

A New Tomographic Method of Staging/Classifying Keratoconus: The ABCD Grading System

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ABSTRACT

Purpose: To incorporate advanced corneal imaging into a new keratoconus classification system that utilizes posterior curvature, thinnest pachymetry, and best-corrected distance vision (CDVA) in addition to standard anterior parameters.

Materials and methods: A total of 672 eyes of 336 normal patients were imaged with the Oculus Pentacam HR. Anterior and posterior radius of curvature measurements were taken using a 3.0 mm zone centered on the thinnest area and corneal thickness was measured at the thinnest point. Mean and standard deviations were recorded and anterior data were compared to the existing Amsler-Krumeich (AK) classification.

Results: A total of 672 eyes of 336 patients were analyzed. Anterior and posterior values were 7.65 ± 0.236 mm / 6.26 ± 0.214 mm respectively and thinnest pachymetry values were 534.2 ± 30.36 μ m. Comparing anterior curvature values to AK staging yielded 2.63, 5.47, 6.44 standard deviations for stages 1, 2, and 3 respectively. Posterior staging uses the same standard deviation gates. Comparative pachymetric values yielded 4.42, and 7.72 standard deviations for stages 2 and 3 respectively.

Conclusion: A new keratoconus staging system incorporates posterior curvature, thinnest pachymetric values, and distance visual acuity in addition to the standard anterior curvature and consists of stages 0 to 4 (5 stages), closely matches the existing AK classification stages 1 to 4 on anterior curvature. The new classification system by incorporating curvature and thickness measurements based on the thinnest point, as opposed to apical, better reflects the anatomic changes in keratoconus.

Keywords: Classification, Ectasia, Keratoconus, Radius of curvature.

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INTRODUCTION

Keratoconus was first described in detail in 1854 as a chronic, noninflammatory ectasia of the cornea that is characterized by corneal steepening, visual distortion, apical corneal thinning and central corneal scarring. The disease typically begins at puberty and shows progression until the third-fourth decade of life. The corneal thinning induces irregular astigmatism, myopia and corneal protrusion, leading to mild to marked impairment in the quality of vision, and often has a significant impact on patient's quality of life.¹⁻³ Keratoconus is a relatively uncommon condition with a reported annual incidence of 2 per 100,000 and prevalence of 54.5 per 100,000 though rates vary greatly in different geographic regions.⁴⁻⁷ Keratoconus typically affects both eyes, although only one eye may be affected initially. The disease may be highly asymmetric^{8,9} and symptoms are variable. Early in the disease, and in subclinical keratoconus, there may be minimal or no symptoms, whereas in advanced disease there is significant distortion of vision accompanied by profound visual loss.¹⁰

Several classification systems for keratoconus have been proposed in the literature.¹¹⁻¹⁹ The Amsler-Krumeich (AK) system (Table 1) is among the oldest and still the most widely used. In the AK system, the severity of keratoconus is graded from stage 1 to 4 using spectacle refraction, central keratometry, presence or absence of scarring and central corneal thickness.²⁰ Others have used this system with various modification and additions in an attempt to better diagnosis or characterize the severity of disease.^{21,22} Other studies, such as the collaborative longitudinal evaluation of keratoconus (CLEK) Study sought to describe the clinical course of keratoconus and to identify both risk and protective factors that influence its severity and progression.²³ The CLEK study used changes in vision, anterior corneal curvature based on keratometry, biomicroscopic signs, corneal scarring, and vision-specific quality of life, as measures to define stage and severity of disease in keratoconic patients. Topographic analysis was not used in either the AK or CLEK classifications.²³

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Table 1: Amsler-Krumeich classification

Stage I	Eccentric steepening Myopia/astigmatism < 5.00 D Mean K < 48.0 D
Stage II	Myopia/astigmatism > 5.00 D but < 8.00 D Mean K < 53.0 D Absence of scarring Minimal apical corneal thickness > 400 μ m
Stage III	Myopia/astigmatism > 8.00 D but < 10.00 D Mean K > 53.0 D Absence of scarring Minimal apical corneal thickness < 400 μ m but > 300 μ m
Stage IV	Refraction not possible Mean K > 55.0 D Central corneal scarring Minimal apical corneal thickness < 300 μ m

Other imaging techniques have been studied to try and establish a more comprehensive staging system for keratoconus. Alio et al proposed new classification using videokeratoscopy to measure anterior corneal surface higher order aberrations as a tool to detect and grade keratoconus.¹⁵ Numerous other approaches have been described.^{15,16,18,19,22-28} Each of these methods, however, has its limitations. It is the limitations of these staging systems that discourage their widespread acceptance and clinical utility. None of the commonly used systems incorporate posterior corneal data or analyze the full corneal thickness map.²⁹ According to the Global Consensus on Keratoconus and Ectatic Diseases (2015), due to the limitations of the various staging methods in use in clinical practice, there is currently no clinically adequate classification system for keratoconus.³⁰ The most widely used AK system fails to make use of current information and technological advances in corneal imaging. Specifically, the posterior corneal surface and full pachymetric data, which holds significant diagnostic value, is not utilized in the AK classification.³¹⁻³⁴

Older staging systems, based solely on the anterior corneal surface, appear inadequate as newer treatment modalities, such as cross-linking, may be utilized earlier in the disease process and at times prior to clinical changes on the anterior corneal surface.³⁵ This paper will propose a new method of describing or staging keratoconus which utilizes tomographic data and better reflects both the anatomical and functional changes in ectatic disease.

