CORNEAL STROMA

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PREVALENCE OF KERATOCONUS

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Purpose: To determine the prevalence of keratoconus (KC) in a "normal" population and to provide statistical reference values for corneal topography numerical descriptors.

Methods: All 670 subjects, age 18 to 22 y., from a single class were examined during the military duty incorporation process in the French Armed Forces. Objective and manifest refraction were determined with an autorefractometer and corneal topography was assessed using a TMS-1 system. Data were analyzed using 13 descriptors of corneal powers distribution from the new TMS-1 statistical software and an artificial intelligence based keratoconus prediction index (KC%, version 1.5).

Results: 22 eyes (from 21 subjects) in 1340 had power distribution and KC% index compatible with the diagnosis of keratoconus. Qualitative analysis of color coded maps authentified "true" KC in 9 eyes (including a bilateral case), all with KC% greater than 30%. However further topographical evaluation of these cases revealed 3 pseudokeratoconus, associated to mire reflection defects, leaving 5 subjects affected with authentic KC.

Mean (± SD) values of main numerical descriptors for normal corneas)Gaussian distribution) were:

SRI=0.7±0.86	SAI=0.34±0.47	simK1=43.5±1.70	simK2=42.8±1.91
DSI=1.97±1.76	SDP=0.85±1.97	OSI=0.92±1.44	CSI=0.26±0.38
IAI=0.37±1.12	KPI=0.20±0.07	AA=70.7±10.6	CYL=0.70±0.90

Conclusion: The prevalence of true keratoconus in this "normal" population was 0.75%, while 1.94% of subjects were identified as having a "keraconus-suspect" condition.

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INTERLEUKIN-1 AND INTERLEUKIN-6 LEVELS IN CORNEAS OF KERATOCONUS PATIENTS

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Purpose of the study was to compare clinical and histological findings and Interleukin levels in corneas of keratoconus patients. Keratoconus is characterized by a cone-shaped protrusion of the stroma caused by thinning of the stromal layers. The weakening of the corneal stroma is suggested to be a result of abnormalities in the synthesis, turnover or composition of the stromal matrix. In cultures of keratoconi more collagenolytic activities were found than in cultures of normal corneas. In *in vitro* studies corneal fibroblasts from keratoconus patients contained four times more interleukin-1 binding sites than fibroblasts of normal corneas. Normal corneal fibroblasts produced more collagenase if Interleukin-1 was added to the culture medium.

Methods: We investigated 18 corneas of keratoconus patients. The corneal buttons were snap frozen immediately after explantation. Cryosections were prepared for histological examination. The frozen specimens were processed under liquid nitrogen and the water-soluble protein fraction was measured for protein content, interleukin-1 and -6 concentrations. The interleukin levels were calculated from the protein content.

Results: A wide scatter of interleukin-1 (1,2-170 pg/mg protein) and Interleukin-6 (0,06-20,4 pg/mg protein) values were found. Highest levels were found in the same corneas for interleukin-1 and interleukin-6 but no correlation could be seen in low interleukin levels. Histological observation showed no correlation of inflammatory cellular reactions and increase of interleukin levels.

Conclusions: Corneas from keratoconus patients seems to be a more heterogeneous group regarding interleukin values than it appears from the histological and clinical point of view. Interleukins normally involved in inflammatory and/or immune reactions also may be involved as mediators in "non inflammatory" processes like keratoconus.

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LYSOSOMAL ENZYME ACTIVITIES IN CONJUNCTIVAE AND SKIN BIOPSY SPECIMENS OF PATIENTS WITH KERATOCONUS

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Purpose Keratoconus is a corneal disease characterized by thinning and scarring of the central portion of the cornea. We have shown previously in corneas obtained from patients with keratoconus that lysosomal enzyme levels are elevated, especially in the epithelial layer. In this study, we examined the lysosomal enzyme activities in the conjunctival tissues and skin biopsy specimens of keratoconus patients.

Methods Conjunctival tissues and skin biopsies collected from patients with keratoconus and normal subjects were fixed and processed for frozen sections. Histochemical staining for two lysosomal hydrolases - acid esterase and acid phosphatase -- were performed.

Results The epithelium of conjunctival as well as the epidermal and dermal layers of skin specimens all stained positively for acid esterase and acid phosphatase. The staining intensity in the conjunctival tissues of keratoconus patients was much more prominent than that seen in specimens from normal controls. The staining in the skin dermal and dermal cells of keratoconus patients was within the normal range.

Conclusions It appears that the conjunctival epithelium may also be involved in keratoconus. The biochemical abnormality is however not manifested in the skin.

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AQUIRED KERATOGLOBUS AND EHLERS-DANHLOS SYNDROME : A CLINICAL AND HISTOPATHOLOGICAL STUDY

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Purpose
To investigate the fine structure of a unique corneal sample with initial clinical features suggestive of posterior polymorphous dystrophy (PPD), then characterized as a keratoconus (KC), and finally as a true keratoglobus (KG), treated successively by epikeratophakia, then by penetrating keratoplasty (PKP).

Methods
A patient with Ehlers Danhlos Syndrome (Marfanoid phenotype with diffuse arterial dysplasia, diaphragmatic hernia, megadolichocolon and phymosis), born in 1973 was followed up every year from 1979 to 1995. At age 6. (1979), visual acuity was - 3(-1.5x10) 20/200 OD and - 1.5(- 1.25x80) 20/50 OS, and cornea presented endothelial abnormalities most suggestive of PPD. A age 10. (1983), the refraction was - 7.5(+5x130) OD and - 4.5(+2.5x120) OS, and a keratoconus was diagnosed, in association with an increased corneal diameter (13.5 mm OD and OS). At age 15. (1988), the diagnosis of keratoglobus was patent and a cornea-scleral epikeratoplasty was performed in OD (Dr. KH). Surgical removal of an epithelial invasion was required in 1989, limiting the visual acuity to - 4(-4x45) 20/200 OD. A 7 mm diameter PKP was performed in 1993 in OD and the corneal button was processed for transmission electron microscopy (TEM).

Results
Optical and TEM analysis revealed typical lesions of KC in Bowman's layer and stroma, while lesions more suggestive of KC were observed in the posterior stroma and Descemet's membrane.

Conclusion
This unique clinico-pathological observation may help to clarify the relationship between KC and aquired KG as opposed to congenital KG.