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ORIGINAL ARTICLE

Keratoconic Cone using its Keratometry, Decentration, and Thickness as Staging Parameters

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ABSTRACT

Aim: To propose a new system of keratoconus staging using a set of parameters describing the keratoconic cone.

Materials and methods: Retrospective case series study of 101 keratoconic eyes of 58 patients was undertaken. They all had complete eye examination including corneal topography (Oculus Pentacam). K_{mean}, K_{max}, higher order aberrations (HOAs) root mean square (HOARMS) value, pachymetry at thinnest point and steepest corneal meridian were obtained from Pentacam. Apex to thinnest pachymetry distance (D) was calculated using trigonometry. Pearson correlation coefficients between K_{max} and HOARMS, between D on the one hand and the adjusted angle of steepest meridian, K_{mean} and K_{max} respectively on the other, were calculated.

Results: There is a statistically significant positive correlation between K_{max} and HOARMS ($p < 0.00001$). There is a negative correlation, a "horizontalization," of the steep meridian with D increase, although it fell short of statistical significance ($p = 0.07$). D and K_{mean} ($p = 0.003$), and D and K_{max} ($p = 0.005$) are significantly negatively correlated.

Conclusion: K_{max} correlates with significant changes in HOAs. D correlates with corneal astigmatic meridian change and has a divergent path to K_{mean} and to K_{max}. We propose a new keratometry, decentration, and thinnest pachymetry staging using the parameters K_{max} (K), distance from the corneal apex to the thinnest pachymetry point (D), and corneal thickness at its thinnest point (T) to give a better, detailed description of a keratoconic cornea which could lead to improvements in assessment of its severity and treatment outcomes.

Keywords: Decentration, Keratometry, Thickness.

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INTRODUCTION

Keratoconus is a noninflammatory, ectatic corneal disorder, characterized by corneal protrusion and thinning, leading to corneal irregularity and decreased visual acuity.[1] Estimated prevalence is between 50 and 230 cases per 100,000,[2] and although unilateral cases have been described, it is most commonly a bilateral, usually asymmetric disease.[3,4]

Diagnosis of keratoconus is based on clinical findings (corneal protrusion, corneal thinning, Vogt's striae, Fleischer ring, anterior stromal scars, or corneal hydrops) along with keratometry readings, keratoscopy, and corneal topography. Videokeratographic indices suggested by Rabinowitz for keratoconus diagnosis are central corneal power greater than 47.2D, inferior–superior (I–S) asymmetry over 1.2D, Sim-K astigmatism greater than 1.5D, and skewed radial axes (SRAX) greater than 21°.[5,6]

The global consensus on keratoconus and ectatic diseases concluded in 2015 that there was no clinically adequate classification system for keratoconus.[7]

Amsler–Krumeich (A-K) classification remains the most well-known keratoconus staging system.[8] More recently, Alió and Shabayek[9] investigated corneal higher order aberrations (HOAs) in keratoconic patients, proposing a new system to quantify and grade keratoconus. Both classifications take into account K_{mean}, presence or absence of scarring, and corneal thickness at the thinnest point, along with refractive error in A-K classification, whereas corneal HOAs are incorporated in Alió–Shabayek (A-S) classification. In both schemes, stage is determined by the most advanced parameter. It means that for a given keratoconic cornea classified as a particular stage, it is not possible to know if this stage is due to the K_{mean}, thinnest pachymetry, scarring or the HOA (in A-S classification), or the refraction (in A-K classification).

A new multiparameter staging scheme incorporating posterior curvature, thinnest pachymetry, and distance visual acuity in addition to the standard anterior curvature (both anterior and posterior curvatures were taken using 3.0 mm zone centered on the thinnest area) had been devised recently. Curvature and thickness measurements are based on the thinnest point better reflecting the anatomical changes in keratoconus.[10]

We investigated the feasibility and advantages of using three topographic parameters describing the main features of the sine qua non of keratoconus – the

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keratoconic cone, its steepening, decentration, and thinning – and propose a new system of keratoconic staging using them.

