

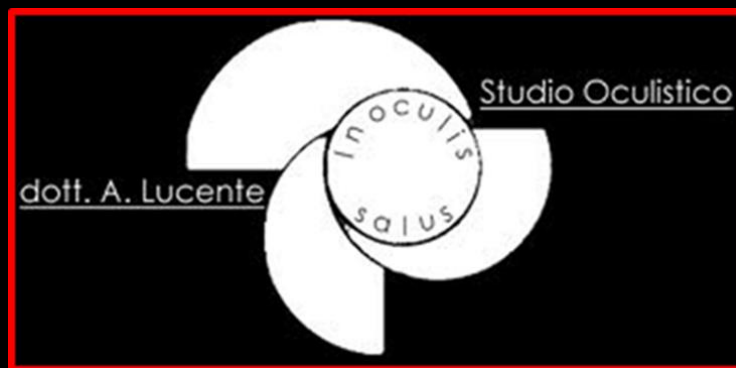
# Nuove Frontiere in diagnostica e chirurgia oftalmica

27/28 maggio 2016 – **Alghero**

Sessione Patologia della cornea e del limbus: Gestione dei casi complessi

Moderatori: **Pierangelo Pintore** - **Francesco Zanetti** - **Alain Serru**

## *Cheratocono ed altre ectasie corneali diagnosi clinica e strumentale*



[www.amedeolucente.it](http://www.amedeolucente.it)

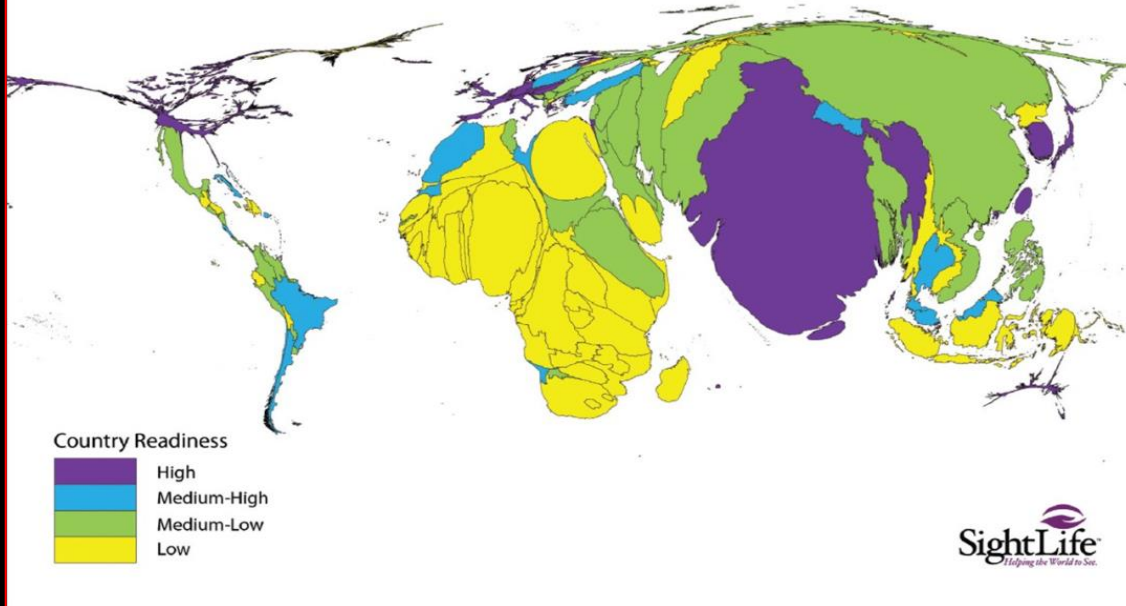
# *Disclosure*

Consulting Free:

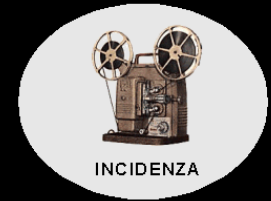
- *Carl Zeiss Meditec*
- *Alfa Intes*

## Corneal Blind Estimates and Corneal Transplantation Readiness

Size of countries reflects the population of corneal blind.  
Colors represent readiness for eye banking and corneal transplantation.



