicine, Indianapolis, IN, USA

Erin D. Stahl, MD Department of Ophthalmology, Children's Mercy Hospital, Kansas City, MO, USA

Steffany M. Straight, MD Department of Ophthalmology, Munson Healthcare Cadillac Hospital, Cadillac, MI, USA

Daniel Straka, MD The Ohio State University, Eye Center of Columbus, Nationwide Children's Hospital, Columbus, OH, USA

Christina Szperka, MD Department of Neurology, University of Pennsylvania, Pediatric Headache Program, Children's Hospital of Philadelphia, Philadelphia, PA, USA

Elias I. Traboulsi, MD, MEd Cleveland Clinic, Cole Eye Institute, Cleveland, OH, USA

Rupal H. Trivedi, MD Department of Ophthalmology, Storm Eye Institute, Charleston, SC, USA

Stephen H. Tsang, MD, PhD Department of Ophthalmology, New York Presbyterian-Columbia, New York, NY, USA

Virginia Miraldi Utz, MD Abrahamson Pediatric Eye Institute, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA

Department of Ophthalmology, University of Cincinnati College of Medicine, Cincinnati, OH, USA

Gonzalo Vike Vicente, MD Georgetown University Hospital, Washington, DC, USA

Chevy Chase, MD, USA

Palak B. Wall, MD Pediatric Ophthalmology and Strabismus, Lexington, KY, USA

Sarah B. Wierda, BA, MD Pediatric Ophthalmology Associates, Omaha, NE, USA

M. Edward Wilson, MD Department of Ophthalmology, Storm Eye Institute, Charleston, SC, USA

Kathryn P. Winkler, MD Department of Ophthalmology, Kresge Eye Institute, Detroit, MI, USA

Michael B. Yang, MD Abrahamson Pediatric Eye Institute, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA

Department of Ophthalmology, University of Cincinnati College of Medicine, Cincinnati, OH, USA

Brian G. Zamora, MD, PhD Department of Ophthalmology, University of Cincinnati College of Medicine, Cincinnati, OH, USA

Sandy Zhang-Nunes, MD KECK Hospital, LAC+USC Medical Center, USC Eye Institute, Los Angeles, CA, USA

Part I

The Pediatric Clinical Examination

Approach to Visual Acuity Assessment and Strabismus Evaluation of the Pediatric Patient

1

Cindy Pritchard and George S. Ellis Jr.

Abstract

Children present unique ophthalmologic considerations and require an examination approach that corresponds to his or her level of visual and psychosocial development. Employment of age-related, patient-specific strategies may be utilized to maximize the information obtained from the clinical examination as well as make the examination both enjoyable and rewarding for the patient, family, and practitioner. This chapter will discuss the approach to the ophthalmologic examination in pediatric patients including visual assessment, motility, strabismus, and motor fusion.

Keywords

Motility • Cover-uncover test • Cover test • Visual acuity • Strabismus • Fusion

Introduction

Examination of the pediatric patient can be very challenging for even the most experienced clinicians. This is especially true for strabismus evaluations. Many other pediatric eye conditions such as glaucoma and retinal, corneal, and optic nerve diseases can be assessed under anesthesia, if necessary. Strabismus, however, is a dynamic condition that requires a cooperative and alert subject if an accurate, detailed examination and useful data are to be obtained. Although there are obvious limitations to how much control the examiner has over the level of cooperation or state of alertness of the child, there are strategies that can enhance cooperation or state of alertness of the child, hence the quality of the exam and the ease with which information is obtained.

Approach to Examining the Pediatric Patient

Optimizing Cooperation

The most significant barrier to obtaining the child's cooperation is fear. The fearful child often closes his eyes tightly or continuously turns his face away from the examiner, not allowing instruments or the examiner's hands near his face. Conversely, the comfortable child who feels safe will often interact with the examiner, optimizing the opportunity for adequate ocular assessment. Therefore, establishing a good initial rapport is an essential element to the examination process, especially when the child is very young. This is best accomplished by maintaining a calm, pleasant demeanor. The examiner should interact/play with/entertain the child while obtaining needed information from the parents. For example, one can show the very young child a small toy that will be used later as a fixation target, talking to them about the toy and allowing them to touch it if he shows interest. With young children who are verbal, engaging them in casual

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C. Pritchard, CO (✉)
Ophthalmology Department, Children's Hospital of New Orleans,
200 Henry Clay Ave, New Orleans, LA 70118, USA
e-mail: cindyhpritchard@gmail.com

G.S. Ellis Jr., MD
Ophthalmology Department, Children's Hospital of New Orleans,
200 Henry Clay Ave, New Orleans, LA 70118, USA

Tulane University School of Medicine,
200 Henry Clay Ave, New Orleans, LA 70118, USA

Louisiana State University Health Science Center,
200 Henry Clay Ave, New Orleans, LA 70118, USA

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conversation can be disarming. One might ask them about the superhero or Disney princess on an item of clothing that they are wearing or tell them about some of the toys that the examiner will show them during the exam. If they are sitting in a parent's lap, another strategy for creating a feeling of safety is to involve the parent in the exam by having them hold the fixation target or by holding a prism or trial lens to the parent's eye to show the child that, although the examiner's hands and tools are very close to their face, they will not hurt them. When these strategies are successfully employed, the child can actually enjoy having his eyes examined and look forward to return visits.

Optimizing Alertness

Since visual attention is necessary for accurate ocular motility evaluation, the sleeping, unarousable child can be difficult or impossible to examine. This occurs more commonly with infants. When the infant cannot be suitably awakened by removing him from a comfortable lying position, the examiner can try talking to him somewhat loudly but calmly while gently moving his/her arms and legs in a playful manner. If the baby remains asleep or too drowsy to be attentive to your targets, one can use the visit as an opportunity to gather information that is available in the sedate state such as pupil reaction, retinoscopy, tonometry when appropriate, and funduscopy. The doll's head maneuver can be used to determine if ocular rotations are full. With the lids manually lifted, the examiner can rotate the infant's head side to side and up and down. The vestibular system will drive the eyes into peripheral gazes allowing the examiner to rule out any limitation of movement. For example, if an infant has a left type I Duane's syndrome, one will see that the left eye fails to fully abduct as the head is rotated to the infant's right. With an unarousable infant, the remainder of the motility exam can be obtained at a subsequent visit. The parents should be instructed to avoid an appointment at nap time and to avoid feeding the child shortly before the visit. A hungry child is more alert, but also could be more irritable. The infant can be given a bottle during the exam to keep him calm while motility testing is performed.

