

Pemphigus of the skin and mucous membrane of the mouth, associated with "essential shrinking" and pemphigus of the conjunctivae / by A. Hill Griffith.

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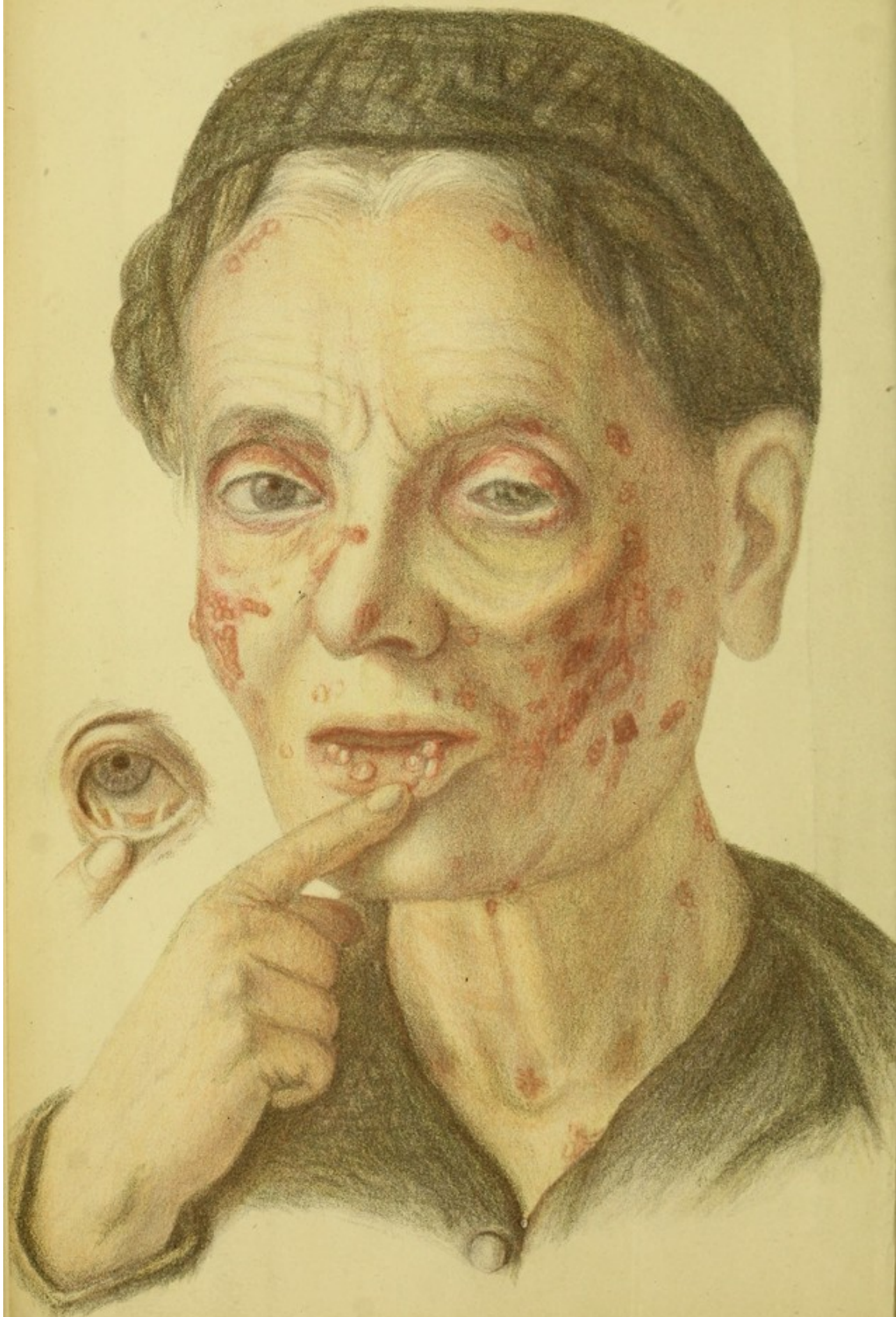
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M^r Malcolm Morris's case of Pemphigus Conjunctivæ, etc.