BACKGROUND

Prior to the emergence of refractive surgery there was little need to identify individuals with subclinical (early) disease as treatments (e.g. contacts, penetrating keratoplasty) were instituted when there was reduced best-corrected vision which closely followed changes on the anterior corneal surface.³⁵ In as such, the AK

classification, which is based on keratometry, central corneal thickness and the degree of myopic astigmatism tended to mimic the decrease in best corrected visual acuity and as such had clinical utility.^{2,3,12,21} In spite of significant advances in corneal imaging, the AK classification is still the most commonly used, albeit outdated, system. Refractive surgery put greater demands on identifying early or subclinical disease as the tissue removal associated with laser ablative surgery could cause a corneal biomechanical failure (i.e. postrefractive ectasia) in otherwise totally asymptomatic individuals.³⁶ Newer imaging technologies [e.g. Scheimpflug, optical coherence tomography (OCT)] are capable of measuring the posterior corneal surface in addition to the anterior cornea.²⁹⁻³⁴ With both anterior and posterior corneal surfaces identified, a full corneal thickness map could be generated.^{29,37} It is possible to have significant posterior ectatic changes in spite of a normal anterior surface. This is called subclinical disease since visual acuity is typically normal and patients often unaware of their disease (Fig. 1).^{10,38-40}

The AK classification fails to recognize any changes other than on the anterior corneal surface. Full corneal thickness maps have also shown the limitations of relying on a single apical measurement.²⁹ Differences between an apical reading, as would be typical with ultrasonic pachymetry, and the true thinnest point can vary greatly particularly in keratoconic corneas where the cone is often displaced (Fig. 2).^{21,29,37,41} While the clinical utility of posterior corneal data and a full thickness map are evident, there is currently no accepted classification/staging system incorporating this information.

ENHANCED REFERENCE SURFACE

The additional information available from anterior segment tomographic devices lead to the development of various refractive surgery screening programs.^{10,26-28,32,33,38-44} Once such program is the Belin-Ambrosio Enhanced Ectasia Display (BAD) (Fig. 3).

The BAD display (available on the Pentacam, Oculus GmbH, Wetzlar, Germany) utilizes both anterior and posterior elevation data and pachymetric data to screen for ectatic change. It displays the elevation data against the commonly used best-fit-sphere (BFS) taken from the central 8.0 mm zone, but also uses a newly developed reference surface called the 'Enhanced Reference Surface'.⁴⁴⁻⁴⁸

The concept behind the 'Enhanced Reference Surface' is to generate a surface that more closely resembles the patient's more normal peripheral cornea, as this will further magnify any existing ectatic pathology. A small diameter optical zone centered on the thinnest portion



