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Little, Brown and Company Boston

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First Edition

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Library of Congress Catalog Card No. 81-80542

ISBN 0-316-74894-3

Printed in the United States of America

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To my wife Lucy and our children Henry, Erica, and William

### **Preface**

In recent years pediatric ophthalmology has become well established as a subspecialty of ophthalmology. An American Association for Pediatric Ophthalmology has been formed and lists nearly 200 members. The Journal of Pediatric Ophthalmology and Strabismus is gradually attracting papers of good quality, and increasing numbers of young ophthalmologists are choosing to pursue a career in the pediatric end of their specialty. This increasing attention to children's eye problems has resulted in growth and clarification of the body of medical knowledge bearing on these problems. There has been in the field a sense of excitement and purpose that is inherent in any actively growing area of medicine. Ophthalmology for the Pediatric Practitioner was written in an effort to share this crystallization of knowledge in the field of pediatric ophthalmology with pediatricians and general physicians, medical practitioners who are concerned on a daily basis with the detection and management of pediatric eve problems. It is important for these physicians to know what is happening in a medical specialty that so closely touches their patients. Communication of medical knowledge is not a luxury but an obligation that cannot be ignored when medicine is practiced in such a diversified fashion as it is in this country today. The book may also be useful to students of ophthalmology, at various levels, who wish to have an abbreviated review of pediatric ophthalmology as a place to begin. For a thorough understanding of any given subject there is, of course, no substitute for study of original articles, and to this end a large number of references are included in the text. These references are also intended to lead the general medical reader to ophthalmologic sources that may be as yet unfamiliar.

My inclination from the start was to write the book as a single author, without imposing my own editorial preferences and time schedule on a group of collaborators. Therefore, any limitations in scope or inaccuracies in content are my own. It is hoped that some uniformity of style and singularity of purpose will outweigh whatever shortcomings have resulted from this approach. Although no formal contributions from medical colleagues are included, I must quickly express indebtedness to a small group of superb ophthalmologists with whom I have worked at Children's Hospital over the past 15 years: Robert A. Petersen, M.D.; David S. Walton, M.D.; Anne B. Fulton, M.D.; William P. Boger III, M.D.; and Frederick J. Elsas, M.D. All have contributed ideas and clinical material in one way or another. I am also indebted to two longtime colleagues who have contributed to the preparation of this book in a very tangible

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way. Richard T. Sullivan prepared the photographic illustrations, mostly from color transparencies, with great patience and skill, and Laurie S. Rachins typed and edited the manuscript with her usual blend of enthusiasm and tact. The graphic illustrations were expertly done by Jean Kansky of The Children's Hospital Department of Visual Education. Many others, who will be glad to see this project completed, have contributed with encouraging words or by taking on added responsibilities while I was preoccupied with preparation of the manuscript. To all of them I am grateful.

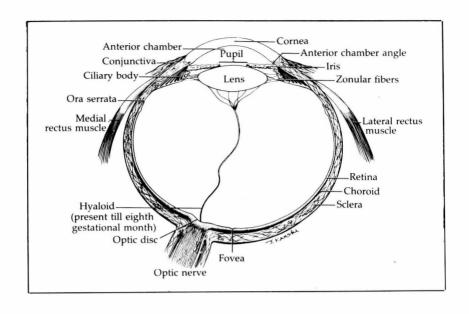
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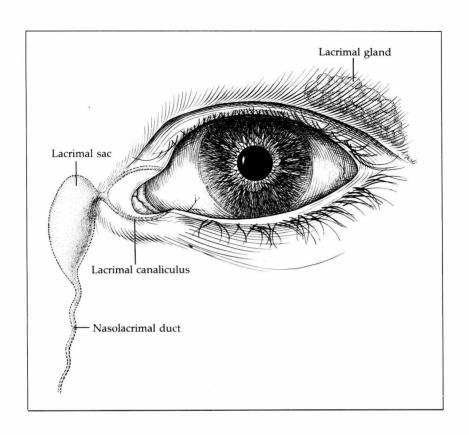
#### Contents

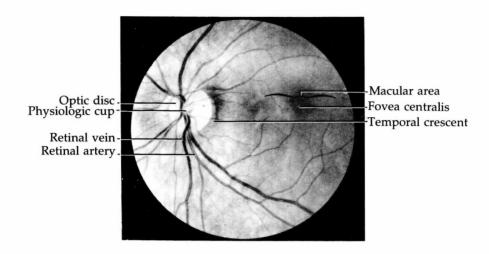
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- 1. The Eye Examination in Infants and Children 1
- 2. Detection and Management of Strabismus 21
- 3. Cataracts and Lens Dislocation 37
- 4. Childhood Glaucoma 45
- 5. Retinoblastoma, Retrolental Fibroplasia, and the White Pupil 53
- 6. Ocular Infections and Inflammations 63
- 7. Developmental Abnormalities of the Eye 75
- 8. Blepharoptosis and Orbital Neoplasms 101
- 9. Ocular Manifestations of Pediatric Systemic Disease 107
- 10. Pediatric Neuro-Ophthalmology 133
- 11. Ocular Trauma 151

Index 155







### 1. The Eye Examination in Infants and Children

The pediatrician or family physician is in an ideal position to identify eye problems in infants and children. Although definitive management of the problems often may require referral to an opthalmologist, there is much to be gained by early detection. If early examination results in recognition of strabismus or discovery of congenital glaucoma, the gain may be conservation of sight. If it allows recognition of retinoblastoma, the result may be preservation of life. This kind of early diagnosis requires not so much that pediatricians and general physicians master difficult techniques of eye examination but that they be alert to the possibility that a problem exists. The diagnostic maneuvers themselves are far from insurmountable.