## Background



PRACTICAL OBSERVATIONS  
ON  
CONICAL CORNEA,  
AND ON THE  
SHORT SIGHT,  
AND  
OTHER DEFECTS OF VISION CONNECTED WITH IT.

BY  
J. NOTTINGHAM, M.D.,

FELLOW OF THE ROYAL COLLEGE OF PHYSICIANS OF ENGLAND, CORRESPONDING  
MEMBER OF THE MEDICAL SOCIETY OF EXETER AND OF PARIS, OF THE ROYAL  
MEDICAL SOCIETY OF BRISTOL, OF THE SOCIETY OF MEDICINE AND  
SURGERY OF BOSTON AND BIRMINGHAM, AND OF THE ACADEMY  
OF NATURAL SCIENCES OF BRUSSELS. LONDON: BY  
JOHN STURROTT AND CO. LONDON, 1854.

- **incidenza:** eventi/tempo
- **prevalenza:** eventi/popolazione

- **Nottingham J.** *Practical Observation on Conical Cornea: and on the Short Sight, and Other Defects of Vision Connected with it.* London: J Churchill; **1854**
- Corneal Steepening + visual distortion + apical corneal thinning + central corneal scarring
- **Apical Corneal Thinning: Typically Inferotemporal, central, superior**
- Usually becomes apparent second decade of the life, normally during puberty
- **Progresses until the fourth decade of life when it usually stabilized**
- **Annual Incidence of 2/100.000, Prevalence of 54,5/100.000**

# Amsler-Krumeich AK

1-4 stage

Spectacle Refraction

Central Keratometry

Presence or absence of scarring central

Corneal Thickness

# Classification AK

## Stage I

- Eccentric steeping.
- Myopia and/or induced astigmatism  $<5.00$  D
- Mean central K readings  $<48.00$  D.

## Stage III

- Myopia and/or induced astigmatism from 8.00 to 10.00 D.
- Mean central K readings  $>53.00$  D.
- Absence of scarring.
- Minimum corneal thickness 300 to 400  $\mu\text{m}$ .

## Stage II

- Myopia and/or induced astigmatism from 5.00 to 8.00 D
- Mean central K readings  $<53.00$  D
- Absence of scarring.
- Minimum corneal thickness  $>400$   $\mu\text{m}$

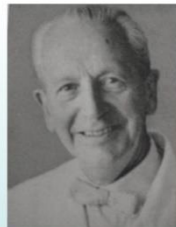
## Stage IV

- Refraction not measurable.
- Mean central K readings  $>55.00$  D.
- Central corneal scarring.
- Minimum corneal thickness 200  $\mu\text{m}$ .

Amsler M. Keratocone classique et keratocone fruste; arguments unitaires.

