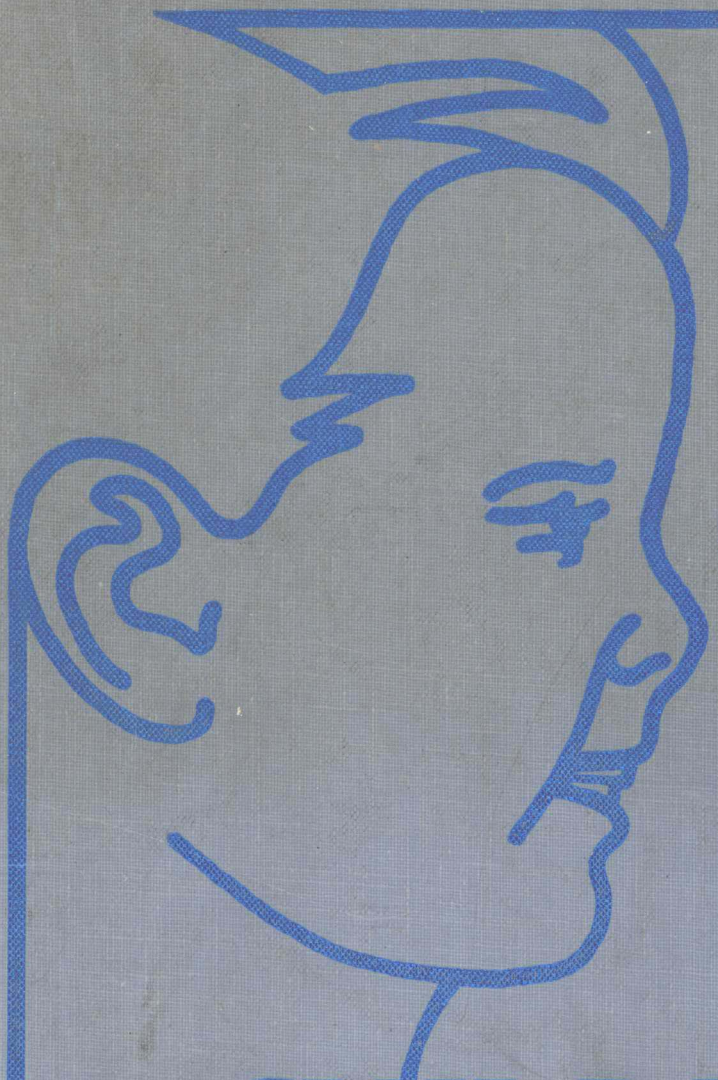

THE EYE IN CHILDHOOD

Edited by
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and
J. Donald Morin, M.D.

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This book is dedicated to all the children.

Foreword

Pediatric ophthalmology has come of age, and it is only appropriate that a textbook of this size, coauthored by subspecialists, be dedicated to this subject. This volume covers a variety of topics, including strabismus, plastic surgery, genetics, ocular oncology, drug therapy, anesthesia, and contact lenses—all of which pose special problems that are unique to children. The authors invariably give excellent advice on the diagnosis and management of pediatric ophthalmologic disorders based on their long-standing personal observations. Such insights make this book good reading regardless of one's level of expertise. The book is aimed at the general ophthalmologist, the ophthalmology resident, the orthoptist, the ophthalmic nurse, and the pediatrician. Given the excellent clinical skills of the authors as well as the scope of the book, I am sure it will be of significant help to pediatric ophthalmologists as well.

The book originated as a manual for residents in ophthalmology. Although it has mushroomed to an extensive textbook, it has maintained the individuality of the authors, who give coherence to the book by offering practical tips in every chapter. It is not a book that includes only results obtained through controlled studies, rather each chapter is a personal testimony to the authors' skills and joy in taking care of children. It is therefore a lively book to read and should be of significant help to anybody involved in the diagnosis and management of a child with a visual problem.

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Preface

Pediatric opthalmology has been one of the fastest growing ophthalmic specialties ever since the days of Dr. Frank Costenbader, who is the acknowledged founder of the speciality. Advances in understanding pediatric ocular disease and newer techniques of treatment necessitate a constant updating of knowledge of all aspects of pediatric ophthalmology by textbooks such as *The Eye in Childhood*.