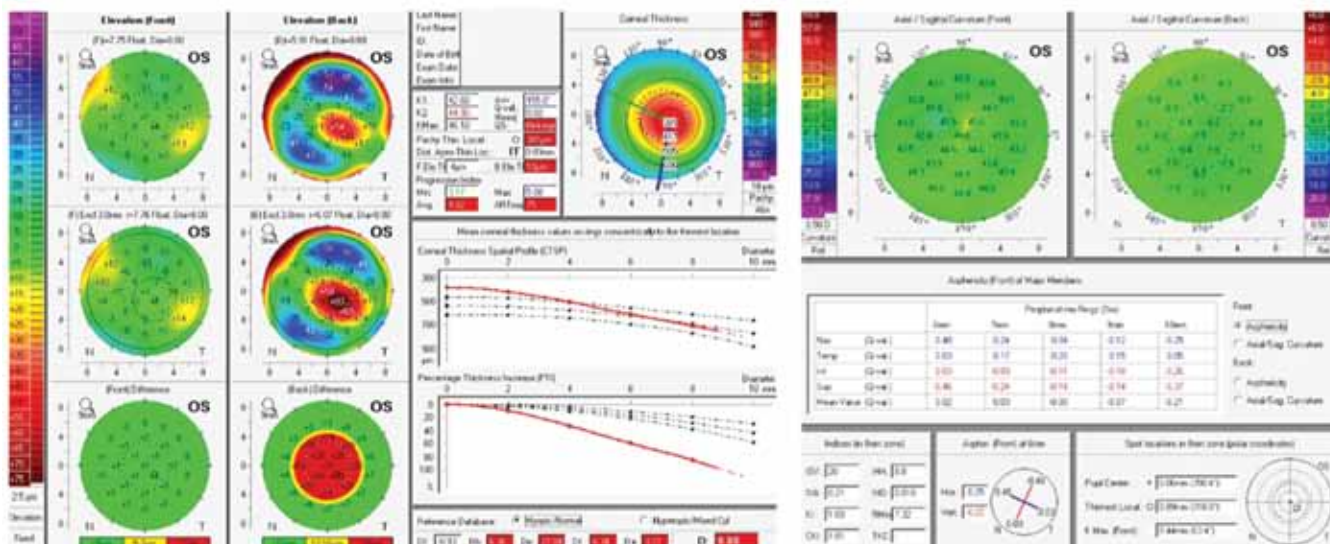


Fig. 1: An example of subclinical keratoconus. The BAD display on the left shows advanced keratoconus with a prominent posterior ectasia, highly abnormal pachymetric progression and a final 'D' > 8 standard deviations from the norm in spite of a normal anterior surface and normal anterior indices shown on the right topometric display

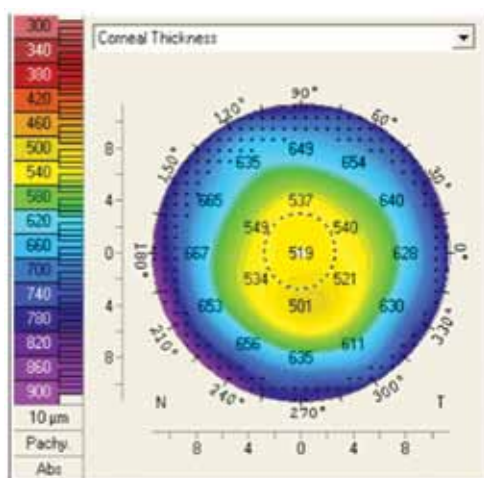


Fig. 2: Full corneal thickness map showing a significant difference between the apical reading of 519 and the thinnest point which is located inferotemporal

of the cornea is excluded from the standard 8.0 mm BFS reference shape calculation. The new 'enhanced BFS' utilizes all the valid elevation data from within the 8.0 mm central cornea, and outside the exclusion zone (Fig. 4). The exact size of the exclusion zone varies between 3.0 and 4.0 mm based on a proprietary algorithm, but typically 3.0 mm in keratoconic corneas.

The resulting new reference surface (Enhanced Reference Surface) more closely approximates the more normal peripheral cornea and exaggerates any conical protrusion (Fig. 5).⁴⁵⁻⁴⁸

With a conical cornea, excluding this small zone from the BFS calculation eliminates the cone or steep portion of the cornea and results in a significantly flatter BFS that is based more on the normal peripheral cornea. The resulting elevation maps show a significant difference as

the conical portion of the cornea is now more pronounced (i.e. easier to identify) (Fig. 6).

Because normal eyes are only minimally prolate, excluding this zone from normal eyes has little effect on the elevation maps. The elevation maps using the standard BFS and the enhanced BFS will look remarkably similar (Fig. 7).⁴⁵

Not only was the enhanced reference surface useful qualitatively in visualizing subtle or early ectatic change, but the elevation difference between a standard BFS and the enhanced reference surface proved to be highly significant quantitatively in separating normal eyes from those with ectatic change (Table 2).⁴⁵

The enhanced reference surface works because the exclusion zone centered on the thinnest point incorporates the major ectatic region. Excluding this zone from the standard 8 mm BFS results in a reference surface that closely mimics the more normal portions of the cornea.^{39,45-48}

'One man's trash is another man's treasure'—Unknown

A similar concept can be used to stage or classify keratoconus. As opposed to excluding the 3.0 to 4.0 mm

Table 2: Elevation change from standard BFS to enhanced BFS

	Normal	Keratoconus	p-value
Anterior elevation change apex	1.86 ± 1.9 μm	20.4 ± 23.1 μm	0.0001
Max anterior elevation change	1.63 ± 1.4 μm	20.9 ± 21.9 μm	0.0001
Posterior elevation change apex	2.86 ± 1.9 μm	39.9 ± 38.1 μm	0.0001
Max posterior elevation change	2.27 ± 1.1 μm	45.7 ± 35.9 μm	0.0001

