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Keratoconus: The ABCD Grading System

Keratokonus: Das ABCD-System zur Stadieneinteilung

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Abstract

Purpose: To propose a new keratoconus classification/staging system that utilises current tomographic data and better reflects the anatomical and functional changes seen in keratoconus.
Method: A previously published normative database was reanalysed to generate both anterior and posterior average radii of curvature (ARC and PRC) taken from a 3.0 mm optical zone centred on the thinnest point of the cornea. Mean and standard deviations were recorded and anterior data were compared to the existing Amsler-Krumeich (AK) Classification. ARC, PRC, thinnest pachymetry and distance visual acuity were then used to construct a keratoconus classification.
Results: 672 eyes of 336 patients were analysed. Anterior and posterior values were 7.65 ± 0.236 mm and 6.26 ± 0.214 mm, respectively, and thinnest pachymetry values were 534.2 ± 30.36 µm. The ARC values were 2.63, 5.47 and 6.44 standard deviations from the mean values of stages 1–3 in the AK classification, respectively. PRC staging uses the same standard deviation gates. The pachymetric values differed by 4.42 and 7.72 standard deviations for stages 2 and 3, respectively.
Conclusion: A new keratoconus staging incorporates anterior and posterior curvature, thinnest pachymetric values, and distance visual acuity and consists of stages 0–4 (5 stages). The proposed system closely matches the existing AK classification stages 1–4 on anterior curvature. As it incorporates posterior curvature and thickness measurements based on the thinnest point, rather than apical measurements, the new staging system better reflects the anatomical changes seen in keratoconus.

Zusammenfassung

Zielsetzung: Es wird hier ein neues Klassifizierungssystem bzw. ein neues System zur Stadieneinteilung bei Keratokonus vorgestellt, das auf aktuellen tomografischen Daten beruht. Das neue Klassifizierungssystem reflektiert die mit der Keratokonus-Erkrankung verbundenen anatomischen und funktionellen Veränderungen der Hornhaut besser.
Methode: Eine bereits zuvor veröffentlichte normative Datenbank wurde erneut analysiert, um Durchschnittswerte für die vorderen und hinteren kornealen Krümmungsradien (R_v und R_h) zu bekommen. Die Messungen wurden in einer 3,0 mm optischen Zone vorgenommen, welche die dünnste Hornhautstelle als Mittelpunkt nahm. Mittelwerte und Standardabweichungen wurden aufgezeichnet, und die neu ermittelten Daten für den vorderen Krümmungsradius wurden mit der bestehenden Klassifikation nach Amsler und Krumeich (AK) verglichen. R_v, R_h, Pachymetrie-Werte für die dünnste Stelle und Sehschärfe im Fernbereich wurden dann verwendet, um ein neues Keratokonus-Klassifikationssystem zu entwickeln.
Ergebnisse: Es wurden 672 Augen von 336 Patienten analysiert. Der Durchschnittswert der vorderen und der hinteren Krümmungsradien war 7,65 ± 0,236 mm bzw. 6,26 ± 0,214 mm, und der dünnste Pachymetrie-Wert war 534,2 bzw. 30,36 µm. Verglichen mit den auf der AK-Klassifizierung basierenden Einteilungsstadien 1–3 wichen die R_v-Werte jeweils um 2,63, 5,47 und 6,44 Standardabweichungen vom Mittelwert ab. Die auf R_h-Werte beruhende Stadieneinteilung verwendet denselben Rahmen für die Standardabweichung. Ein Vergleich der Pachymetrie-Werte ergab eine Standardabweichung von 4,42 bzw. 7,72 für das Stadium 2 bzw. 3.
Fazit: Das neue System zur Stadieneinteilung bezieht die vordere und die hintere Krümmung, die

jeweiligen an der dünnsten Stelle gemessenen Pachymetrie-Werte und die Sehschärfe im Fernbereich mit ein, und besteht nunmehr aus den Stadien 0–4 (insgesamt 5 Stadien). Das hier vorgeschlagene System hat große Ähnlichkeit mit den bereits existierenden AK-Klassifizierungsstadien 1–4 für die vordere Krümmung. Weil aber das neue System auch die hintere Krümmung einbezieht und die Messung der Hornhautdicke an der dünnsten Stelle anstatt am Apex durchgeführt wird, trägt das neue System den anatomischen Veränderungen im Keratokonus besser Rechnung.

