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AN ATLAS OF
Diseases of the Eye

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Foreword

There is no doubt that the only satisfactory way to learn medicine is by the repeated study day after day and month after month of large numbers of patients in a hospital under the guidance of a wise teacher. Thus the student first finds his way around the medical world; but as speciality succeeds speciality in the medical curriculum and when studentship is transformed into practice, the distinctness of the memory of clinical pictures necessarily becomes blurred. Particularly is this so in a subject such as ophthalmology, for in most medical schools the study of diseases of the eye receives less attention than it deserves, crowded out as it is by the multitude of subjects which appear to have greater immediate interest. This is indeed unfortunate – to the general practitioner, because the importance of recognizing ocular conditions in general practice is often crucial, particularly the differentiation of what is trivial from what may be of serious import; and to the specialist in many fields, as well as to the general practitioner, in so far as an understanding of vascular and other changes as seen in the eye, is frequently of immense value in establishing the diagnosis and assessing the prognosis of a host of general systemic diseases.

How is the position most easily remedied? There is no doubt that a pictorial representation of pathological conditions is much more meaningful and valuable for this purpose than a host of printed words, and an Atlas supplemented by a text the best substitute for the patient himself. This volume, much of which has previously been published in the form of booklets, is an attempt to fulfil this want, to provide the student, the general practitioner and the non-ophthalmological specialist with a short guide to the more common and important ocular disorders. For this purpose no trouble has been spared.

All the illustrations have been specially selected or prepared in order that a representative collection could be made widely available. In the case of fundus appearances use has been made of both colour photography and drawing; the selective hand of the artist serves to underline the important features of any given condition, whereas photography more nearly reproduces a single ophthalmoscopic view with its profusion of distracting detail: some care in interpretation is therefore necessary.

The external appearances have mainly been recorded in colour by the camera, supplemented where necessary by the artist whose particular contribution is in the synthesis of microscopic views seen by slit-lamp illumination. Particular attention has also been paid to the photo-mechanical method of reproducing the originals, and by means of offset lithography the ideal has largely been attained.

There is no doubt that the excellence of modern techniques of reproduction makes many of these conditions live. But these techniques are expensive; and the cost of an Atlas of this type, if it were to be published at its economic price would be prohibitive. Its publication, however, at a relatively low cost, has been made possible by the generosity and public spirit of the firm of Roche Products Limited. One of the features of modern medicine is the partnership that is nowadays frequently formed between it and the large pharmaceutical firms, many of which have done much to further the therapeutic aspects of medicine by the immense effort they expend in

research and the happy collaboration they offer to academic laboratories; the subsidy of an Atlas of this type by Roche Products Limited so that it may become available to the profession at about one-tenth of its cost price is a novel contribution to the dissemination of knowledge. For this, and in so far as the Atlas may aid in the treatment of our patients, we are grateful to them.

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STEWART DUKE-ELDER

Acknowledgements

It is clear that a compilation of this character can never be complete and must always depend to some extent upon the willing co-operation of many people.

In particular, members of the medical staff of Moorfields Eye Hospital (incorporating the Royal London Eye Hospital, the Royal Westminster Ophthalmic Hospital and the Central London Ophthalmic Hospital) have made the greatest contribution by making their records so freely available. Dr Norman Ashton of the Institute of Ophthalmology has been kind enough to provide pathological material to supplement purely clinical appearances and Mr E. F. Fincham was responsible for producing the special cataract photograph. Certain gaps have been nobly filled by other hospitals including Guy's Hospital, St Bartholomew's Hospital, Westminster Hospital and the Hospital for Sick Children, London. Mr D. P. Greaves of University College Hospital has also been most helpful in providing specific examples required for this atlas.

Most of the pictures themselves have been prepared by the staff of the Medical Illustration Department of the Institute of Ophthalmology amongst whom we are pleased to name Mr N. Jeffreys, F.I.B.P., and Mr T. R. Tarrant, M.M.A.A. The work of other illustrators is also represented and we are glad to acknowledge the following: Miss J. Trotman, Mr E. R. Alexander, Mr A. W. Head, Mrs L. Geddes and Theodore Hamblin, Ltd.

