

# SYSTEM OF OPHTHALMOLOGY

Edited by  
SIR STEWART DUKE-ELDER

VOLUME XIII  
THE OCULAR ADNEXA

PART II  
LACRIMAL, ORBITAL  
AND PARA-ORBITAL DISEASES

# SYSTEM OF OPHTHALMOLOGY

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SIR STEWART DUKE-ELDER

VOL. XIII

## THE OCULAR ADNEXA

By

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*WITH 1,300 ILLUSTRATIONS AND 6 COLOURED PLATES*

PART II

LACRIMAL, ORBITAL AND PARA-ORBITAL DISEASES

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## CHAPTER IX

# DISEASES OF THE LACRIMAL GLAND

TWO NAMES connected with diseases of the lacrimal gland will always be remembered—Otto Schirmer and Henrik Sjögren.

OTTO SCHIRMER [1864–1918] (Fig. 644) was born in Greifswald and, after studying in the university of that city as well as in Munich, Freiburg, Königsberg and Halle, succeeded his father, Rudolf Schirmer, to the Chair of Ophthalmology in Greifswald in 1896; he subsequently went to Kiel (1907) and Strassburg. His contributions to ophthalmology were considerable and varied, the best known being his work on the histology and biochemistry of cataract, his original description of the corneal complications of vaccinia of the eyelids, the first detailed study of rosacea keratitis, and his elaborate description of sympathetic ophthalmitis in the second edition of the *Graefe-Saemisch Handbuch*. His name, however, is most frequently remembered for his work on the lacrimal passages which included the elegant and simple test for the estimation of lacrimation which bears his name (1903); this was followed by his treatise on the microscopic anatomy and physiology of the lacrimal system in the second edition of the *Graefe-Saemisch Handbuch* in 1904, a hereditary task since his father had written on the diseases of the lacrimal apparatus in the first edition of this work (1877). In 1909 he left Strassburg and emigrated to New York where he joined the staff of the New York Postgraduate Medical School, the Herman Knapp Memorial Eye Hospital, and St. Marks and the Bronx Hospitals.

HENRIK SJÖGREN [1899—] (Fig. 645) was born in Köping in Sweden and studied medicine at the Karolinska Institutet in Stockholm. After working in the ophthalmic clinics of two hospitals (Serafimer and Sabbatsberg) in Stockholm he brought out his classical Thesis on keratitis sicca associated with atrophy of the lacrimal gland at the Karolinska Institutet in 1933. Thereafter he became ophthalmic surgeon at Jönköping (1936–67), receiving a doctorate in ophthalmology at the University of Göteborg in 1957 and an honorary professorship in 1961. His extremely detailed and elaborate studies on the local and systemic manifestations of the condition now generally known as Sjögren's syndrome have been universally acclaimed as a model of clinical and pathological investigation and have well earned him a quiet retirement in Lund.

A considerable amount of information on the lacrimal apparatus has already been incorporated in other Volumes of this *System*—its evolution and characteristics in different species of animals,<sup>1</sup> its anatomy,<sup>2</sup> embryology,<sup>3</sup> physiology,<sup>4</sup> congenital anomalies,<sup>5</sup> the neurological conditions that may affect it<sup>6</sup> and injuries.<sup>7</sup> In this Volume we shall confine ourselves to the diseases to which it is subject.

<sup>1</sup> Vol. I, pp. 345, 349, 367, 375, 425, 437, 441, 493.

<sup>2</sup> Vol. II, p. 562.

<sup>3</sup> Vol. III, p. 237.

<sup>4</sup> Vol. IV, p. 410.

<sup>5</sup> Vol. III, p. 911.

<sup>6</sup> Vol. XII, p. 957.

<sup>7</sup> Vol. XIV, pp. 306, 434, 443, 652.

