Global Consensus on Keratoconus and Ectatic Diseases—Edition 2

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Purpose: Ten years after the first Global Consensus on Keratoconus and Ectatic Diseases, this updated edition revisits key issues, incorporates advances in new technologies, and integrates the expertise of corneal and refractive surgeons from 12 international societies. This initiative aimed to establish a consensus among global ophthalmology experts, including corneal and refractive specialists from 6 continents, on the definition, diagnosis, staging, clinical management, and surgical treatment of keratoconus and ectatic corneal diseases.

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J. A. P. Gomes, F. Hafezi, and R. Ambrósio contributed equally to this work and should be considered joint first authors.

J. A. P. Gomes, F. Hafezi, and R. Ambrósio Jr. led the project design, coordinated the Delphi process, and co-wrote the manuscript. C. J. Rapuano co-wrote the manuscript and developed the flowchart. Topic coordinators were responsible for designing the round questionnaires, moderating the panels, analyzing the results, contributing to the in-person meeting in Chicago, and drafting the manuscript. Panelists contributed by completing group-specific questionnaires, discussing topics after the round results, and participating in the face-to-face meeting. M. Hillen co-wrote and reviewed the manuscript. Intrials Scientific Team (Celine Pompeia, Matheus José Barbosa Moreira, Regina Sider, and Rodrigo Pereira) contributed to the statistical analysis, methodological design, and drafting of the manuscript. All authors critically revised the manuscript for important intellectual content and approved the final version. The funding body had no role in the design, implementation, or interpretation of the results of this project. This funding defrayed the costs of statistical analysis, 4 rounds of the Delphi Panels, printed materials, the portfolio, supplements, questionnaires, and travel expenses for the Intrials teams to attend the face-to-face meeting in Chicago. All corneal societies contributed to both logistical support and publication expenses. The Cornea Society funded part of the face-to-face meeting in Chicago.

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Methods: The Delphi method consisted of 4 rounds of questionnaires, supplemented by a face-to-face meeting. A total of 128 ophthalmologists participated, with 3 main coordinators and 125 keratoconus experts distributed across 7 panels: definition/diagnosis/staging, clinical treatment and noninvasive visual rehabilitation, cross-linking for progression, and invasive visual rehabilitation (including therapeutic approaches, improved corrected distance visual acuity, keratoplasty techniques, and cataract surgery in keratoconus). The consensus threshold was defined as at least two-thirds agreement.

Results: A significant consensus was reached on topics such as definitions, diagnostic and progression criteria, and management strategies for keratoconus and other ectatic corneal disorders. A comprehensive approach was outlined, encompassing both non-surgical and surgical treatments, organized in a group-based approach. In addition, major agreements and disagreements across the 7 subcommittees are outlined in individual tables.

Conclusions: This updated Global Consensus provides revised definitions, expert statements, and recommendations for diagnosing and managing keratoconus and other ectatic corneal diseases. It benefits from broader participation, with more refractive surgeons and greater representation from major ophthalmic societies across 6 continents, capturing a wider range of global clinical perspectives than the prior report.

Key Words: keratoconus, corneal ectasia, consensus, corneal cross-linking, corneal transplantation, intraocular lenses

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Although keratoconus and other ectatic corneal diseases were first described nearly 2 centuries ago, it was only a decade ago that prominent corneal scientific societies convened to combine their expertise and formulate the first "Global Consensus on Keratoconus and Ectatic Diseases."[1–3] Its primary objective was to integrate the extensive clinical experience of specialists with the rapidly expanding knowledge generated by advances in diagnostic technologies, such as corneal topography and tomography, and treatments including corneal cross-linking (CXL) and intrastromal corneal ring segments (ICRS).[4]

Over the past decade, new diagnostic and therapeutic tools have been introduced, alongside a proliferation of research addressing the challenges faced by corneal and refractive surgeons worldwide.[5] Registries have enabled the collection of large amounts of clinical data on keratoconus, including patient-reported outcomes.[6] These developments have substantially advanced ectasia screening and early detection, including the ability to characterize intrinsic susceptibility to the disease. Diagnostic modalities have expanded to support staging, prognosis, monitoring, and individualized management of corneal ectatic diseases. Modern multimodal diagnostic approaches now go beyond Placido disk–based topography to include Scheimpflug three-dimensional tomography, layered epithelial and segmental mapping with anterior segment optical coherence tomography (AS-OCT), digital very high-frequency ultrasound (dVHF-US), and ocular wavefront analysis.[5,7–9] In addition, confocal microscopy enables in vivo cellular evaluation, whereas advances in cell and molecular biology and genetic testing provide unprecedented insights into the underlying pathophysiology and disease predisposition.[10] Artificial intelligence (AI) has been a key enabler of this shift, integrating and interpreting vast amounts of clinical data, enabling clinicians to synthesize structural, biomechanical, optical, cellular, and molecular information for more accurate and timely clinical decisions.[7,8,11] This comprehensive, multidimensional approach has enabled detailed assessment of anterior and posterior corneal architecture, cellular health, and the molecular landscape with a level of precision previously unattainable.

Crucially, in vivo biomechanical assessments, such as high-speed dynamic Scheimpflug imaging with the Corvis ST and Brillouin microscopy, now allow measurement of corneal mechanical properties and resilience, potentially enhancing the ability to identify susceptibility and preclinical stages of ectasia.[12–15] The combination of these advanced diagnostic tools, increasingly supported by AI, has strengthened both screening of refractive surgery candidates and guidance of personalized treatment strategies, while also refining the classification of keratoconus and related ectasias based on current knowledge and available technology.[5,10,16]

At the same time, clinical management of keratoconus and other ectatic diseases has advanced considerably, particularly through interventional and surgical approaches. Building upon the original "Dresden" CXL protocol, innovations, such as accelerated CXL and transepithelial (epi-on) CXL, have shortened procedure times and enhanced the safety profile of the procedure.[17] Other advances, such as enhanced riboflavin formulations, pulsed UV-A light delivery, and oxygen supplementation, have significantly improved the efficacy of these newer accelerated and epi-on CXL protocols, enabling earlier intervention and more effective disease stabilization. Together, these developments support the continued use of CXL as the standard of care for disease stabilization across all stages of keratoconus.[18] Invasive visual rehabilitation procedures—including photorefractive keratectomy (PRK), phakic intraocular lenses (IOLs), ICRS, and corneal allogenic intrastromal ring segments (CAIRS)—and elective refractive and cataract surgery, have evolved to support individualized treatment and improve visual quality with greater safety.[19–21] Keratoplasty, in the form of deep anterior lamellar keratoplasty (DALK), is considered the preferred technique when the endothelium is healthy, with surgeons adopting improved strategies to perform the procedure, manage complications, and reduce the need for conversion to penetrating keratoplasty (PK).[22,23] With this background, and in line with the Delphi panel methodology of updating the consensus every 10 years, we convened the "Global Consensus on Keratoconus and Ectatic Diseases—Edition 2."