Introduction

Recent advances in the diagnosis and treatment of ectatic disease have increased the necessity of having a staging or classification system that accurately reflects the physical changes seen. Additionally, newer imaging modalities (e.g. Scheimpflug, Ocular Coherence Tomography [OCT]) provide additional anatomical information previously not available. Several classification systems for keratoconus have been proposed in the literature [1–10]. The Amsler-Krumeich (AK) system (**Table 1**) is amongst the oldest and still the most widely used. The AK system grades keratoconus into 4 stages based on spectacle refraction, central keratometry, presence or absence of scarring, and central corneal thickness [11–14].

Other studies such as the Collaborative Longitudinal Evaluation of Keratoconus (CLEK) Study used changes in vision, keratometry, biomicroscopic signs, corneal scarring, and vision-specific quality of life, as measures to define stage and severity of disease. Topographic analysis was not used in either the AK or CLEK classifications [15].

None of the commonly used systems incorporate posterior corneal data or analyze the full corneal thickness map [16]. According to the recently published *Global Consensus on Keratoconus and Ectatic Diseases* (2015) there is currently no clinically adequate classification system for keratoconus [17]. The most widely used AK system predates modern imaging. 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 [16]. Differences between an apical reading and the true thinnest point can vary greatly particularly in keratoconic corneas where the cone is often displaced (**Fig. 1**) [14,16,18,19]. 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.

The additional information available from anterior segment tomographic devices lead to the development of various refractive surgery screening programs [1,19–30]. Once such program is the Belin-Ambrosio Enhanced Ectrasia Display (BAD) (**Fig. 2**). 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 and also uses a reference surface called the "*Enhanced Reference Surface*" [30–34]. The "*Enhanced Reference Surface*" eliminates a small diameter optical zone (3.0–4.0 mm) centered on the thinnest point from the standard 8.0 mm BFS computation. 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 flatter reference surface that more closely reflects those portions of the cornea less typically altered by keratoconic ectatic changes and results in greater separation between the ectatic region and the reference surface (**Fig. 3**) [26,31–35].

As opposed to excluding the 3.0 to 4.0 mm zone centered on the thinnest point, that zone can be utilized as it better reflects the major ectatic region than a single point parameter such as Kmax or maximal elevation [33]. 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:

1. Lack of posterior data,
2. Relying on apical corneal thickness as opposed to thinnest point,
3. Lack of visual acuity considerations,
4. Different parameters may fall into different stages,
5. Poor differentiation of normal from abnormal.

Table 1 Standard Amsler-Krumeich keratoconus 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 µm
Stage III	Myopia/astigmatism > 8.00 D but < 10.00 D Mean K > 53.0 D Absence of scarring Minimal apical corneal thickness < 400 µm but > 300 µm
Stage IV	Refraction not possible Mean K > 55.0 D Central corneal scarring Minimal apical corneal thickness < 300 µm

Methods

The study population was a previously described normative database of 682 eyes/341 patients [35]. All files were reanalyzed with Pentacam software version 6.08r13. The newer software has stricter criteria than the original and subsequently 5 eyes were flagged as not acceptable quality. If any single eye was flagged, both eyes were removed from analysis resulting in 672 eyes/336 patients. Special software was developed to compute the 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. ARC and PRC from the 3.0 mm zone centered on the thinnest point is not cur-