## DISORDERS OF SECRETION

It will be remembered that the secretion of tears is of two types, basic and reflex; the former, derived mainly from the tarsal and conjunctival glands and the accessory glands of Krause and Wolfring as well as the glands of Zeis and Moll, should maintain the normal hydration of the outer surface of the eye and produce the components of the pre-corneal film; the reflex secretion, derived from the main and accessory (palpebral) lacrimal glands, is excited mainly through its parasympathetic nerve supply by several factors.<sup>1</sup> The basic secretion is present in all terrestrial vertebrates (and the whale), occurs during sleep, is the only lacrimal secretion in the infant during the first weeks of life, and thereafter remains relatively constant until about the age of 60 when it gradually decreases; the reflex secretory type is stimulated by sensory, retinal (light) and psychogenic (central) stimuli.

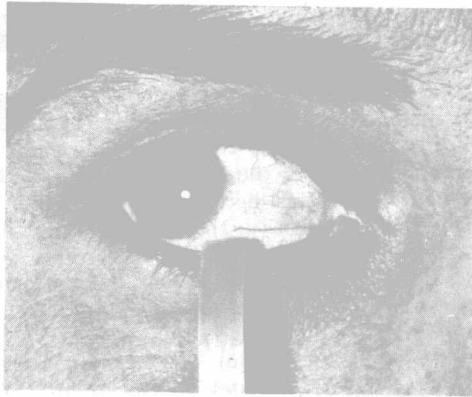


FIG. 646.—SCHIRMER'S FILTER-PAPER TEST.

For measuring the amount of lacrimal secretion; see text.

A useful clinical method of investigation of the total lacrimal secretion is *Schirmer's filter-paper test* (1903). A strip of filter-paper (0.5 cm.  $\times$  3.5 cm.) is folded 5 mm. from one end so that the short flap can be inserted into the lower fornix and the fold overlaps the ciliary margin (Fig. 646); when the patient is in subdued light the normal secretion should moisten at least 1.5 cm. of the strip as measured from the fold at the ciliary margin in 5 minutes. The test suffers from the disadvantage that over-secretion may occur owing to the conjunctival irritation caused by the filter-paper (a very variable factor) but, although it has no pretence to accurate assessment, it provides an indication of excessive lacrimation or hyposecretion.<sup>2</sup>

If hyposecretion is found to exist, the basic secretion test of Jones (1966) can analyse its nature. Schirmer's test is repeated in very subdued light or in the dark after the conjunctiva has been anaesthetized by the instillation of a topical anaesthetic enhanced by placing a cotton-tipped applicator soaked in 5% cocaine hydrochloride for 30 seconds to the part of the conjunctiva which will be in contact with the filter-

<sup>1</sup> Vol. IV, p. 419.

<sup>2</sup> de Röttg (1941), Gifford *et al.* (1943), Eisner (1960), Wright and Meger (1962), Jones (1966), Jones and Linn (1969).

paper. Reflex secretion excited by light and irritation is thus eliminated and the basic secretion alone occurs; it should produce 10 mm. of wetting on the filter-paper. If the basic secretion is defective the absence of its constituents results in a considerable amount of irritation; if excessive lacrimation occurs in these circumstances, it is due to an increased reflex secretory output (a *pseudo-epiphora*, Jones, 1966).

If the basic secretion is normal and the reflex secretion is apparently defective, *Schirmer's No. 2 test* (1903) should be applied wherein this element in lacrimation is abnormally stimulated through the nasal mucosa by smelling ammonia or the homely onion or by a cotton-tipped applicator placed in the region of the anterior end of the middle turbinate and moved about. If the filter-paper is moistened within 2 minutes there is a "fatigue block" for stimuli from the conjunctival but not from the nasal sensory nerves; if it is not moistened a complete failure of the secretory mechanism is present.