The task of preparing the manuscript for publication has fallen to Miss J. Richards and Miss B. Bate. This work has involved many revisions and their help in this respect has been invaluable. Miss M. H. T. Yuille has also kindly assisted in checking proofs.

We should like to thank Sir Stewart Duke-Elder who was responsible for the original plan; both he and Mr R. C. Davenport have made many helpful suggestions at various stages of production. Mr Frank Law and Mr E. F. King have both carefully appraised the pictures and text; as a result of their helpful criticisms several changes have been made.

Special equipment has been used in production of many of the illustrations and reference should be made to Messrs Clement Clarke Ltd, for making certain technical facilities available to us.

More than a word of recognition is due to the printers – Messrs W. S. Cowell Ltd of Ipswich. They have lavished unusual care and attention on the reproduction of originals which, in many instances, contained very fine detail. So much has depended on their efforts and the consistent results which they have obtained from different types of material are a singular achievement.

Lastly, we should like to thank Roche Products Limited for their generous support and co-operation throughout the preparation of the volume.

E.S.P.
P.H.

CONTENTS

PART IV *The Fundus in Systemic Disease*

PART V *The Fundus in Local Disease*

The Normal Eye

ALTHOUGH A DETAILED KNOWLEDGE of the normal anatomy of the eye and orbit is not necessary for an understanding of most of the conditions illustrated in this Atlas, some reminder of the gross anatomy and its terminology may be helpful as an introduction.

The Eyelids The eyelids consist essentially of a plate of condensed fibrous tissue (the tarsal plate) lined internally by conjunctiva and covered externally by the orbicularis muscle and skin. The Meibomian glands are embedded in the tarsal plate and open on the free margin of the lid very close to its posterior border.

The palpebral fissure is the almond-shaped space formed when the lids are open. It will be seen from Fig. 1 that normally the upper lid cuts across the upper part of the cornea, while the lower lid margin is related to the junction between cornea and sclera – the limbus.

The inner and outer angles of the palpebral fissure are known as the inner and outer canthi and at the inner canthus can be seen the caruncle and plica semilunaris. On the lid margin by the plica semilunaris is a small elevation known as the papilla lacrimalis in the centre of which is a hole, the punctum lacrimalis through which the tears flow. The punctum lies in close apposition to the globe and cannot normally be seen unless the lid is everted.

The Globe The eyeball is so positioned in the orbit that the anterior surface of the cornea is just in line with the superior and inferior orbital margins – a useful relation in the assessment of proptosis.

The cornea joins the sclera at the limbus, the corneal epithelium becoming continuous with the epithelium of the conjunctiva which is adherent here to the underlying episcleral tissue. Elsewhere the conjunctiva forms a loose covering for the globe and extends peripherally to form two pockets – the upper and lower fornices, before continuing on to the posterior surface of the lids (Fig. 3).

The anterior chamber is the space enclosed by the cornea anteriorly and the lens and iris posteriorly. The pupillary margin of the iris is in constant contact with the anterior surface of the lens, although aqueous humour is able to flow from the posterior chamber (the small space between the periphery of the iris and the lens) through the pupil into the anterior chamber, from which it drains through the trabeculae into Schlemm's canal.

The iris, ciliary body and choroid form a continuous structure called the uveal tract which is derived embryologically in part from the anterior portion of the optic cup and in part from the surrounding mesoderm.

The choroid is a purely vascular structure which supplies the outer one-third of the retina with blood. Fig. 3 is a diagrammatic sagittal section through the eye and orbit, showing the gross relations of the bony orbit, lids, globe and intra-ocular structures.

The Slit-Lamp Microscope Several conditions in this Atlas are illustrated by drawings of slit-lamp appearances and for those unfamiliar with the apparatus a brief description is included here.

