MANAGEMENT OF KERATOCONUS

An M.S. Essay.

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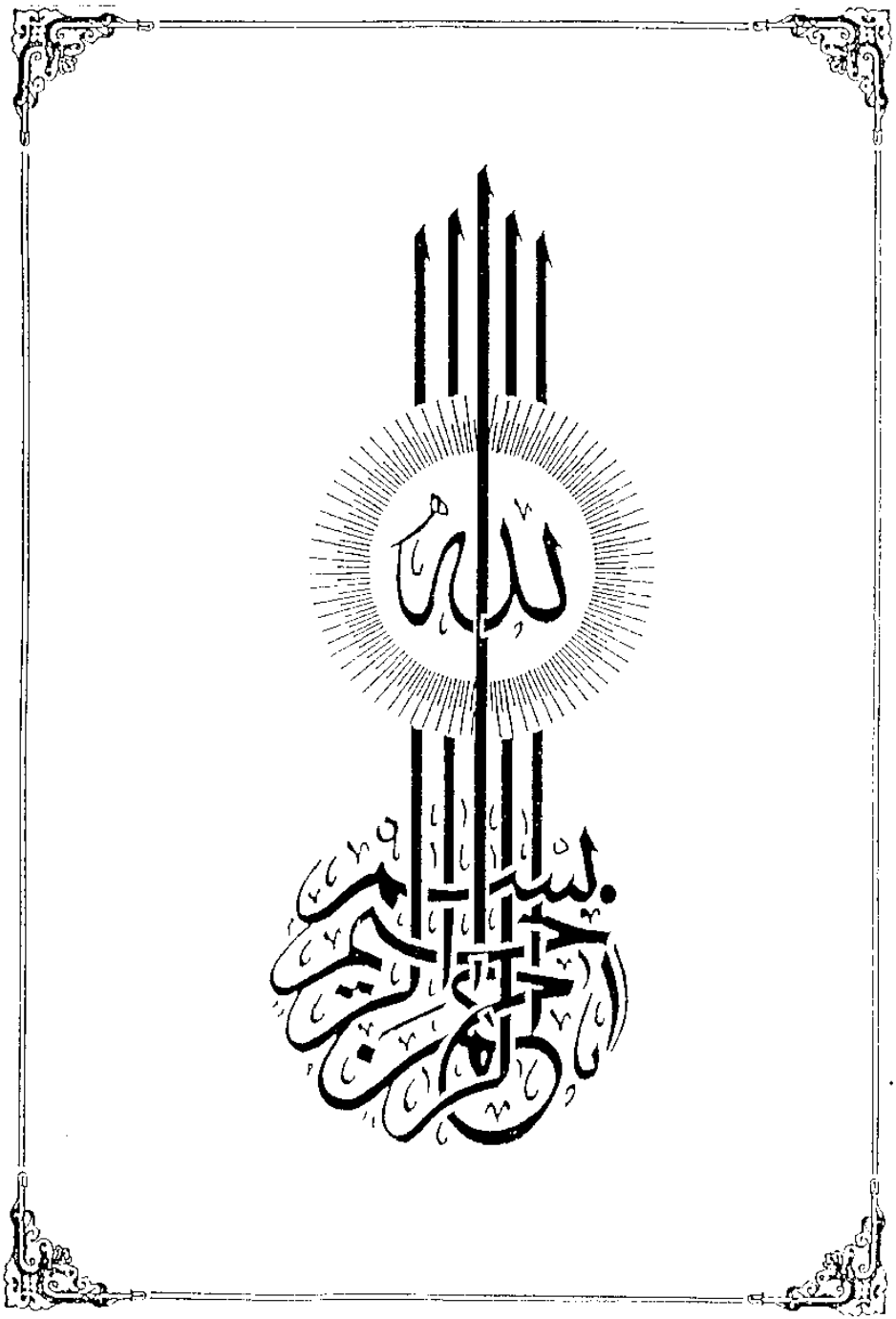

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AETIOLOGY

Keratoconus is a clinical term used to describe a condition in which the cornea assumes a conical shape because of thinning and protrusion.