This book sets out the practical experience of the staff of The Hospital for Sick Children, Toronto, with reference to current ideas from the world literature when appropriate. Emphasis is given to surgical techniques and treatment protocols that have given the best results, and there has been no attempt to include all the known methods.

There are special chapters on embryology, childhood diseases with significant ocular involvement, radiology and pediatric ophthalmology, contact lenses in children, and management in education of the visually impaired child. This wide range of subjects embodies the scope of pediatric ophthalmology.

We owe a great debt to earlier authors in the field of pediatric ophthalmology, as well as to the following institutions and individuals, without whose capable assistance this project could not have been completed:

1. *The Hospital for Sick Children, Toronto.* The Departments of Pediatrics, Surgery, and Radiology provided valuable advice and criticism. The Department of Bacteriology, especially Drs. P. C. Fleming and R. M. Bannatyne, and the Department of Virology, especially Dr. P. Middleton, were great assets. We are also grateful to the secretarial staff of the Eye Clinic, especially Connie Campbell, Helen Gigas, Deborah Doherty, and Audrey Hodgson.
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Contents

1

Normal and Abnormal Development of the Eye

Carlton G. Smith
Brenda L. Gallie
J. Donald Morin

A knowledge of the successive steps in the development of the eye is essential in interpreting anomalous developmental conditions and in fixing the time during gestation when a disturbing influence produces its effect. Although the sequence of steps in development does not vary, the time of completion of each step may vary. It is possible, for practical purposes, however, to divide the stages in the development of the eye into four periods of intrauterine life. The duration and major events of each stage and the associated abnormalities are outlined in Table 1-1. The first stage in development is usually completed in the first lunar month; the second stage in the second and third months; the third stage in the fourth, fifth, and sixth months; and the fourth stage in the last four lunar months of intrauterine life.

STAGE I

In the first fetal month the optic cup, the lens, the vitreous, and the blood vessels begin to develop.

Optic Cup

The eye begins as an evagination of the anterior ventral part of the floor of the third ventricle (Fig. 1-1). This evagination takes the form of a vesicle with a stem that elongates until the vesicle reaches the deep surface of the skin. At this stage the brain and the optic vesicle and its stalk are embedded in mesenchyme, a loosely constructed embryonic connective tissue. The skin is a thin layer of ectodermal epithelium. As the vesicle approaches the skin, it collapses to form the dorsal half of a two-layered cup. The inner layer goes on to develop into the light-sensitive part of the retina, the neural layer.

The outer layer acquires pigment during the fourth week to become the pigment epithelium of the retina. The critical role played by the microenvironment in maintaining differentiation is demonstrated by attempts to grow pigment epithelium *in vitro*. Without basement membrane, Type III collagen, and an epithelial growth factor, the cells continue to grow but lose pigment. The space between the two layers of the optic vesicle will be occupied ultimately by the

Table 1-1.
Stages in the Development of the Eye

Stage	Lunar Month	Major Events	Major Abnormalities
I	1	Optic vesicle, cup	Anophthalmos
		Primary lens fibers	Congenital cystic eye
		Primary vitreous	Congenital nonattachment of retina
		Hyaloid artery	Colobomata
			Primary aphakia
			Central cataract
			Optic nerve aplasia
II	2, 3	Optic cup neural proliferation	Cataract, rubella
		Secondary vitreous	Optic nerve hypoplasia
		Secondary lens fibers	Persistent hyperplastic primary vitreous
		Annular vessel, greater arterial circle	
		Long posterior ciliary arteries	
		Choroid	
		Cornea, sclera	
III	4–6	Neural differentiation	Coloboma ciliary body iris
		Vascular proliferation	Aniridia
		Lesser arterial circle	Coloboma lens
		Pupillary membrane	Dislocated lens
		Retinal vessels	Anterior chamber cleavage syndrome
		Short posterior ciliary arteries	Glaucoma
		Choroid	Foveal hypoplasia
		Ciliary processes and muscle	Retinal dysplasia
		Iris sphincter	
		Secondary and tertiary vitreous, cornea, sclera	
IV	7–10	Macular differentiation	Microphthalmos
		Pupillary dilator	Cryptophthalmos
		Atrophy of hyaloid vessels	Superficial lens opacity
		Atrophy of pupillary membrane	Absent pupillary dilator
		Anterior chamber	Albinism
			Glaucoma
			Retinal foveal hypoplasia