Additional Strategies

The examination is initiated at first sight of the child. As one talks to the parents, one watches the child for nystagmus, ptosis, head thrusting, or obvious strabismus. If the child appears to be maintaining an abnormal head posture, the examiner can casually encourage fixation away from his preferred gaze looking for nystagmus or strabismus. If the child has an obvious strabismus, the preferred fixating eye is noted. By observing the child gaze around the room, it is sometimes possible to gain useful information regarding ocular versions. For example, as the child looks to the side, one might notice a hyperdeviation suggesting oblique dysfunction. It is important to recognize that attention and cooperation might be limited to the first few minutes after meeting the child. Sometimes the period of time *before* the actual examination begins is the *only* time that the child is calm enough to make these important observations. Often the young patient remains calm until the examination begins, and attention is drawn to him. The child may become fearful, and cooperation is subsequently lost. One should begin the examination with the least threatening tests. The farther away one is from the child, the less threatened the child will feel. Remote, nonthreatening testing can begin while one is talking to the parents. For example, dry retinoscopy screening (without lenses) performed 2–3 ft away from the child will enable the practitioner to determine if the media is clear centrally, if there is any significant refractive error or anisometropia, if corneal light reflexes suggest the presence of manifest strabismus, or if there is anisocoria. The Bruckner simultaneous red reflex test can be performed by comparing the red reflexes between the two eyes as the pupils are viewed simultaneously with the direct ophthalmoscope. Any asymmetry in the color, brightness, or size of the two red reflexes indicates a potential amblyogenic condition such as media opacity, strabismus, or asymmetric refractive error. One can test versions by moving a small toy into various gazes without getting too close to the child. These strategies allow the examiner to obtain important information before the actual hands-on examination even begins. The examiner should be aware and relay to the parents that infants, very young children, and children with significant developmental delays may require more than one visit to obtain a full ocular assessment.

Vision Assessment

The method used to assess visual acuity will be dictated by the child's level of development and degree of cooperation. Prior to the child reaching the stage at which Snellen acuity can be obtained, there are several approaches to obtain an estimate or measure of visual acuity; some are quantitative, others qualitative.

Preverbal Child

Quantitative Methods

Most quantitative preliterate vision tests employ the forced-choice preferential looking method in which the child is presented a target that has gratings, forms, or contours on one

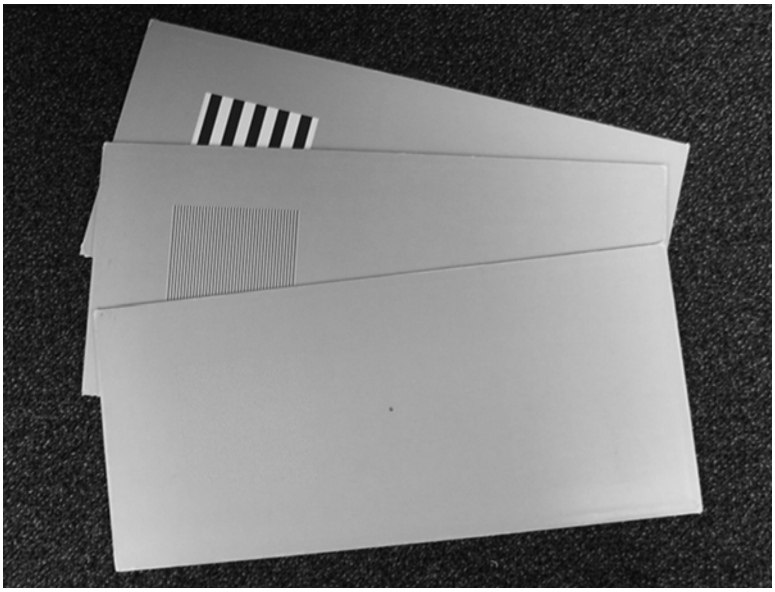


Fig. 1.1 Teller acuity cards. Sample of three Teller acuity cards with spatial frequency gratings. Sets of Teller acuity cards contain 12 cards with a gratings ranging in spatial frequency from 0.23 to 38 cycles per centimeter

half and is blank on the other half. The Teller acuity cards were the first of these tests to be fairly commonly used in clinical practice (Fig. 1.1). If the child looks toward the side with the gratings when presented the Teller acuity card, it is presumed that the child can see them. Cards with progressively higher spatial frequency gratings are presented until the child randomly looks to either side of the card, implying that the child cannot see the gratings. The grating acuity can then be translated to estimated logarithm of minimal angle of resolution (logMAR) Snellen acuity by referring to a chart provided with the test [1–3]. Testing distance varies by age: infant births to 6 months (38 cm), 7 months and 3 years (55 cm), and >3 years (84 cm). Patients with poor visual acuity may require an unusually close distance (19 and 9.5 cm). Other forced-choice preferential looking tests have subsequently been developed for quantifying infant acuity [4–6]. Visual evoked potentials have also been used to quantify infant acuity [7, 8]. With this technique, electrodes are placed over the occipital lobe, and the patient is presented with a series of bar or grid patterns. If the pattern is observed, the impulse will be recorded by the electrodes. The smallest grating pattern that stimulates an impulse is an estimate of visual acuity.

Qualitative Methods

Central, Steady, Maintained

A commonly used qualitative method of preverbal estimation of vision involves describing fixation for each eye monocularly, then comparing the subjective data between eyes. A small toy is often used as the test target. It is best to use one's hand as an occluder for monocular testing. A standard occluder is more threatening to the child and can be a distraction. The target should be slowly moved during testing, watching for appropriate pursuit eye movements and quality of fixation. Monocular fixation is described as central (C) if the child appears to be fixating with his/her fovea. If fixation is grossly eccentric in which case the child does not appear to look directly at the target with his fovea, the vision would be noted as not central (nC).

This method also assesses fixation in terms of steadiness (S) or not being steady (nS). Fixation in which the eye does not remain steadily on the target (nS), rather displays unsteady searching movements, implies poor vision. Nystagmus also prevents steady fixation on the target and may be recorded as CnS in each eye.