Ophthalmologica

1946



Marc Amsler

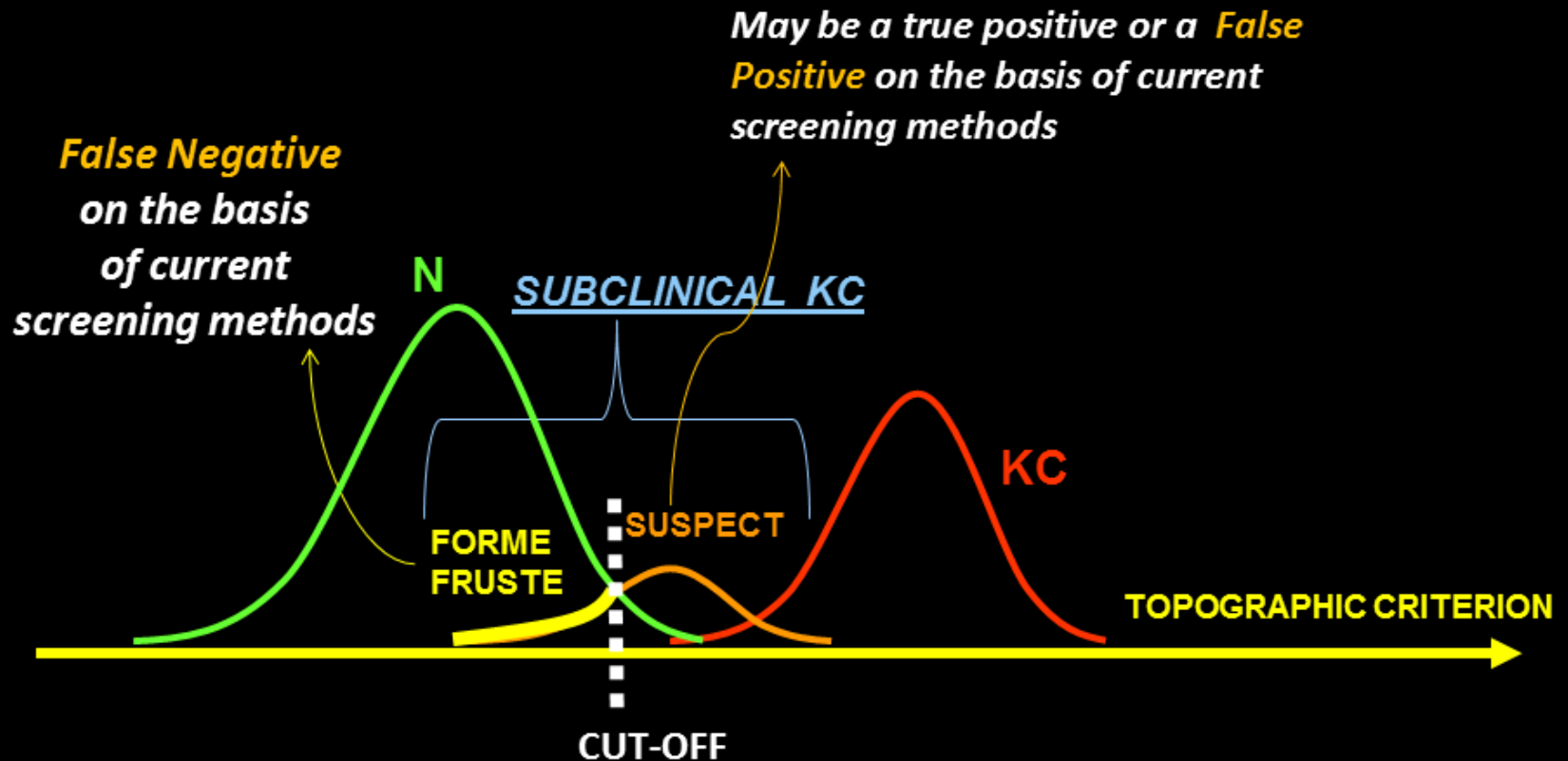
Swiss ophthalmologist, born 1891, died 1968.



Jorg H. Krumeich

# KERATOCONUS: SIGNS

- SUSPECT vs FORME FRUSTE vs SUBCLINICAL KC ...



## ***Univariate quantitative detection systems***

- Simulated keratometry (SIMK)
- Surface asymmetry index (SAI)
- Surface Regularity index (SRI)
- Central Keratometry (K Central)
- Inferior-Superior Value (I-S)
- Average corneal power (ACP)
- Topographic irregularity (IT)
- Skew of steepest radial axis (SRAX)
- Corneal uniformity index (CU)
- Irregular astigmatism index (IAI)
- Calossi-Foggi Top-Bottom Index

- Apex curvature (AK)
- Asphericity coefficient (Q)
- Effective refractive power (EffRP)
- Corneal Irregularity Measurement (CIM)
- Analyzed area (AA)
- Mean Toric Keratometry (MTK)
- Centre Surround Index (CSI)
- Different Sector Index (DSI)
- Opposite industry Index (OSI)
- Orbscan surface irregularity

## ***Multivariate quantitative detection systems***

- KISA% (calculated from four indices: Central K, SIMK, I-S and SRAX)
- Belin/Ambrósio Enhanced Ectasia Display III (BAD III)
- Keratoconus severity index (KSI) Smolek-Klyce method
- Keratoconus Index (KCI) Klyce-Maeda method
- Keratoconus prediction index (KPI)
- PathFinder Corneal Analysis
- Rabinowitz and McDonnell Index