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PEMPHIGUS OF THE SKIN AND MUCOUS MEMBRANE OF THE MOUTH, ASSOCIATED WITH "ESSENTIAL SHRINKING" AND PEMPHIGUS OF THE CONJUNCTIVÆ.

BY MALCOLM MORRIS, F.R.C.S.E.,

AND

H. LESLIE ROBERTS, M.B.

MARY G., spinster, aged 60, came as an out-patient to St. Mary's Hospital early in September 1888.

Family History.—Both parents are dead. The mother died at the age of 76, of pneumonia; the father at 75 of cancer of lower jaw. There were ten children in all: five died in childhood; the eldest brother died at the age of 41 of consumption; another, a soldier, died abroad, the cause being unknown. There are three sisters remaining. The eldest, aged 71, is described as healthy; the second, aged 68, is not very strong.

Personal history and that of present disease.—When a child she had small-pox, measles, and "cholera." About the age of 21 she suffered from axillary abscesses, which burst. Matter discharged, and healing took place spontaneously. Between the ages of 40 and 50 she received burns upon her legs from her clothes catching fire. Between 50 and 60 had two attacks of rheumatic fever, one in two successive years. She

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was five weeks ill during the first, and eight weeks during the second of these attacks, and was not treated professionally.

She has sustained innumerable small burns and accidents. The patient cannot give a connected account of the course of her present illness. The pemphigus of the skin appears to have started some five years ago, during her recovery from the second attack of rheumatic fever. It had been recurrent and subject to acute exacerbations when the patient took cold. The eye affection had begun with epiphora, and they had, soon afterwards, become inflamed.

She has been a laundress all her life, and during the summer of 1888 had suffered from much domestic hardship, with insufficiency of food.

Present state.—On November 7th, 1888, her condition was as follows:—General bodily and mental state feeble. Has some dysphonia. The skin of almost every part of her body is marked by scars of various ages and appearances. That of both cheeks is converted into glossy, scar-like tissue, of a mottled, purplish colour. The auricles of both ears on their inner surface and the forehead presents similar scar-like patches. Both upper extremities, on their flexor and inner aspects, are covered with scars. Most of these have a purple colour, and are raised, and when grasped between the fingers feel thicker than the normal skin. Others, much smaller, are thin, and have a pearly lustre, the skin between them being deeply pigmented. On the chest, abdomen, back, thighs, and legs are scars quite similar in appearance to those on the arms; some of them are covered with broken, dead epidermis. Many of these scars the patient attributes to burns and other accidents; only those on the arms and face seem due to pemphigus.

On the mucous membrane of the hard palate is a white shrunken patch about 1 cm. broad, and surrounded by a dark red areola.

The left eye.—The palpebral fissure is narrowed transversely and vertically. Both lids are of a red colour, and the upper one partially droops. On depressing the lower lid the *cul-de-sac* is seen to be nearly obliterated, and several bands or bridges of conjunctiva, pale and thickened, pass from the palpebral to the ocular surface. The ocular conjunctiva is

congested, and the cornea is nearly opaque. The iris is muddy and the pupil rigid. Vision is nearly lost, but the perception of light remains. Movements of the globe are restricted in the following directions—outwards, upwards, and downwards.

The right eye.—This is not so badly affected as the left. There is some shrinking of the lower *cul-de-sac*. The cornea is clear. The pupil reacts to light and during accommodation. Vision is limited to large type at near distances.

No bullæ or vesicles are visible on any part of the skin, mucous membrane, or conjunctivæ.

Progress of the case. November 26th.—The remains of a recent bulla exists on the right temple. On the skin of the lower lip is a clear vesicle, about 3 mm. broad. On the mucous membrane of that lip, of the floor of the mouth, and of the hard palate, are a few unbroken vesicles.

December 3rd.—She entered the hospital as an in-patient. The shrinking of the conjunctiva of the right eye has made considerable progress.

January 1st, 1889.—The condition is about the same as on Dec. 3rd. There are no fresh vesicles, but the old vesicles still exist in the mouth, and are unbroken. The dysphonia is more marked, the patient speaking in whispers.

Jan. 4th.—An unbroken vesicle was seen on the lower part of the conjunctiva bulbi of left eye.

