HANDBOOK OF PEDIATRIC OPHTHALMOLOGY

EDITORS STEPHEN S. FEMAN, M.D.
ROBERT D. REINECKE, M.D.

HANDBOOK OF PEDIATRIC OPHTHALMOLOGY

Editors

Stephen S. Feman, M.D.
Associate Professor
and Director of Retinal Services
Department of Ophthalmology
Vanderbilt University
Nashville, Tennessee

Robert D. Reinecke, M.D.
Professor and Chairman
Department of Ophthalmology
Albany Medical College
Albany, New York

Grune & Stratton
A Subsidiary of Harcourt Brace Jovanovich, Publishers
New York San Francisco London

LIBRARY OF CONGRESS

CATALOG CARD NO.: 78-71861

Feman, Stephen S. and Robert D. Reinecke
Handbook of Pediatric Ophthalmology.

New York, N.Y.: Grune & Stratton
200 p.
7812 780905

Grune & Stratton, Inc.
111 Fifth Avenue
New York, New York 10003

Distributed in the United Kingdom by
Academic Press, Inc. (London) Ltd.
24/28 Oval Road, London NW 1

Library of Congress Catalog Number 78-71861
International Standard Book Number 0-8089-1115-5

Printed in the United States of America

Preface

As the medical specialties have become more fully developed and the subspecialties have become better defined, the volume of information involved has rapidly enlarged. This phenomenon has caused the production of encyclopedic textbooks in all fields. The reader can easily get lost in the morass of technical material. This handbook is intended to supply the reader with a broad overview and a basic understanding of the field of pediatric ophthalmology. It is our hope that this will facilitate more detailed pursuit of selected topics. Most of the references are to textbooks that describe each of these subjects in elaborate detail. Thus the reader can progress to any subject that is of specific interest and can study it in depth.

The authors would like to thank the third-year students at Albany Medical College, as well as our colleagues and friends. Their numerous questions and their requests for abbreviated crib-side consultations led us to an awareness that a "field guide" to children's eyes was needed. Gratitude must also be expressed to the many members of the faculty at the Albany Medical College who aided in the production of various portions of this book. In addition, we are most grateful to our wives, Edith and Mary, for their understanding and patience.

Stephen S. Feman, M.D.
Robert D. Reinecke, M.D.

List of Contributors

Stephen S. Feman, M.D.
Associate Professor and Director of Retinal Services
Department of Ophthalmology
Vanderbilt University
Nashville, Tennessee

John Griffin, M.D.
Assistant Professor of Ophthalmology
Albany Medical College
Albany, New York

Pei-Fei Lee, M.D.
Associate Professor of Ophthalmology
Albany Medical College
Albany, New York

Ian Porter, M.D.
Professor of Pediatrics
Albany Medical College
Albany, New York
and Director, Birth Defects Institute
State of New York

Robert D. Reinecke, M.D.
Professor and Chairman, Department of Ophthalmology
Albany Medical College
Albany, New York

Richard S. Smith, M.D.
Professor of Ophthalmology
Albany Medical College
Albany, New York

HANDBOOK OF
PEDIATRIC OPHTHALMOLOGY

Contents

Robert D. Reinecke, M.D.

1
Vision, Amblyopia, and Strabismus

The purpose of this chapter is to suggest an orderly approach in dealing with children suspected of having visual defects or misaligned eyes. In any such examination vision must be a primary concern, and only by notations of impressions of visual status can appropriate referral or reassurance be accomplished. In general, if a visual defect is suspected, the earlier it is investigated the better is the potential for successful therapy.

A brief review of what constitutes visual acuity should serve as an appropriate background. Visual acuity (normally a test for central vision) is the ability to detect two lines separated by a space equal to the width of the line. This is called minimum separable acuity and is the type of acuity used in the construction of all visual acuity charts. Obviously this is considerably different from minimal detectable acuity, which can be likened to the ability to see a white spot on a dark background or a star in the sky. Such types of acuity testing are difficult to use, although occasionally they must be employed. Examples of such instances include having a child follow a rolling marble on the floor or follow a moving flashlight. Obviously, we are testing rather crude visual acuity when we resort to such determinations. As each age group is considered, the most appropriate visual acuity test will be mentioned.