# Ectatic Corneal Disease

## “Global Consensus of Keratoconus and Ectatic Disease” (April 2015)

### 4 Supranational Corneal Societies:

- **Asia Cornea Society** (Asia)
- **Cornea Society** (USA and international)
- **EuCornea** (Europe)
- **PanCornea** (Latin America, United States, Canada)

- Each Society: **9 experts**  
- Total : **36 participants**  
**plus coordinators**

- **Keratoconus**
- **Pellucid marginal degeneration PMD**
- **Keratoglobus**
- **Post-refractive disorders (Ambrósio et al, 2014)**

**Not be classified as ectatic diseases but rather as thinning disorders**

- *Terrien’s marginal degeneration*
- *Dellen*
- *Inflammatory melts*

Currently, there is no clinically adequate classification system for **keratoconus**

- Keratoconus and PMD are different clinical presentations of the same disease
- Keratoglobus and keratoconus are different clinical entities
- True unilateral keratoconus does not exist
- Posterior corneal elevation abnormalities must be present to diagnose early or subclinical keratoconus
- Central pachymetry is the least reliable indicator (determinant) for diagnosing keratoconus
- Keratoconus can be present in a cornea of normal central thickness

# *Diagnostic Imaging Technology for Corneal Analysis*

- **Corneal Topography**

(Placido, Cassini multicolored-spot reflection topographer, US FDA approved, 700 LED)

- **Corneal Tomography**

(Slit Scanning-Orbiscan, Scheimpflug imaging-Pentacam, Scheimpflug/Placido-Galilei, Sirius, TMS5 Tomey)

- **AS-OCT, SD-OCT/SS-OCT**

- **Ultrasound Biomicroscopy**

(UMB 35/50MHz, High Frequency Ultrasound Scanner-50MHz ArcScan Artemis 3)

- **Scheimpflug Noncontact Tonometry**

(ORA, Corvis)

- **Confocal Microscopy**



# Best parameter for documenting disease progression

- **Parameter 1: Anterior Corneal Surface**  
"Will not show changes on the anterior surface until later the disease process"
- **Parameter 2: Epithelial Thickness**  
"Is no published literature exploring the progression of disease with relation to epithelial thickness"
- **Parameter 3: BCVA**  
"Due to this variability, BCVA is not a reliable measure of keratoconus progression"
- **Parameter 4: Corneal Thickness**  
"A full corneal thickness map does have good potential to document progression"
- **Parameter 5: Posterior Surface**  
"Has strong potential for measuring disease progression"

BY **MICHAEL BELIN**,  
CATARACT & REFRACTIVE SURGERY  
TODAY EUROPE **JULY/AUGUST 2014**

**The BAD Berlin/Ambrósio Display for Pentacam**  
sensitivity 99,59% and specificity 100%



R. Ambrósio



M. Berlin

# ABCD Grading System

## ABCD Grading System

### Criteria A and ARC (3.0mm Zone)

Anterior Radius of Curvature. Average curvature in the 3.0mm zone centered on the thinnest location of the cornea.

### Criteria B and PRC (3.0mm Zone)

Posterior Radius of Curvature in the 3.0mm zone. Average curvature in the 3.0mm zone centered on the thinnest location of the cornea.

### Criteria C and Thinnest Pachymetry

Thinnest pachymetry in  $\mu\text{m}$

### Criteria D and DCVA

"Distance Best Corrected Visual Acuity"

DCVA is not generated by the Pentacam software. It should be input manually by clicking left mouse into the corresponding field.

The input value will be saved to the examination.

Whole number stages are rounded down from the number with decimal place.

