PEDIATRIC EYE DISEASE

COLOR ATLAS & SYNOPSIS

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To my wife Gloriann

Only her talent and energies as a loving mother, teacher, and child-care professional surpass her daily support of my personal and professional passions. She is an inspiration to me and a role model for any young woman who desires to overcome the complicated and diverse demands required by our times.

RWH

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PREFACE

The idea for this atlas originated with the generous gift from David B. Schaffer, M.D., of his 30-year collection of clinical photographs to me upon his retirement. His attention to detail in the collection, categorization, and preservation of this material inspired me to keep this material from collecting dust for the next 30 years. Using these photographs only for selected lectures was not doing them justice. I felt that this collection could be better used in publication form for a wider audience. With the aid and support of Ms. Darlene Cooke of McGraw-Hill, the proposal for this atlas became more purposeful and began to take on life. As the idea for this atlas took shape, the other editors and I decided that participation of other eye care professionals would provide a diversity of opinion and experience that truly represents the needs and eye diseases of infants and children. The added benefit of this participation was the consolidation of their figures, photos, and thoughts with those of the editors, thus providing a more complete compilation of materials.

There are multiple purposes to this atlas. Our first goal was to provide a varied, visual representation of the more common eye diseases that present in infancy and childhood. We understand that no book can show the hundreds of diseases presenting in hundreds of ways; however, with the support of the publisher we have been able to publish about 450 photographs (about 400 in color), which illustrate the variation in appearance of many childhood eye diseases. We organized the text so as to highlight definitions of terms, differential diagnosis, workup, and major treatment options. We do not wish to replace the many excellent texts covering the diagnosis and treatment of pediatric eye diseases and strabismus, but we do wish to photographically enhance these texts. We have created this atlas to serve as a reference material to be used by the many people involved with infants and children—pediatric physicians and nurses, family care personnel, child day care centers, emergency rooms, schools, optometrists, opticians, ophthalmologists, and those who come in daily contact with infants and children.

We have divided this atlas into five major parts, which reflect, as much as possible, those diseases that more commonly present in neonates (Part I), infants (Part II), toddlers (Part III), school-age children (Part IV), and other common childhood eye problems (Part V). We understand that there may be considerable overlap in the age of presentation of these eye diseases, but we felt that this format would allow the busy professional examining a child the most direct route of photographic reference. Neonatal eye diseases include ophthalmia neonatorum, TORCH syndromes, cataracts, glaucoma, retinopathy of prematurity, and congenital anomalies of the lids and orbit. Infantile diseases include strabismus, genetic and craniofacial syndromes, vitreoretinal diseases, optic nerve anomalies, nonaccidental trauma, and nasolacrimal duct obstruction. Problems in toddlers include amblyopia and strabismus, infections and inflammations, ptosis, and orbital and eye tumors. Diseases in school-age children include uveitis, nystagmus and anomalous head positions, vision

development, testing and visual screening, and refractive errors. Other common problems include accidental trauma and spectacles in infants and children.

We feel that the specific addition of chapters on nonaccidental trauma (abuse and neglect) vision testing and screening and spectacles in children provide unique additions to an atlas of this type. The illustration of these common eye conditions and childhood needs should provide any professional caring for infants and children special insights. These may be particularly useful for the non-eye care professional in that they emphasize salient clinical characteristics and testing procedures encountered in common childhood eye situations.

We understand that the rapidly changing technology, transfer of information, and almost daily acquisition of new knowledge create challenges to the medical educator. This is true whether one is educating health care professionals, students, or patients and families. This pressure to provide "up-to-date" information is a prevalent part of our times and drastically complicates the publishing process. This atlas is unique and timely in that, while our understanding and treatments of eye diseases in infants and children are constantly changing, the appearance of these diseases is almost timeless. We believe that this timeless appearance of eye diseases in infants and children will be the foundation of this atlas as a long-standing reference for those who have the fortune and pleasure of working with children.