NEWBORNS

To make a successful visual examination of a newborn, it is necessary that the child be in a receptive mood, that is, awake but not crying or feeding. Awakening a newborn from sleep prevents any estimation of visual acuity. A

newborn who is awake and receptive should easily be able to follow the movement of the examiner's face above the crib. As a flashlight is brought from either side, the child will tend to look toward the light. Moving objects are more attractive to the child than stationary objects. Since one of the principal objectives of all vision testing is to determine the visual ability in each eye independently, it is necessary to use some means of covering each eye sequentially. This does not mean that a hand must be put close to the child's face; interception of the pupillary axis is all that is necessary. Occasionally, a hand can be held over one eye at close range, but in most cases occlusion at a considerable distance from the eye will be less threatening to the child. Before doing that, it is a good idea to assess (as far as possible) the ability of the child to look about. Then one should carefully occlude each eye without disturbing the child, taking note of any change in the reaction. Most children will tolerate this quite well because they can see equally well with either eye. If a change in performance is detected as one eye is covered, this should be interpreted as poor visual acuity, and further tests are in order.

When there is difficulty in covering the eyes, it is appropriate to have a nurse place an adhesive patch over one eye prior to the arrival of the examiner in the nursery. The nonoccluded eye should be checked on that particular visit; if necessary, the opposite eye can be occluded during a similar sleeping period so that the other eye can be tested on the next visit to the nursery.

The use of small test objects is, of course, inappropriate, since it is impossible for a child to respond in any fashion other than simply to seek out a flashlight or wiggling fingers. The optokinetic drum is occasionally useful, since if the child does have detectable optokinetic nystagmus (see Chapter 6), use of the drum will confirm that the child has vision in that eye. Since most optokinetic drums have extremely large stripes, this confirms no more than crude vision.

The next step is to look into each eye to determine if the child will fixate with the fovea. This is easy to do with newborns if they are examined at the appropriate time. The direct ophthalmoscope is used with the aperture reduced so that a small circle of light is all that the child and the examiner can see. By looking into the child's eye from very close range, the examiner will be able to see the foveal area. Usually the child will cooperate, and the disk can be examined as well. Children of this age are usually extremely cooperative; there is no excuse for not examining the fundus in detail. Should any questions remain concerning the child's vision, the pupils should be dilated for a complete examination.

The quality of the foveal reflex is not good in any child, and one must be careful not to equate an absence of shiny foveal reflex with poor visual acuity. The foveal area is slightly granular, and the entire fundus typically will have a blond appearance in most children. Even in a black child the fundus will be remarkably pale, as compared to fundi in black adults.

In the newborn child who has nystagmus, it is well to note carefully any instability of fixation as well as transillumination of the iris. One should not be alarmed by the amplitude of the nystagmus; typically, it will decrease with time. It should also be noted that the newborn's optokinetic nystagmus may be rather poor in quality. Overconcern with this poor response should not lead the examiner into reporting poor vision to the parents.

If iris transillumination is present, complete albinism may be diagnosed. Ocular albinism can be difficult to detect at birth, since typically the iris does not transilluminate. As the child is followed along, by 6–10 years of age the iris will begin to transilluminate in a detectable fashion. Ocular albinism, which may be incomplete as far as lack of pigment is concerned, shows slightly reduced visual acuity, but most such children will do relatively well in the long run.

Newborns may have misaligned eyes (strabismus). The eyes may turn in, out, or up. Any type of misalignment should cause concern to the examiner, for it may indicate poor vision in one eye at that time or that amblyopia is developing secondary to the strabismus. Although it is true that in rare instances children will have misaligned eyes that subsequently will become straight, in most instances this is a myth; it should be quickly dispelled in favor of the more reasonable deduction that the child is likely to have permanent strabismus. Hence it is important that vision in these children be followed carefully to be certain that an amblyopia is not developing.

In conclusion, the newborn who has misaligned eyes is a real cause for concern, and the child who does not see well with each eye should be suspected of having a serious problem.

CHILDREN 3 MONTHS TO 1 YEAR OF AGE

Children 3 months to 1 year of age are more difficult to examine than newborns, but the same parameters are applicable. By this time it is usually easier to determine whether or not strabismus is present. By far the most helpful feature is whether or not some member of the family has detected a turned eye. If the parents report that the eye is turned in, but the examiner is unable to detect esotropia (by alternately covering one eye at a time and noting whether or not the eye has to move out to pick up fixation), the concern is probably not valid. On the other hand, if the parents report that the eyes turn out, but examination does not confirm exotropia, it is best for the examiner to believe the parents and proceed on the basis that the child does have intermittent exotropia. Any serious suspicion of a misaligned eye should once again trigger a careful follow-up to be sure that amblyopia does not develop. Certain tests have been developed (e.g., the use of smaller and smaller marbles rolled across a dark carpet) to assay the amount of vision in this age group. Most of

these tests are not very satisfactory; clinical impression of the child's performance as each eye is occluded is a much better assay. Should the examiner have further concern, it is appropriate to give the parents some adhesive eye patches and have them cover one eye at a time (for an hour or so) to observe the child's performance. The child should perform the same with either eye covered.