The instrument consists of two parts; a low-power binocular microscope mounted horizontally and an illumination system which provides a bright slit of variable width focusing sharply at the point of focus of the microscope. It is designed primarily to examine the transparent media of the eye; i.e. the cornea, aqueous humour, lens and vitreous. The narrow slit beam gives the effect of an optical section in transparent or semi-transparent structures and clearly demonstrates differences in optical density due to anatomical structure or pathological change.

As can be seen in the drawing of a normal eye (Fig. 4) the anterior and posterior surfaces of the cornea show clearly, the corneal stroma reflects some light but the normal aqueous humour is optically empty. The anterior surface of the lens shows well and the discontinuity of the layers of the lens substance can be seen.

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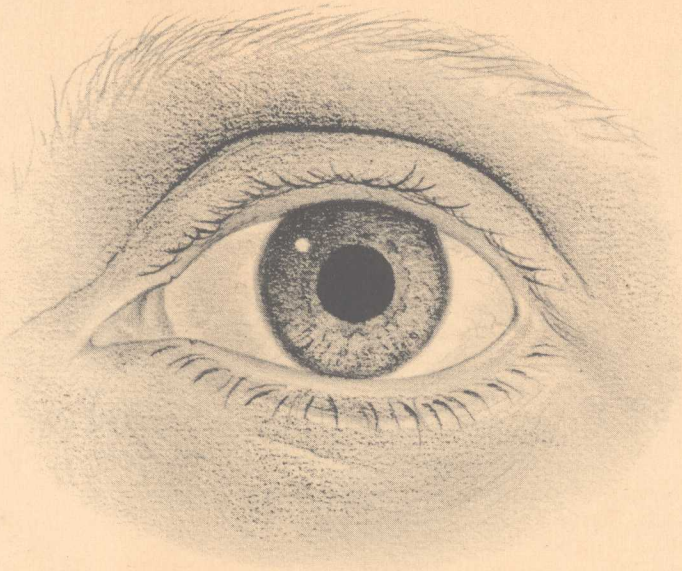

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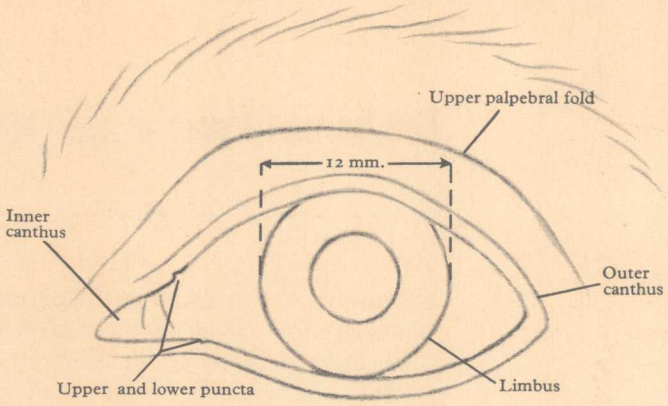

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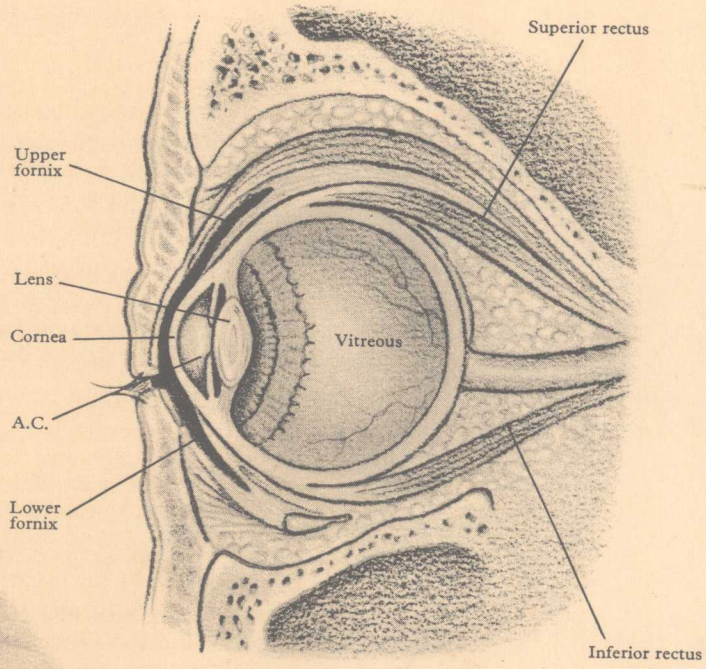
FIG. 1. } EXTERNAL APPEARANCE OF NORMAL EYE
FIG. 2. }