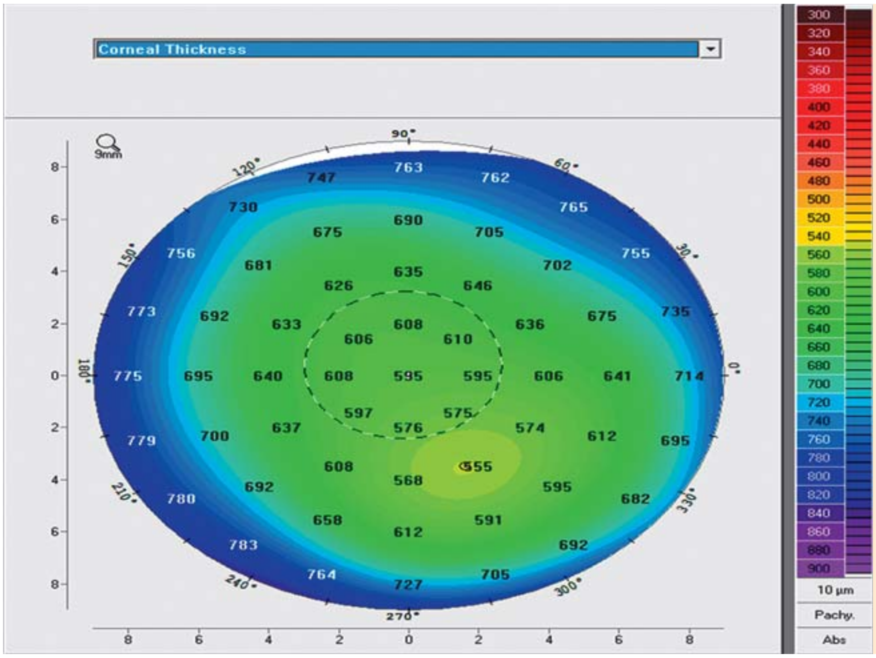

Fig. 1 Full corneal thickness map showing a significant difference between the apical reading of 595 µm and the thinnest point of 555 µm which is located inferiorly.

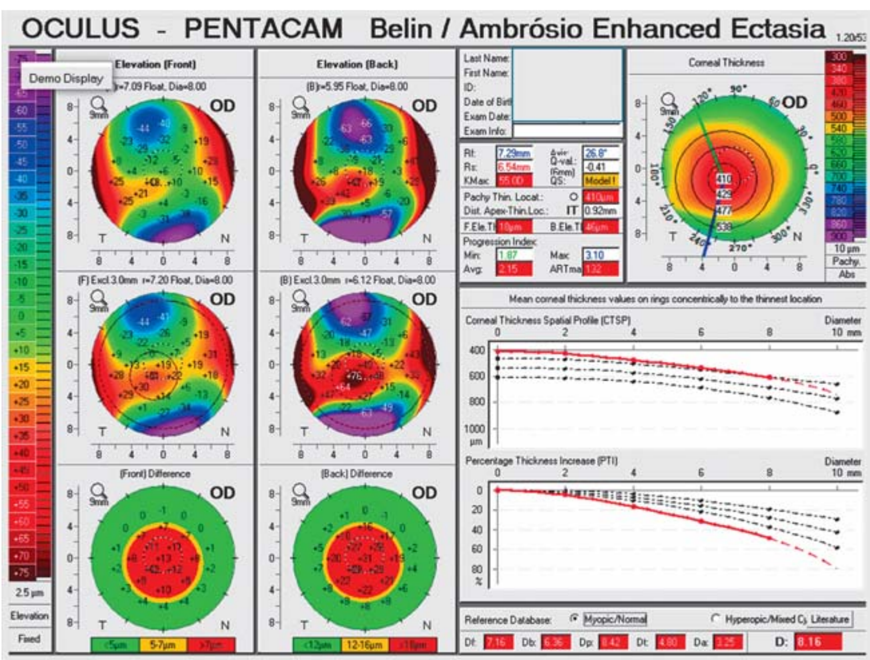

Fig. 2 Sample of the Belin/Ambrosio Enhanced Ectasia Display. The left side of the map shows anterior and posterior elevation with a standard 8.0 mm BFS and the enhanced reference surface. The right side shows the corneal thickness analysis and the pachmetric progression graphs.

rently available on the Pentacam. The data generated from this database was used to develop a classification system that approximated stages 1–4 on the AK system for anterior data and corneal thickness, but added a fifth stage (stage 0), representing values more typically seen in normal eyes. Similar gates based on the standard deviations derived from the anterior surface were utilized for the posterior surface.