METHODS

Design and Organization (Coordinators and Societies)

As in the first "Global Consensus on Keratoconus and Ectatic Diseases," a modified Delphi panel method (designated

"Delphi +1") was used to obtain updated expert consensus on key aspects of keratoconus and other corneal ectasias. The process involved 4 structured questionnaire rounds, including a face-to-face meeting to address unresolved issues.[1,24–26] The project was conducted by 3 main coordinators (J.A.P.G, F.H., and R.A.Jr.) who were experienced vision scientists and cornea or refractive surgery specialists with expertise in designing, conducting, and publishing expert panel studies. Their responsibilities included the following:

1. Evaluating the literature review and identification of appropriate peer-reviewed journal articles to provide as references for the panelists;
2. Designing the methodology;
3. Drafting the final manuscript; and
4. Project oversight.

A contract research organization (Intrials Clinical Research, São Paulo, Brazil) provided logistical, methodological, and statistical support for this project and was responsible for the data collection and independent statistical analysis.

Twelve of the most representative national/supranational societies, which include specialists in keratoconus and ectatic diseases from all regions of the world, were invited and accepted to participate in the Consensus project (see list below).

1. Asia Cornea Society (Asia and Oceania);
2. The Cornea Society (USA and International);
3. EuCornea (Europe);
4. PanCornea (Latin America, the United States, and Canada);
5. The Keratoconus Experts Meeting (International);
6. The International Keratoconus Society (International);
7. Middle East Africa Council of Ophthalmology (Middle East and Africa);
8. The College of Ophthalmology of Eastern Central and Southern Africa (Africa);
9. The South African Society of Cataract and Refractive Surgery (Africa);
10. Australian and New Zealand Cornea Society (Oceania);
11. International Society of Refractive Surgery (International); and
12. Refractive Surgery Alliance (International).

Considering the newness related to the scope of this Consensus, the rapidly evolving nature of the topic, and the large variety of themes to cover, the main coordinators formed 7 distinct panels according to the following major topics of interest:

1. **Definition/diagnosis/staging**: Covered clinical features distinguishing keratoconus from other ectatic diseases, consensus diagnostic criteria, risk stratification, staging systems, and pediatric screening recommendations.
2. **Clinical treatment and noninvasive visual rehabilitation**: Focused on conservative management strategies, including the use of contact lenses, management of inflammation and allergy, eye-rubbing avoidance, and treatment of acute corneal hydrops, alongside recognition of systemic risk factors (eg, oxidative imbalance).
3. **CXL for progression**: Addressed indications, protocol selection, and outcomes for CXL in keratoconus, with particular emphasis on pediatric cases, thin corneas, and evolving approaches, and the role of adjunctive pharmacotherapy.
4. **Invasive visual rehabilitation:** Examined therapeutic approaches to improve corrected distance visual acuity (CDVA) in contact lens–intolerant patients, including PRK, ICRS, phakic IOLs, and emerging approaches such as CAIRS and stromal lenticule addition keratoplasty (SLAK), emphasizing individualized treatment planning.
5. **Keratoplasty approaches**: Evaluated indications for DALK versus PK. The remit of the consensus included developing a standardized nomenclature for big-bubble techniques, with recommendations for surgical planning, conversion criteria, and postoperative management.
6. **Elective refractive surgery (to improve uncorrected distance visual acuity [UDVA] and/or corrected distance visual acuity [CDVA])**: Focus on screening, safety, and suitability of refractive surgery procedures in keratoconus and ectasia-susceptible eyes, emphasizing the role of tomography, biomechanics, and epithelial mapping, and outlining appropriate use of PRK, phakic IOLs, and combined procedures.
7. **Cataract surgery in keratoconus**: Guided preoperative stability assessment, timing after CXL, ICRS, or CAIRS, and complex IOL selection strategies in irregular corneas. This included the use of new biometric formulas (eg, Barrett True-K), toric, extended depth of focus (EDOF), and pinhole IOLs, and approaches for managing residual refractive error.

Selection of Expert Panel

Each participating society proposed a list of candidates who fulfilled the following selection criteria:

1. Ophthalmologists with experience in the management of keratoconus and corneal ectasias;
2. Authorship of scientific publications in high-impact, peer-reviewed medical journals;
3. Wide recognition by the specialized medical community;
4. Willing to participate in the question rounds, face-to-face meeting, project design, and comply with the timeline requirements.

The selection process was designed to ensure a global geographic representation and proportional participation from all 12 societies. Each society appointed between 1 and 4 topic coordinators and 2 to 14 panelists, depending on the society's size and the availability of experts (Table 1). Topic coordinators were responsible for reviewing the recommended reference materials, meeting with their co-coordinators, and

formulating specific questions that reflected real-world challenges encountered by ophthalmologists in clinical practice, in accordance with the Delphi panel methodology. The design stipulated that after each round of questionnaires, the topic coordinators would have access to the analysis of panelists' responses, which would inform the development of questions for the subsequent round. Panelists were responsible for answering the consensus questions for their group, using both recommended reference materials and their own clinical and scientific judgment. As with all Delphi-based consensus initiatives, the recommendations presented reflect expert agreement rather than uniform evidence derived from randomized or prospective studies. Where available, published literature informed the statements; where evidence is limited or evolving, expert clinical judgment was used.