#### ASSESSMENT OF VISION

Any eye examination should begin with an attempt to quantitate vision. In infancy, this is usually based on observations of visual behavior since verbal responses are lacking. An infant's ability to fix and follow an object of visual interest is present at birth [1], but, in an office testing situation, it becomes more obvious at 6 weeks to 2 months of age [2]. The object of visual interest can be a face, a flashlight, or a brightly colored toy. Although adding sound to the test object theoretically might compromise its purity as a visual stimulus, in practice a light that rattles or a toy that squeaks is often more effective in attracting an infant's attention. The size of the object and its distance from the patient are not critical since the examiner is not trying to estimate visual acuity. The object is moved back and forth in front of the child while the examiner watches to see if the eyes turn toward the object and follow its movement in the visual field. If visual fixation and following are not present by 4 months of age, further evaluation certainly is indicated. Following movements should be smooth and free of nystagmus. The eyes should appear straight and well aligned on the target. If a light is used for the target, its reflection from the eye, the corneal light reflex, should be centered in the pupil of each eye. Although momentary misalignment of the eyes may occur in infancy, any sustained misalignment when the object of regard is known and controlled is abnormal. Such misalignment is called *strabismus* or simply *squint*. The use of the cover test for detection of strabismus is discussed in Chap. 2.

Another important clinical sign in the assessment of vision is the pupillary reaction to light. This reaction is present at birth [3], but it must be looked for carefully because an infant's pupils are rather small normally, and further constriction to light may be inapparent.

Absence of the pupillary reaction to light usually indicates a severe abnormality of the retina or optic nerve. The presence of a pupillary reaction is reassuring, but of course it does not guarantee that vision is normal.

There are numerous ways to estimate an infant's visual acuity in a more quantitative fashion than is possible with fixation and following responses and pupillary reactions. Preferential looking tests may be used (Fig. 1-1). They are based on an infant's preference for a patterned visual stimulus over a plain stimulus when the two are presented simultaneously [4]. This preference disappears when the pattern is too fine to be resolved by the infant's visual system. With this technique, the vision of a normal one-month-old infant has been estimated to be 20/300, and that of a 6-month-old infant approximately 20/100. This numerical expression of vision, the Snellen notation, is explained on p. 6.

Optokinetic nystagmus, jerk nystagmus induced by rotating before the eyes a drum with alternating black and white stripes (Fig. 1-2), has also been used to estimate visual acuity at an early age [5]. The limit of acuity is the stripe width at which nystagmus is no longer induced. Unfortunately, many normal infants are insufficiently attentive to the rotating stripes to develop nystagmus at any given time, and a negative response to this test is difficult to interpret.

Evoked occipital potentials have also been used as a parameter of visual function [6]. These electrical potentials, representing brain activity in the visual cortex, are induced by repetitive light stimuli. The relatively slight individual responses are summated by computer and can be observed to vary with the check-size of the patterned stimulus, thus allowing an estimate of visual acuity. The elaborate equipment needed for this kind of testing obviously limits its clinical usefulness. However, visually evoked potentials occasionally are helpful in assessing vision when other behavioral manifestations of visual awareness are lacking.

#### SUBJECTIVE TESTING OF VISUAL ACUITY

The subjective testing of vision is usually first possible between 21/2 and 3 years of age. Schematic picture cards, such as those designed by Allen [7], are convenient symbols to use (Fig. 1-3). These are presented for identification at increasing distances from the patient, and the visual acuity is recorded as the ratio of the farthest distance (in feet) at which the cards can be identified to an assumed normal distance of 30 feet for an adult, e.g., 15/30. The eyes are usually tested separately with the aid of an occluder or an eye patch. Some examiners prefer to use the so-called STYCAR test (Screening Test for Young Children and Retardates), in which the letters T, H, V,

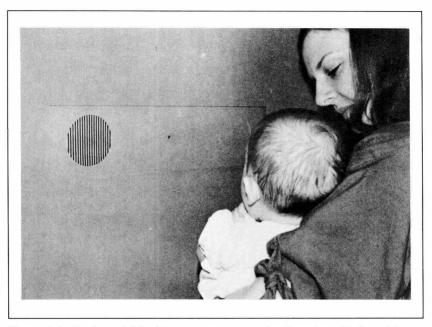


Figure 1-1. Preferential looking test to assess visual acuity of infant. The test is based on an infant's preference for a vertically striped disc over a plain one when the two are presented simultaneously. The observer on the other side of the screen behind a peephole records the infant's fixation pattern.

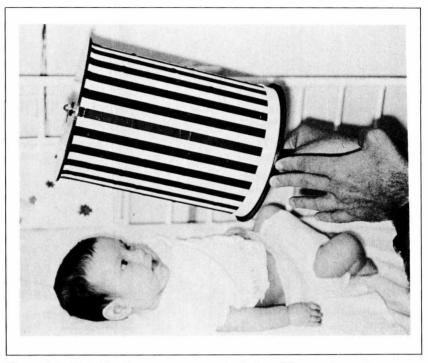


Figure 1-2. Optokinetic nystagmus can be induced by rotating a drum with alternating black and white stripes.