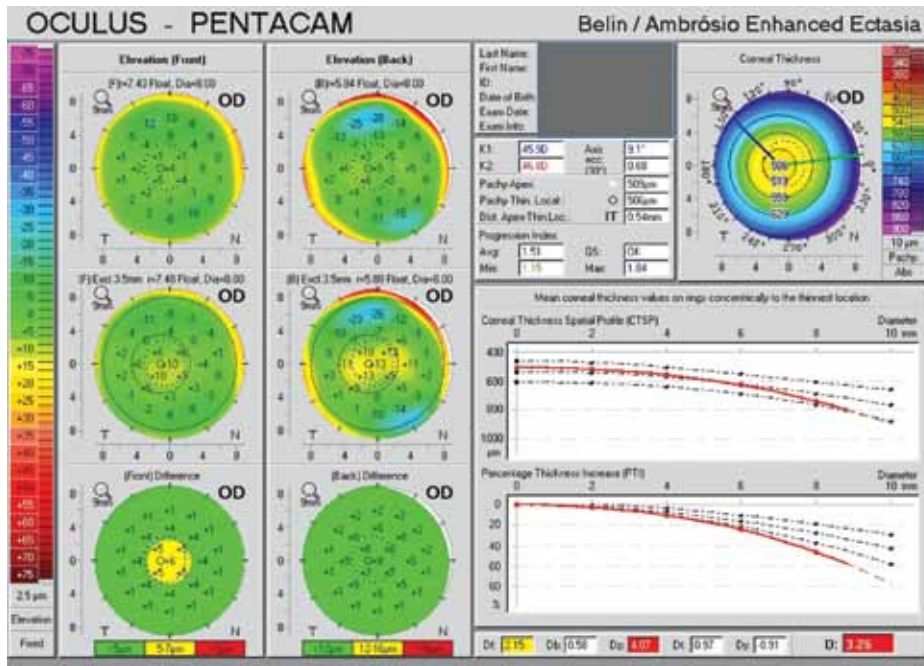


Fig. 3: Sample of the Belin/Ambrosio enhance ectasia display. The left side of the map shows anterior and posterior elevation with a standard 8.0 mm BFS and the enhanced reference surface. The right side shows the corneal thickness analysis and the pachymetric progression graphs

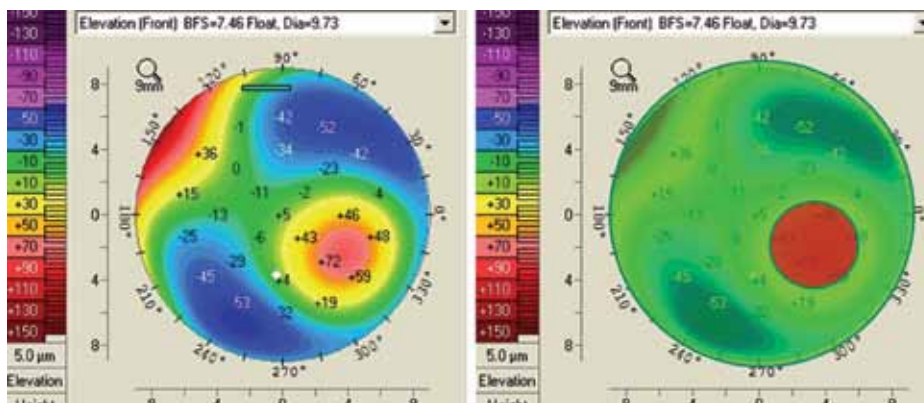


Fig. 4: Standard anterior elevation map on the left in the cornea with a prominent ectasia and the depiction of the exclusion zone on the right (in red)



Fig. 5: Schematic drawing of the enhanced reference surface which more closely follows the more normal corneal periphery resulting in a flatter reference surface. The flatter reference surface allows for greater separation between the enhanced reference surface and the ectatic region

zone, we should look at the exclusion zone centered on the thinnest point as this area represents the ectatic region better than a single point parameter, such as K_{max} or maximal elevation.⁴⁷ The goal was to develop a classification/staging system that had some similarities to the AK system for anterior data, but addressed the following deficiencies:

- Absence of posterior data.
- Relying on apical corneal thickness as opposed to thinnest point.
- Failure to distinguish normal from possible pathology.
- Inability to classify a cornea when different parameters fall into different stages.
- Lack of visual acuity considerations.

MATERIALS AND METHODS

The study population was a previously described normative database of 682 eyes/341 patients.⁴⁹ Each patient had at least 3 years of uneventful follow-up. All files were previously examined by two fellowship trained, experienced refractive surgeons (MWB, RA). All files were reanalyzed with Pentacam software version 6.08r13. The newer software has more strict criteria than the original and subsequently 5 eyes were flagged as not acceptable