The third part of this qualitative test is performed under binocular conditions. The acuity in one eye is indirectly compared to that of the fellow eye. If manifest strabismus is present, this part of the test can be performed by evaluating for a fixation preference for one eye. As an example, a child with a 30^{Δ} esotropia will view the target with either the left eye or the right eye, with the fellow eye in an obvious deviated position. The examiner covers the fixing eye, forcing the child to take up fixation with the deviated fellow eye. While performing this test, the examiner can assess fixation in the previously deviated eye in terms of C or nC and S or nS. When the cover is removed, the examiner notes whether the child immediately reverts to fixation with the eye that was under cover or if the child maintains fixation with the eye that was originally the deviated eye. If the child maintains fixation with the previously deviated eye under binocular conditions, vision is recorded as maintained (M) for each eye. Therefore, a child with central and steady fixation without a preference for one eye would have acuity documented as CSM for each eye. If, however, during the binocular part of the assessment the child reverts to fixation with the eye

Table 1.1 Gradations of visual acuity using the CSM method as compared to the fellow eye

Notation	Interpretation
nCnSnM	Unsteady and eccentric or wandering fixation, very poor visual acuity; if eccentric fixation, can estimate degree of eccentricity for subsequent comparison
CnSnM	Central but unsteady fixation will not hold fixation under binocular conditions, implies poor visual acuity
CSnM	Central and steady fixation but will not continue to fixate with that eye under binocular conditions, implies visual acuity is poorer than fellow eye
CSM briefly[a]	Briefly holds fixation under binocular conditions, implies fairly good visual acuity but not as good as fellow eye, rarely vision equal to fellow eye
CSM to a blink[a]	Holds fixation under binocular conditions but reverts to preferred fellow eye following a blink, implies good visual acuity but still not equal to fellow eye
CSM through a blink[a]	Will continue to hold fixation with non-preferred eye beyond a blink, implies good visual acuity, better than CSM to a blink but still not equal to fellow eye
CSM prefers right or left eye[a]	Implies almost equal visual acuity
CSM alternates	Implies equal visual acuity

[a] Although visual acuity might be equal, implying the absence of amblyopia, in these states, the preference for fixation with one eye can lead to amblyopia in the fellow eye. Treatment might be indicated to prevent the development of amblyopia depending upon the strength of the preference and the age of the patient

under cover when the cover is removed, acuity for the non-preferred eye would be noted as not maintained (nM). A child with mild or moderate amblyopia, for example, might have central and steady fixation with the amblyopic eye but will not maintain fixation with that eye under binocular conditions because the child sees more clearly with the sound eye. Their vision would be recorded as CSM for the sound eye and as CSnM for the amblyopic eye.

When a fixation preference for one eye exists, it is useful to describe the strength of the preference. For example, when the cover is removed, the child might continue to maintain fixation briefly before returning fixation to the dominant eye. A comment such as "holds briefly" should be added to CSM for that eye. A blink can be used to further describe fixation preference. The child might maintain fixation with the non-preferred eye under binocular conditions until he/she blinks, at which point he shifts back to fixation with the dominant eye. In this case, a comment such as "holds to a blink" would be added to CSM for the nondominant eye. Another possibility is that he maintains fixation beyond a blink but eventually returns to fixating preferentially with the dominant eye. A comment would then be added "holds through a blink." By adding these comments, vision can be compared from visit to visit (Table 1.1). A child that is being treated for amblyopia might begin as CSnM, then progress with treatment to holding to a blink, later through a blink, then ideally begin to alternate fixation without a preference suggesting equal vision.

If the eyes are well aligned, a large prism can be used to simulate manifest strabismus for the "maintained" (M or nM) portion of vision assessment. For example, the 20^{Δ} base-down prism test involves placing a 20^{Δ} prism base down in front of one eye, optically inducing vertical strabismus. This will force the child to fixate with one or the other eye. If the child's eyes appear to be viewing above the target with the prism in place, he is fixating with the eye in front of which the prim is being held. If, however, the eyes appear to be looking straight at the target, the child is viewing with the eye that does not have the prism in front of it (Fig. 1.2). One might see vertical saccades equal in amplitude to the size of prism used. When this occurs, the child is freely alternating between eyes suggesting equal vision. In this case, each eye would be recorded as CSM. If a preference for fixation for one eye is detected with the prism in place, the examiner can proceed to determination of the strength of the preference with the prism-induced simulated strabismus in the same manner as described above for the child with true manifest strabismus.

A significant limitation to the CSM method of vision assessment is that central and steady fixation does not necessarily equate with normal vision. For example, even an uncorrected high myope has central and steady fixation. The strength of this test, however, is that it is somewhat reliable in detecting amblyopia [9–11]. Most conditions that limit acuity can be detected without subjective responses (refractive errors, media opacities, retinal, and optic nerve diseases). On the other hand, although *amblyogenic* conditions can be detected objectively, amblyopia itself is diagnosed by either subjective response or, in preverbal children, by comparing vision between the two eyes during the "M" portion of CSM testing. If one eye is preferred over the fellow eye, it implies unequal vision. If the disparity in vision cannot be explained by findings on the physical exam, amblyopia should be suspected and treated.