FIG. 3. SAGITTAL SECTION THROUGH ORBIT

FIG. 4. THE EYE AS SEEN BY SLIT-LAMP ILLUMINATION



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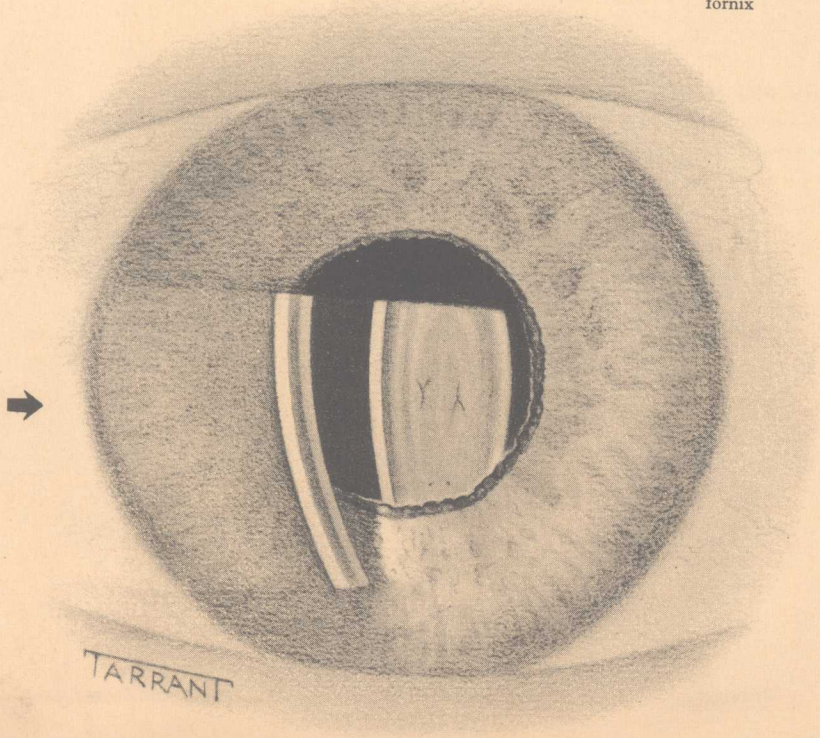

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Congenital Defects of Lids and Orbit

Coloboma of the Lids A notch in the lid margin is not uncommonly found as a congenital defect. Although the term coloboma is used to describe this condition, it is in no way analogous to the coloboma found in the iris or choroid and described later in this series. The defect may appear as a small notch in the upper lid margin or may extend to involve the whole thickness of the lid, as can be seen in Fig. 1. This photograph also shows the corneal scarring which may occur due to the exposure of the cornea by the lid defect. The pathogenesis of the condition is not entirely clear, as in no stage of development does the lid normally have a cleft. It seems probable that the defect is due to pressure of amniotic bands during development of the embryo.

Epicanthus The term epicanthus is used to describe the vertical skin folds at the inner canthi, seen in the photograph, Fig. 2. Such folds are normal during foetal development from the third to the sixth month but, in the Caucasian races, they have normally disappeared by birth. In the Mongolian races, on the other hand, the condition persists into adult life, giving rise to the typical Mongolian eye. When it persists in the Caucasian races the child is seen to have a broad flat nose with widely separated eyes and often an apparent convergent squint. Careful examination, however, will show that the eyes are actually straight and the epicanthal folds and the apparent squint can frequently be made to disappear by pinching up the loose skin over the bridge of the nose. Many mild cases cure themselves when the nose develops normally at puberty or before. In more severe cases operative procedures are necessary to remove the skin folds.