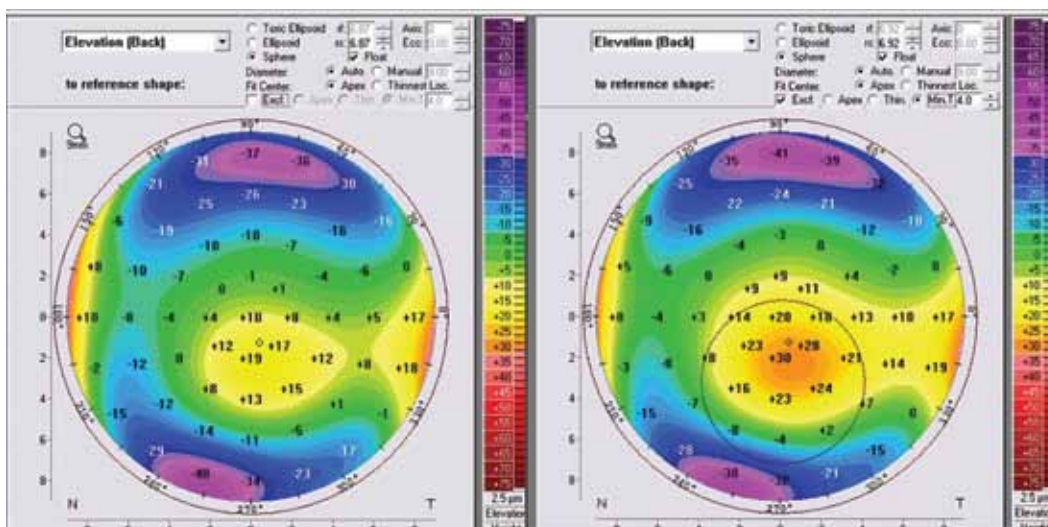


Fig. 6: A comparison of the posterior elevation maps in an eye with moderate keratoconus. The map on the left uses the standard BFS, while the map on the right uses the enhanced reference surface. The enhanced reference surface map shows significantly greater elevation values

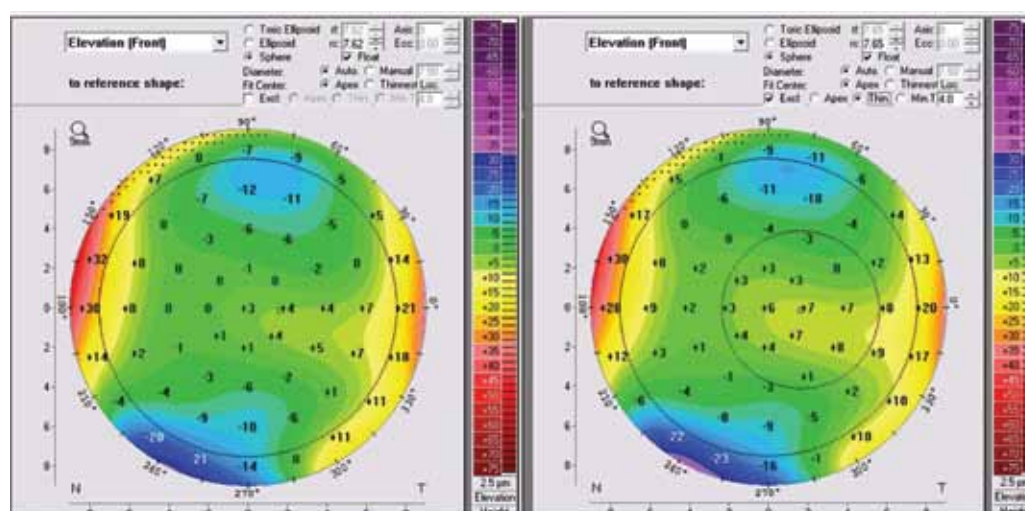


Fig. 7: A comparison of anterior elevation in a normal eye with the standard reference surface (left) and the enhanced reference surface (right). There is minimal elevation difference between the maps

quality. If any single eye was flagged, both eyes were removed from analysis resulting in 672 eyes/336 patients. As opposed to the previously described normative data which was based on the standard 8.0 mm BFS, the new data reported corneal thickness at the thinnest point and radius of curvature for the anterior surface (ARC) and posterior surface (PRC) for a 3.0 mm zone centered on the thinnest point. The 3.0 mm zone was chosen as this is the exclusion zone size utilized in the BAD software for most keratoconic corneas. The normative data generated from this database was then used to develop a classification system which approximated stages 1 to 4 on the AK system for anterior data and corneal thickness, but added a stage 0, representing more 'normal' values. Once the anterior data gates were established, similar gates based on the standard deviations derived from the anterior surface were utilized for the posterior surface.

RESULTS

A total of 672 eyes of 336 'normal' patients were analyzed. There were 52% females/48% males with an average age of 44.9 years (25–75). Anterior and posterior ROC values were 7.65 ± 0.236 mm/ 6.26 ± 0.214 mm respectively and thinnest pachymetry values were 534.2 ± 30.36 μ m (Table 3). Comparing anterior curvature values to AK staging yielded 2.67, 5.47, 6.44, >6.44 standard deviations for stages 1 to 4 respectively. Comparative pachymetric values yielded 4.42, 7.72, >7.72 standard deviations for stages 2 to 4 respectively (AK criteria has no pachymetric value for stage 1). Posterior staging uses the same standard deviation gates as generated for the anterior data (Table 4).