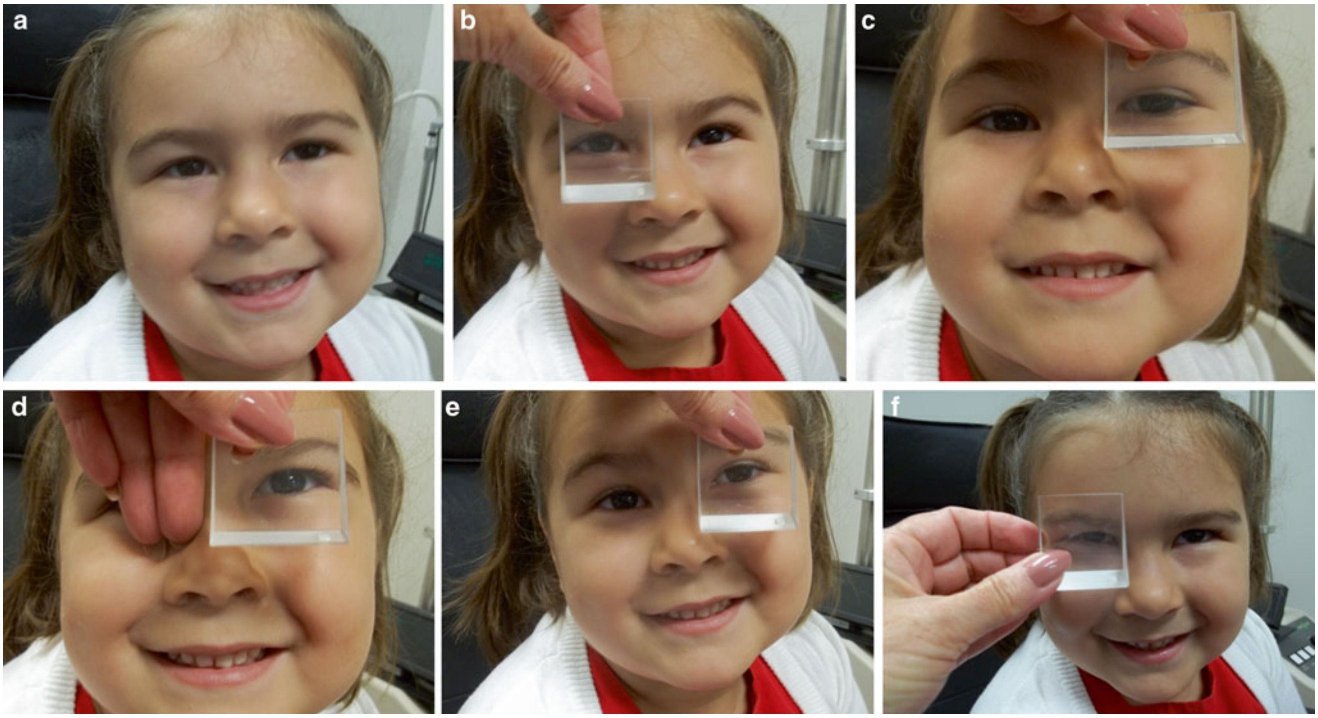


Fig. 1.2 20Δ base-down prism assessment of vision. (**a**) Child without strabismus has central and steady fixation in both eyes. (**b**) 20Δ base-down prism test is employed to compare visual function between eyes. With prism over right eye, visual axes shift up indicating child is fixating through prism with right eye. (**c**) With prism over left eye, no shift in visual axes indicates child continues to fixate with right eye. (**d**) With right eye covered, child fixates through prism with left eye. (**e**) With right eye uncovered, child returns to right eye fixation. (**f**) Prism placed again over right eye. Upward shift of visual axes confirms right eye preference. Outcome of test reveals vision as central, steady, and maintained (CSM) right eye and central, steady, non-maintained (CSnM) left eye

Poor Vision/Visual Attention

A child with poor vision or poor visual attention might not fixate on any target. In this case, it can be useful to test for optokinetic nystagmus (OKN) using an OKN drum. The OKN response consists of a series of reflexive eye movements induced by a repetitive pattern of lines or objects moving across the frontal field of vision (slow phase or pursuit movements and reflex saccadic movements in the opposite direction). If an OKN response is observed, this indicates that the vision is probably better than 20/400. Importantly, pursuit movements do not develop until approximately 3–6 months of age. If there is no response to the OKN drum, infants can be held up by the examiner and rotated, looking for an OKN response with the exam room serving as a full-field OKN stimulus. If vision is present, a few beats of OKN will likely be observed. If, however, the child has extremely poor vision, there will be an absence of optokinetic nystagmus. The vestibular system will cause the eyes to move in the opposite direction that the child is being rotated. For example, if a child with extremely poor vision is being rotated clockwise, the eyes will drift to the child's right and remain in right gaze until rotation is stopped or direction of rotation changes. One can also test for suppression of the vestibular input by attempting to get the child to look at the examiner's face while being rotated. Suppression of the vestibular ocular reflex suggests that the child does have some vision. A child can also be tested for light perception by looking for any reaction to changes in room lights or reaction to a bright light source such as that used for indirect ophthalmoscopy.

Preliterate Verbal Child

The most commonly used preliterate vision charts are the E game, LEA symbols, HOTV chart, Allen figures, and Landolt rings. These charts either require the child to verbalize or to indicate a response and to understand verbal instructions.

The distance HOTV chart has the letters H, O, T, and V as the optotypes and requires the child to understand the concept of matching. A card with large print letters H, O, T, and V is held by the child. As the examiner points to letters on the distance chart, the child is instructed to point to the same letter on the card they hold.

Landolt ring and E game charts involve having the child indicate which direction the E or the gap in the Landolt ring is pointing. Developmentally, children are often able to name or match figures before they are able to reliably indicate the direction of E's and Landolt rings.

Allen and LEA symbols require the child to name the figures. Some children are able to identify the figures but refuse to speak during the exam. In such cases, the test can be adapted to be performed in a similar matching manner as the HOTV chart.

Special Considerations

Linear optotype monocular visual acuity cannot always be obtained, even when the child is developmentally ready for such testing. Some children are distracted by occlusion to the point that they will not cooperate for monocular vision testing. In such cases, obtaining a binocular acuity can suffice for that visit. As the child becomes more comfortable with the testing process, monocular testing can be performed at subsequent visits. The parents can take home photocopies of the test figures to practice the testing process with the child. Familiarity with the figures will enhance cooperation at the subsequent examination. In addition, when a child is first being tested with preliterate symbols, presenting isolated symbols can be less confusing to the child than rows of symbols. One must keep in mind, however, that the diagnosis of amblyopia can be missed or depth of amblyopia misjudged under isolated symbol testing conditions. One of the characteristics of amblyopia is the "crowding phenomenon" or contour interaction, where optotypes become more difficult to recognize when they are surrounded by similar forms secondary to abnormalities in spatial summation in the amblyopic eye [12, 13]. The visual acuity for amblyopic eyes can be several lines poorer when the symbols are presented in a row (crowded) as compared to isolated symbols. The examiner should use crowding bars that surround isolated targets when they are available and progress to linear testing with rows of symbols as soon as the child is able to adequately cooperate and to understand such testing.