It has been known for over 200 years as quoted in Duke-Elder and Leigh's system of ophthalmology (1965). Mauchart (1748) and Taylor (1766) were the first to refer to keratoconus.

Keratoconus is found in different races in different parts of the world. A thinning and anterior protrusion of the cornea causes a visual defect ranging in severity from mild dysfunction to a severe threat to vision.

It commonly appears in early adulthood and is arrested over the years.

Aetiology

The exact aetiology of keratoconus is not known. Many factors are to be blamed for the condition.

Some authors believe that it is developmental in origin. Duke-Elder (1965) stated that: Salzman (1907) believed that keratoconus is a primary anomaly of growth, like the stretching of the posterior pole associated with myopia. Greene (1945) attributed the condition to delayed separation of the lens from the cornea while Politzer (1952) believed that keratoconus occurs due to lack of development of corneal mesenchyme (Duke Elder,1965). Keratoconus was observed in association with other congenital anomalies;

aniridia,blue sclera, Marfan's syndrome,ectopia lentis,Ehler-Danlos and congenital catarct. Further association of the nature are mongolism, mental defects and pigmentary retinopathy (Duke-Elder,1965). Yet, the progressive nature of the disease limits the possibility of keratoconus being congenital (Appelbaum,1976). Hereditary and familial background were observed .Von Ammon (1830) was the first to find occasional keratoconus cases in the same family ,while dominant inheritance was first described by Vacher (1909) and Coppey (1909) (Cited by Duke-Elder and Leigh,1965).

The aetiological role of heredity has been verified in a number of investigations, including twin studies (Franceschetti et al.,1958;Hammerstein,1972). Waardenburg et al.,(1961) reported on families in which the disease was assumed to be recessively inherited due to consanguinity of the parents. Various cases of keratoconus suggestive of autosomal dominant inheritance have been reported (Falls and Allen,1969). Regarding keratoconus as an autosomally dominant disease Hammerstein (1971;1974) admits that the possibility of X-chromosomal inheritance still remains, while Hullerman and Wilson (1977) considered the disease multifactorial,but could not exclude a sporadic occurrence of both recessive and autosomal cases.

Disturbance in nutrition is considered by some authors an important factor in the development of keratoconus. As the conrnea is avascular,it would naturally suffer greatly

from lowered general nutrition. Jackson (1917) obserbved that keratoconus follows anaemia,measles,scarlet fever, typhoid fever and other acute infections,from this observation Appelbaum (1938) concluded that the condition must arise from the inability of the cornea to withstand the normal intra-ocular pressure at a time when toxins of an infected disease have impaired its nutrition and the power to resist pressure (Appelbaum,1936).

Ellis (1924) observed that women who had keratoconus were chronic sufferers from menstrual disturbances, such as dysmenorrhoea and menorrhgia. (Appelbaum,1936) made use of this observation and expressed his beleif that menstrual disorders are at least a significant factor in the causation of keratoconus. This is supported by the fact that the incidence of keratoconus is a little higher in females, usually about puberty.

Allergic symptoms related to keratoconus have been noted on several occasion in the literature. Gonzales (1920) reported several cases in which keratoconus followed spring catarrh. He expressed the belief that the circumcorneal vegetations occuring in spring catarrh exert a deleterious influence on the nutrition of the cornea. So as to diminish its resistance and cause protrusion. (Appelbaum,1936).

Ridley (1957,1967) stated that an undue proportion of patients with keratoconus are allergic subjects. Hay fever, asthma and atopic dermatitis,the proportion being roughly three times that of the normal population for atopy as a whole but ten times the average for atopic dermatitis in

particular. Less than one third of all patients with keratoconus suffer from some form of eczema. A selection of sufferers from keratoconus and random controls were assessed in order to substantiate claims that there existed a significant evidence of patients with both keratoconus and a raised serum level of immunoglobulin E. The results appeared to confirm a high incidence of raised total serum IgE levels in patients with keratoconus and also indicated that the additional measurement of serum specific IgE was more sensitive than total IgE. This observation therefore offers the possibility of an immunological marker for keratoconus (Kemp and Lewis,1982). Keratoconus patients have a tendency to rub their eyes and this rubbing may be a factor in the development of the disease. Eye rubbing may be brought on by itching from allergy (Dohlman,1964).