STAGING

The goal of our study was to propose a new classification/staging system that both addressed the deficiencies of the

Table 3: Values for anterior and posterior radius of curvature and thinnest point

	Anterior radius of curvature (mm)	Posterior radius of curvature (mm)	Corneal thickness at thinnest point (µm)
Mean	7.65	6.26	534.2
Median	7.64	6.25	533
STD	0.236	0.214	30.36
Range	6.89–8.66	5.61–6.93	454–614

AK system, utilized current imaging capabilities, be independent of a specific imaging device, be clinically user friendly, convey meaningful anatomic and functional information and, in the future, may have the potential to be utilized to determine ectatic progression. To this end, we propose a new staging system that is similar to the approach taken by the Spaeth angle classification where each anatomic component is independently graded.⁵⁰ The new grading system named ABCD looks at the anterior radius of curvature (A), posterior radius of curvature (B for back surface), corneal pachymetry at thinnest (C), Distance best-corrected vision (D), and adds a modifier (-) for no scarring, (+) for scarring that does not obscure iris details and (++) for scarring that obscures iris details (Table 5).

This grading system is relatively simple to use and has the advantage of grading each component independently, recognizing subclinical disease, and adding a stage 0 to better reflect an absence of possible disease. The grading system is dependent on tomography to produce both

posterior data and thinnest point pachymetry, but this information could be available from any commercial tomographic unit (i.e. Scheimpflug, slit-scanning, OCT). What is not currently available is the radius of curvature at a specific diameter (3.0 mm) surrounding the thinnest point. We feel this is critical to better reflect the true ectatic region, and this software modification can easily be added to other systems other than the one used in our study (Oculus Pentacam).

The greatest hindrance to a clinical adoption is the lack of familiarity ophthalmologist have in using radius of curvature instead of diopters. Radius of curvature is typically used in contact lens fitting and spectacle design but fewer ophthalmologists fit contacts than in the past. Radius of curvature was selected to allow the same measurement of both the anterior and posterior surfaces as radius of curvature is independent on index of refraction. The posterior corneal surface is a negative lens with a low power due to the cornea/aqueous interface. Reporting the true dioptric power of the posterior cornea would be even less intuitive. For ease of adjustment, the posterior surface power is shown as an anterior power equivalent using the same radius to diopter conversion commonly used for anterior surface keratometry.

$$\text{Diopters} = 337.5/\text{radius of curvature (mm)}$$

A sample application of the new ABCD grading system is shown in Figure 8. The BAD display shows mild to moderately advanced keratoconus with a final 'D' of 4.88. The anterior changes are relatively minor with an

Table 4: Comparable values for anterior and posterior radius of curvature and thinnest point

	AK criteria	Comparable ARC	Comparable PRC	Comparable thickness
Stage I	Avg K < 48.0 D	<2.63 STD >7.05 mm	>5.70 mm	
Stage II	Avg K < 53.0 D	<5.47 STD	>5.15 mm	4.42 STD
	Apical thickness > 400 µm	>6.35 mm		>400 µm
Stage III	Avg K > 53.0 D	<6.44 STD	>4.95 mm	7.72 STD
	Apical thickness > 300 µm	>6.15 mm		>300 µm
Stage IV	Avg K > 55.0 D	>6.44 STD	<4.95 mm	<300 µm
	Apical thickness < 300 µm	<6.15 mm		

Table 5: ABCD keratoconus classification

ABCD criteria	A	B	C	D	Scarring
	ARC (3 mm zone)	PRC (3 mm zone)	Thinnest pach. µm	BDVA	
Stage 0	>7.25 mm (<46.5 D)	>5.90 mm (<57.25 D)	>490 µm	≥ 20/20 (≥ 1.0)	-
Stage I	>7.05 mm (<48.0 D)	>5.70 mm (<59.25 D)	>450 µm	<20/20 (<1.0)	-, +, ++
Stage II	>6.35 mm (<53.0 D)	>5.15 mm (<65.5 D)	>400 µm	<20/40 (<0.5)	-, +, ++
Stage III	>6.15 mm (<55.0 D)	>4.95 mm (<68.5 D)	>300 µm	<20/100 (<0.2)	-, +, ++
Stage IV	<6.15 mm (>55.0 D)	<4.95 mm (>68.5 D)	≤ 300 µm	<20/400 (<0.05)	-, +, ++

Scarring – clear; no scarring (-); scarring, iris details visible (+); scarring, iris obscured (++); Diopters shown for anterior radius of curvature; anterior equivalent diopters shown for posterior radius of curvature