Other techniques have been suggested based on the introduction of dyes into the conjunctival sac and the determination of the rate at which the colour fades in the fluid, which may be done by photometry (Norn, 1965-66; Mishima *et al.*, 1966; Maurice, 1967). It is interesting that the average results of the tear-flow thus obtained approximate those of Schirmer, averaging 0.5 to 0.75 g. in 16 waking hours, although higher figures have been obtained (Thaysen and Thorn, 1954; Norn, 1965).<sup>1</sup>

It should be noted that *hyperlacrimation* must be differentiated from *epiphora* due to blockage of the lacrimal passages. This will be discussed in the following Chapter, but a useful clinical test to determine their patency depends upon the passage of a dye such as fluorescein from the conjunctival sac into the nose. This is verified by the presence of the dye after 2 minutes on a wisp of cotton wool wound round an applicator of thin flexible wire inserted 1½ inches into the inferior meatus of the nose (the *primary dye test* of Jones, 1962). If the test is positive there is no obstruction in the lacrimal passages and tearing is due to hypersecretion.

#### LACRIMAL HYPERSECRETION

Excessive lacrimation is not nearly so common as epiphora nor does it usually give rise to so much distress. It frequently occurs intermittently in paroxysms in which case it produces no ill-effects apart from annoyance and social or cosmetic embarrassment. A constant oversecretion of tears, on the other hand, as a constant epiphora, tends eventually to produce a chronic irritation and maceration of the lower lid leading to a lacrimal conjunctivitis with hypertrophic changes in the mucosa (Capolongo, 1950),<sup>2</sup> a chronic blepharitis<sup>3</sup> and sometimes palpebral eczema. Organic complications of this kind, however, are relatively rare.

The *aetiology* of lacrimation is varied; the causes may be classified as primary (due to a disturbance of the lacrimal gland itself), central or psychic (including hysteria), neurogenic (brought about by reflex processes), irritative (as by lacrimatory gases) and symptomatic (occurring in the course of certain diseases). All of these except the first have been discussed in other Volumes where the relevant bibliographies will be found.<sup>4</sup>

**PRIMARY LACRIMATION.** that is, *lacrimation due to direct disturbance of the lacrimal gland*, is rare and is not a marked clinical symptom. It may be

<sup>1</sup> Vol. IV, p. 424.

<sup>2</sup> p. 208.

<sup>3</sup> Vol. VIII, p. 81.

<sup>4</sup> Vol. XII, p. 959; Vol. XIV, p. 1153.

produced *pharmacologically* by strong parasympathomimetic drugs; thus mecholyl (acetyl- $\beta$ -methyl choline chloride) injected intraperitoneally in rats induces a flow of tears after 2 minutes which continues for about 5 minutes (Winbury *et al.*, 1949). This type of lacrimation may be inhibited by parasympatholytic drugs (atropine, etc.). A similar excessive lacrimation follows the administration of powerful inhibitors of cholinesterase such as tetra-ethyl-pyrophosphate (TEPP) (Grob and Harvey, 1949). Lacrimation may also occur in *diseases of the lacrimal gland*. It is thus seen particularly in the early stages of the development of cysts and tumours or Mikulicz's syndrome; indeed, the occurrence of lacrimation may be a sign of value in the diagnosis of a swelling in the lacrimal gland fossa suggesting that the gland itself is probably involved.

It may be convenient to summarize the other factors causing lacrimation even although they have been discussed elsewhere.

CENTRAL (PSYCHIC) LACRIMATION, which occurs only after the first few months of life, is associated with emotional states, but may be exaggerated to occur with relatively small stimuli.

NEUROGENIC LACRIMATION results from the stimulation of any branch of the trigeminal nerve but most profusely from those supplying the eye itself. Stimulation of the endonasal mucosa by touching with a probe the inferior margin of the middle turbinate and the tuberculum septi also produces a persistent lacrimation (the *naso-lacrimal reflex*).

Stimulation of the cervical sympathetic may provoke lacrimal secretion but the effect is inconsequential and inconstant.

A reflex lacrimation accompanies such physiological actions as laughing or vomiting, while a similar reaction has been said to be associated with a hyperactive carotid sinus reflex or with the pressure of enlarged lymphatic nodes on the vagus nerve.