Ptosis A drooping lid on one or both sides is a common congenital defect. The degree of ptosis varies from a hardly perceptible narrowing of the palpebral fissure to a complete paralysis of elevation of the upper lid, which hangs down obscuring the pupil, Fig. 3. The patient attempts to remedy the condition by raising the eyebrow, by contracting the frontalis muscle and tilting the head back, producing a very typical appearance. The absence or weakness of the levator palpebrae superioris muscle, which is the cause of the ptosis, may be associated with weakness or absence of the superior rectus muscle as evidenced by poor elevation of the globe on the affected side.

Dysostosis of the Skull There are several conditions in which too early fusion of the cranial sutures results in deformities of the skull. Oxycephaly, Crouzon's disease and hypertelorism are clearly recognizable clinical types and a photograph (Fig. 4) of a child with oxycephaly has been chosen to represent the group. The abnormal development of the orbits causes proptosis, and stretching of the optic nerve may result in papilloedema and optic atrophy, causing defective vision.

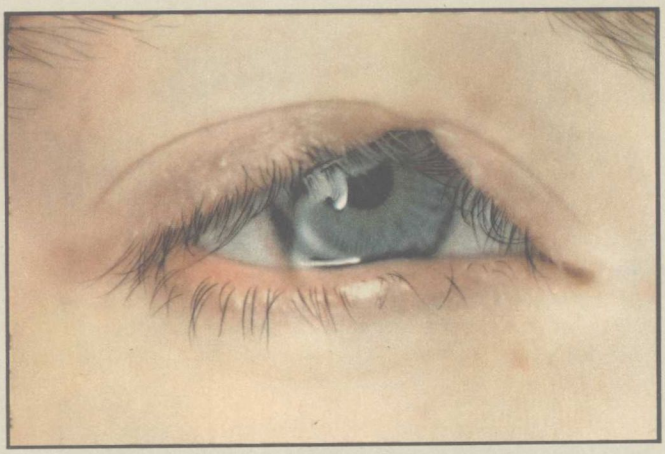

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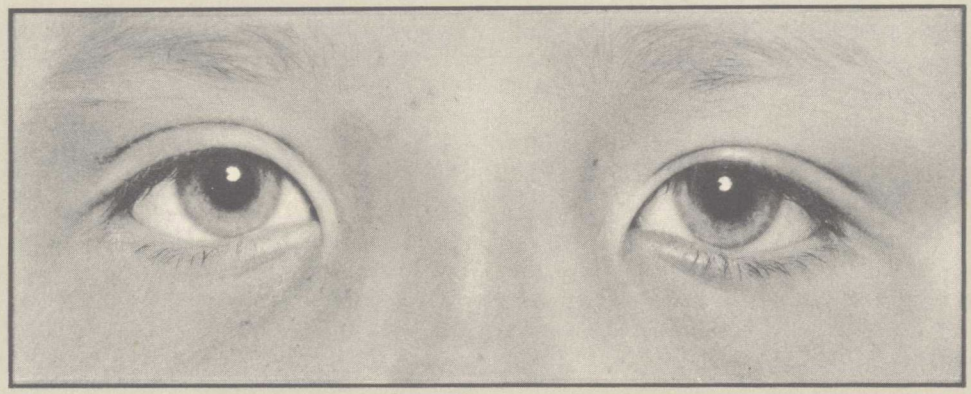
FIG. 1. COLOBOMA OF THE LID WITH CORNEAL SCARRING