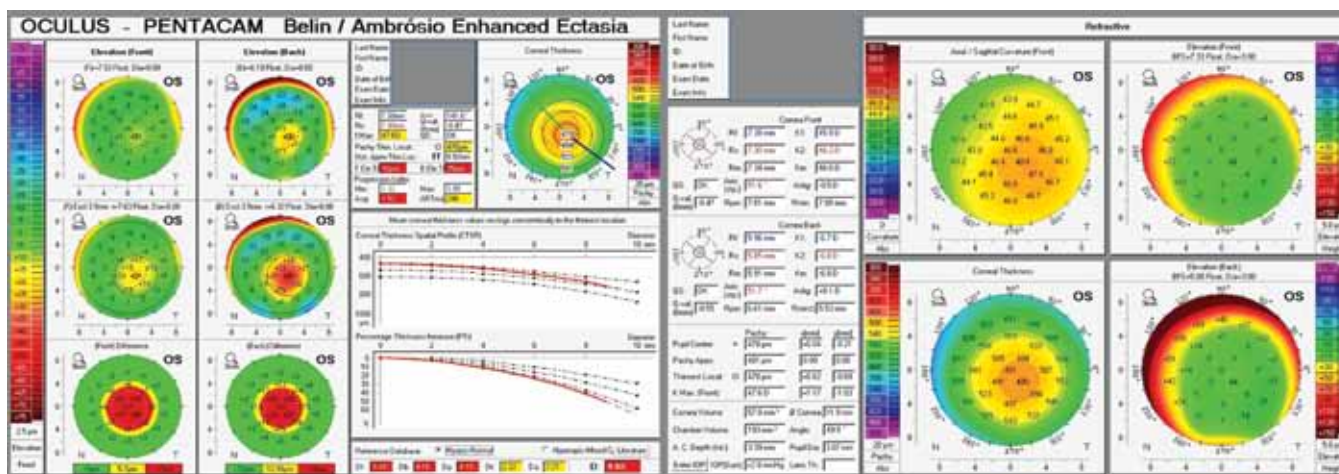


Fig. 8: Sample of the ABCD grading in mild to moderate keratoconus. The ARC is 7.34, the PRC 5.88 and the thinnest pachymetry 470. There is no corneal scarring and the visual acuity is 20/30+. The final staging is A0/B1/C1/D1-

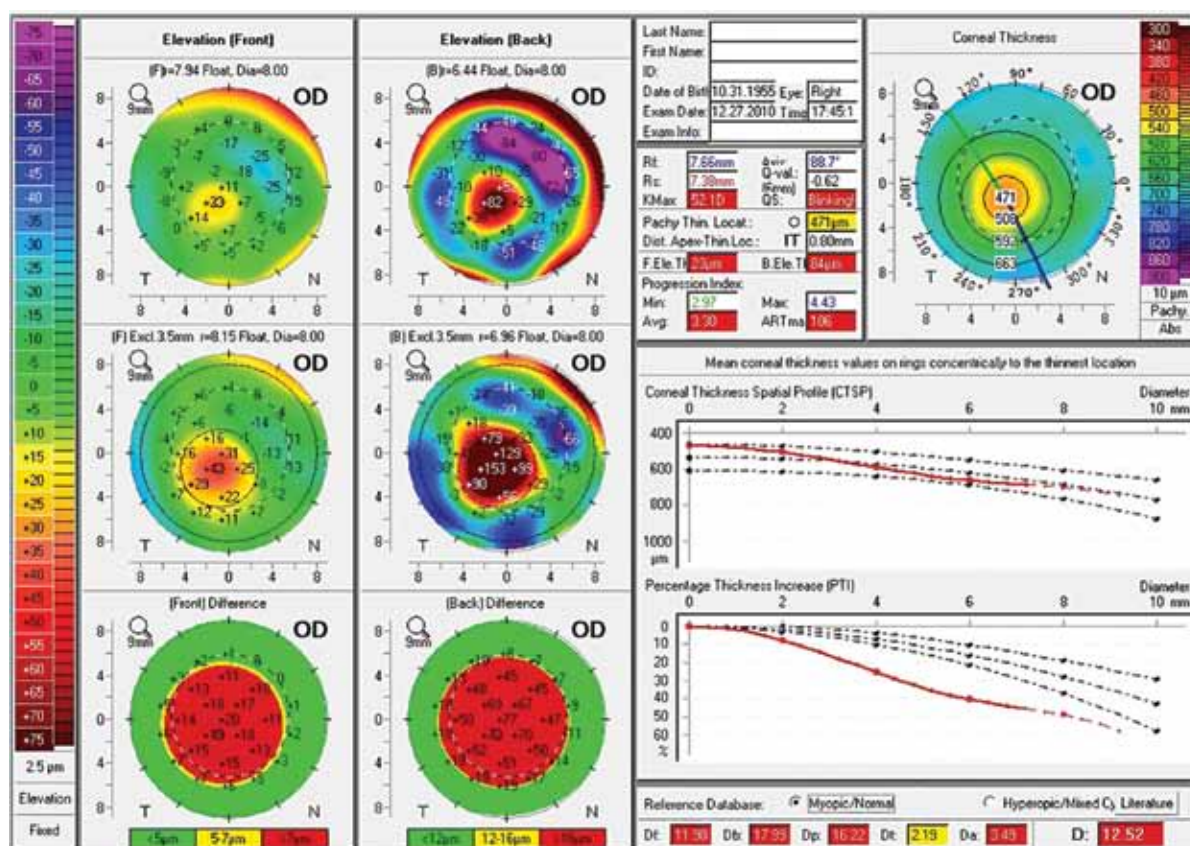


Fig. 9: Sample of moderately advanced keratoconus. The visual acuity is decreased to 20/40+ and there is not corneal scarring. The final classification is A2/B2/C1/D1-