When assessing a child's vision, one should observe for and document any abnormal head postures. The child should be tested in his preferred head position. This is especially important for children with nystagmus as they will position their gaze to utilize a null zone if one exists. The null position or zone is the position of gaze in which the nystagmus dampens, and visual acuity is maximized. Children with infantile strabismus often have an associated latent nystagmus. Manifest nystagmus can have a latent component, as well. When one eye is covered, the latent component causes an increase in nystagmus intensity, hence a decrease in monocular acuity. Latent nystagmus dampens in adduction. The best acuity will be obtained with the eye in an adducted position. Latent nystagmus can also be dampened by using the blur of a high plus lens instead of an opaque occluder to "cover" the non-tested eye. In addition to testing visual acuity monocularly, children with latent nystagmus and children with manifest nystagmus should have visual acuity tested under binocular conditions. Binocular testing will eliminate or minimize latent nystagmus and can result in a significant improvement in visual acuity when compared to that obtained monocularly. The binocular acuity is, in fact, more representative of the child's functional vision since under ordinary viewing conditions, the child has both eyes open simultaneously.

Detecting and Measuring Strabismus

Fusion State

Strabismus can be manifest or latent. When strabismus is manifest, normal fusion is not present, and the visual axes are not aligned under binocular conditions. Strabismus is designated as latent if fusion allows the visual axes to become aligned under binocular viewing conditions. A latent deviation is referred to as a phoria, while a manifest deviation is referred to as a tropia. It is important to determine whether a deviation is latent or is manifest as this can impact management decisions and assessment of treatment response. In some cases, deviations are sometimes latent, at other times manifest, presenting as a combination of both. This is most commonly seen with exodeviations and is discussed further in Chap. 51.

Herring's and Sherrington's Laws

Whether a deviation is latent or manifest, it is important to understand that the deviation involves both eyes. This fact should be communicated and explained to families. The bilaterality of strabismus is due to Herring's law of equal innervation. There are special forms of strabismus that appear to violate Herring's law. Dissociated strabismus, discussed in Chap. 56, is an example of a deviation that initially appears to violate Herring's law. Herring's law states that when the muscles of one eye contract or relax, the yoke muscles in the fellow eye will receive an equal change in innervation (Fig. 1.3). Sherrington's law of reciprocal innervation states that as a muscle is stimulated to contract, its antagonist receives an equal reduction in innervation allowing it to relax. For example, if a subject with a blind left eye performs a leftward saccade with his sighted right eye, the right medial rectus muscle (RMR) will receive the amount of innervation needed to move the eye to the intended target, while according to Sherrington's law of reciprocal innervation, the antagonist, the right lateral rectus muscle (RLR), will receive an equal reduction in innervation to allow the RLR to relax the appropriate amount. The yoke of the contracting RMR is the left lateral rectus muscle (LLR). According to Herring's law, the LLR will receive the same amount of innervation as its yoke, the RMR. The antagonist of the LLR, the left medial rectus muscle, receives an equal reduction in innervation according to Sherrington's law. The result is that the blind eye moves in harmony with the sighted eye just as the eyes of

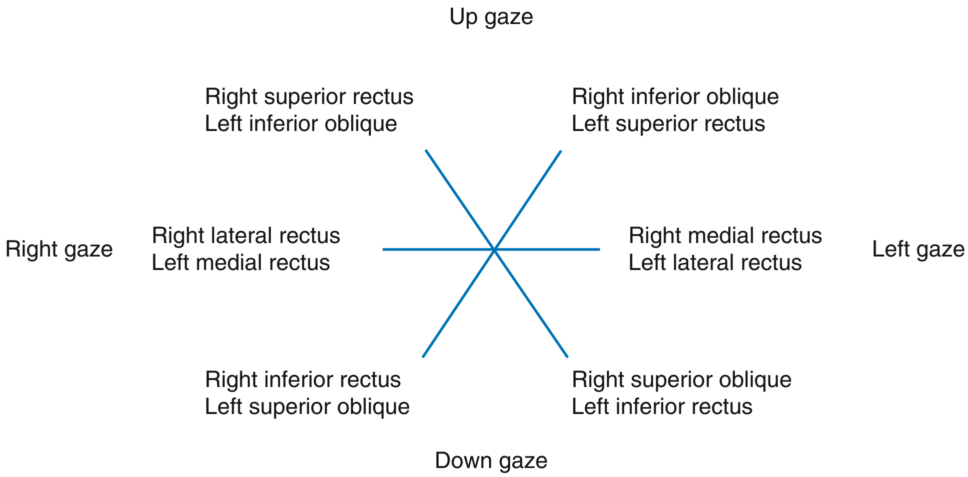


Fig. 1.3 Yoked muscle pairs in gazes for comparing muscle function. Cardinal positions of gaze are essential for isolating field of action for each muscle. If versions are abnormal, then duction testing should be performed

normally sighted subjects move together whether they are aligned or misaligned, following Herring's and Sherington's laws, respectively.

Cover Testing

Strabismus and motor fusion state can be evaluated by cover testing. Cover testing requires that the child be able to fixate centrally on a discrete target. Ideally, the fixation target should have detail that requires appropriate accommodation for clarity. Controlling accommodation is important, especially for horizontal strabismus. When accommodation fluctuates, changes in accommodative convergence can cause variability in horizontal alignment. When accommodation is not appropriate for the testing distance, measurement of the deviation can be inaccurate. The relationship between accommodation and ocular alignment will be discussed in Chaps. 47 and 51 and will be discussed in more detail later in this chapter.

Cover testing is typically performed with an occluder. Young children, however, are often distracted or feel threatened by the occluder. In such cases, using one's hand in place of the occluder is advisable and will enhance cooperation and attention to the fixation target. Accurate cover testing requires that fixation on the target be well-maintained. The younger and more active the child, the more encouragement they will need to maintain visual attention. The examiner should utilize near targets that the child will find interesting when possible. Ideally, the targets should have details that require appropriate accommodation to be visible. It is necessary to draw the child's attention to the target. This can be accomplished by asking the child questions about the target. For example, if the near target is a cartoon character sticker on a tongue depressor or a small toy, one asks the child questions about the toy or character (e.g., What color is Mickey's shirt?). For distance testing, one can have the parent take a toy to the end of the room. The parent can encourage the child to fixate on the toy. Changing to a new target when the child becomes disinterested is critical—"one toy, one look." Having a variety of interesting targets on hand will provide more opportunities to obtain the needed information. For toddlers and older preliterate children, movies or cartoons playing on a monitor or screen at the end of the room are useful targets to control fixation and allow qualitative evaluation and quantitative measurements of strabismus. Some of the computerized visual acuity systems are programmed to include movies or other animations for distance fixation targets. Most children are excited to be given the opportunity to "watch TV" while at the doctor's office, enabling them to relax while motility tests are performed. Accommodation must be controlled for clear viewing of the movie allowing for accuracy when measurements are obtained. Having the child describe what he is seeing on the monitor ensures attention and controlled accommodation. In addition, the sound from the movie helps maintain attention to the target. For older children, some projectors or computerized systems allow a single 20/60 optotype (or smaller) to be placed on cycle mode, and distance fixation can be assured by encouraging the child to verbalize the letter as it changes. When cycling mode is not available, presenting three full lines of letters will assist in maintaining visual attention at distance. The examiner can change the letters as needed. When vision or attention is extremely poor, accurate cover testing will not be possible, and other methods for strabismus evaluation involving corneal light reflexes will be necessary.