FIG. 2. EPICANTHUS

FIG. 3. CONGENITAL PTOSIS

FIG. 4. OXYCEPHALY



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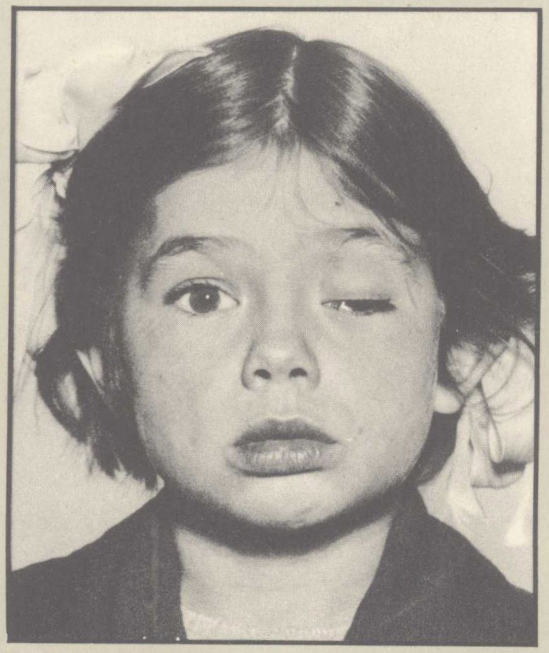

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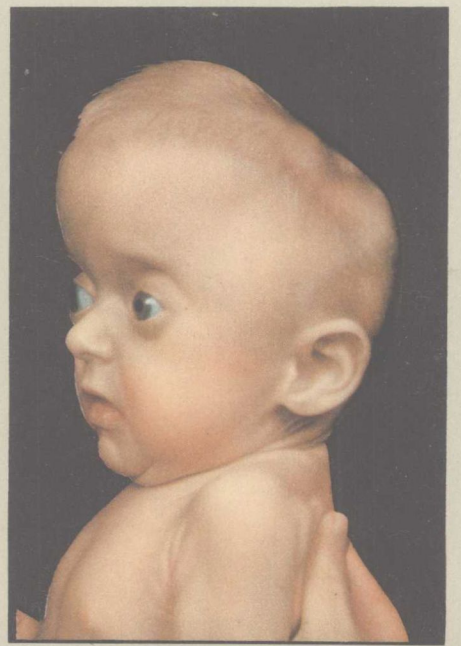

(*Hospital for Sick Children, London*)

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Inflammatory Lesions of the Lids

Hordeolum or Stye This well known and common condition is essentially a staphylococcal infection of a lash follicle and corresponds to a boil of the skin elsewhere. It starts as a painful swelling of the whole lid so that at first it may be difficult to find a localized lesion, but soon one area becomes more swollen and, as pus forms, a yellow point associated with an eyelash can be seen near the lid margin, Fig. 1.

The differential diagnosis is from an acute inflammation of the Meibomian glands – the so-called hordeolum internum. A stye is in the skin and always associated with the lashes, while a Meibomian infection is in the tarsal plate and the skin is not primarily involved. Examination of the conjunctival surface of the lid in hordeolum internum will show a red velvety area with a central yellow spot, through which pus will later discharge. As the Meibomian glands are embedded in tough fibrous tissue, pain and reaction may be more severe than in an ordinary stye. These acute infections of the lid may be associated with acne, general debility or such conditions as diabetes.

Local treatment is by heat until the abscess points, when it may be opened to allow drainage of the pus. Removal of the affected lash is frequently sufficient in a hordeolum externum. If the infection is severe, systemic treatment with antibiotics should be considered.

Chalazion This is a chronic affection of the Meibomian glands. A painless firm lump appears in the lid and slowly increases in size, Fig. 2. Symptoms are few, although occasionally pressure on the cornea may produce some astigmatism.

Frequently called a Meibomian cyst it is, however, not truly cystic but a chronic granuloma caused primarily by the retention of the secretion of the gland. The skin moves freely over the swelling and if the lid is everted a grey spot surrounded by inflamed conjunctiva will be seen at the site of the lesion. Treatment is incision and curettage through the conjunctival surface of the lid.