average K of 7.34 mm (46.0 D). The corneal thickness map shows a thinnest reading of 470 μm with a slight inferior-temporal displacement. Both the posterior surface and pachymetric progression show greater change with a central posterior radius of curvature of 5.91 μm. The cornea exhibited no scarring and the patients BDVA OS was 20/30+. The ARC and PRC taken from the 3 mm zone centered on the thinnest point were 7.34 and 5.88 respectively. The ABCD classification (Table 5) for this cornea would be A0/B1/C1/D1-.

Figure 9 depicts a more advanced cone, but again the posterior surface changes are more severe. Visual acuity is 20/40+ and there is no visible corneal scarring. The ABCD classification in this example would be A2/B2/C1/D1-.

The third example (Fig. 10) is a case of markedly advanced keratoconus with mild scarring and a best corrected vision of 20/200. The classification would be A4/B4/C2/D3+. The '+' indicating mild corneal scarring.

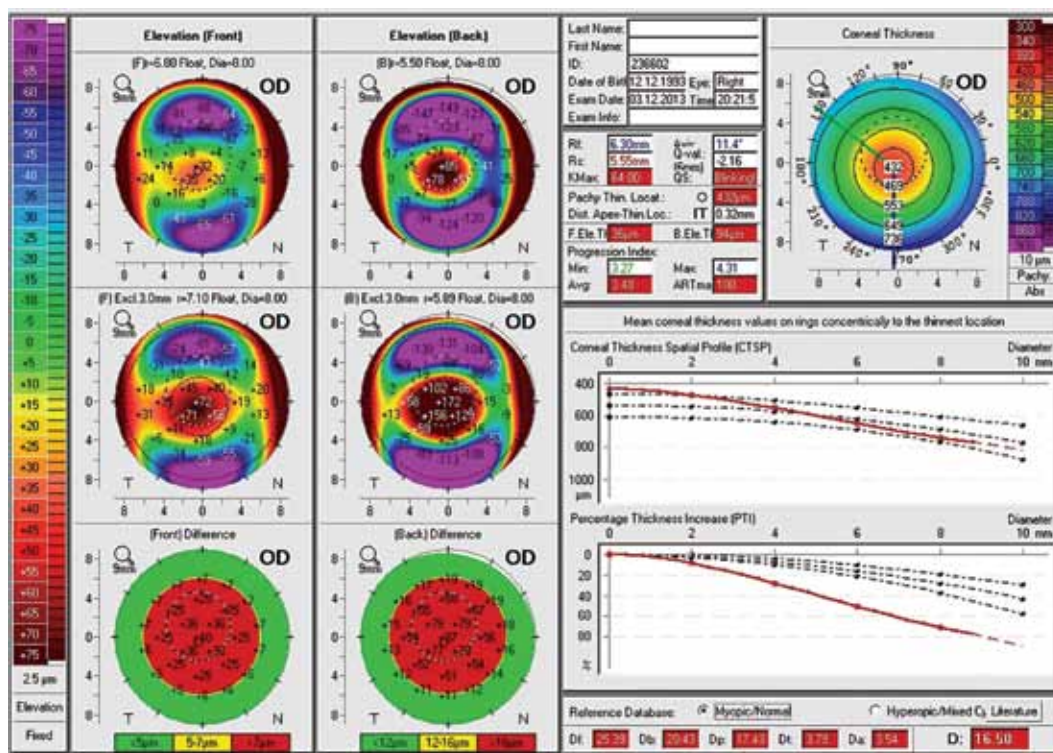


Fig. 10: Sample of markedly advanced keratoconus with a best-corrected visual acuity of 20/200 and mild apical scarring. The final ABCD grade would be A4/B4/C2/D3+

CONCLUSION

The proposed new classification system conveys both anatomical and functional data that are missing from the AK classification. It conveys information on both anterior and posterior corneal surfaces, is centered on the thinnest point which is typically the region of the cone and adds a visual acuity measurement as well as an indication of corneal scarring. The new ABCD classification allows for a much improved description of the keratoconic cornea than was previously possible. It also may allow for more tailored treatment plans as different surfaces of the cornea may be more amenable to different medical or surgical intervention.

The new ABCD classification system should be available in the near future on the oculus pentacam. The classification display will compute the ARC and PRC at the 3 mm zone centered on the thinnest point, and the thinnest pachymetry. The operator would need to add the distance visual acuity and the presence or absence of scarring and the display would automatically classify the cornea according to the ABCD criteria. Other tomographic systems may similarly develop a comparable classification display.

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