outer segments of the rods and cones. The ventral half of the optic cup begins to develop by the addition of cells to the inner and outer layers of the upper half of the cup along its inferior margins. As the margins approach each other, the narrowing interval between them is called the choroid fissure (Fig. 1-1). When the margins meet to complete the cup, the inner and outer layers unite in such a way that no evidence of the line of fusion persists. The proximal end of the choroid fissure remains open in the optic stalk as a tubular passage for the hyaloid artery.

As the choroid fissure closes and the outer layer of epithelium becomes pigmented, the inner or neural layer begins to differentiate (Fig. 1-2). At the end of the first month, the inner layer consists of the germinal zone of closely crowded nuclei and a layer of developing nerve fibers, called the marginal zone, which is adjacent to the developing vitreous.

Lens

The lens develops as a plate-like thickening of the surface ectoderm, in response to the approach of the optic vesicle (Fig. 1-3). This plate of epithelium bulges into the optic cup and is eventually pinched off from the skin to form a vesicle with a wall one cell thick. At this stage the deep half of the lens vesicle occupies almost all the space in the optic cup. As the lens continues to develop, the cells of the deep half of the vesicle begin to elongate, encroach on, and eventually obliterate its cavity. These elongated cells are

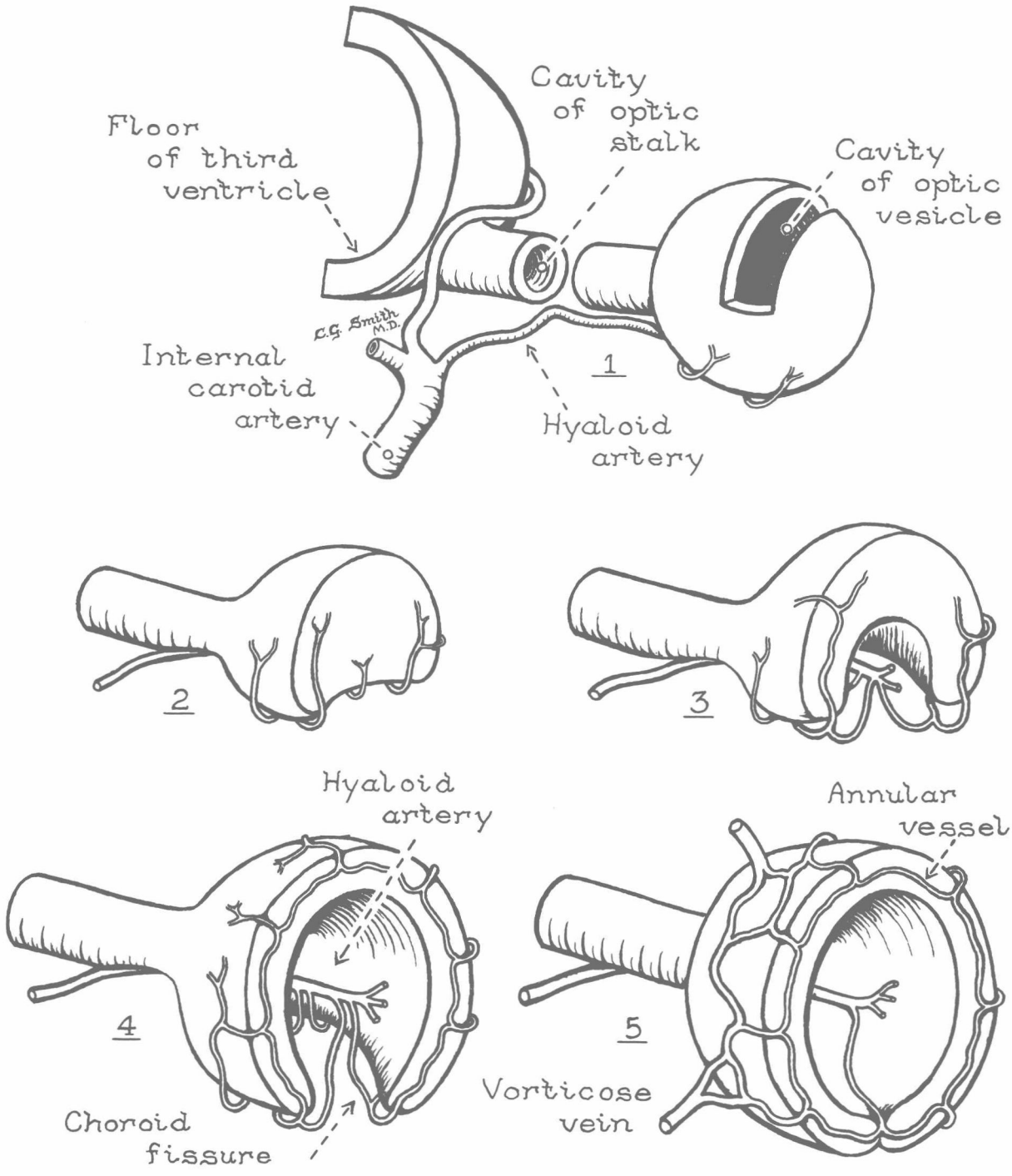

Figure 1-1. Five steps in the development of the optic cup during the first month of fetal life "stage I."

the primary fibers of the lens. They lengthen until they reach the anterior cuboidal epithelium of the vesicle and then stop growing (see Fig. 1-3). Hence, their maximum length in the mature eye will be equal to the diameter of the lens vesicle at this early stage, that is, about 1 mm or less. To mark the end of this first month of development, the cells of the lens secrete the basement membrane that forms its capsule (Fig. 1-2). Thus, at the end of the first month, the lens has acquired the anterior layer of cuboidal epithe-