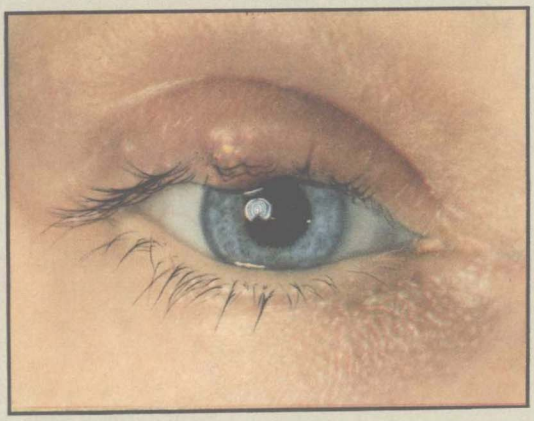
Blepharitis Chronic inflammation of the lid margins is a very common and distressing condition. The inflammation may be mild and consist simply in a hyperaemia of the lid margin with scaling of the skin (squamous blepharitis) or more severe and affect the lash follicles, leading to destruction or distortion of the lashes and deformity of the lid margin (ulcerative blepharitis).

Both types are commonly associated with seborrhoea of the skin. Attention to the general health is of great importance in the treatment and local applications to the lids will not prevent recurrence unless the underlying cause is removed. Fig. 3 shows a long-standing case of ulcerative blepharitis in which the lid margins are deformed, many lashes are missing and others are distorted and turn in to rub on the cornea.

Acute Dacryocystitis Although acute inflammation of the lacrimal sac is not a lid condition it has been included here because it may have to be considered in the differential diagnosis of inflammatory swellings at the inner canthus.

Acute dacryocystitis may be an incident in chronic infection of the lacrimal sac or may be an initial infection. Fig. 4 shows the typical appearance of the swelling below the medial canthal ligament. Lacrimal drainage is blocked, thus causing epiphora. Pressure over the sac (if this is not too painful) may cause regurgitation of pus through the puncta.

Treatment consists of local heat and systemic antibiotics but incision may be required.



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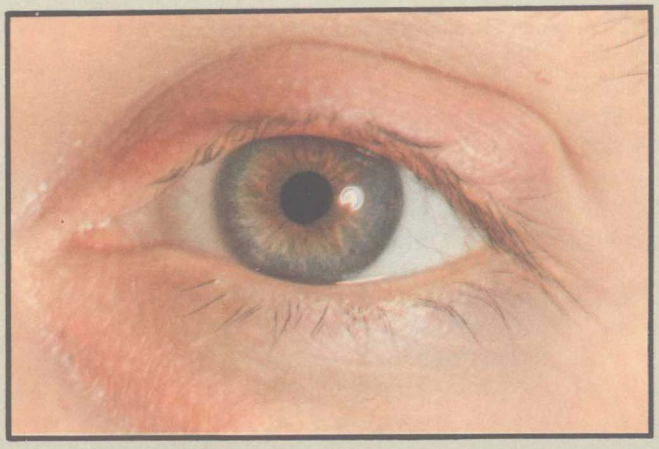

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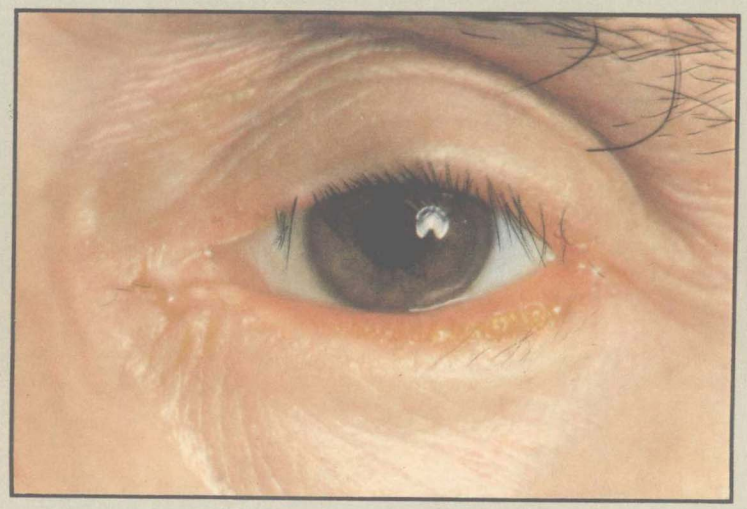