CHILDREN 1–2.5 YEARS OF AGE

During the period from 1 year to 2.5 years of age strabismus, should it develop, is most likely to be noted. Hence, careful strabismus history from the parents is quite important. The previous notations in regard to parents' history are applicable in this age group. It is tempting to try "illiterate E" testing, Landolt ring testing, and other specific visual acuity tests during this period of time. Generally these are unsuccessful, and it may be best not to burden the routine examination with such attempts. It is better to be certain that the eyes are aligned, which may be done by alternately occluding each eye and determining any refixation movement, and similarly noting whether or not the child has difficulty in performance with either eye covered. Should any strabismus be detected, it is essential that the child be followed carefully to ensure appropriate refractions and glasses (if necessary), appropriate patching for amblyopia, and surgery should other methods fail to align the eyes. Data have shown that early approaches to misaligned eyes give much better long-term effects.

CHILDREN 2.5–4.5 YEARS OF AGE

Children 2.5–4.5 years of age generally cooperate sufficiently well in the office that visual acuity testing in a conventional manner may be carried out if sufficient effort is made. However, a screening test is more appropriate. For the pediatrician's office, the screening test we recommend is that for stereopsis. Although it does not allow determination of visual acuity in the usual sense, a high level of stereopsis confirms good and equal vision. There are a number of tests available, but generally the random-dot E test is by far the best. Polaroid glasses are placed in front of the child, and three cards are used to determine the stereopsis. Since there are no other clues available that allow the random-dot E cards to give false readings of good stereopsis, the test has an appropriate referral rate and is very successful. Instructions come with the test and need not be detailed here.[1,2]

In the event that such a test is not available, one must employ the visual acuity tests in the usual fashion. Kits of figures measurable to 20/30 called Allen figures are helpful in this early age group. Unfortunately, the tumbling

E blocks and picture cards give what is known as isolated visual acuity, i.e., looking at an isolated letter as opposed to a full line of print. Isolated visual acuity is typically better than linear acuity; indeed, it may mask amblyopia. Hence, care must be taken in interpreting isolated visual acuity as being equal to full-line visual acuity.

It may be helpful at this point to review briefly what is meant by the Snellen equation. For practical reasons the Snellen eye charts are constructed so that *each detail* (i.e., each line and each space between two lines) of the letter or figure on the 20/20 line will subtend a 1-minute angle when viewed at 20 feet. The notation 20/20 means that a distance of 20 feet the eye can resolve a target the size of which subtends an angle equal to a total height of 5 minutes of arc. Most adults with normal eyes achieve this 20/20 vision. The notation 20/30 means that letters that subtend a total arc of 5 minutes at a distance of 30 feet can be visualized at only 20 feet. When the distance is 20 feet but the height of the letter is that which a 5-minute arc would subtend at 200 feet, the designation is written 20/200. Conversion to the metric scale is easy because 6/6 (meters) equals 20/20 (feet). Hence, 6/12 would equal 20/40. Many times the testing distance in the examiner's office will be shorter than the distance required for the standard acuity chart. Thus, if there is a 20/20 line, but it is viewed at 10 feet, that should be noted as 10/20; it is, of course, equivalent to 20/40. If there is a 20/10 line on the visual acuity chart and it is measured at 10 feet, then this should be recorded as 10/10, which is equivalent to 20/20.

Many children are remarkably advanced in knowledge of the alphabet by virtue of watching many hours of television. Hence, it is appropriate to try the standard Snellen acuity chart before resorting to any other test, other than the stereopsis screening test. If at all possible, the complete line should be used for testing, rather than isolated letters. It is also important, of course, to carefully occlude each eye independently as vision is tested. Appropriate eye treatment too often is delayed because the child has peeked from under a cover during vision testing. Every effort should be made to avoid that pitfall.

The concerns previously mentioned in determining visual acuity also apply to alignment of the child's eyes. If the eyes are not aligned, further investigation into the possible cause is extremely important. One cannot emphasize the cardinal rule too often that the earlier strabismus and amblyopia are treated, the better the result.