Feb. 28th.—Fresh bullæ have formed, one on the front of the neck, two others on the mucous membrane of the lower lip. At the outer angle of right eye the conjunctiva has shrunk into narrow bands, which pass in a vertical direction from one lid to another (see plate, in which the right eye has been drawn separately). The ciliæ of both lids are curved inwards, and brush against the cornea and the sclerotic conjunctiva. On the right eye only a few ciliæ of upper lid at the inner angle are misdirected, the remaining ones being normal.

March 10th.—Fresh vesicles continue to make their appearance upon the mucous membranes of the lower lip and the floor of the mouth.

REMARKS.—We have collected and arranged in the form of a table all the cases of pemphigus conjunctivæ, which have been published up to the present time. We have chosen this

method of arranging them, as it offers greater facility for comparing the leading features of the disease, and any uniformity which may exist will thus be readily apparent. Twenty-eight cases are on record, and though others have been seen, they have either not been recorded, or merely briefly referred to. Of these twenty-eight cases three are by French, seventeen by German, three by American, and five by English observers. As regards the sex, excluding three cases described by Klemm as "children," twelve were females, and thirteen males. Their ages varied from infancy to the age of 76, the average being about 40 years. The general previous health is described in some as "good," and in others as "bad." Some were in a condition of ill-health and of emaciation at the onset of the disease, or when coming under observation; while others are described as being in excellent health, and well nourished and developed. In only one case was there a history of syphilis, and that was ten years before the commencement of the pemphigus.

Out of the twenty-eight cases sixteen began on the skin as pemphigus vulgaris or foliaceus, four on the mucous membranes, and eight on the eye. One eye always became affected before the other, but the lapse of time before the second became affected varied from a few days to two years. In twelve out of the whole number of cases vesicles or bullæ were observed upon the conjunctiva either in an unruptured state or soon after bursting. In some cases a diphtheritic pseudo-membrane is described as forming upon the conjunctiva. Reference to the column in the table headed "Mode of Termination," will show how uniform are the changes which are ultimately produced in the conjunctiva. These changes manifest themselves by a characteristic shrinking of the conjunctiva of the lids and of the globe. The result of this is twofold: first, a deformity of the lids, by which the ciliæ are directed against the corneal and scleral conjunctiva, and secondly the irritation of these structures, which results from this entropion. Sooner or later the cornea of the affected eye becomes cloudy, and the sclerotic conjunctiva thickened. They finally become dry, a condition which some observers have compared to xerophthalmia. Vision may be

impaired even before the cornea becomes affected, but then it is amenable to treatment. Generally vision in one or both eyes is lost, but the perception of light always remains. Perforation of the cornea, prolapse of the iris, and total destruction of the globe have been recorded.*

With reference to the treatment of pemphigus conjunctivæ unfortunately little can be said. There are two broad lines upon which it has hitherto been attempted: (1) the operative, (2) the palliative. The former includes operations for entropion, epilation of lashes, transplantation of rabbit's mucous membrane. Of these, the operations for entropion have given temporary relief, but the transplantation experiments have completely failed. The palliative measures consist in improving the general health, in alleviating the dryness of the eye with glycerine or mucilaginous lotions, and in the application of soothing remedies to allay the inflammation.

But in spite of all known treatment the shrinking maintains a progressive and uninterrupted course till total blindness results. In one case only, that of Samelsohn, mentioned by him at the 12th Congress of the Ophthalmological Society at Heidelberg, 1879, is a more favourable termination recorded. Here the cornea cleared up, and the trichiasis almost disappeared, while the conjunctiva appeared shrunken to only a trifling extent.

There is no question about the rarity of this affection, as the following facts will show. While Arlt affirms that in an experience of forty years he had only seen one case of pemphigus conjunctivæ in the practice of Professor Stellwag, in the Ophthalmic Hospital in Vienna, Hebra, out of 200 cases of pemphigus of the skin had not seen a single case. Steffan, of Frankfurt, amongst 84,000 patients had not seen (up to 1884) more than one in which vesicles formed on the conjunctiva in association with shrinking. Horner, out of 70,000 patients, had seen only three cases of shrinking of the conjunctiva. Professor Gräfe, at the congress referred to above, mentioned having seen four cases which he described as *essentielle Schrumpfung der Binderhaut*.