The *treatment* of lacrimal hypersecretion from any cause has already been discussed in detail with the relevant bibliography.<sup>1</sup> It will be remembered that if the causal factor cannot be removed so that the excess of tears is eliminated, the classical method of treatment by excision of the orbital or preferably the palpebral lobe of the gland is dangerously drastic, certainly if the conjunctiva is diseased and sometimes also when it is healthy; it runs the risk of changing a wet eye into a dry eye and converting a relatively harmless but annoying functional malady into an organic and incapacitating disease (Sjögren and Kronning, 1951); at the same time palpebral dacryoadenectomy has been done with good results in a large series of patients with the development of only one case of mild xerophthalmia (Taiara and Smith, 1973). Sectioning or diathermic coagulation of the lacrimal ducts, a procedure which can be done in stages, is safer (Wiedersheim, 1928; Jameson, 1937). The alternatives of partial destruction of the palpebral lobe by x-rays or the injection of alcohol are unpredictable in their results, as also is section of the various nerves exciting lacrimation—the sphenopalatine, the greater superficial petrosal, the tympanic branch of

<sup>1</sup> Vol. XII, p. 963.

the glosso-pharyngeal or the vidian. The use of astringent drops or lotions is usually ineffective. On the whole, treatment should be undertaken reluctantly and in stages.

#### LACRIMAL HYPOSECRETION

While lacrimal hypersecretion involves no serious ill-effects apart from its annoyance and discomfort, a serious diminution or lack of tears may result in the development of *kerato-conjunctivitis sicca*—a chronic intractable, irritable conjunctivitis unrelieved by treatment, characterized by a ropy mucoid discharge and often associated with epithelial roughening and the development of fine corneal erosions and filamentary keratitis—a condition which may give rise to a considerable amount of distress and may ultimately involve the development of corneal infiltrates which affect the vision.<sup>1</sup> It is to be remembered also, although it is of less dramatic importance, that the lack of tears involves an absence of the lacrimal lysozyme.

Hyposecretion is not uncommon and appears in several conditions which may be classed as *primary* (due to disease of the gland), *paralytic* (due to paresis of one or other of the nerves supplying the gland) and *toxic*; its occurrence as a *congenital defect* has been noted in another Volume.<sup>2</sup>

**PRIMARY HYPOSECRETION.** Acute diseases of the lacrimal gland do not as a rule seriously affect the secretion of tears, although a diminution may be noted in the later stages of tumours of the gland—a point which may be of diagnostic importance—and is frequently seen in developed cases of Mikulicz's syndrome associated in this case with a failure in salivary secretion. In the atrophic stages of severe inflammatory or other diseases of the gland, however, deficiency of lacrimation is common.

*Atrophy of the lacrimal gland* is associated with a diminution of secretion. This occurs typically in two conditions—*senile atrophy*,<sup>3</sup> and *idiopathic atrophy* (*Sjögren's syndrome*)<sup>4</sup>—both of which are discussed later.

*Excision of the lacrimal gland* or, which amounts to the same thing, of the palpebral lobe, in the great majority of cases has no serious ill-effects, sufficient lubrication being obtained by the basic secretion derived from the accessory glands of Krause and the mucous secretion of the conjunctiva; indeed, Calderaro (1917) found that there was a compensatory increase in the secretion of conjunctival mucus. As a rarity, however, a troublesome kerato-conjunctivitis sicca develops, and these exceptions, although admittedly few, should give pause to the too light-hearted undertaking of the operation particularly in cases in which chronic disease has diminished the function of the conjunctival glands (Wagenmann, 1893; Avizonis, 1928; Weve, 1928; Engelking, 1928; Knapp, 1929; Sjögren and Kronning, 1951).

*Obstruction of the lacrimal ductules* at their conjunctival orifices may

<sup>1</sup> p. 628; Vol. VIII, pp. 128, 694.

<sup>2</sup> Vol. III, p. 913.

<sup>3</sup> p. 625. <sup>4</sup> p. 626.