CHILDREN 4.5 YEARS OF AGE AND OLDER

Testing visual acuity in children 4.5 years of age and older is usually the same as testing an adult. Charts that have full lines of letters should be used. Careful notation of the testing distance should be made, and the same attention should be given to preventing the child from peeking out from under the

cover during the testing period. The stereopsis screening test is equally valid for all age groups and is particularly appropriate for the preschooler, since it is easily and quickly administered.

If poor vision is detected, referral is usually the logical course; however, should the presence of pathology be a concern, use of the pinhole effect is one of the handiest means of differentiating organic from refractive visual problems. Defective vision, if optical in origin, will be corrected by use of the pinhole. Hence, simple visual acuity measurements done monocularly and then again with a pinhole will confirm whether the poor visual acuity is due to some organic defect or is due simply to the need for an optical correction. It is often helpful to show myopic children how to simulate a pinhole (by curling the index finger into a small tunnel) so that those who lose their gasses can see to get about.

We may summarize visual acuity and strabismus testing as follows: Always look for the fixation response to be certain that the eye is capable of seeing. Whenever possible, use visual acuity test targets that are measurable, with careful notation as to the distance they are held from the patient. Any discrepancy from one eye to the other should be cause for referral. Above all, do not succumb to the myth that eyes will straighten if given sufficient time. Turned eyes mean that a problem exists, and appropriate attention should be given immeditaely.

STRABISMUS

There are several types of strabismus and various etiological factors involved, this section will attempt to outline some of the common ones.[3]

Esotropia

Esotropia (crossed eyes) is one of the more common problems in children. Indeed, about 3–4 percent of patients will have esotropia of some variety. From a screening point of view, this is an exceptionally rewarding area to cover. Some children will have esophoria rather than an esotropia. In esophoria there is a tendency for the eyes to turn in, but the child is able to straighten the eyes when given the opportunity. If the child is tested carefully, the tendency to turn in can be noted. As the cover is removed from one eye and the child is allowed to view things binocularly, the eyes will suddenly become straight. These children have a tendency to deteriorate from esophoria to esotropia. Thus, early attention should be given them whenever possible. Children who are extremely farsighted fall into a similar category. As they make an attempt to see objects clearly, the eye has to accommodate an exceptional amount, and the accommodative-convergence reflex turns the

eyes in excessively. Hence, when the child is relaxed, the eye may be straight, but when the child attempts to look carefully at an object, *intermittent esotropia* will be noted.

Since one of the basic determinations in esotropia is refractive error, as mentioned previously, and since the eyes often can be straightened with glasses alone, accurate assessment of the optical defect is essential. This assay, high on the ophthalmologist's priority list, is accomplished by cycloplegic refraction.

Many ophthalmologists use atropine, and the dosage is rather high considering the size of the patient. Hence, it is not uncommon for a mildly toxic reaction to be reported by the parents. This toxic reaction will result in the child becoming febrile and having a flushed appearance with dilated pupils. This should not be confused with any illness. It is logical to inquire whether a child who is febrile has been to an ophthalmologist recently. The toxicity usually is not great; the best advice to the parents is to cease administration of the atropine and simply take the child's temperature at reasonable intervals. If no significant hyperthermia develops, then they should keep the next appointment with the ophthalmologist. In rare instances patients may develop hyperthermia or other significant side effects; then appropriate antidotes may be given.

More commonly used are the shorter-acting cycloplegic agents. They also may cause flushing of the skin and may even give rise to mild disorientation and drowsiness. The latter signs are usually seen in younger children or fair-complexioned children. Drowsiness typically lasts 2–3 hr. Hence, reassurance can be given about such children. Occasionally a patient will have a short convulsion, but treatment with barbiturates is seldom necessary.

Often the ophthalmologist may use an objective means of measuring the child's optical error. This is called retinoscopy and is quite precise, so that glasses may be prescribed for the child of any age, from a few days on.

Esotropia also may be caused by sixth nerve paralysis. Any time esotropia develops suddenly, concern regarding the numerous possibilities that can give rise to a defective sixth nerve should always be at the back of one's mind. Fortunately, lateral rectus paralysis secondary to sixth nerve interruption is not common, and in the majority of cases it may be self-limited. The reader is referred to Chapter 6 for further details.

A and V patterns are common in esotropia; these are descriptive terms related to the vertical position of greatest esotropia. The V pattern means that when the child looks down, the eyes become crossed; when the child looks up the eyes may be quite straight. Since most of the time when a person looks down the eyelids cover the eyes, esotropia in downgaze is not easily noted at an early age. As a result, these often are not detected until the child becomes older and someone is in a position to see the eyes in downgaze. Conversely, the child who has A esotropia has a greater crossing when looking up. In other

positions of gaze, the eyes may be quite straight. Indeed, many of these children do not need treatment, but they may be a cause for concern to the parents until the children grow tall enough to look the parents straight in the eye.