Detecting a Manifest Deviation

The cover test used to detect the presence of a manifest deviation (tropia) is referred to as the single cover test or the cover-uncover test. As with all cover testing, the child must be encouraged to maintain fixation on the target throughout

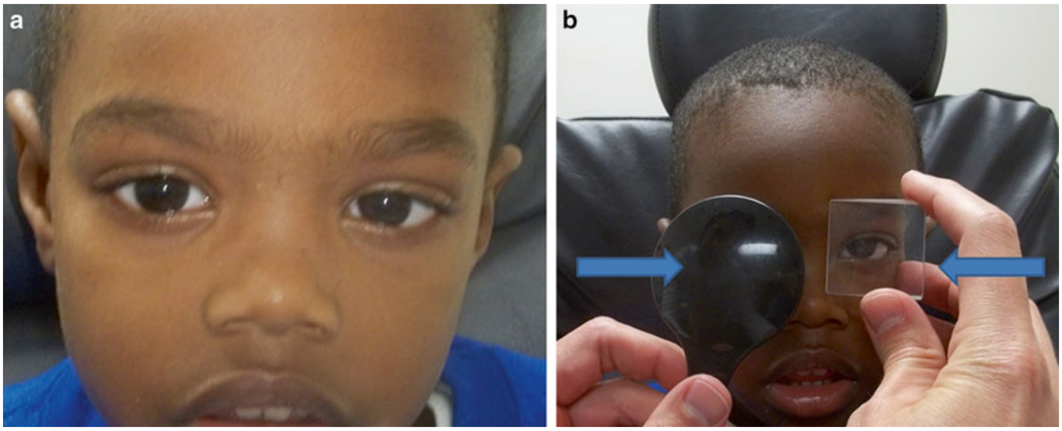


Fig. 1.4 Simultaneous prism and cover test. (**a**) Small angle left esotropia can be seen by corneal reflections. (**b**) Occluder is placed over fixing right eye as base-out prism in an amount equal to the estimated size of esotropia and is placed over the deviated left eye. Occluder and prism are simultaneously placed in front of each eye. Test is repeated with adjustments in prism power until left eye does not shift as prism, and occluder is simultaneously introduced. *Blue arrows* denote the simultaneous placement of cover and prism when testing

testing. To perform the single cover test, one eye is covered while watching for a shift in position of the fellow eye. The occluder is then removed, allowing the child to regain binocular vision. The test is repeated, now covering the other eye, looking for a shift in the fellow eye as the cover is introduced. If there is no shift when either eye is covered, no tropia is present. If, however, there is a shift when one eye is covered, it indicates that the fellow eye is deviated and that a tropia is present. This shift reflects the movement required for a deviated eye to achieve fixation on the target. If an inward deviation is present, the examiner will see an outward shift in position as the fellow eye is covered. This indicates that the child has an esotropia. If the shift in position is in a nasal direction, it indicates that the uncovered eye was deviated temporally. The child therefore has an exotropia. A vertical shift indicates that there is a hypertropia.

Often the single cover test is repeated several times to ensure accurate interpretation or to see if one eye is consistently the deviated eye when a tropia is detected. Some patients will alternate their fixation allowing either eye to be the deviated eye. In such cases, the child maintains fixation with the formerly deviated eye, with the fellow eye now deviated. When repeat testing is conducted, it is very important to allow binocular viewing between each test.

Measuring a Manifest Deviation

The size of the manifest deviation can be measured by performing the simultaneous prism and cover test (SPCT). The examiner first estimates the size of the shift detected on the single cover test. The SPCT will be performed using a prism equal in power to the estimated size of the deviation. The prism will be "simultaneously" placed over the deviated eye as the occluder is placed over the fixing eye. The test is repeated, adjusting the prism power until no shift is seen as the prism and cover are simultaneously placed in front of the child's eyes. No shift indicates that the deviation has been successfully neutralized by the prism. If, for example, a 12^Δ base-out prism is placed over the left eye as the right eye is covered, the child would have 12 prism diopters of left esotropia by SPCT (Fig. 1.4). Conventional nomenclature for this deviation would be 12^Δ LET. The ET refers to esotropia; the L indicated that it was the left eye that was deviated. This can also be documented as ET with strong right fixation preference. When measuring deviations, whether it be by SPCT or by alternate cover testing discussed in the next section, prisms should be held with the apex pointing in the direction of deviation. For example, for an outward deviation (exotropia), the apex of the prism should be temporal with the base placed nasally ("base in"). If the right eye is hypertropic, deviating in an upward direction, the prism should be held with the apex up, base down over the right eye. When performing the SPCT, the prism must be held over the deviated eye.

It is not necessary to measure manifest deviations in all cases. The SPCT is most often performed when the manifest deviation appears to be significantly smaller than the combined manifest and latent deviation.