MATERIALS AND METHODS

The retrospective case note review study was undertaken at the Stanley Eye Unit, Abergele Hospital, Wales, United Kingdom. A total of 101 keratoconic eyes of 58 patients were included. Bilateral data from 43 patients were recorded. Unilateral data from another 15 patients were recorded; 2 cases had unilateral corneal graft, 4 cases had unilateral keratoconus, and 9 patients were bilateral keratoconic patients who did not wish to remove the contact lens 2 weeks previous to corneal topography from both eyes, but just from one eye, as they could not manage with spectacle correction. A total of 42 patients were male, whereas 16 patients were female. Ages ranged from 15 to 73 years (mean of 38.6 years). A complete, thorough eye examination for all the patients was performed, including best corrected visual acuity, refraction, slit-lamp biomicroscopy, applanation tonometry, fundus examination, and corneal topography (Pentacam HR, Oculus GmbH, Wetzlar, Germany). For our study, corneal topography findings were the main interest. K_{mean}, K_{max}, and steepest corneal meridian were obtained from corneal topography. The steepest corneal meridian was then adjusted where necessary to obtain the degree of deviation from the horizontal meridian. For example, a steep meridian at 135° is adjusted as 45° (180 – 135 = 45) from the horizontal meridian. Distance from apex to the thinnest point in millimeters (decentration – D) was calculated using trigonometry. For a right triangle, the hypotenuse can be obtained as the square root of the sum of the square value of each leg. D is calculated as the square root of the sum of X coordinate squared plus Y coordinate squared:

$$D = \sqrt{(X^2 + Y^2)}$$

Corneal higher order aberrations (HOAs) root mean square (HOARMS) values were obtained from Zernicke decomposition up to the 8th order in the central 6 mm cornea by Pentacam software. We then analyzed the relationships between K_{max} value (diopters) with HOARMS (µm), and between D and the steep meridian. Also correlations between D on the one hand and K_{mean} and K_{max} on the other were calculated.

Statistical Package for the Social Sciences (SPSS) version 17.0 Chicago, Inc was used for this purpose.

Using the three parameters relating to the keratoconic cone – K_{max} value in diopters (K), decentration in mm of the thinnest pachymetry point from the apex (D) and the thinnest pachymetry reading (T) – a multiparameter KDT staging system is proposed (Table 1).

Table 1: KDT staging for keratoconus

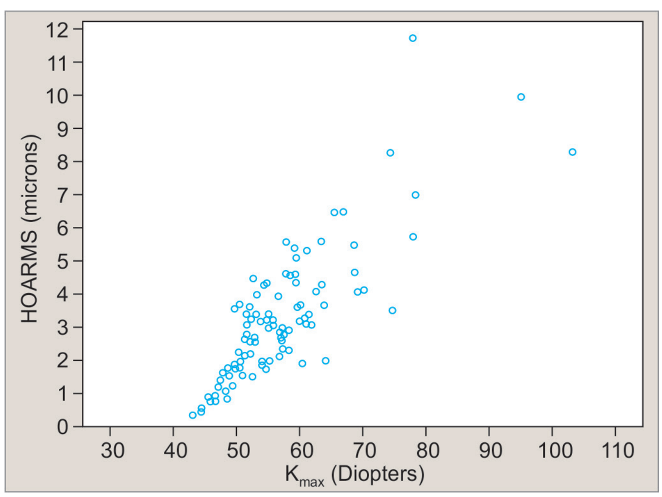
Staging parameter		*Characteristics*
K_{max} (Diopters)	K1	< 50
	K2	from 50.1 to 60
	K3	from 60.1 to 65
	K4	> 65
Decentration of thinnest point (mm)	D1	< 0.5
	D2	> 0.5 to 1.0
	D3	> 1 to 1.5
	D4	> 1.5
Thinnest pachymetry (µ)	T1	> 450
	T2	From 400 to 450
	T3	From 200 to 399
	T4	< 200