In March 2024, the list of experts nominated by each society was completed and sent to the main coordinators and Intrials Clinical Research. Subsequently, an invitation email was sent to the selected experts outlining the study's objectives, key topics to be addressed, methodology, and inviting their involvement as topic coordinators and panelists. Each participant was assigned to 1 of the 7 groups listed above. There was no overlap in topic coordinators or panelists between groups.

Detailed Step-by-Step Process

Each panel consisted of 3 coordinators and 14 or 15 expert panelists, totaling 128 participants (3 general coordinators, 21 theme coordinators, and 104 panelists). A systematic review was conducted using major databases (MEDLINE and Web of Science), with the search strategy for each theme and the respective cutoff date outlined in Supplemental Appendix, Supplemental Digital Content 1, http://links.lww.com/ICO/B995. The identified articles were evaluated by the coordinators, resulting in a recommended bibliography tailored to each group, which was then provided to the panelists as reference material.

The first, second, and third rounds of questions occurred between May and September 2024. A face-to-face meeting was held in October 2024 during the American Academy of Ophthalmology annual meeting in Chicago, USA. The meeting involved open discussions, and where necessary, experts revisited and discussed items from previous rounds. Each session was moderated by 3 coordinators, who remained impartial and refrained from influencing the experts' opinions or responses as they presented and debated the results from their respective groups. This was achieved by displaying the statements on-screen and revising them until no further comments were made. Controversial points, considered relevant by the majority, were recorded, and the coordinators then formulated questions for additional rounds. A fourth and final round, incorporating feedback from the face-to-face meeting, took place in December 2024 to finalize the key topics discussed. A detailed depiction of this step-by-step process is presented in Figure 1.

To minimize attrition, a short turnaround time was maintained between rounds, with personalized emails and regular reminders sent to experts who did not respond. To reduce bias that can arise from seniority or dominant personalities, experts remained anonymous to one another throughout the first 3 rounds. The topic coordinators reviewed the comments and suggestions from panelists between rounds, incorporating relevant issues into subsequent rounds as new questions.

Intrials Scientific Team acted as independent methodologists, overseeing the entire process to ensure each round and group followed the defined procedures. They maintained panelist anonymity within each group and ensured coordinators remained unaware of their group's results until the face-to-face meeting. After the meeting and the fourth round of questionnaires, the topic coordinators drafted a manuscript summarizing the results. This draft was shared with all coordinators for review and feedback, with revisions made accordingly.

Data Collection and Analysis

All questions were formulated based on the literature review and contributions from all topic coordinators. Each questionnaire was hosted on a secure Web site, with access credentials provided exclusively to panelists. During each round, only the independent statistics team had access to the results. Topic coordinators received a compiled summary of results for their respective groups only after all panelists had submitted their responses.

The consensus threshold was defined as when at least two-thirds of the panel selected the same option, consistent with the cutoff point used in the I Global Consensus.[1] All questions were closed-ended; however, experts were allowed to provide additional comments in the free-text fields if desired.

After each round, descriptive statistics were used to summarize numerical, ordinal, and categorical responses, which were subsequently reviewed by the coordinators. Items lacking consensus or deemed unclear or confusing based on panel feedback were reformulated for subsequent rounds. Descriptive statistics included percentages for categorical questions and means or medians for numerical and ranking questions. The statistical analysis was performed using SAS version 9.4 (SAS Institute Inc, Cary, NC).

RESULTS

Group 1: Definition, Diagnosis, and Staging

Keratoconus was defined as a primary, bilateral, typically asymmetrical corneal ectasia, characterized by progressive steepening of the cornea and a cone-apex–centered thinning. The major diagnostic criteria for clinical keratoconus are an abnormal steepening of the anterior and/or posterior corneal surfaces, an abnormal corneal thickness profile, classic slit-lamp biomicroscopic signs, and a decline in CDVA. Although these 4 criteria were considered "major criteria," the last 2 are not mandatory for the diagnosis of keratoconus, especially in the early stages of the disease (Tables 2 and 3).

TABLE 1. Main Topics and the Corresponding Coordinators and Panelists

Definition, Diagnosis, and Staging	Clinical Treatment and Noninvasive Visual Rehabilitation	Corneal Cross-linking for Progression	Invasive Visual Rehabilitation
Coordinators:	**Coordinators:**	**Coordinators:**	**Coordinators:**
Maria Henriquez, MD, PhD (Peru)	John Nkurikiye, MD (Rwanda)	Cosimo Mazzotta, MD, PhD (Italy)	José Guell, MD, PhD (Spain)
Mauro Campos, MD, PhD (Brazil)	Shizuka Koh, MD, PhD (Japan)	Emilio Torres-Netto, MD, PhD (Switzerland)	Luis Izquierdo, MD, PhD (Peru)
Shigeru Kinoshita, MD, PhD (Japan)	Stephanie Watson, MD, PhD (Australia)	Michael Belin, MD (USA)	Salah Mahjoub, MD (Tunisia)
Panelists:	**Panelists:**	**Panelists:**	**Panelists:**
Anthony Aldave, MD (USA)	Adel Barbara, MD (Israel)	Elsie Chan, MD (Australia)	David Lockington, MD, PhD (UK)
Bruce Allan, MD (UK)	Alvin Young, MD (Hong Kong)	Federico Cremona, MD (Argentina)	David Touboul, MD, PhD (France)
Cynthia Roberts, PhD (USA)	Andreia Rosa, MD, PhD (Portugal)	Ingemar Gustafsson, MD, PhD (Denmark)	Francisco Sánchez Leon, MD (Mexico)
Damien Gatinel, MD, PhD (France)	Angela Maria Gutierrez, MD (Colombia)	Lim Li, MD (Singapore)	Heykel Kamoun, MD (Tunisia)
Dan Reinstein, MD (UK)	Bennie Jeng, MD (USA)	Marjan Farid, MD (USA)	John Kanellopoulos, MD (Greece)
Elisante Muna, MD (Tanzania)	César Lipener, MD, MSc (Brazil)	Mashep Alexandria, MD (Kenya)	José Manuel Vargas, MD (Venezuela)
Enrique Graue, MD, MSc (Mexico)	Christian Becker, MD (Peru)	Michael Mrochen, MD, PhD (Switzerland)	Kazutaka Kamiya, MD, PhD (Japan)
Francis Price, MD (USA)	David Smadja, MD (Israel)	Mouhcine Elbakkali, MD (Morocco)	Paulo Ferrara, MD (Brazil)
Fung-Rong Hu, MD (Taiwan)	J.C Bett, MD (Kenya)	Pierre Fournie, MD (France)	Pepe Alfonso, MD (Spain)
Marcony Santiago, MD, PhD (Brazil)	Koji Kitazawa, MD, PhD (Japan)	Rodrigo Donoso, MD (Chile)	Roberto Albertazzi, MD (Argentina)
Mukharram Bikbov, MD, PhD (Russia)	Nicolás Alejandre, MD, PhD (Spain)	Rohit Shetty, MD, PhD (India)	Roger Zaldivar, MD, MSc (Argentina)
Prema Padmanabhan, MD (India)	Rashid Alsaidi, MD (Oman)	Samar Al-Swailem, MD (Saudi Arabia)	Shady Awwad, MD (Lebanon)
Sebastian Siebelmann, MD (Germany)	Roland Hollhumer, MD, PhD (South Africa)	Shihao Chen, MD, MSc (China)	Soosan Jacob, MD, MS (India)
Wuqaas Munir, MD (USA)	Ximena Nunes, MD (Colombia)	Vishal Jhanji MD (USA)	Uri Soiberman, MD (USA)
	Zeba Syed, MD (USA)	William Trattler, MD (USA)	Zhou Xingtao, MD, PhD (China)