One of the more interesting features of these patients with A and V patterns is the tendency of the oblique extraocular muscles to overact. When such a child looks to one side, the adducting eye (the eye looking toward the nose) may suddenly rotate up or down. The cosmetic effect may be traumatic to parents and child alike, but the implication is not important other than in the treatment of the A and V patterns.

Exotropia

Exotropia (walleye or turned-out eye) typically develops as a child gets older (i.e., 6–10 years of age) but it may present at any time, even in the neonatal period. The majority of these patients demonstrate an intermittent tendency that often is not noticeable until the child becomes older. One of the early symptoms the parents will report is that on going outside the child will close one eye. One reason for this is that in bright light the eyes have a tendency to turn out, and the exotropia becomes manifest; the child then closes one eye in order to avoid diplopia. Nearsightedness (myopia) is sometimes associated with the exotropia and will be suitably corrected on referral. The exotropia, because it is intermittent, allows development of good stereopsis at near vision; hence, the eyes work well together most of the time. Because of this, treatment for exotropia is not as aggressive from a surgical standpoint as is treatment for esotropia. An esotropic patient has a great tendency toward not using the two eyes toegher, whereas the exotropic patient does well in this regard, and surgery may be delayed until the exotropia becomes more constant. However, if exotropia is present in extremely young children, surgery often is indicated earlier, since these children will not have straight eyes for any significant time during a 24-hr period.

A and V patterns are noted among exotropic patients, with A patterns somewhat more common than V patterns (i.e., when the child looks down, the eyes may turn out to a greater extent). The overacting obliques also may be noted in exotropia; hence, the child looks rather bizarre when attempting to look to either side, when the adducting eye may turn up or down excessively.

Refractive Errors

Throughout the preceding discussion we mentioned the need for glasses and the essential nature of refraction in all patients with strabismus or amblyopia. One of the reasons for this is that the eyes may have different refractive errors; hence, the image in one eye may be blurred while the image in the opposite eye may be clear. The child tends to suppress this blurred

image, causing amblyopia. Eyes may be nearsighted or farsighted or may have astigmatism. Astigmatism simply means that the cornea of the eye is shaped more like the surface of a lemon, as opposed to the usual spherical shape of most corneas. This irregularity can be easily corrected with appropriate spectacles.

A few words of explanation about refractive disorders may be helpful in explaining these problems to parents.[4]

Myopia or nearsightedness. A positive family history typically is present in the child who is found to be nearsighted. Often the parents are extremely concerned about the child having to wear nearsighted glasses, as they have had to do throughout their lives. Unfortunately, there is no means of prevention of nearsightedness, but parents should be reassured that the vision will be as good as theirs or perhaps better and that appropriate contact lenses can be prescribed any time that the child's age permits or the parents deem them appropriate. Whereas it is true that some forms of nearsightedness become so extreme that vision can be poor, the majority of patients will have good vision, and glasses are an appropriate solution to the problem. If the myopia is severe, yearly examinations are in order, since these large eyes are more susceptible to retinal detachments.

Farsightedness. The average child is farsighted at birth, and this gradually becomes less severe as the child's eye grows and becomes normal in size. Mention has already been made of the association between farsightedness and esotropia. Suffice it to say that whenever amblyopia or esotropia is suspected, the child obviously must be refracted; if hyperopia is present, that must be fully corrected. If the hyperopia is not severe or if bifocals are required, drugs may sometimes be used in place of glasses to control the excessive convergence that some of these patients manifest. This is termed a high accommodative-convergence/accommodation ratio. Miotics such as echothiophate iodide (Phospholine Iodide) may be used that cause miosis and cause the eye to react as if it has already accommodated; hence, it will not require as much accommodative convergence. Often these drugs are remarkably successful in controlling esotropia, thus avoiding the need for surgery. Since these are long-acting miotics, it is important to be aware that they can cause side effects, including gastrointestinal upsets. Anesthesia can be somewhat hazardous by prolonging the effect of succinylcholine if it is used. Thus it is mandatory that whenever anesthesia is advocated, inquiry be made as to whether or not the child has been receiving eyedrops.

Treatment of Amblyopia

In addition to the use of glasses to correct any refractive error, patching is the basis for all amblyopia treatment. The good eye is patched in such a way