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This section allows a contemplative assessment of where one is how one got there. For me this is not a recitation of places but of people. I would like to thank all the families that trusted us with their most precious gift, their children. If not for these patients, this book would not have been possible. I would next like to thank all the contributing authors, who interrupted their busy lives and provided a unique dedication to their authorship task. Throughout my personal life and professional career, there have been people who have believed in my ideals, supported my plans, encouraged my passion for ophthalmology and guided my sometimes wayward energies. All these people are fundamentally responsible for the completion of this book. My parents Ann and Richard and brothers and sister, Steve, Jim, and Sue provided me with the values of a strong work ethic, loyalty to family and friends and honesty at work and home. My stepchildren Jessica and Jamie have given me the privilege of being a parent. The late Malio Cascardo was there during my tumultuous teen years and provided a professional and personal sanctuary. As an undergraduate student in Columbus, Ohio, Robert Hagman II of Ohio State Optical Co. and Gary L. Rogers, M.D., each gave me opportunities to shape my professional work in eye care and medical research. During my medical training in Northeastern Ohio, the late Hayes Davis, M.D., was a powerful mentor to me, as he was for many students, and Scot E. Lance, M.D., became my friend for life. Mohammed Ashrafzadeh, M.D., practically "gave" me all his surgical skills and was a beacon of light and calm during the storm of residency in Boston. Louis F. Dell'Osso, Ph.D., has been, and remains, a mentor and friend. David B. Schaffer, M.D., Glen A. Gole, M.D., James A. Katowitz, M.D., and Arthur J. Jampolsky, M.D., took the time to show me the skills needed to become an expert in pediatric ophthalmology and strabismus. In recent years Marshall M. Parks, M.D., has embraced me and provided community support for my pediatric ophthalmology clinical research service at the National Eye Institute. Last, I would like to thank the Residents in Ophthalmology at The University of Pennsylvania, The Walter Reed Army Medical Center, and The Washington Hospital Center as well as those Fellows in pediatric ophthalmology who have worked with me over the years. Their constant questioning, desire for knowledge and dedication to the care of children have forced me to stay sharp. Those previous Fellows include David B. Granet, M.D., Joseph A. Napolitano, M.D., Sule Ziylan, M.D., Gary D. Markowitz, M.D., Martin C. Wilson, M.D., Yvette M. Jockin, M.D., Sepidah L. Rousta, M.D., Darron A. Bacal, M.D., Mitra Maybodi, M.D., and Michael Schaffer, M.D.

Richard W. Hertle

CONTENTS

PART I

NEONATAL EYE DISEASE

CHAPTER 1

OPHTHALMIA NEONATORUM

RICHARD W. HERTLE

DEFINITION OF TERMS

Ophthalmia neonatorum (ON) is an inflammatory disease of the ocular surface occurring in the first 28 to 30 days of life. It is usually caused by *Neisseria gonorrhoeae* infection. In the 1880s Crede's use of silver nitrate as prophylaxis against ON was a remarkable medical and public health achievement, because it decreased the incidence of newborn conjunctivitis from 10% to 0.3%. The Centers for Disease Control (CDC) reported 900,000 cases of ON in 1986.

DIFFERENTIAL DIAGNOSIS

The most common causes reflect the infectious nature of this condition and include the following:

1. *N. gonorrhoeae*
2. *Chlamydia trachomatis* (about 20% of neonates will suffer from associated systemic involvement)
3. Bacterial conjunctivitis (staphylococci, streptococci, *Haemophilus* species, coliform groups)
4. Viral conjunctivitis and keratitis (most commonly herpes simplex types I and II).
5. Chemical conjunctivitis (silver nitrate)
6. Ocular trauma
7. Congenital glaucoma
8. Congenital lacrimal system obstruction

WORKUP

1. Parental history of sexually transmitted diseases
2. Complete ophthalmic examination
3. Cytologic analysis of ocular discharge (Gram stain, Giemsa stain, Wright stain)
4. Immunofluorescent antibodies (*Chlamydia* infection)
5. Fluorescein-conjugated antibodies of McCoy cell cultures (*Chlamydia* infection)
6. Cultures (aerobic, anaerobic, and viral infections)
7. Immunocytochemical diagnosis (viral infections)

TREATMENT (See Table 1-1 on page 4)

1. Topical and systemic antibiotics (various combinations of penicillins, erythromycin, cephalosporins, and antiherpetic agents)
2. Irrigation of the ocular surface
3. Treatment of parents and their sexual contacts

CONCLUSIONS

The impact of 100 years of prophylaxis in preventing and decreasing the prevalence of blinding disease from ON in neonates must not lead to complacency. Until predisposing conditions are eliminated, this remains a potentially significant medical problem.