RESULTS

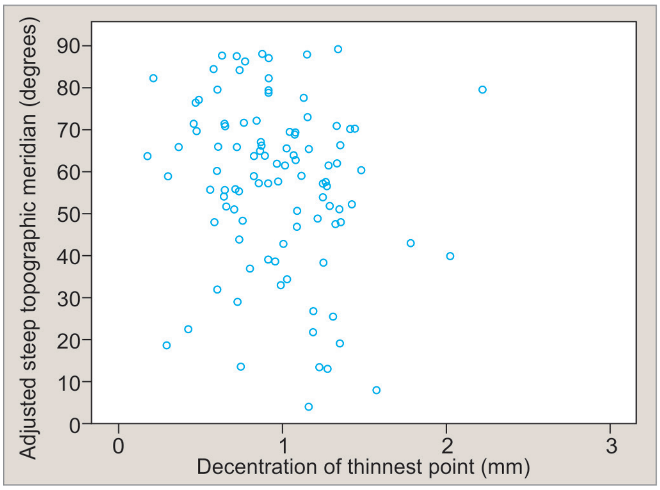
The descriptive data for the eyes/corneas examined were as follows: K_{max} ranged from 43 to 103 diopters (mean 57.04D, standard deviation (SD) 9.64). Decentration ranged from 0.18 to 2.22 mm (0.96 mm, SD 0.36) and thinnest pachymetry ranged from 193 to 557 µm (mean 440.22 µm, SD 64.83). Mean sphere in our sample was –1.79D (SD 4.97) with a range from –20.78 to +5.75 and cylinder ranged from 0 to 13.00D (mean 3.29D, SD 2.83).

Pearson correlation coefficient was calculated for the following parameter pairs. The relationship between K_{max} value and HOARMS was statistically significant ($r = 0.751$; $p < 0.00001$) (Graph 1), showing an increase in corneal HOA with the increase of K_{max} dioptric power.

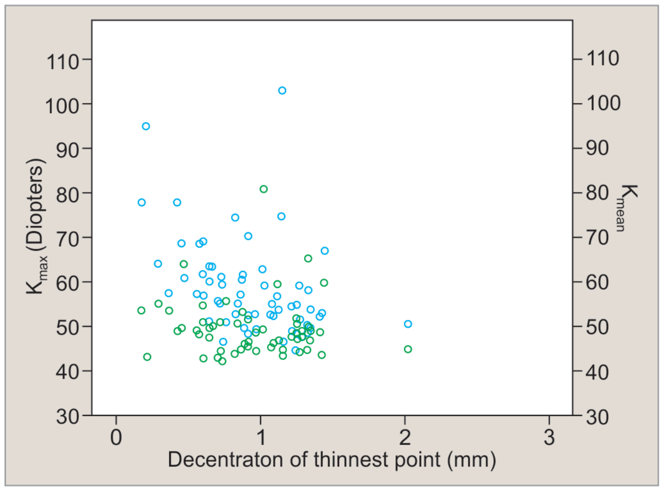
There was a negative correlation between D and the steep meridian, suggesting that the more decentred the thinnest point becomes, the smaller the angle between the steep meridian and the horizontal meridian, i.e., the corneal astigmatism becomes more against the rule. In other words, changes in thinnest point location (cone decentration) influence corneal shape change. However,



Graph 1: Scattergram showing relationship between K_{max} dioptric power and corneal HOARMS



Graph 2: Scattergram showing relationship between distance from apex to thinnest point – decentration (D) – and the steep topographic meridian. Note that for the eyes having the steepest axis between 90° and 180°, an adjustment was made. As e.g., a cornea having a steep meridian at 135° was adjusted to have the steep meridian at 45° from the horizontal meridian



Graph 3: Double Y-axis scattergram plotting decentration (D) in mm against K_{mean} (Green) and K_{max} (Blue)

Table 2: Comparison of staging by Krumeich and KDT parameters

Stage	*Krumeich*	*K*	*D*	*T*
1	44	21	10	51
2	25	54	55	31
3	16	13	31	18
4	16	13	5	1
Total corneas	101	101	101	101

results fell short of statistical significance ($r = -0.179$; $p = 0.07$) (Graph 2).

There are statistically significant negative correlations between D and K_{mean} ($r = -0.292$; $p = 0.003$) and D and K_{max} ($r = -0.279$; $p = 0.005$).

Graph 3 is a double Y-axis scattergram plotting D in mm against K_{mean} and K_{max} in diopters. It shows a negative correlation for D *vs* the other two parameters. The distribution of the 101 corneas by Krumeich classification and by each of the KDT parameters is shown in Table 2.