ABCD Criteria	A ARC (3 mm Zone)	B PRC (3 mm Zone)	C Thinnest Pach $\mu\text{m}$	D BDVA	Scarring
STAGE 0	> 7.25 mm ( $< 46.5$ D)	> 5.90 mm ( $< 57.25$ D)	> 490 $\mu\text{m}$	= 20/20 (= 1.0)	-
STAGE I	> 7.05 mm ( $< 48.0$ D)	> 5.70 mm ( $< 59.25$ D)	> 450 $\mu\text{m}$	< 20/20 ( $< 1.0$ )	-, +, ++
STAGE II	> 6.35 mm ( $< 53.0$ D)	> 5.15 mm ( $< 65.5$ D)	> 400 $\mu\text{m}$	< 20/40 ( $< 0.5$ )	-, +, ++
STAGE III	> 6.15 mm ( $< 55.0$ D)	> 4.95 mm ( $< 68.5$ D)	> 300 $\mu\text{m}$	< 20/100 ( $< 0.2$ )	-, +, ++
STAGE IV	< 6.15 mm ( $> 55.0$ D)	< 4.95 mm ( $> 68.5$ D)	= 300 $\mu\text{m}$	< 20/400 ( $< 0.05$ )	-, +, ++

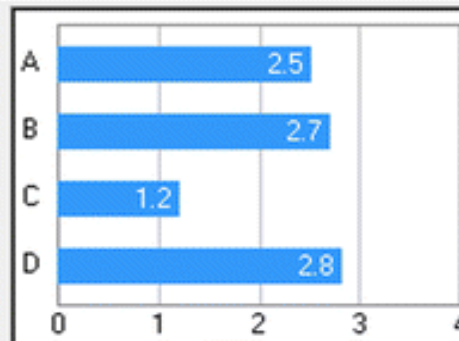
## Belin ABCD Keratoconus Staging

ARC (3mm Zone): 6.64 mm

PRC (3mm Zone): 5.26 mm

Thinnest Pachy: 480  $\mu\text{m}$

DCVA: 20/80



A2 B2 C1 D2

Klin Monbl Augenheilkd. 2016 Jan 20  
Keratoconus: The ABCD Grading System  
Belin MW, Duncan JK

## 4 Criteria

"A" for **anterior**, ACR

"B" for **back** surface, PCR

"C" for **corneal** thickness

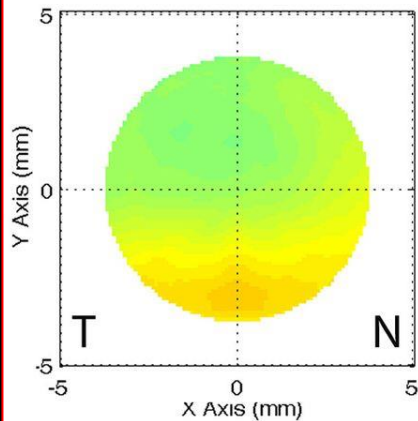
"D" for **distance** visual acuity

672 eyes of 336 patients were analysed

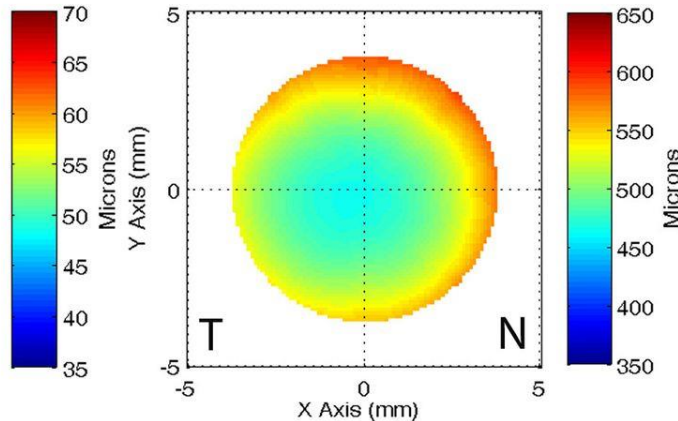
ARC, PRC (anterior and posterior radius of curvature), thinnest pachymetry and distance visual acuity