SUGGESTED TREATMENT APPROACH FOR NEONATAL CONJUNCTIVITIS*

Diagnosis (by History and Physical and Laboratory Evaluations)	Treatment
1. *Chlamydia trachomatis* (see Figs. 1-5–1-7)	Ocular—Erythromycin ointment O U qid × 2 weeks Systemic—Erythromycin 30–40 mg/kg/d po × 2 weeks
2. *Neisseria gonorrhoeae* (see Figs. 1-1–1-4)	Ocular—Erythromycin ointment O U qid × 2 weeks and topical ocular irrigation Systemic—Aqueous penicillin G, 100,000 U/kg/d I V qid × 7 days or ceftriaxone, 28–50 mg/kg/d I V q8–12 h × 7 days
3. Herpes simplex (see Figs. 1-8–1-10)	Ocular—Trifluorothymidine (Viroptic) 9 times qd × 14 days Systemic—+/– Acyclovir (Zovirax) IV solution
4. Other viral or epidemic (see Figs. 1-12–1-13)	Ocular—Cool compresses, +/– erythromycin ointment or polymyxin B solution qid × 7 days Systemic—Supportive treatment
5. Chemical (see Fig. 1-11)	Ocular—Self-limited, observation only

*If a corneal ulcer is suspected or diagnosed (topical fluoroscein staining of the cornea is positive), prompt ophthalmic referral is indicated.

Key: +/–, means no problem using or not using —usually used when there are signs of secondary bacterial involvement.

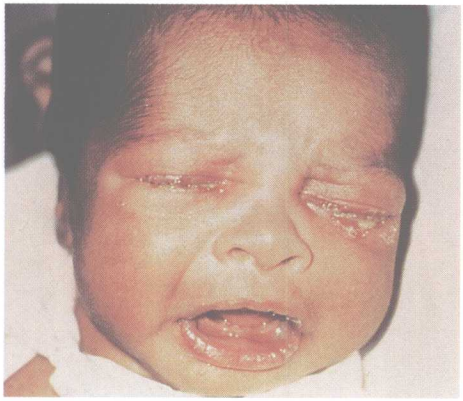
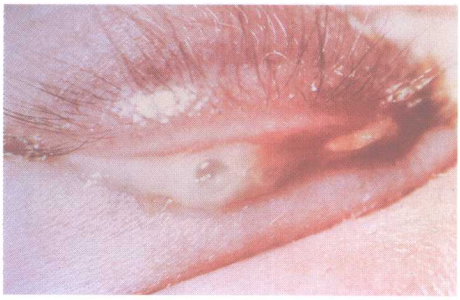


Figure 1-1



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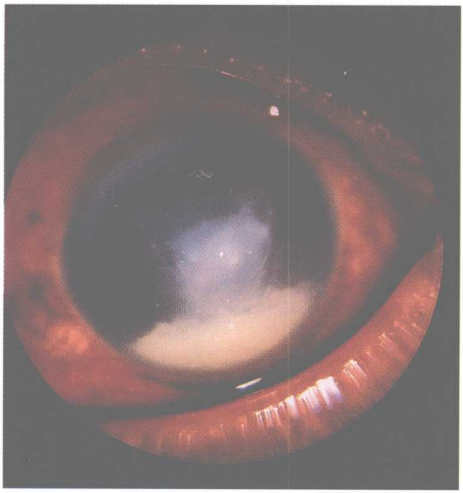

Figure 1-3

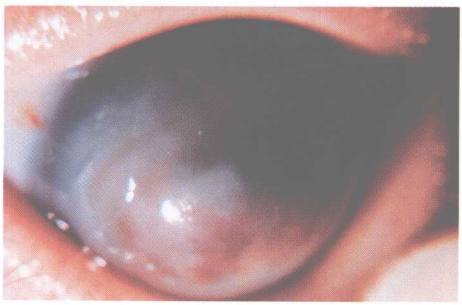

Figure 1-4

Figures 1-1 to 1-4 *These figures depict the spectrum of involvement of the conjunctiva, corneal surface, and eye in* Neisseria gonorrhoeae *infection. Figures 1-1 and 1-2 show the copious mucopurulent conjunctival discharge that is a clinical characteristic of this infection. Figure 1-3 shows progression of the infection to include the cornea, producing ulcerative keratitis and a hypopyon ("pus" in the anterior chamber). Figure 1-4 shows end-stage panophthalmitis with infection of the intraocular contents with secondary corneal thinning and ectasia with the possibility of prolapse of intraocular contents through the cornea.*