Keratoplasty Approaches	Elective Refractive Surgery	Cataract in Keratoconus
Coordinators:	**Coordinators:**	**Coordinators:**
Namrata Sharma, MD (India)	Daniel Scorsetti, MD, PhD (Argentina)	Béatrice Cochener-Lamard, MD, PhD (France)
Mario Nubile, MD, PhD (Italy)	Jod Mehta, MD, PhD (Singapore)	Rajesh Fogla, MD (India)
Vincenzo Sarnicola, MD (Italy)	Riccardo Vinciguerra, MD (Italy)	W Barry Lee, MD (USA)
Panelists:	**Panelists:**	**Panelists:**
Alaa Eldanasoury, MD (Saudi Arabia)	César Carriazo, MD (Colombia)	Helen Wu, MD (USA)
Berthold Seitz, MD (Germany)	David Lin, MD (Canada)	Jesper Hjortdal, MD, PhD (Sweden)
Boris Knyazer, MD (Israel)	Deep Dhaliwal, MD (USA)	Jorge Alió, MD, PhD (Spain)
Christopher Rapuano, MD (USA)	Doyle Stulting, MD, PhD (USA)	Mazen Sinjab, MD, PhD (UAE)
Cor van Zyl, MD (South Africa)	George Kymionis, MD, PhD (Greece)	Menen Ayalew, MD (Ethiopia)
Ernesto Otero, MD (Colombia)	Gustavo Tamayo, MD (Colombia)	Michael Taravella, MD (USA)
Harminder Dua, MD, PhD (UK)	Guy Kezirian, MD (USA)	Mor Dickmann, MD, PhD (Netherlands)
Juan Guillermo Arbelaez, MD (Oman)	Luis Rodríguez, MD (Venezuela)	Mo Ziaei, MD (New Zealand)
Luigi Fontana, MD, PhD (Italy)	Marcus Ang, MD, PhD (Singapore)	Rahul Tonk, MD (USA)
Massimo Busin, MD (Italy)	Miltos Balidis, MD, PhD (Greece)	Samar Basak, MD (India)
Mohamed Hosny, MD, PhD (Egypt)	Mohamed Shafik Shaheen, MD, PhD (Egypt)	Sergio Kwitko, MD, PhD (Brazil)
Nicolás Pereira, MD, PhD (Brazil)	Nicholas Davey, MD (South Africa)	Shigeto Shimmura, MD, PhD (Japan)
Takefumi Yamaguchi, MD, PhD (Japan)	Paolo Vinciguerra, MD (Italy)	Sorcha Ní Dhubhghaill, MD, PhD (Belgium)
Virender Sangwan, MD (India)	Sri Ganesh, MD, MS (India)	Sonia Yoo, MD (USA)
Yonas Tilahun, MD (Ethiopia)	Suphi Taneri, MD (Germany)	Valeria Sánchez, MD (Mexico)

NZ, New Zealand; UAE, United Arab Emirates; UK, United Kingdom; USA, United States of America.