Results

▼

672 eyes of 336 "normal" patients were analyzed. There were 52 % females/48 % males with an average age of 44.9 years (range 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 µm (**Table 2**). Comparing anterior curvature values to AK staging yielded 2.67, 5.47, 6.44, > 6.44 standard deviations for stages 1–4 respectively. Comparative pachymetric values yielded 4.42, 7.72, > 7.72 standard deviations for stages 2–4, 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 3**).

Discussion

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The proposed new grading system named **ABCD** looks at the **A**nterior radius of curvature (**A**), Posterior radius of curvature (**B** for

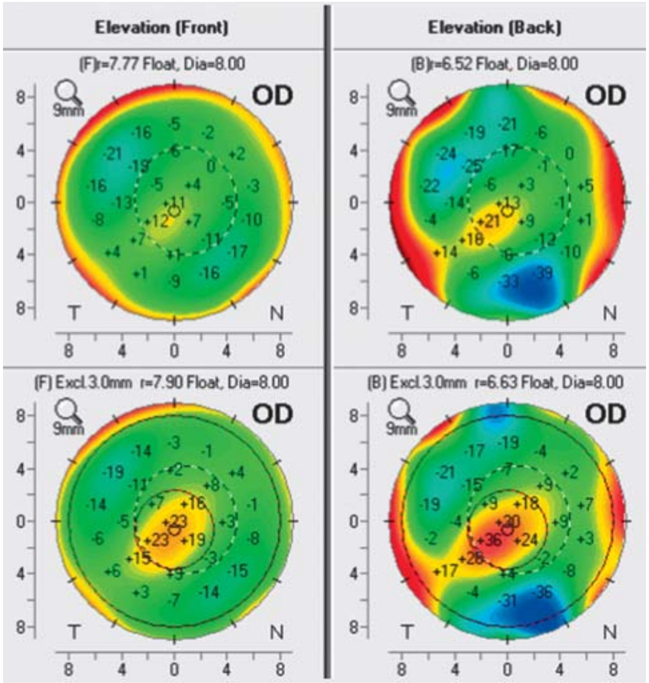

Fig. 3 A comparison of anterior and posterior elevation with the standard BFS (upper left and upper right) with the enhanced reference surface (lower left and lower right). Both anterior and posterior elevation show increased values with the enhanced reference surface.

Table 2 Study population values (mean, median, standard deviation and range) for anterior and posterior radius of curvature and corneal thickness at the 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

back surface), **C**orneal pachymetry at thinnest (**C**), **D**istance 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 4**).

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 probable 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).

A sample application of the new **ABCD** grading system is shown in **Fig. 4**. The BAD display shows early or subclinical keratoconus with a final "D" of 2.37. The accompanying topometric map displays classic anterior curvature based indices which are all

	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 Apical thickness > 400 µm	< 5.47 STD >6.35 mm	> 5.15 mm	4.42 STD >400 µm
Stage III	Avg K > 53.0 D Apical thickness > 300 µm	< 6.44 STD >6.15 mm	> 4.95 mm	7.72 STD >300 µm
Stage IV	Avg K > 55.0 D Apical thickness < 300 µm	> 6.44 STD <6.15 mm	< 4.95 mm	< 300 µm

Table 3 Equivalent values for the study population based on comparable anterior curvature and pachymetry values from the Amsler-Krumeich classification for anterior radius of curvature (ARC) and corneal thickness at the thinnest point. Posterior radius of curvature values (PRC) used the same standard deviations as the ARC.

Table 4 Proposed ABCD keratoconus grading system. Stages (0 to IV) are based on anterior and posterior radius of curvature (ARC, PRC), thinnest pachymetry, best corrected distance visual acuity (BDVA) and a modifier for the presence of corneal scarring.

