

Keratoconjunctivitis Sicca**Sjögren's Syndrome****DEFINITION**

1. The surface of the eye, particularly the cornea, is normally lubricated by a thin tear film produced by the lacrimal gland. When there is instability or reduction of the tear film, **dry eye** is the general term applied.
2. **Keratoconjunctivitis sicca** is a particular form of dry eyes which may occur alone or as part of Sjögren's syndrome.

CLINICAL MANIFESTATIONS

3. The most common symptoms are irritation, a foreign body sensation, presence of a stringy mucous discharge and transient blurring of vision. Complaints also include itching, photophobia and a tired feeling of the eyelids. Where there is associated severe keratitis, individuals complain of severe pain on blinking.
4. The symptoms of dry eyes are exacerbated by exposure to conditions associated with increased tear evaporation, such as air conditioning and central heating, wind, or prolonged reading, when the blink reflex is reduced. Closing the eyes may improve the symptoms.
5. The symptoms of dry eyes may be mimicked by chronic blepharoconjunctivitis due to staphylococci, rosacea or allergic conjunctivitis. These diagnoses must be excluded before a diagnosis of dry eyes is made.

AETIOLOGY

6. There are two main groups of conditions causing dry eyes.

- 6.1. **Atrophy and fibrosis**

- 6.2. **Miscellaneous causes**

Atrophy and fibrosis

7. Atrophy and fibrosis of lacrimal tissue occurs as the result of an autoimmune destructive infiltration by monoclonal cells, which causes a deficiency of tears.
8. There are two forms of dry eyes which fall into this category, **keratoconjunctivitis sicca** and **Sjögren's syndrome**.
9. **Keratoconjunctivitis sicca** is the form in which the lacrimal glands only are involved.

10. **Sjögren's syndrome** is a chronic disease which was originally described as the triad of dry eyes, dry mouth and rheumatoid arthritis and characterised by inflammation and destruction of exocrine glands.
11. The principal manifestations are due to involvement of the lacrimal and salivary glands, to give keratoconjunctivitis sicca and xerostomia respectively. Other mucous membranes may be involved.
12. The condition is now classified as either primary or secondary. When the features of keratoconjunctivitis sicca and xerostomia occur in isolation, it is referred to as **primary** Sjögren's syndrome. When these features are associated with other disease such as rheumatoid arthritis, systemic lupus erythematosus, systemic sclerosis, polymyositis, psoriatic arthritis, Hashimoto's thyroiditis and primary biliary cirrhosis, it is called **secondary** Sjögren's syndrome.
13. Autoimmune diseases are those in which the body reacts against its own constituents. However, the precise mechanism of the disease process is not known. Factors which have been postulated as triggers for the autoimmune process include genetic, hormonal and viral. A role for viruses has been extensively investigated but has not been confirmed.
14. A hypergammaglobulinaemia and the presence of antinuclear antibody are common findings, as is the presence of HLA-DR3 in primary Sjögren's syndrome.

Miscellaneous causes

15. **Destruction of lacrimal tissue:** tumours, sarcoidosis, chronic inflammation, trauma to the lacrimal glands.
16. **Meibomian gland dysfunction** which destabilises the tear film.
17. **Absence of the lacrimal gland:** congenital or after surgical removal.
18. **Blockage of the excretory ducts** of the lacrimal gland due to severe conjunctival scarring (as a result of trauma or infection) or trauma to the lacrimal ducts.
19. **Neurological lesions** such as the Riley-Day syndrome.
20. **Altered corneal surface:** irregularities of the corneal surface due to past corneal disease.
21. **Eyelid paralysis:** failure of blinking and closure results in poor tear film distribution and tear evaporation respectively.
22. **Degenerative changes** in the structures that produce and maintain the tear film. These changes are the commonest cause of dry eyes in the elderly.
23. **Environmental:** hot or dry environments alone may result in dry eyes. Removal from or reversal of the adverse environmental conditions will, in the absence of other factors, result in the re-establishment of a normal tear film, with the relief of symptoms.

CONCLUSION

24. **Keratoconjunctivitis sicca** and Sjögren's syndrome are autoimmune conditions resulting in dry eyes. Several factors have been suggested as being involved in the aetiology.
25. There is a miscellaneous group of causes for dry eyes. The aetiology of these types is that of the underlying cause.

REFERENCES

Kanski Jack J. Clinical Ophthalmology. 3rd Ed. 1994. Oxford. Butterworth Heinemann. Disorders of the Conjunctiva. p4.92-4.96.

Miller Stephen J H. Parson's Diseases of the Eye. 18th Ed. 1990. Edinburgh. Churchill Livingstone. Disorders of the Lacrimal Apparatus. p32.372-32.373.

Venables P J W. Sjögren's Syndrome. In: (Eds) Weatherall D J, Ledingham J G G and Warrell D A. Oxford Textbook of Medicine. Oxford. Oxford Medical Publications. 3rd Ed. 1996. p3036.

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