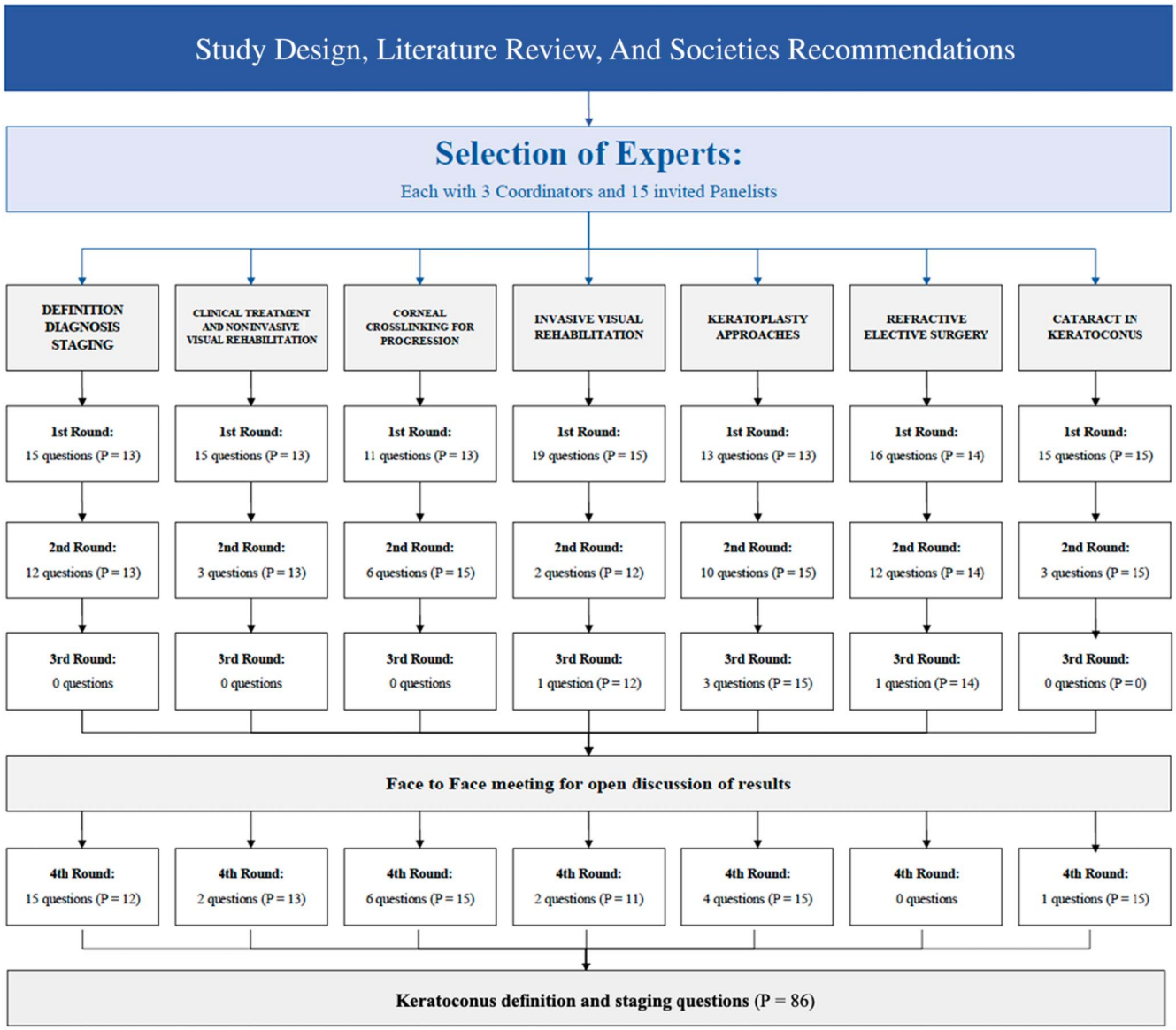

FIGURE 1. Step-by-step process flowchart. P, number of panelists who answered the questionnaire.

Keratoconus is classified as a primary ectasia. Secondary ectasia can result from biomechanical trauma and/or corneal inflammation. Distinguishing between primary and secondary forms can be challenging with current diagnostic tools. Among the primary ectasias, keratoconus is differentiated by its characteristic pattern of central or paracentral thinning, in contrast to the peripheral thinning seen in pellucid marginal degeneration or the diffuse thinning in keratoglobus.

Comprehensive assessment of a patient with keratoconus begins with a detailed clinical history, slit-lamp biomicroscopy, and visual acuity assessment. Diagnostic accuracy can be enhanced by taking a multimodal imaging approach, incorporating technologies such as Scheimpflug tomography and AS-OCT. The integration of AI for analyzing these datasets may further refine diagnostic precision in the future.

Consensus was reached to continue using the terms "subclinical" and "forme fruste" keratoconus. Both entities are considered earlier stages of clinical keratoconus. Usually "subclinical keratoconus" refers to a progressive disease, whereas "forme fruste keratoconus" characterizes a nonprogressive disease. The evaluation of corneal biomechanical properties may aid in detecting ectasia susceptibility during the preclinical stage. Historically, the terms "forme fruste keratoconus," "subclinical keratoconus," and "keratoconus suspect" have been used inconsistently, often reflecting the diagnostic capabilities available at the time. Earlier literature relied predominantly on Placido disk topography, whereas contemporary definitions increasingly incorporate corneal tomography and biomechanical assessment. The present consensus integrates this historical context with current diagnostic paradigms.

TABLE 2. New Proposed Keratoconus Severity Staging, Based on the Belin ABCD Criterion

Keratoconus Severity*	Belin ABCD Criterion
Mild	All criteria are 0, or all criteria are 0 except for 1 criterion that is 1
Moderate	Any combination that does not meet the criteria for being mild or severe
Severe	At least 1 criterion is 4

A, anterior curvature; B, posterior curvature; C, thinnest pachymetry; D, corrected distance visual acuity;

*Staging using the Belin ABCD system assumes a confirmed diagnosis of keratoconus.

Suspicious keratoconus was defined as an eye (or both eyes) showing some clinical features suggestive of keratoconus, but without sufficient anterior or posterior corneal curvature abnormalities to establish a definitive diagnosis. Slight abnormalities may also be present in the corneal thickness profile (including epithelium and/or stromal thickness), corneal aberrations, corneal biomechanics, or corneal biomarkers. Notably, in this case, the fellow eye does not exhibit clinical signs of keratoconus. Patients classified as having "suspicious keratoconus" should be closely monitored for potential progression because of the progression risks inherent with clinical keratoconus. This is particularly relevant in very young patients because of increased risk of rapid disease progression, and in refractive surgery candidates because of their elevated risk of developing postsurgical ectasia. Ectasia susceptibility in suspicious keratoconus is often assessed thoroughly through the use of Scheimpflug tomography, corneal biomechanics assessments, and epithelial thickness mapping.

Keratoconus screening in children: Complementary examinations (beyond clinical history, slit-lamp examination, and visual acuity assessment) should be considered, particularly in children aged between 6 and 18 years who present with astigmatism greater than 2 D in subjective refraction, taking into account local health care resources, access to care, and socioeconomic factors. There was consensus that children with corneal astigmatism exceeding 2 D should undergo Placido disk topography and/or corneal tomography (Scheimpflug imaging or AS-OCT) for further evaluation.

Classification of Keratoconus: The panelists agreed that a practical and universal classification of keratoconus severity is needed. The proposed new severity system is based on the Belin ABCD staging, which uses data from the anterior radius of curvature at the thinnest point (A), the posterior radius of the corneal surface at the thinnest point (B), thinnest pachymetry (C), and visual acuity (D).[27] This classification is increasingly available on Scheimpflug and AS-OCT devices, regardless of the manufacturer. The proposed new categorization of keratoconus into "mild," "moderate," or "severe" disease based on the Belin ABCD display criteria (Table 2).