DISCUSSION

Keratoconus causes thinning and conical protrusion of the cornea and is progressive to varying degrees in different sufferers of the condition. The keratoconic cone has traditionally been divided into three morphologic types – nipple, oval, and global, mainly based on slit-lamp appearances. The advent and sensitivity of computerized videokeratography, and later tomography and wavefront aberrometry, allows more detailed analysis of corneal shape in these eyes and one of their main uses has been detecting or screening for the presence of keratoconus.[11-13]

It also became possible to accurately track the shape change over time of the cornea. Longitudinal analyses of corneal topography in suspected keratoconus[14] or in established keratoconus[15,16] using a variety of parameters (keratometric, refractive, aberrometric) have been undertaken.

K_{max} (keratometry at the steepest point over the entire scanned anterior corneal surface on Pentacam HR topographer) represents cone steepening, not central corneal steepening, which K_{mean} represents. We therefore, suggest it is a better marker for keratoconic progression. As a parameter derived from axial curvature topography map, it is less sensitive to localized curvature changes[17] but allows the broader keratoconic cone to be taken into account, not just its pinnacle. It can be obtained easily from Pentacam topographer. In this study, we analyzed the relationship of K_{max} and corneal HOA and found it to be very highly significant, a finding previous studies noted as well.[14] As such, we think that K_{max} is a very good proxy for HOA as well as being a good marker for keratoconic cone steepening. K_{max} has been recommended as an indicator for keratoconic progression and a 1.5D increase within a year's interval is suggested as a trigger for corneal collagen cross-linking.[18] However, it has not been suggested previously as a parameter to grade or classify keratoconus.

A cohort study of progressive keratoconic patients found that topographic cone location as defined by the K_{max} (steepest corneal location on sagittal curvature map) – apex distance increases with increasing age.[19] We decided to use D (distance from apex to the thinnest point) instead of the steepest corneal location, as the former is an anatomical landmark. D is negatively correlated with steep topographic astigmatic meridian, i.e., the (adjusted) steep meridian "horizontalizes" as D increases. This relationship between D and corneal astigmatic meridian change may be what underlies the SRAX parameter used in detecting keratoconus as described by Rabinowitz.[5] We think that our use of average steep topographic meridian, as provided by Pentacam, instead of the superior hemi-meridian, which is usually closer to the horizontal meridian, probably underplayed the horizontalizing effect of decentration and resulted in the negative correlation between the two parameters falling short of significance. It is an interesting finding nonetheless and points to the need for further, more detailed studies.

There are also statistically significant negative correlations between D and K_{mean} ($r = -0.292$; $p = 0.003$) and D and K_{max} ($r = -0.279$; $p = 0.005$), and Graph 3 shows the relationship between those three parameters graphically. In Krumeich classification, central (average) keratometry is used as a staging parameter, i.e., the higher its value the more advanced the keratoconic stage is. The negative correlations of the thinnest point decentration (which we use as the marker for keratoconic cone decentration) to keratometry (central K_{mean} as well as K_{max}) in our study indicate that decentration and keratometry follow divergent paths, i.e., they do not increase in tandem. It follows that using keratometry only (whether it be K_{mean} or K_{max}) without using a decentration parameter as well would result in an incomplete picture and therefore, a poorer staging scheme.

The explanation for our finding that cone decentration leads to reduced central corneal steepening may lie in Smolek and Klyce's[20] idea that any stretching that occurs in keratoconus could be described as a form of warpage, rather than a true stretching process. Steepening in one place (the cone) may be at the expense of flattening at another.

The relevance of decentration can be further discerned by the finding that pericentric cones and displaced cones probably respond differently to corneal collagen cross-linking.[21]

For all these reasons, we think that D should be a separate parameter in staging keratoconus.

Corneal thickness spatial profile and percentage increase in thickness have been found to differ between keratoconic corneas and normal corneas,[22] indicating that corneal thinning is related to keratoconus development. Since when corneal tomography allowed corneal pachymetry measurements, keratoconus staging systems have included thinnest pachymetry as an important parameter.[8,9] The fact that more recent treatment modalities use it as a demarcating criterion for stratifying management choices has added weight to its importance.[23]

Both A-K and the A-S classifications treat the parameters as distinct and disparate. The bald statement of the stage a cornea is in, in these schemes, gives no clues as to which parameter/characteristic actually determined the stage for that particular cornea.