The pathology of this disease is still obscure. As yet we

* See Steffan's case, No. 17 in Table.

have the records of only one post mortem examination, and of three microscopical investigations. Unfortunately these investigations do not throw much light upon the etiology of the disease, neither are they uniform in their results. In Gelpke's case,* the autopsy showed internally nothing specially abnormal beyond extreme anæmia of organs and general atrophy. Gelpke,† Bäumlér,‡ and Sattler§ have examined the altered conjunctiva and sclerotic microscopically. From Sattler's preparation it appeared that the swelling of the conjunctivæ bulbi was not due to infiltration of conjunctiva with lymphoid cells, but partly by a great swelling of the connective tissue bundles, and partly from the expansion of the fissure-spaces which were filled with liquid. The subepithelial layer of the conjunctiva bulbi formed a specially modified membrane of about 30–40 μ . broad. It was much more turbid than the rest of the tissue, being almost opaque, and running parallel with the surface of the conjunctiva, and covering the greater part of the cornea. This subepithelial turbid layer might easily be mistaken for the epithelium itself at first sight. The entire stroma of this layer was pervaded by granular bodies, which did not stain with bismarck brown or hæmatoxylin. The blood vessels of conjunctiva were very numerous, and much congested. They were not, however, apparent on a superficial view of the conjunctiva, being concealed by the condition of the epithelium. This epithelium had lost totally its normal character. From the deepish layers to the most superficial it consisted of horny cells. It was 30 μ . thick, and separated easily from the underlying conjunctiva. Bäumlér admits that the description of his preparations does not differ essentially from that of Sattler. He failed, however, to find the subepithelial turbid layer which the latter observes and describes. On the other hand, the preparations of Sattler did not show the hypertrophy and the papillary formation of the epithelium, which Bäumlér had described. For further details we must

* Zehenders klin. Monatsblätter f. Augenheilkunde, vol. xxiii. p. 199.

† *Ibid.* p. 206.

‡ *Ibid.* p. 341

§ Rep. of 12th Congress of Ophthal. Soc. at Heidelberg (Zeh. Monatsbl. Augenhilke., vol. xvii.



TABLE OF RECORDED CASES OF PEMPHIGUS CONJUNCTIVÆ.
CHRONOLOGICALLY ARRANGED.