Keratoconus progression: The main criteria for defining keratoconus progression include significant, consistent, measurable changes in anterior curvature, posterior curvature, corneal thinnest pachymetry (corneal thinning), and deterioration in visual acuity. Although these 4 criteria were considered as "major criteria," the deterioration in visual acuity is not mandatory for the diagnosis of progression. A substantial majority of panelists (92%) agreed that Kmax has low repeatability and reproducibility in evaluating keratoconus progression. Nevertheless, 75% of panelists still use Kmax in their clinical practices for this purpose, and 67% consider Kmax as a relevant parameter for evaluating progression.

When asked whether alternative keratometric indices—such as steepest K, mean K, radius of curvature, or zonal keratometry—offered better performance, only the anterior radius of curvature achieved consensus as a superior and more reliable metric.

Panelists further recommended that all curvature-derived metrics, such as the anterior and posterior radii of curvature and zonal K, be automatically included in the outputs of Scheimpflug imaging and AS-OCT devices to enhance future progression analyses. They emphasized that curvature variability increases with the severity of keratoconus, a factor that must be considered when evaluating progression.

The role of the corneal epithelium was also discussed. Panelists agreed that although epithelial maps support the diagnosis of keratoconus, specific quantitative epithelial cell data remain lacking to reliably diagnose, classify, or monitor disease progression.

Group 2: Clinical Treatment and Noninvasive Visual Rehabilitation

Relatively few publications addressing clinical treatment and noninvasive visual rehabilitation in keratoconus were identified.

The first round achieved excellent consensus among the experts. Specifically, there was a full consensus that keratoconus may be associated with low serum levels of vitamin D, zinc, copper, and selenium, and an imbalance in the oxidant–antioxidant status. Further, inflammation was thought to have a role in the pathogenesis of keratoconus (100% consensus). There was also 100% consensus that oral riboflavin supplementation is not a substitute for conventional CXL performed with topical riboflavin and UV-A. In addition, panelists reached 100% agreement on the role of various contact lens modalities in managing irregular astigmatism in keratoconus, including corneal rigid gas-permeable (RGP) lenses, hybrid lenses, piggyback systems, and scleral lenses.[28–30]

Concerning keratoconus progression, panelists unanimously agreed that wearing RGP contact lenses does not halt keratoconus progression. In addition, 69% of the panelists also agreed that RGP lenses do not accelerate the progression of keratoconus. Consensus also emerged around nonsurgical measures to reduce the risk of progression, including the use of topical antiallergic medications, lubricants to alleviate itch and improve comfort, and patient education to prevent eye rubbing. In Round 2, the recommendation to avoid eye rubbing achieved complete consensus (100%). However, the use of goggles to avoid eye rubbing did not reach a consensus, with only 46% of panelists supporting their use.

TABLE 3. Group 1 Summary of Recommendations

Definition, Diagnosis, and Staging

Main findings:

- **Keratoconus definition:**
  - Keratoconus is a primary, bilateral, typically asymmetrical corneal ectatic disease characterized by progressive steepening of the cornea and a cone-apex–centered thinning.
  - The major diagnostic criteria for clinical keratoconus are as follows:
    - Abnormal steepening of the anterior and/or posterior corneal surface;
    - Abnormal corneal thickness profile;
    - Classic slit-lamp biomicroscopic signs; and
    - Decline in CDVA.
    - Although these 4 criteria were considered "major criteria," the last 2 are not mandatory for the diagnosis of keratoconus, especially in the early stages of the disease.
  - Secondary ectasia can occur because of secondary biomechanical trauma, corneal inflammatory diseases.
  - The pattern of thinning distinguishes keratoconus from pellucid marginal degeneration and keratoglobus.
  - Current diagnostics make it challenging to distinguish primary from secondary ectasia using only current diagnostic modalities.
- **Keratoconus staging criteria**:
  - Patients who meet the definition of keratoconus can be stratified into mild, moderate, and severe disease categories as shown in the new proposed keratoconus severity staging (Table 2), which is based on the Belin ABCD classification.
- **Keratoconus terminology:**
  - The terms "subclinical keratoconus" and "forme fruste keratoconus" (FFKC) should continue to be used.
    - Both subclinical keratoconus and FFKC are early stages of clinical keratoconus.
    - Subclinical keratoconus is characterized by altered biomechanics and is usually considered a progressive disease.
    - FFKC may display altered biomechanics and is usually considered a nonprogressive disease.
- **Presumed/suspicious keratoconus is defined as the following:**
  - An eye (or both eyes) with some clinically distinctive keratoconus signs in the anterior or posterior corneal curvature that are insufficient to confirm keratoconus.
    - Abnormalities in the posterior cornea, corneal thickness profile (and/or epithelial/stromal thickness), corneal aberrations, corneal biomechanics, and corneal biomarkers can also be present.
  - There is no evidence of keratoconus in the fellow eye.
  - Patients classified under "suspicious keratoconus" should be monitored closely for potential progression of clinical keratoconus. Especially close monitoring should be considered for younger patients who are at higher risk for rapid progression of keratoconus and for those who are candidates for refractive surgery, because of the risk of developing postsurgical ectasia.
- **Keratoconus assessment:**
  - A comprehensive assessment of keratoconus patients must begin with the following:
    - Comprehensive clinical history/anamnesis.
    - Slit-lamp biomicroscopic examination.
    - Visual acuity measurement.
  - A multimodal diagnosis that uses different technologies (e.g. Placido disk topography, Scheimpflug imaging, and/or AS-OCT) increases diagnostic accuracy and improves staging.
  - AI may improve the accuracy of keratoconus diagnosis in the future.
  - Evaluation of corneal biomechanical weakness can help detect ectasia susceptibility in a preclinical stage of keratoconus (e.g., forme fruste, subclinical, or suspicious keratoconus).
  - Advanced diagnostic tools such as corneal tomography, corneal biomechanics assessment, and epithelial thickness mapping are essential for assessing ectasia susceptibility.
  - A morphological classification of the astigmatism pattern in keratoconus remains valuable, primarily for determining the most appropriate treatment plan.
  - Multimodal diagnostics extend beyond corneal imaging and refractive characterization to incorporate molecular biology and genetic information, with the goal of enhancing diagnostic accuracy.
- **Keratoconus in children:**
  - Complementary examinations should be considered, particularly in children aged between 6 and 18 years with astigmatism greater than 2 D in subjective refraction, taking into account local health care resources, access to care, and socioeconomic factors:
    - Placido disk topography
    - Scheimpflug imaging
    - AS-OCT
- **Keratoconus progression:**
  - Although Kmax continues to be used in clinical practice for evaluating keratoconus progression, it has low repeatability and reproducibility.
  - The anterior radius of curvature demonstrates better reproducibility and repeatability than Kmax.