## Normal Average

Epithelial Thickness

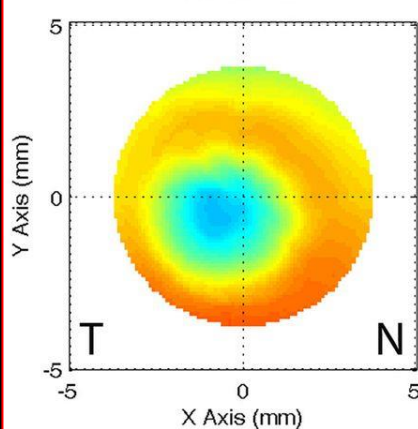


Stromal Thickness

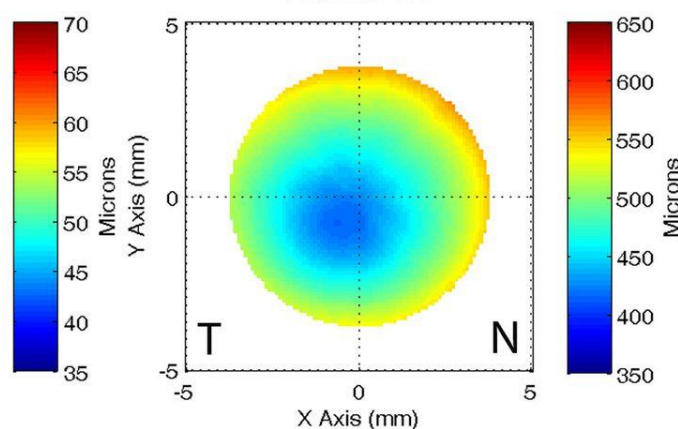


## Keratoconus Average

Epithelial Thickness

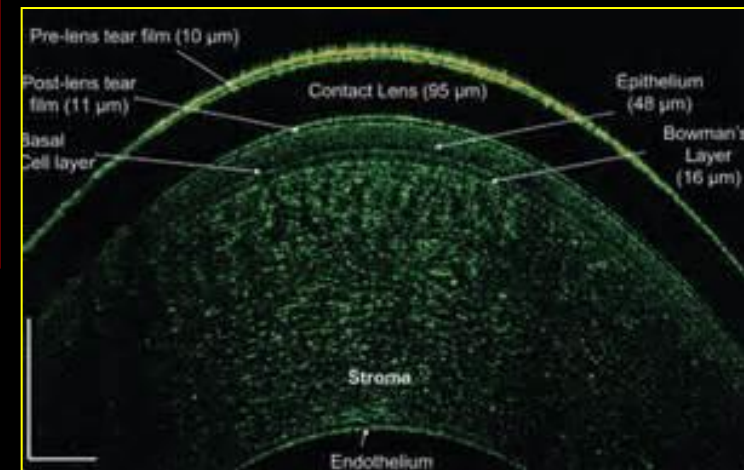


Stromal Thickness



## Epithelial Thickness Mapping *ETM*

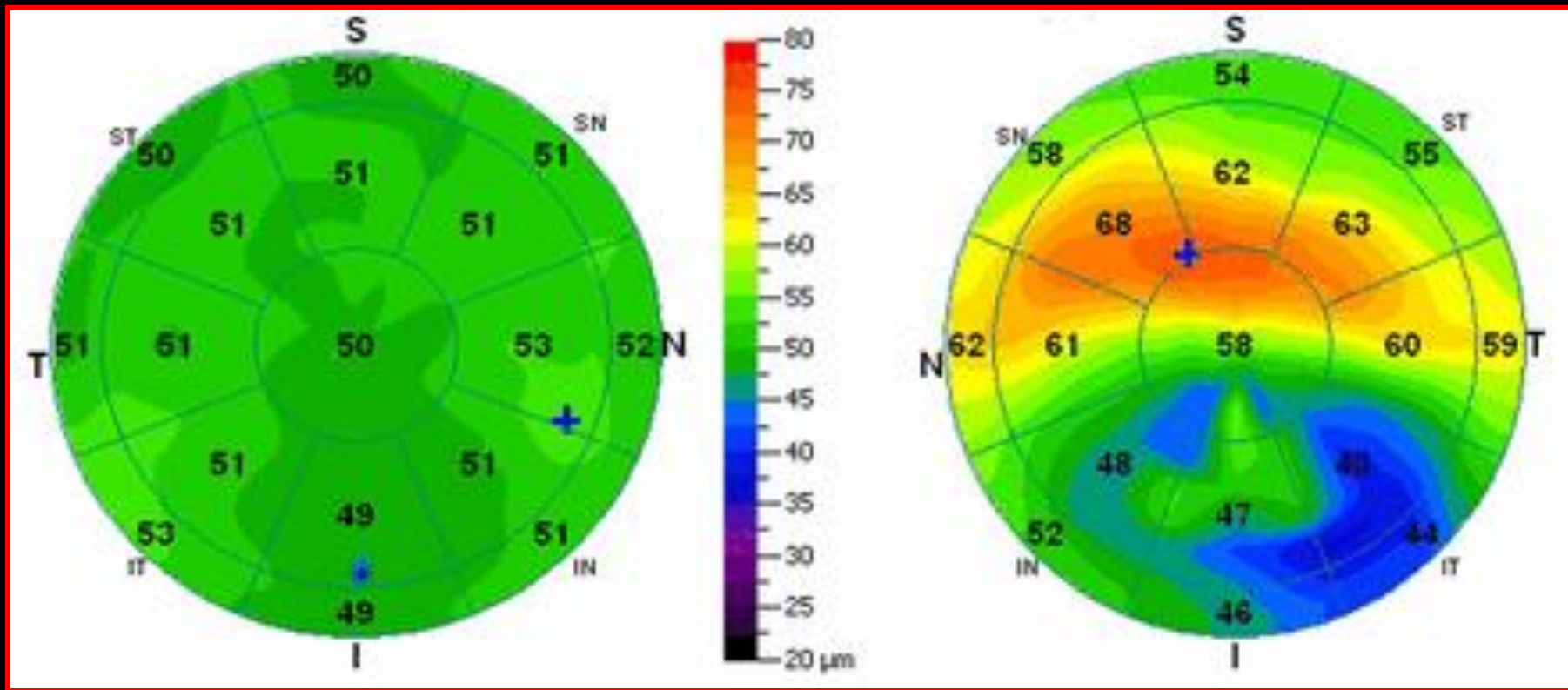
Distribution of epithelial maps has been studied in **373 normal subjects** using **AS-OCT imaging**. Epithelial pachymetry shows a nearly normal distribution with a mean thickness of  **$53.28 \pm 3.34$  microns (range 45-60  $\mu\text{m}$ )**, while the topographical variation (**variability**) is  **$1.78 \pm 0.78$  microns**. **Kanellopoulos A.** Cornea. 2013 Nov