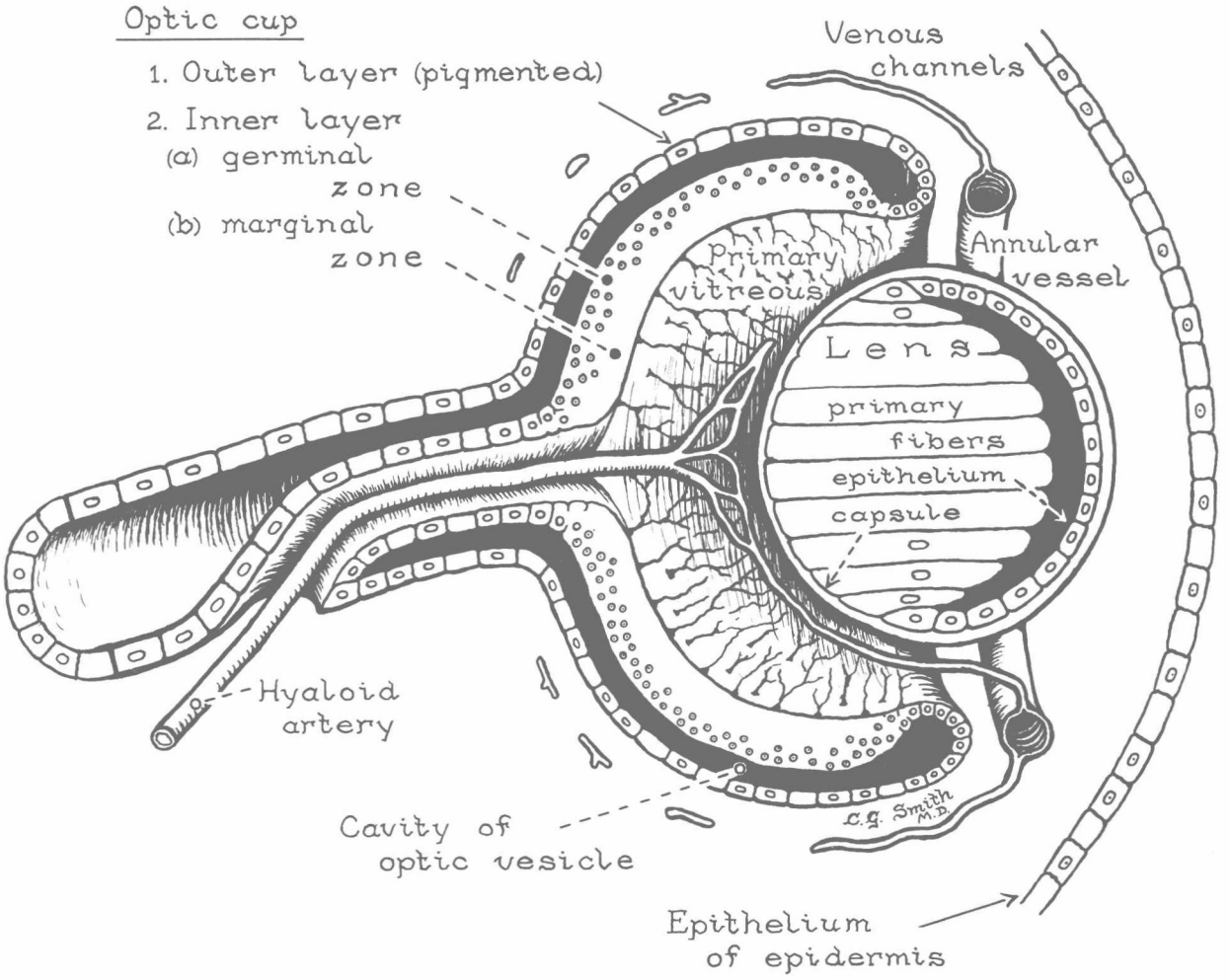

Figure 1-2. The structure of the eye at the end of first month of fetal life.

lium, the nucleus of primary lens fibers, and the hyaline capsule.

Vitreous Body

The primary vitreous is elaborated during the first month (Fig. 1-2) as interlacing strands that extend from the cells of the optic cup and from the cells of the lens vesicle until the hyaline capsule of the lens cuts off this source. This primary vitreous contains the branches of the hyaloid artery and thus differs from the avascular secondary vitreous that develops after the first month.

Blood Vessels

As the optic cup develops, endothelial-lined spaces containing fetal erythrocytes develop in the surrounding mesoderm. A branch of the internal carotid artery, the future ophthalmic artery (known at this stage as the hyaloid artery), establishes connections with the ventral capillary network (Fig. 1-1). This capillary network sinks into the cup and as the margins of the cup come together, the connections with the vessels on the outside are pinched off. The only part of the fetal fissure that persists is located in the optic stalk close to the cup and is kept open by the hyaloid artery. The hyaloid artery at the end of the first month supplies the vitreous aspect of the lens and has branches to the vitreous. The blood thus brought into the cup is drained by a single vessel that passes below the lens to the rim, where it divides T fashion into two branches that course along the margin of the cup to meet and anastomose, forming the annular vessel. This vessel has connections with the developing veins on the outside of the cup.