TABLE 3. (*Continued*) Group 1 Summary of Recommendations

Definition, Diagnosis, and Staging
o The variability of curvature measurements increases as the severity of keratoconus worsens. Therefore, cutoff points for evaluating progression should be adjusted based on the severity of the keratoconus.
o Important values related to corneal curvature, such as zonal Kmax, and anterior and posterior radii of curvature at the thinnest point of the cornea should be displayed on every Scheimpflug or OCT imaging device for more accurate future assessments of keratoconus progression.
o Despite its contribution to keratoconus diagnosis, specific quantitative epithelial data remain insufficient for diagnosis, classification, and assessment of progression.

ABCD, anterior curvature (A), posterior curvature (B), corneal thickness (C), corrected distance visual acuity (D); AI, artificial intelligence; AS-OCT, anterior segment optical coherence tomography; FFKC, forme fruste keratoconus; Kmax, maximum keratometry.

Consensus was also sought on risk factors for keratoconus, with eye rubbing (100%), family history (100%), allergic eye diseases (92%), and atopy (84%) reaching consensus. However, asthma reached only 69% consensus in Round 1 and declined to 38% in Round 2, failing to reach consensus. Dry eye was also considered a potential risk factor, but did not reach consensus, with only 53% agreement.

Regarding the management of acute corneal hydrops, several nonsurgical interventions were evaluated, with experts agreeing on their potential use. Some treatments reached consensus in Round 2, including intraocular pressure (IOP) reduction and topical corticosteroid use, both at 76%. Other approaches, such as cycloplegic agents, pressure patching, and intracameral gas injection, were acknowledged as used in practice. However, their use did not achieve consensus. Panelists also noted that treatment practices for hydrops vary across countries and regions. The use of oral corticosteroids was explicitly excluded in the final round (Table 4).

Group 3: Corneal Cross-linking for Progression

There was a unanimous consensus (100%) among panelists that CXL should be performed in cases of confirmed tomographic and/or topographic progression. In addition, a substantial majority (≥86%) supported considering CXL without waiting for progression in pediatric patients (<18 years old) with confirmed keratoconus (ie CXL treatment on diagnosis), because of the increased risk of rapid disease progression in this population.

Panelists also agreed that advising patients to stop eye rubbing is an essential behavioral intervention; however, 73% indicated that this recommendation should neither delay nor replace timely CXL treatment in a progressive disease. In the absence of documented anterior topographic progression, specific corneal changes were considered sufficient to consider or justify CXL, including progressive thinning at the thinnest point, abnormal pachymetric progression profiles, and increased posterior corneal elevation. There was consensus that central corneal pachymetry should not be used as a standalone parameter for assessing progression. Although there was no consensus on epithelial thinning, 60% of panelists considered it relevant to clinical decision making.

Beyond imaging parameters, additional non–imaging-based factors were evaluated as potential indications for CXL. Patient compliance achieved an 80% agreement and was considered a crucial factor in the decision to treat. Conversely, rapid progression in the contralateral eye (60%) and corneal biomechanics (53%) did not achieve consensus, indicating ongoing clinical debate. Other factors, such as severe allergic eye disease/vernal keratoconjunctivitis (40%) and other ocular surface diseases (6%), were not considered relevant for prompt CXL indication. However, because of the greater probability of progression, there was a widespread opinion among the panelists to keep these cases under close observation for treatment, which, however, must not be done a priori.

There was strong consensus (93%) that CXL protocols should be individualized based on patient-specific factors such as age, progression rate, and corneal thickness, underscoring the importance of a personalized approach. Epi-off CXL remained the most commonly used technique. Among panelists using epi-off protocols, the most frequently applied method was accelerated CXL with continuous UV-A irradiation at 9 mW/cm^2 for 10 minutes (40%), followed by the original Dresden protocol (26%), which involves 30 minutes of 3 mW/cm^2 UV-A irradiation; both protocols deliver a UV fluence of 5.4 J/cm^2.

Although its use is increasing, epi-on CXL was used by only 36% of panelists and 80% consensus was reached, reserving epi-on for milder cases or age groups less prone to progression. There was a 93% consensus that treatment with epi-On CXL in young adults was not considered useful before determining whether the disease is progressive or not. In instances of continued progression after initial CXL, 90% of panelists supported CXL retreatment. In cases where the first procedure was epi-on CXL and the disease continued to progress despite initial treatment, panelists preferred (86%) switching to an epi-off protocol for retreatment rather than repeating an epi-on CXL procedure.

There was over 90% consensus that CXL procedure can be safely repeated, and although there is no unanimity on the timing, 40% of panelists recommended retreatment at least 6 months after the initial procedure, and 33.3% preferred to wait at least 12 months, so in case of retreatment, 1 should reasonably wait at least 6 to 12 months. Smaller proportions supported earlier retreatment (<6 months) upon confirmed progression (13%), whereas others favored longer intervals of 24 months (6%) or 48 months (6%), highlighting variability in clinical practice.