Arrangement of Cases in Chronological Order of their publication.	Name of Observer.	Reference to Original Article.	Number of Cases described by each Observer.	Sex and Age of Patient.	General previous Condition.	Part first attacked.	Position in which bullæ were observed.	Cases in which bullæ or vesicles were actually seen on the conjunctiva.	Mode of Termination.
1. 1858	White Cooper	Roy. Lond. Oph. Hospital Rep., Vol. I.	1	F. 24	Bad health.	Skin.	Arms and legs.	On both globes.	Right Eye. Lower lid adherent to globe by bands. Left Eye. Similar adhesions; vision unimpaired.
2. 1868	Wecker	Klinische Monatsblätter f. Augenheilkunde, v. Zehender, Vol. VI, p. 232.	1	M. 68	Good.	Face.	Nose, mouth, and the neighbourhood.	Conjunctiva bulbi of left eye.	Left Eye. Palpebral fissure narrowed, lids almost completely adherent to globe; cornea dry, vision limited. Right Eye. Lids partially adherent to globe, cornea and vision normal.
3 & 4. 1869	Lasègue Bazin Hardy Mahmond	Monographie du Pemphigus, Paris, 1869, par Hassan Effendi Mahmond; cited by Pflüger Zeh. Klin. Monatsbl. f. Augenheilkde. XVI. 1878, p. 1.	1	M. 60	Megrin and indigestion. No syphilis.	Skin of nose.	Pemphigus foliaceus ala nasi, lower lid of right eye, mucous membrane of throat and œsophagus.		Left Eye. Lower lid adherent to globe; vision lost. Right Eye. Conjunctiva not affected.
			1	M. 71	Suffered from pemph. 11 years ago; recovered.	Skin generally.	Skin in all parts; pemphigus foliaceus.	Three bullæ seen on left eye; later two bullæ seen on right eye.	Xerophthalmia of right cornea, left similar; later, death.
5-7. 1870	Klemm	Archiv. f. Klin. Med. Vol. IX., p. 199.	3	Young Children		Skin.	Skin in all parts; mucous membrane of mouth.	Numerous vesicles seen on conjunctivæ in all three.	Violent inflammation of eyes resembling diphtheria. 1. Death. 2. Recovery, very slow.
8. 1875	Kunkel	"Zwei Fälle vom Pemph. foliac.", Internationale homœopathische Presse V. 6, 1875, Kiel; cited by Pflüger, l.c., p. 4.	1	F. 26	Never healthy.	Mucous membrane of throat.	Skin in all parts; mucous membrane of mouth and throat.	On conjunctivæ of lids and globe.	Adherence of palpebral conjunctiva to globe. A membrane formed over cornea; disappeared gradually.
9. 1877	Pflüger	Zehenders Klin. Monatsbl. f. Augenheilkunde. XVI, 1878, p. 1.	1	M. 53	Gastric catarrh frequently during late years; also chron. furunculosis. Has never had syphilis.	Mucous membrane of mouth and throat.	Skin in all parts; mucous membrane of mouth; œsophagus and all alimentary canal; pemphigus foliaceus.	On lower lid left eye.	Right Eye began first, lids adherent to globe, cornea ulcerated and cloudy; ectropion of both lids. Death after 10½ years.
10. 1878	James A. Campbell	Monograph pub. at St. Louis, Mo.; cited by Schweigger in his Handbuch der Augenheilkunde, 4 edit., 1880, p. 390.	1	M. 62		Skin (?)	Skin in all parts; pemphigus vulgaris.	?	Right Eye affected.
11. 1879	Borysiewicz & Arlt	Zeh. Klin. Monatsbl. f. Augenheilkde., Vol. XVII., p. 325.	1	M. 76	Good.	Glans penis.	Skin in all parts; pemphigus vulgaris and foliaceus.	None.	Left Eye. Diphtheritic exudation over palpebral and ocular conjunctiva. Shrinking of both conjunctiva. Right Eye later affected; lids adherent to globe.
12. 1879	Sattler	Rep. of 12th Congress of the Ophthalm. Soc. at Heidelberg, 1879, p. 227.	1	M. 38		Skin.	Skin in all parts.	A few seen on the conjunctiva by his friends.	Right Eye. Palpebral fissure much contracted, conj. sacs almost obliterated, conj. bulbi thickened, entropion of lower lid; sclerotic wax-like appearance, partially ulcerated. Left Eye. Similar, but not so advanced. Death from phthisis pulmonalis.
13. 1881	Arlt	Arlt's Klinische Darstellung der Krankheiten des Auges, 1881, p. 84; cited by Steffan, Klin. Monatsbl. f. Augenheilkde. XXII, p. 271.	1	F. 4	Healthy till 3 years of age.	Skin.	Skin in all parts.	None.	Right Eye. Palpebral fissure could only be slightly opened; conj. palp. adherent to conj. bulbi; obliteration of the conj. sacs. Conj. of lids and globe greyish-red and thickened. Left Eye healthy.
14. 1882	Reich (of Tiflis)	Centrbl. f. prakt. Augenheilkunde, 1882, p. 145; cited by Steffan l.c.	1	M. 19		Skin.	Skin in all parts.		Shrinking of both conjunctivæ.
15. 1882	Schüler	Berl. Klin. Wochenschr., No. 48, 1882, p. 736.	1	F. 8	Good, well-developed.	Skin.	Skin in all parts; mucous membrane of tongue, vulva (and rectum? urethra?).		Shrinking of both conjunctivæ, lids swollen, and red, cilia distorted; thin greyish-white membrane closing left palpebral fissure.
16. 1883	Critchett and Juler	Trans. of the Ophthalm. Society of United Kingdom, Vol. IV., p. 30.	1	F. 50	Good.	Eyes.	Skin of upper lids (?) uvula and larynx (?).	None.	Vision lost in both eyes; perception of light retained; adhesion of lids to globe; shrinking of both conjunctiva. No en- or ectropion.
17. 1884	Steffan	Zeh. Klin. Monatsbl. f. Augenheilkunde, 1884, p. 271.	1	F. 73		Left eye.	Skin of both upper lids some months after onset, mucous membrane of throat, under surface of epiglottis.	Once or twice a vesicle seen on both lower conj. sacs.	2½ years from beginning of disease; total destruction of both sclerotics; perforation; prolapse of iris; both lids on both sides completely adherent to atrophied globes. Blindness, total.
18. 1884	Schweigger	Archiv. f. Augenheilkunde, Knapp and Schweigger, Vol. XIII., 1884, p. 247.	1	M. 70	Had suffered for some years from Pemphigus (?) of mucous memb. of mouth.	Both eyes.	None.	On conj. bulbi of right eye, after bursting.	Shrinking in parts of conjunctiva of both lower lids, with adherence to globe. In right eye one of the points of adhesion corresponded to seat of former vesicle.
19. 1885	Gelpke	Zeh. Klin. Monatsbl. f. Augenheilkunde, Vol. XXIII., p. 191.	1	F. 14 (months)	Poorly nourished, badly developed; a universal	Skin.	Cheeks, chin, palm of right hand, tip of right	On mucous membrane of upper lid, left eye, the	Ulceration and mortification of left cornea. Marasmus, diarrhoea, vomiting, death about 13 days after onset.