We think that tracking the primary characteristic of keratoconus, the cone, the sine qua non of keratoconus, is the way forward in staging it. An index (Cone Location and Magnitude Index) based on the keratoconic cone had been considered previously as a diagnostic tool with high specificity and good sensitivity that is easy to compare and understand and allows clinical interpretation.[24] The investigators of this approach have considered expanding the index to include corneal thickness and posterior surface information, primarily to improve keratoconic detection. They also suggested that the expanded index could have the potential to track progression of the disease.[25] Our approach to staging the cone described below is intuitive and simple with easy applicability and comparability.

The ABCD grading scheme proposed by Belin et al shares our emphasis on the keratoconic cone.[10] However, it includes a visual acuity parameter; we feel it confuses the issue. Visual acuity assessment in keratoconic patients is fraught with difficulty and unpredictability.[26] Moreover, it does not include the decentration parameter, which for the reasons we have discussed should be considered an integral part of keratoconic cone description. Posterior curvature of the keratoconic cone should be more properly considered as the (initial) diagnostic criteria or as one of the parameters for progression.[27-29] The ABCD scheme could be considered as a multiparameter system, but the authors still apparently harbor ambition to have an overarching stage, 0 to 4.[8] We are of the opinion that an unabashedly multiparameter staging scheme is the most intuitive and transparent option.

We propose a new multiparameter keratoconus staging scheme based on the above topography-derived physical characteristics – $K_{(max)}$, D and T – of the keratoconic cone.

Our suggestions for the stage-defining limits for each of our parameters are set in Table 1.

For K_{max} (K) the upper limits for stages 2, 3, and 4, we propose, should be set respectively, at 50D, 60D and 65D. For decentration (D) from the apex to the thinnest point, we propose the corresponding upper limits at 0.5, 1, and 1.5 mm. We think the upper pachymetry limits (T) for stages 3 and 4 should remain the same as for A-K and A-S schemes, i.e., 400 and 200 μ respectively. We, however, propose thickness stage 2 for pachymetries between 400 and 450 μ and stage 1 for pachymetries over 450 μ, as this is the minimum thickness (including the epithelium) generally accepted to be required for standard corneal collagen cross-linking.

Comparison of Krumeich and KDT staging of the study corneas in Table 2 suggests that KDT parameters can better indicate and differentiate earlier changes in keratoconic cone protrusion and decentration taking place in these corneas. Another obvious advantage of the KDT staging is that one can, by looking at the T stage a cornea is at, immediately know if that cornea is within the criteria for collagen cross-linking treatment (T1), or if, in T2 stage, consideration can still be given for cross-linking, possibly using hypotonic riboflavin solution.

Staging keratoconus is important for the study of its natural history, the likelihood of success and advisability of any new and emerging treatment modalities, formulation of nomograms for those modalities, and outcome comparisons between them. Keratoconic cone features, i.e., steepening, decentration, and thinnest pachymetry, though undoubtedly interrelated, may need to be teased out individually, as far as is practicable, to focus the effect each has on the outcome of different treatment modules. We believe our staging system could facilitate such studies.

Temporal cones are generally considered much less common than inferior cones, but lack of definitive criteria for designating one has been a disadvantage, possibly affecting proper assessment of effective treatment options for them.[30] Attempts have been made to tailor treatment to the perceived type of cone.[31] They could be designated (adding a *t* to the decentration parameter) as those with thinnest point within 45° of the horizontal axis and with 0.5 mm or more of decentration. Superior cones are very rare and probably would need to be described specifically.[32]

Corneal central scarring is one of the parameters for Krumeich classification,[7] its presence denoting stage 4. In light of current treatment modalities where a superficial nonhydrops central corneal scar could still afford big bubble technique of deep anterior lamellar keratoplasty, keratoconic scars need to be assessed individually.

Further refinements are inevitable with advances in knowledge and technology, but the proposed staging system is a useful tool to studying keratoconus and its treatments. Additional characteristics/parameters of the keratoconic cornea, e.g., corneal biomechanical properties, may be incorporated (or even replace some of the proposed parameters) as we gain better understanding of their effect on keratoconic progression.

In conclusion, a new, more descriptive, and clinically useful multiparameter staging system for keratoconus progression that takes its natural history into account can be achieved by using the cone's keratometry, decentration, and thickness.

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