Ridley (1967) noticed this phenomenon of eye rubbing in keratoconus. He analysed some 600 cases and found that excessive eye rubbing is present in 70% of the cases and he found out that in some cases this rubbing had been present before keratoconus being ever suspected.

Trauma as a factor that may play a role in the development of keratoconus,is not a recent belief. Grisp (1919) described a case in which the condition resulted from trauma. Appelbaum (1936) mentioned that MacKenzie and Nottingham had entertained the belief that trauma is an important aetiologic factor. He added that indirect trauma,such as the tension exerted on the bulbus oculi by

the extraocular muscles,and eye strain,with its accompanying severe and long continued spasm of accomodation,may be a factor in the development of protrusion in a weakened ,undeveloped or poorly nourished cornea. Recently Hartstein (1968) published a report on four wearing corneal contact lenses,who developed keratoconus that was not present prior to the use of contact lenses.He aroused the question;can contact lenses exert pressure or interfer with corneal metabolism and may actually induce keratoconus ?. Hartstein and Becker (1970) continued the research and they included the factor of ocular rigidity and concluded that at least one type of keratoconus may be related to the long term wearing of corneal contact lenses on eyes that have unusually low ocular rigidity. Other believe that it is just a coincidence and keratoconus was going to develop regardless the use of contact lenses (Bier and Lowther,1977)

Patients who develop this syndrome (corneal warping and contact lens induced keratoconus) have usually adapted well to contact lenses. After three or more years corneal curvature increase,corneal astigmatism increases,mires observed during keratometeric readings become distorted,and spectacle-corrected visual acuity shows a significant decrease. However,this type of contact-lens-associated keratoconus rarely progress to the point at which keratoplasty is required (Nauheim and Perry,1985).

So there is some controversy whether rigid contact lenses cause keratoconus. Most authorities do not support this hypothesis because if it is true the incidence should

be much higher than it is in general population. The starting age of a contact lens patient and the age of a keratoconus patient are similar. Also,in the early phases of keratoconus the person may be merely myopic,and so it is quite natural to assume that one cause the other. There are however some who believe that a rigid lens may accelerate the development of early forms of keratoconus. The diverse association of keratoconus suggest that it is the end result of different pathological processes. Therefore,it may be regarded as a heterogenous group of conditions with identical clinical signs but with varied pathogenesis (Karseras and Ruben,1976).

HISTOPATHOLOGY

Histopathology

Hefny and Abdalla in 1970 by the light microscope examination,showed that the striking common characteristic histological findings were:

1- Fragmentation of the basement membrane of the corneal epithelium,fibrillation of Bowman's membrane and the anterior stroma in early stages.

2- Marked thinning and bulging of the central cornea.

3- Wavy appearance of Bowman's membrane with multiple narrow gaps,filled either by newly formed connective tissue,that contains local accumulation of keratoblasts or by epithelium.

4- Folds and buckles in Descemet's membrane and its overlying deep stroma. Sometimes there are ruptures in the memebrane usually in the region of greatest ectasia near the centre of the cone.

Rupture in the Descemet's membrane leading to acute ectasia or hydrops are late changes in advanced keratoconus.

In 1963, Teng,by the electron microscopic study confirmed the histopathological finding that the primary lesion is in the basal epithelial cells. All the other destructive changes can be explained by the release of proteolytic enzymes from the dead and disintegrated cells. The characteristic picture of keratoconus then,is the death of the basal epithelial cells are found mostly in the central area and this change together with the picture of

fragmentation of basement membrane and fibrillation of Bowman's membrane are the key diagnostic point in the morphology.(Fig.1,2,3,4)