					phitis or previous pemphigus.					of crepuscular membrane over conjunctiva; sensation of light retained in both eyes.
23 & 24. 1886	Lang	Trans. of the Ophthal. Society of United King., Vol. VI, 1886, p. 125.	2	(2) F. 9	No evidence of syphilis till 5 years old; quite healthy.	Skin.	Skin in all parts, eyelids, mucous membrane of tongue and mouth.	None.		Right Eye palpebral fissure narrowed, lids shortened, swollen, adherent to globe; cilia turned inwards, sclerotic covered with a membrane. Vision lost. Left Eye not so bad, shrinking commenced, conjunctiva in parts of greyish-white, moist, glistening appearance.
				(1) F. 24	Bad health, suppurating cervical glands.	Skin.	Skin in all parts, mucous membrane of mouth, tongue, pharynx.	None.		After 14 years shrinking of conjunctiva of both eyes; lower conjunctival sacs almost obliterated; vision much impaired; entropion.
				(2) F. 35	Good.	Eyes.	None.	None.		Shrinking of conjunctiva; obliteration of lower cul-de-sacs; entropion of upper cilia; vision almost lost.
25. 1886	Critchett and Juler	Trans. of the Ophthal. Society of United King., Vol. VI, p. 125.	1	M. 50	Syphilis 10 years ago.	Mucous membrane of nose.	None.	None.		Right Eye. Upper and lower cul-de-sacs obliterated, lids adherent to globe, cornea hazy, vision very bad. Left Eye. Cornea clear, lids thickened and brawny, not much shrinking.
26. 1886	Wm. Dickinson	Jour. Am. Med. Assoc., Vol. VII, Chicago, pp. 566-568.	1	F. 56	Good.	Left eye.	On skin of upper eyelids; mucous membrane of throat (?).	Three vesicles on conjunctiva of right eye; four on conjunctiva of left eye; one on margin of upper and lower lids of left eye.		Progressive shrinking of conjunctiva, vision not directly impaired while under observation.
27. 1888	Robert Tulley	Jour. Am. Med. Assoc., Chicago, 1888; also Am. Jour. Ophthal., St. Louis, 1887, Vol. IV, pp. 145-152.	1	M. 12		Eyes.	Skin, on various parts of it, one on mucous membrane of mouth.	None.		Lids adherent to globe, cornea opaque and horny, vision lost, sense of light retained.
28. 1889	Malcolm Morris and H. Leslie Roberts	Brit. Jour. of Derna., London, Vol. I, April, 1889.	1	F. 60	Poor health.	Skin.	Skin of face, mucous membrane of lips, hard palate, and floor of mouth.	A vesicle on conj. bulbi of left eye.		Scarring of skin of cheeks. Left Eye. Lower cul-de-sac obliterated, upper one much shrunken; lids thick, brawny, adherent to globe, some cilia turned inwards, cornea dry, nearly opaque. Vision nearly lost; sense of light retained. Right Eye. Cornea clear, vision improving, some shrinking of lower cul-de-sac, lids thickened and congested.