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

Scarring – clear, no scarring (−), scarring, iris details visible (+), scarring, iris obscured (++); Diopters also shown for anterior radius of curvature

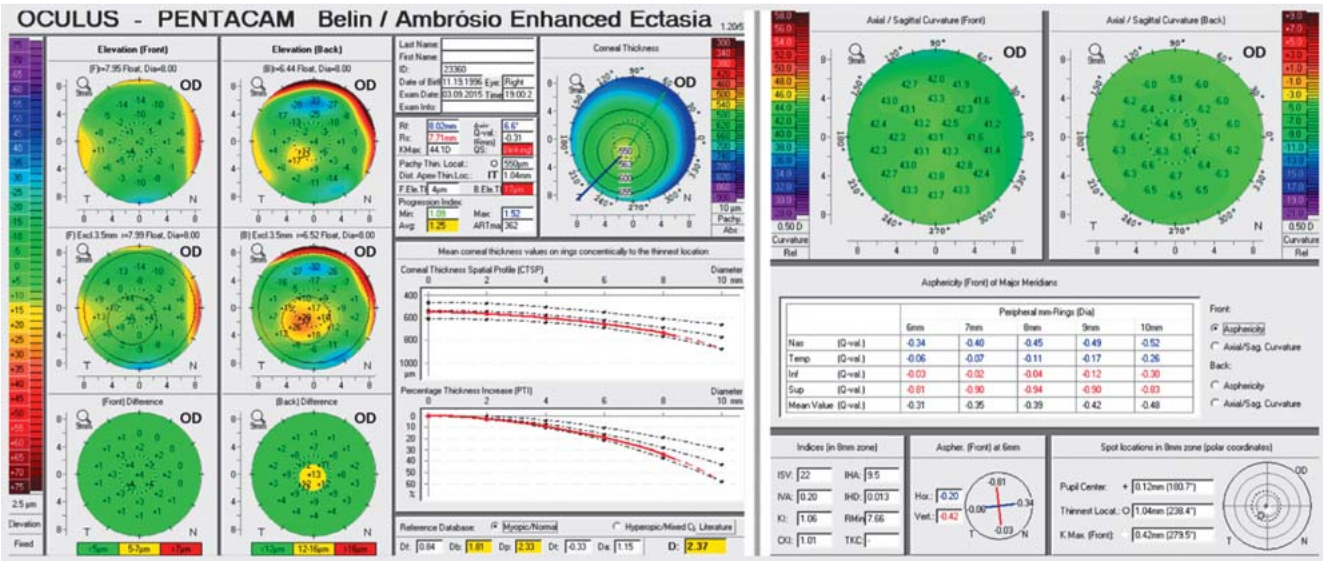

Fig. 4 Comparison of the BAD display (left) showing posterior elevation abnormalities (subclinical keratoconus) to the topometric display (right) which uses only anterior curvature indices, all of which are normal. The ABCD classification would be A0/B1/C0/D0–.