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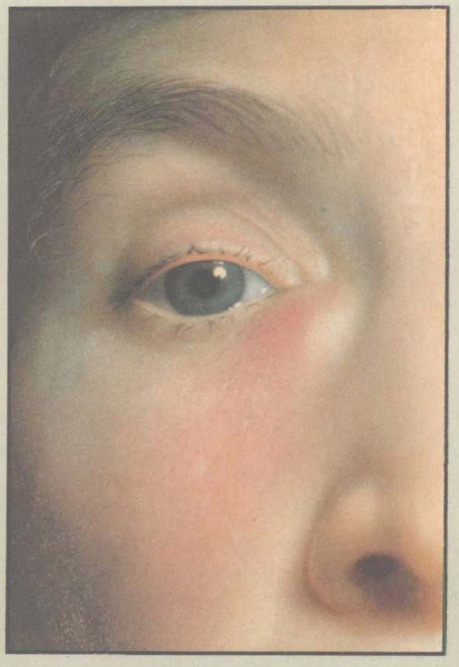

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FIG. 1. STYE (HORDEOLUM EXTERNUM)

FIG. 2. CHALAZION

FIG. 3. ULCERATIVE BLEPHARITIS

FIG. 4. ACUTE DACRYOCYSTITIS

Neoplasms of the Lids

Benign Simple papillomata, particularly along the lid margin as shown in Fig. 1, are common on the lids but require no special treatment except excision for cosmetic reasons. Haemangiomata are less common but of more interest; they are sometimes accompanied by meningeal lesions while a choroidal angioma may occur with other congenital abnormalities of the eyes, causing buphthalmos or infantile glaucoma. Fig. 2 illustrates the typical 'port wine stain' due to a capillary angioma.

Cavernous haemangiomata may occur in the lids and give rise to bluish soft swellings which can be reduced by pressure over them. They are congenital but tend to grow rapidly in the first four years of life and may cause ptosis or exophthalmos. Treatment is by surgical excision, the insertion of radon seeds or the injection of sclerosing fluids into the vessels.

Malignant The lids and skin of the nose near the inner canthus are very common sites for the development of carcinoma in older people. Basal-celled carcinomata or rodent ulcers are more common than the squamous epitheliomata and are characterized histologically by downgrowths of solid darkly staining cells into the dermis. Clinically a rodent ulcer starts as a small nodule in the skin which gradually enlarges and breaks down to form an ulcer with an indurated base and rolled edges. Bleeding from the ulcer is common but any skin nodule which has been present for several months in a patient over the age of forty should be viewed with suspicion. Early removal may save the otherwise inevitable growth of the tumour with much destruction of the lid tissues. However, in the case of large tumours, irradiation may be preferable as excision would leave too big a defect in the lid.

Fig. 3 is a photograph of a typical basal-celled carcinoma of the lower lid. The rolled edges and breaking down base of the ulcer can be clearly seen.

The treatment of choice is excision of the tumour, for although rodent ulcers are very sensitive to irradiation there is some danger to the eye or lacrimal passages in irradiating the lids.

Squamous-celled carcinoma is less common than rodent ulcer but tends to be more malignant and may metastasize to the lymph nodes in the pre-auricular or the submaxillary region. Histologically the tumour shows more resemblance to the general structure of the epidermis. Well-developed prickle cells surround areas of squamous cells which undergo their normal degeneration to form cell nests of acid-staining cornified epithelial cells.

Clinically it appears either as an ulcerated area less symmetrical than a rodent ulcer or as a papillomatous growth. Local extension occurs slowly but relentlessly, eating away the lids, the soft structures of the orbit and even the bone itself if the tumour is left untreated.

Fig. 4 shows a squamous-celled carcinoma which has destroyed a considerable amount of the tissue of the lower lid; in such cases plastic surgery is required to fill in the defect after excision of the growth. Such cases illustrate the importance of early diagnosis and treatment of malignant neoplasms of the lid.

Treatment is the same as that for rodent ulcer, particular care being taken to remove the whole tumour, as the mortality rate is appreciable. Careful follow-up examination is necessary for many years.

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