## Epithelial Thickness Mapping (*ETM*)

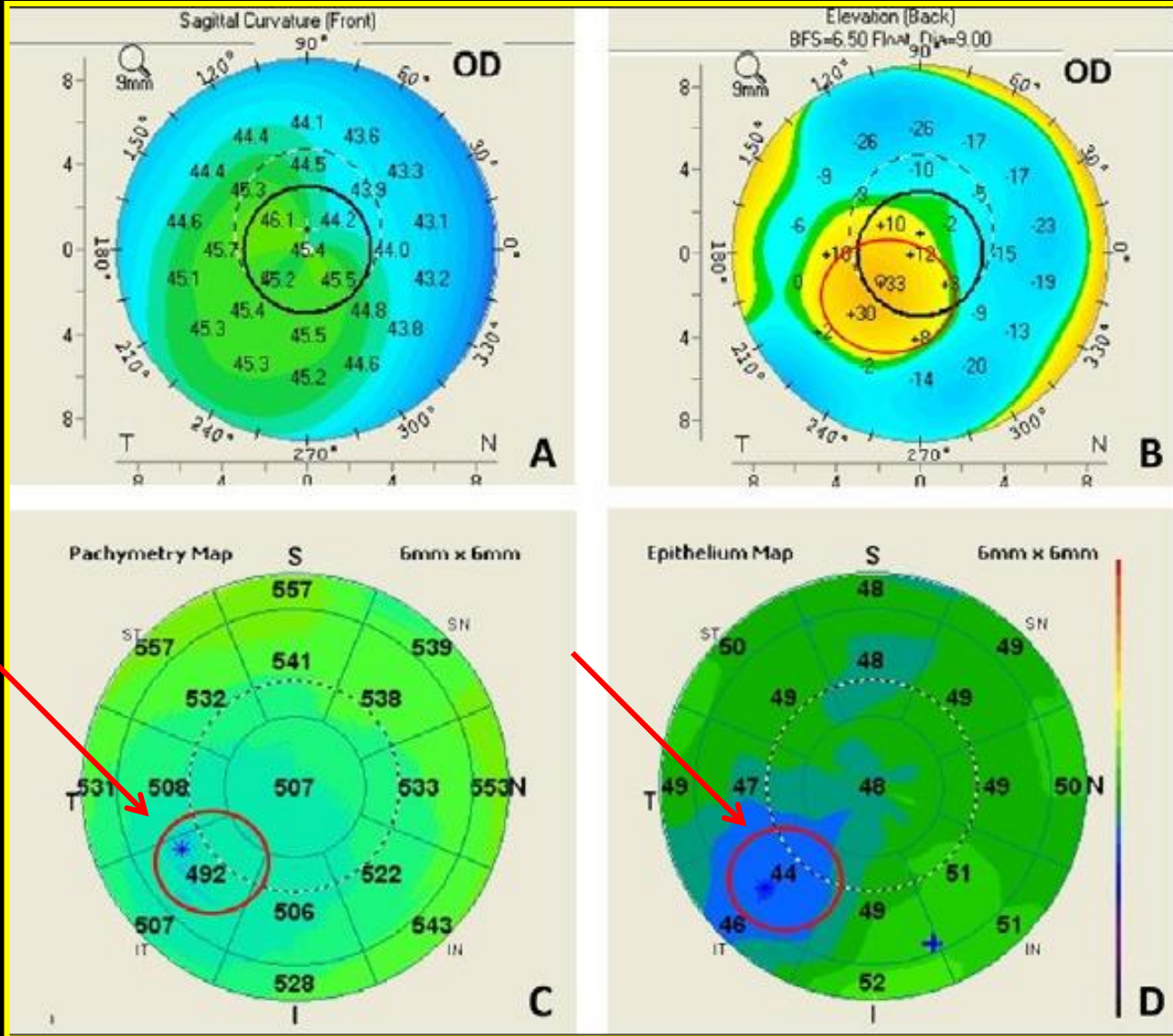
*Optovue SD-OCT Avanti software, color-coded map of the epithelial thickness across the central 6 mm of the cornea.*

# Examples of epithelial mapping of a normal (left) and a keratoconic patient (right) obtained by Optical Coherence Tomography



ARVO Annual Meeting Abstract  
2014 by A.J. Kanellopuolos



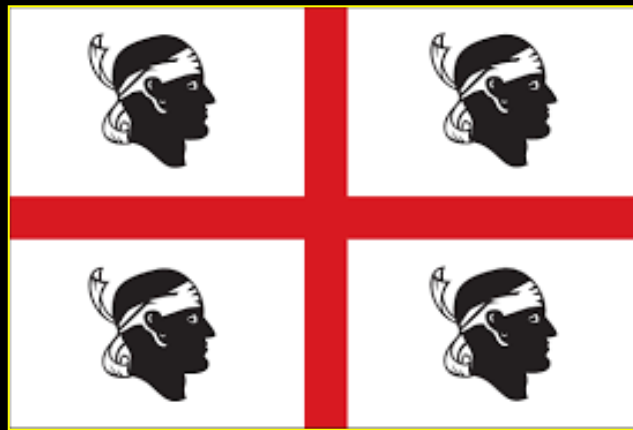


**(A)** Shows axial map of a forme **fruste keratoconus** showing **minimal changes** of keratoconus on the **anterior surface**, **(B)** shows the **posterior elevation** of the same eye showing **significant elevation**, **(C)** shows the total corneal pachymetry on Optovue RTVue OCT along with **(D)** epithelial thickness map, showing **localized epithelial thinning masking the cone**



# Take Message

- *Topography vs Tomography (Scheimpflug camera v/s AS-OCT)*
- *Corneal Thickness mapping*
- *Epithelial Thickness mapping (ultimate goal)*
- *Front and Back Cornea Surface*
- *Belin/Ambrósio Enhanced Ectasia Display BAD III*
- *Corneal biomechanical properties (ultimate goal)*  
(increase in deformation amplitude and oscillation for stiffness)



***Thank you for your kind attention***