Detecting Latent Deviations

The cover test used to detect a latent deviation (phoria) is known as the alternate cover test or the cross cover test. This test is performed by alternating the cover from one eye to the other *without* allowing binocularity as the cover is transferred from one eye to the other. Latent deviations remain latent by means of fusion. Allowing binocular viewing would allow the eyes to possibly realign if fusion ability exists. The alternate cover test is intended to disrupt fusion. When a latent deviation is present, the eyes will deviate while covered. In performing the alternate cover test, the examiner watches the eye that is being uncovered as the cover is transferred to the fellow eye. If the eye was deviated under the cover, there will be a shift in the position of the covered eye as the cover is moved to the fellow eye. Because of Herring's law, the fellow eye will now be deviated under the cover. As described in the section on single cover testing, the direction of movement will indicate the type of deviation present. For example, if a subject has a right hyperphoria, fusion is enabling both eyes to fixate on the target under binocular conditions. In this phoric state, single cover testing will reveal no shift. As the examination proceeds to alternate cover testing, however, fusion will be suspended, allowing detection of the latent deviation (right hyperphoria). When the right eye is covered, it will drift up under the cover. As the cover is moved to the left eye, the right eye will be required to make a downward saccade to take up fixation on the target. This downward movement will be observed by the examiner, indicating the presence of a right hyperphoria. According to Herring's law, as the right eye shifts down to take up fixation, the left eye, now under cover, will also shift in a downward direction. The covered left eye will now be in a hypophoric position beneath the cover. As alternate cover testing continues, the cover is now moved back to the right eye. The examiner will see the hypophoric left eye make an upward saccade to regain fixation on the target. This upward shift of the left eye and downward shift of the right eye will continue as the cover is transferred from one eye to the other. Vertical deviations are defined by the higher eye by convention.

Measuring Latent Deviations

Latent deviations are measured with alternate cover and prism testing. This technique is used whether or not a tropia was present on single cover testing. Prisms are placed in front of one or both eyes with the amount of prism adjusted until it neutralizes all movement as the cover is moved from one eye to the other. Because of Herring's law, when measuring deviations with the prism and alternate cover test, the prism can be placed over either eye. When measuring combined horizontal and vertical deviations, the horizontal prism can be placed on the vertical prism to simultaneously neutralize both the horizontal and vertical deviations. For example, if the subject has an esophoria and right hyperphoria, a base-out prism and a base-down prism can be placed over the right eye while on alternate cover testing to neutralize the deviation. The deviation can also be neutralized by placing a base-out prism and a base-up prism over the left eye. Two prisms with the base in the same direction, however, cannot be placed in front of the same eye. For example, if a subject with a 65^{Δ} exodeviation requires two base-out prisms to neutralize the large deviation, one prism must be placed over one eye, with the other prism placed over the fellow eye. Both prisms cannot be placed in front of the same eye. The effective power of two stacked prisms with the base in the same direction is different than the sum of the power of the two prisms [14].

Latent Versus Manifest Deviations

Deviations can initially present as a tropia or can require alternate cover testing for detection of the deviation in its latent form (phoria). Fusion enables a deviation to present as a phoria. Alternate cover testing, however, disrupts fusion and can cause a latent deviation to become manifest if fusional amplitudes are not sufficient to allow the patient to regain fusion when the cover is removed. When a deviation is at times latent, and at other times manifest, it is designated as an intermittent tropia. Distinguishing between a constant tropia, a phoria, and an intermittent tropia is important as the fusional state can impact management decisions. Nomenclature for documenting direction of the deviation and the fusional state is presented in Table 1.2. It is also helpful to document comments regarding the level of control of intermittent deviations. For example, a patient can be primarily phoric but briefly became tropic during the examination, while another patient might be primarily tropic but fuse (became phoric) briefly during the examination. Both

Table 1.2 Nomenclature for strabismus documentation[a]

Deviation	Phoria	Tropia	Intermittent tropia
Exodeviation (outward)	X	XT	X(T)
Esodeviation (inward)	E	ET	E(T)
Right hyperdeviation[b]	RH	RHT	RH(T)
Left hyperdeviation[b]	LH	LHT	LH(T)

[a] Prime added for near deviation (e.g., RHT′)

[b] Documentation of vertical deviations is based on higher eye by convention

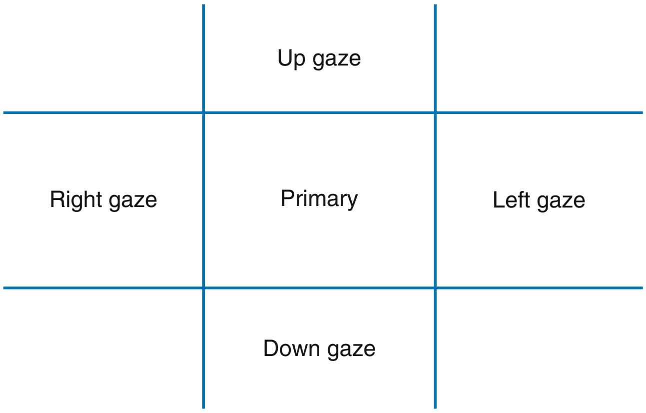
patients would be described as having an intermittent tropia, however, because of the difference in control of the deviation, each might be managed differently.

Incomitance by Gaze

Alternate prism and cover test measurements should be obtained at distance fixation and at near fixation in primary position. Discrepancies between distance and near measurements can impact diagnosis and management. It is also important to measure deviations in peripheral gazes, as this, too, can impact diagnosis and management. When the measurements change with with respect to direction of gaze, the deviation is referred to as incomitant. Ideally, peripheral gaze measurements are obtained with the subject fixing at distance. Measurement in up, down, left, and right gazes is usually sufficient for horizontal deviations. When a vertical deviation is present, measurement in nine fields of gaze and with the head tilted to the left and to the right may be necessary for diagnosis and surgical planning. Gaze measurements are documented as shown in Fig. 1.5.

Incomitance by Fixation

For most forms of strabismus, the deviation measures the same amount whether the left or right eye is fixating. This is because of Herring's law and is the reason that one can hold the prism over either eye when measuring strabismus. The fixating eye is the eye without the prism. For example, if one is measuring a 50^{Δ} esotropia in primary position, and the prism is held over the right eye, the right eye will be viewing in an adducted position as it looks through the base-out prism. It is the left eye that is in primary position as the measurement is taken. Therefore, with the prism held over the right eye, the primary position measurement is by left fixation in this example. If, however, there is a mechanical restriction or a cranial nerve palsy that limits the range of motion of an eye, the deviation can be incomitant by fixation and will measure more with affected eye fixating than with the sound eye fixating. For example, if a subject has a left lateral rectus palsy, the primary position measurement with the sound right eye fixing (prism over the left eye) might measure 30^{Δ} LET, with the left eye in a 15° adducted position under the prism. The deviation with the sound eye fixing is referred to as *the primary deviation*. If the subject is then measured while the left eye fixates and the prism now placed over the right eye, the left eye will be in primary position during the measurement. Because of the left lateral rectus weakness, an excessive amount of innervation to the weak left lateral rectus will be required to move the eye to primary position. Because of Herring's law, the yoke, the right medial rectus muscle, will also receive excessive innervation, driving the right eye further into adduction, increasing the size of the esotropia. The deviation with the affected eye fixating is a larger deviation and is referred to as the *secondary deviation*. A similar phenomenon can be seen when a mechanical restriction causes excessive innervation to the affected eye as the gaze approaches the restricted field. In such cases, the examiner must be alert to which eye is fixing when obtaining measurements.