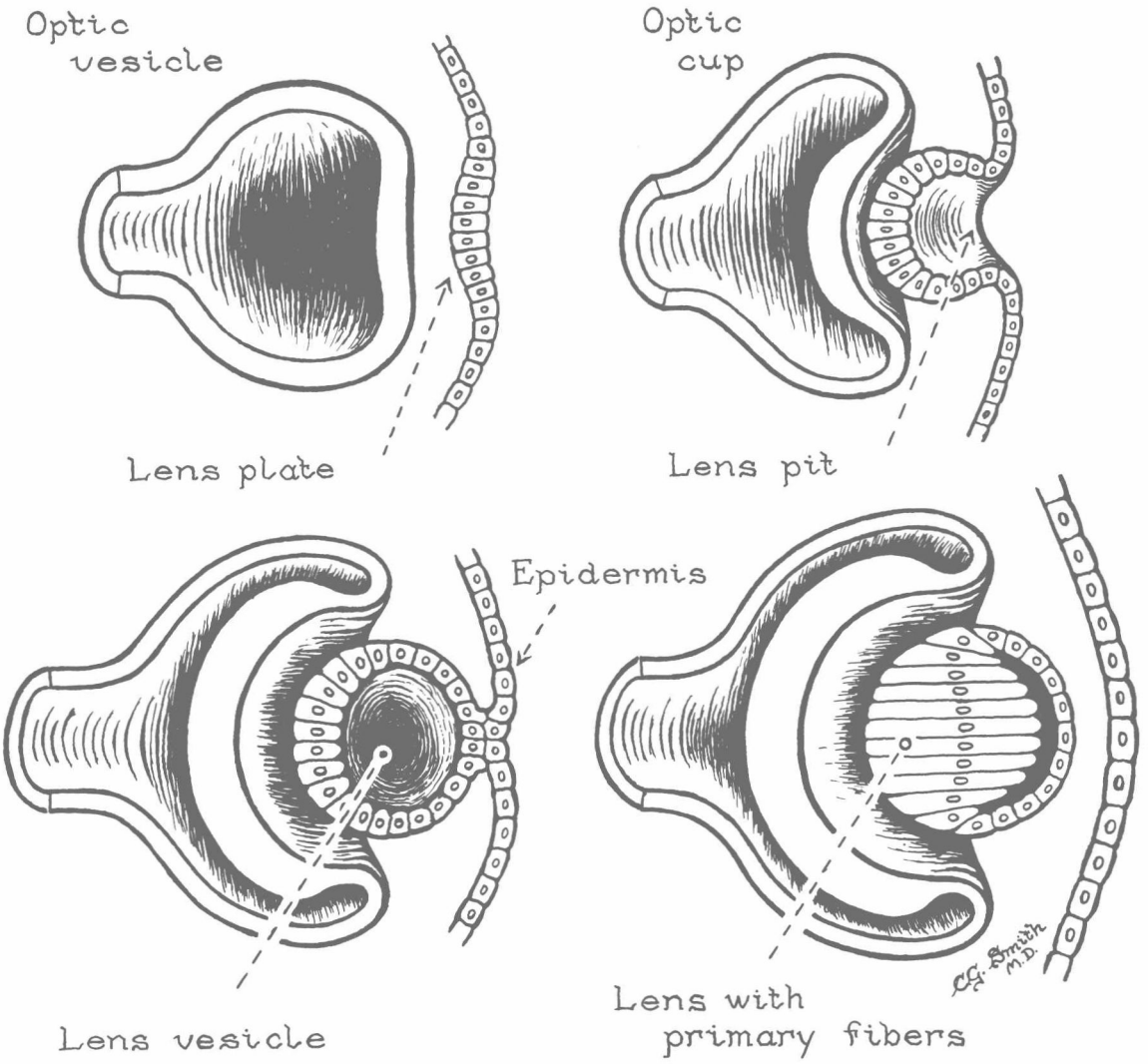

Figure 1-3. Four stages in development of the lens in stage I.

Developmental Abnormalities in Stage I

Anophthalmos

Primary anophthalmos is a sporadic anomaly unassociated with any systemic defect. For some reason the outpouching of the wall of the forebrain has failed to occur and no eye develops, although the orbit and eyelids appear relatively normal. Disorganized mesodermal and ectodermal elements can be seen, but no neuroectodermal elements are present. The central visual pathways are absent or hypoplastic. The diagnosis can often be confirmed only by serial histological sections. Seventy-five percent of cases are bilateral and life expectancy is normal.

Secondary anophthalmos results from suppression of the entire neural tube and is lethal. Consecutive anophthalmos results from degeneration of the optic vesicle after evagination, and some neuroectodermal structures persist. The size of the orbit and conjunctiva sac are frequently reduced in anophthalmos.

Cyclophthalmia or Synophthalmia

Cyclophthalmia, complete fusion of the eyes, is rare. Synophthalmia is generally characterized by complete fusion posteriorly and incomplete fusion anteriorly leading to duplication of the anterior structures. There may be a single optic nerve in the midline and a frontal nasal process forming a single