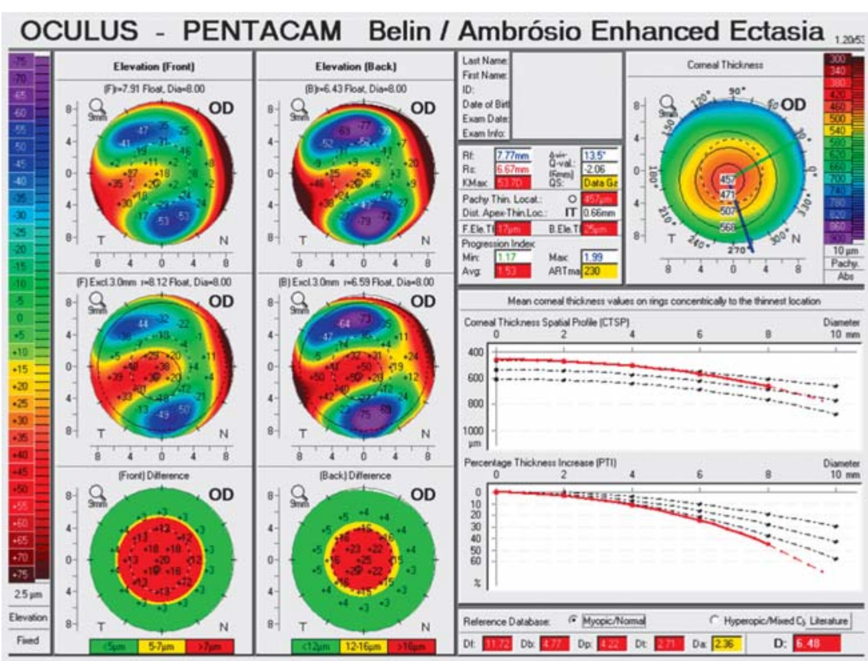

Fig. 5 Sample of moderately advance keratoconus. The visual acuity is decreased to 20/40– and there is no corneal scarring. Thinnest pachymetry is 457, ARC and PRC are 6.22 and 5.00 microns. The final classification is A3/B3/C1/D2–.

normal. The corneal thickness map shows a thinnest reading of 550 µm with a slight inferior-temporal displacement. The cornea exhibited no scarring and the patients BDVA OS was 20/20. The ARC and PRC taken from the 3 mm zone centered on the thinnest point were 7.65 and 5.85 respectively. The ABCD classification (**Table 4**) for this cornea would be A0/B1/C0/D0– reflecting the subclinical nature of the ectasia.

Fig. 5 depicts a more advanced cone. There is no visible scarring and the best corrected vision is 20/40-. The ARC and PRC are 6.22 and 5.00 mm respectively. The ABCD classification would be A3/B3/C1/D2-.

The third example (**Fig. 6**) is a case of markedly advanced keratoconus with mild scarring and a best corrected vision of 20/200. The classification would be A4/B4/C3/D3+. The "+" indicating mild corneal scarring.

Conclusion

The proposed new classification system conveys both anatomical and functional data that is missing from the Amsler-Krumeich 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. It also may allow for more tailored treatment plans as different surfaces of the cornea may be more amenable to different medical or surgi-

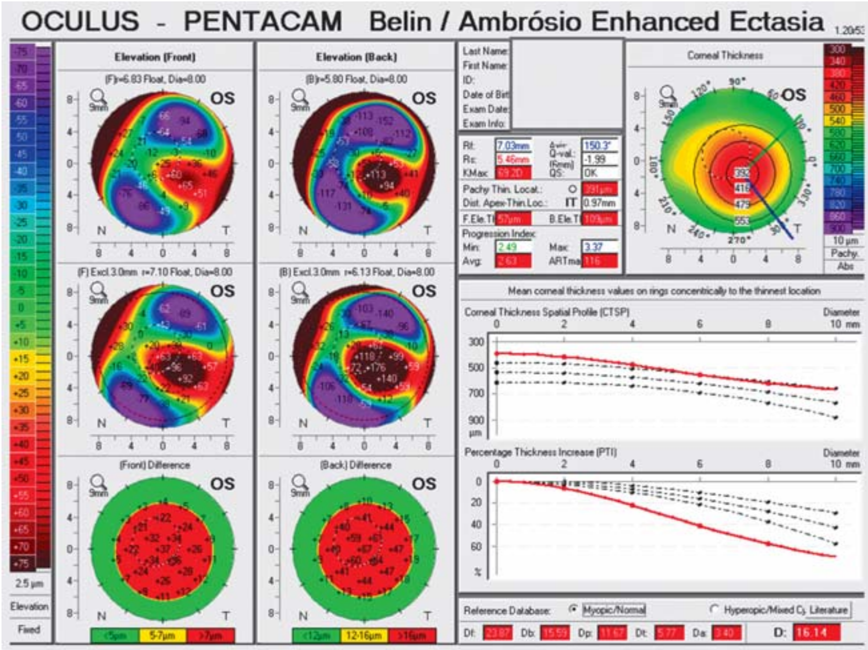

Fig. 6 Sample of markedly advanced keratoconus with a best corrected visual acuity of 20/200 and mild apical scarring. Thinnest pachymetry is < 400 and the final ABCD grade would be A4/B4/C3/D3+.

cal intervention. The new ABCD classification system should be available in the near future on the Oculus Pentacam. A similar grading system could be made available on other tomographic systems.

Conflict of Interest

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Consultant OCULUS GmbH.

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