	Up gaze	
Right gaze	Primary	Left gaze
	Down gaze	

Fig. 1.5 Documentation of gaze measurements

Attention to incomitance by fixation is particularly important when following paretic strabismus for signs of recovery or stability. If on one visit, the deviation is measured with the paretic eye fixing, then measured with the non-paretic eye fixing at the next visit, one could incorrectly interpret the smaller deviation at follow-up as a sign of recovery. In addition, if a patient with incomitance by fixation is going to be managed with a press-on prism, it is important to measure the deviation with the prism held over the eye that will be viewing through the prescribed prism.

Maddox Rod Test

Cover testing is an objective method of detecting and measuring strabismus. Strabismus can also be assessed subjectively with the Maddox rod test. When viewing a point light source through a Maddox rod, the point of light appears as a streak of light. If the rods are held in a vertical orientation in front of the eye, the streak will appear horizontal; with the rods in a horizontal orientation, the streak will be vertical. If a Maddox rod is placed in front of one eye as the subject views a light, one eye will see the light, and the other sees the streak. If the eyes are aligned, the streak will pass through the light. Horizontal alignment is tested with the rods in a horizontal position and vertical alignment with the rods placed in a vertical position.

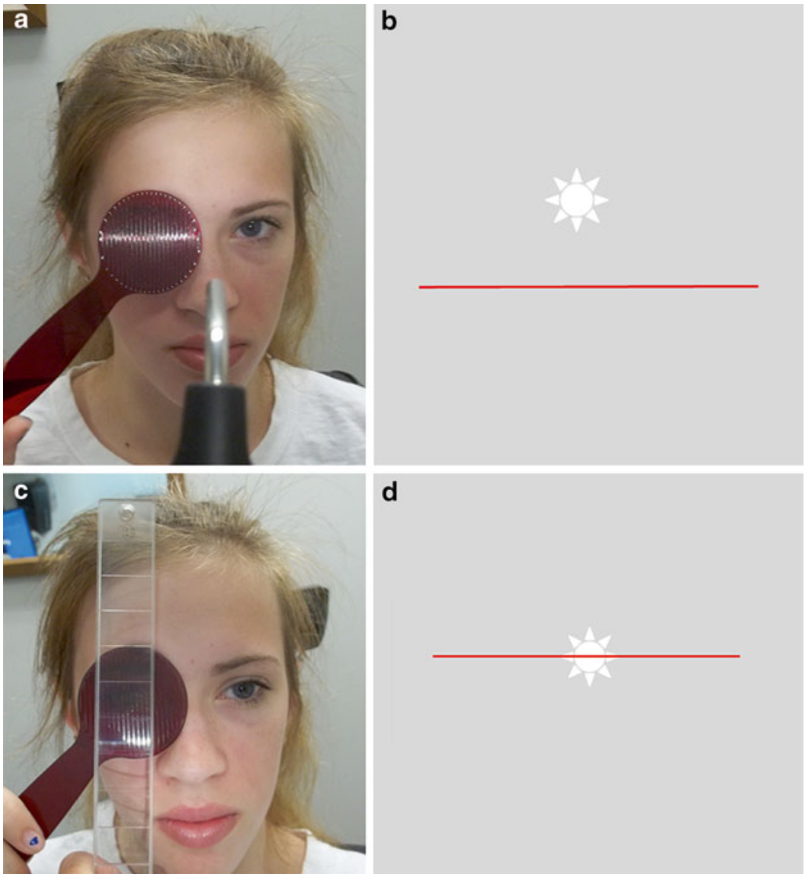


Fig. 1.6 Measuring strabismus with Maddox rod. (**a**) Subject with right hypertropia with Maddox rod over right eye. (**b**) Subject's view. *Red line* induced by Maddox rod (right eye) will be localized below the fixation light (left eye). (**c**) Subject is instructed to describe location of line with respect to the light as base-down prism is introduced to neutralize deviation. (**d**) Subject's view when the correct amount of base-down prism is over the right eye

With the Maddox rod oriented horizontally over the left eye, an esodeviated subject will see a light with a streak to the left (uncrossed localization). An exodeviated subject will see a light with a streak to the right (crossed localization). With the rods oriented vertically over the left eye, a subject with a right hyperdeviation will see a light with a streak above the light. With a left hyperdeviation, the streak will be seen as below the light.

The size of the deviation can be measured with loose prisms or by using a prim bar or rotary prism. When the appropriate amount of prism is in place, the subject will see the streak passing through the light (Fig. 1.6).

One advantage of Maddox rod testing is that it often takes less time than alternate prism and cover testing. It also is useful for detecting very small deviations that are difficult to detect with the cover test. A disadvantage of Maddox rod testing is that for horizontal deviations, accommodation is not controlled. In addition, one must use caution when measuring vertical deviations in head tilt positions in the presence of a horizontal deviation as the results can be inaccurate. Young children often are not good candidates for Maddox rod measurements.

Corneal Light Reflex Assessment

Assessing ocular alignment by evaluating the position of the corneal light reflex is useful when cover testing is not possible due to poor vision in the deviated eye or when cooperation does not allow cover testing or Maddox rod testing. This method cannot be used to detect or measure latent deviations. It only detects manifest deviations. Like the Maddox rod test, corneal light reflex tests do not control accommodation. Although corneal light reflex testing is not as accurate as other methods, it provides an opportunity to quickly obtain measurements in difficult situations.

Angle Kappa

An angle kappa exists if under monocular viewing conditions, the corneal light reflex is nasal (positive angle kappa) or temporal (negative angle kappa) to the center of the pupil. A positive or negative angle kappa can make the results of strabismus assessment by corneal light reflex less accurate. Positive angle kappa simulates exotropia, and negative angle