The use of high-fluence (>5.4 J/cm^2) CXL remained controversial. Only a minority (36%) of non–US-based

TABLE 4. Group 2 Summary of Recommendations

Clinical Treatment and Noninvasive Visual Rehabilitation in Keratoconus
Main findings:
• In keratoconus, the following serological findings outside of those in the cornea may be present:
o Low serum levels of vitamin D, zinc, copper, and selenium.
o Imbalance of oxidant/antioxidant status.
• Inflammation may contribute to keratoconus pathogenesis.
• Oral riboflavin with direct UV-A exposure is not a substitute for conventional cross-linking (topical riboflavin and UV-A).
• Regarding noninvasive visual rehabilitation for keratoconus, the following management options are considered adequate:
o Spectacles
o Soft toric contact lenses to improve the quality of vision in mild disease.
o RGP contact lenses to provide good visual acuity in more advanced diseases.
o Piggyback lenses (combining high-DK silicone hydrogel and RGP contact lenses), for patients who cannot tolerate corneal RGP contact lenses.
o Modern hybrid contact lenses combine the optical quality of corneal RGP lenses with the comfort of soft lenses, but they may be associated with an increased risk of corneal hypoxia.
o Customized high-Dk scleral lenses to provide high-quality vision and comfort tailored to individual patient needs.
• The following approaches can be used to correct irregular astigmatism in keratoconus:
o RGP contact lenses.
o Piggyback contact lens systems.
o Hybrid contact lenses.
o Scleral lenses.
• RGP contact lens wear does not halt keratoconus progression, but its use does not accelerate the progression of the condition.
• The following measures are used in the clinical treatment of ectasias with the aim of halting keratoconus progression:
o Providing verbal guidance to patients regarding the avoidance of eye rubbing.
o Use of topical antiallergic medication for allergic eye disease.
o Use of lubricants to reduce itch and increase ocular comfort.
• The following risk factors observed in daily clinical practice are recognized as concerning for keratoconus:
o Eye rubbing.
o Family history of keratoconus.
o Allergic eye disease.
o Atopy.
• The following approaches are adequate to treat acute corneal hydrops as a nonsurgical management:
o Topical corticosteroids (oral corticosteroids are **not recommended**).
o Reduction in intraocular pressure (IOP).
o Hypertonic saline.

Dk, oxygen permeability; IOP, intraocular pressure; RGP, rigid gas-permeable; UV-A, ultraviolet A.

panelists supported its routine use, and 0.1% riboflavin remained the most widely used concentration for epi-off CXL.

For corneas with a stromal thickness of less than 400 μm, 86% of panelists agreed that CXL should still be performed and that CXL is an appropriate therapeutic option; however, no single approach reached consensus as the preferred technique. Among the methods used, 60% of panelists applied hypoosmolar riboflavin to swell the stroma above 400 μm before UV irradiation, and 53% used the ELZA-sub400 protocol.[28,29]

For pediatric keratoconus with documented progression, epi-off CXL was the preferred approach. However, no consensus emerged for pediatric keratoconus without documented progression: 46% preferred monitoring, 33% favored epi-off CXL, and 20% supported epi-on CXL.

There was a unanimous consensus (100%) that oral riboflavin combined with sunlight exposure cannot replace CXL for stabilizing progressive keratoconus. Regarding adjuvant pharmacological therapies, 73.3% of panelists found current evidence insufficient to support their use. Specifically, oral riboflavin (87%), vitamin D (73%), lysyl oxidase/copper supplementation (87%), and biomarker modulation (87%) were all rejected as viable standalone treatments. However, 93% agreed that if such therapies are used, they should be considered only as adjuncts to CXL, but not as standalone therapies (Table 5).

Group 4: Invasive Visual Rehabilitation

Several invasive approaches are currently available for visual rehabilitation in keratoconus, including PRK, phakic IOLs, ICRS, and CAIRS. A total of 84% of panelists agreed that these procedures should be considered when patients are intolerant to contact lenses. PRK performed with an excimer laser either before or concurrently with CXL was considered by 92% of panelists to be an acceptable technique to provide partial refractive correction. Similarly, 92% of panelists agreed that phototherapeutic keratectomy—performed with an excimer laser after CXL—can reduce higher-order

TABLE 5. Group 3 Summary of Recommendations

Corneal Cross-linking for Progression
Main findings:
• CXL should be offered for patients with definitive topographic and/or tomographic keratoconus progression.
• CXL should be considered upon diagnosis without waiting for progression in pediatric patients with confirmed topographic or tomographic keratoconus.
• Advising patients to stop eye rubbing should not delay or prevent the recommendation of performing a CXL in progressive keratoconus.
• When documented progression in anterior corneal topography/tomography is absent, the following subcriteria should be considered to determine the need for CXL:
o Thinnest corneal pachymetry.
o Pachymetric progression.
o Corneal posterior elevation/surface changes.
o Patient compliance.
• When all CXL protocols are available in your country or region, the choice of protocol should be based on individual patient characteristics, such as age, rate of progression, and corneal thickness.
• Among non-US experts, high-fluence CXL (>5.4 J/cm^2) is not commonly used, either for epi-on or epi-off protocols.
• For epi-off CXL, the most commonly used riboflavin concentration is 0.1%.
• Repeat CXL is considered appropriate for patients experiencing ectasia progression after the initial treatment.
• For corneas with <400 μm of stromal thickness:
o CXL is a valid therapeutic option.
o The following protocols are usually applied: hypoosmolar riboflavin and ELZA-Sub400.
• CXL upon diagnosis, without waiting for progression, is warranted in pediatric patients with topographically/tomographically confirmed keratoconus.
o Epi-off protocols are preferred when progression is present.
o When progression is not present, clinical observation, epi-on, or epi-off procedures can be considered.
• Regarding epi-on CXL:
o Epi-on CXL is not typically used as a first-in-line protocol for most patients.
o It can be considered for less progressive cases and/or milder disease presentations.
• If progression continues despite undergoing epi-on CXL, transitioning to an epi-off CXL protocol is preferred over repeating the epi-on CXL procedure.
• Oral riboflavin and sunlight exposure cannot replace CXL as a treatment for stabilizing progressive keratoconus.
• In cases where any adjuvant pharmacological therapy is indicated, it should be used as a complementary treatment in addition to CXL, never as a sole therapy.

CXL, corneal cross-linking; epi-on, epithelium-on; epi-off, epithelium-off.

aberrations and improve visual quality, often in combination with RGP or scleral contact lenses. However, because both procedures involve stromal ablation, careful case selection is critical, and many corneas may remain unsuitable even after CXL stabilization. In addition, 75% of panelists agreed that mitomycin C (MMC) is contraindicated when PRK is combined with CXL.

ICRS are polymethyl methacrylate (PMMA) arcs, typically 150 to 350 μm in depth, 5 to 6 mm in diameter, and between 90 and 350 degrees in arc length, that are placed in precut stromal tunnels. Unanimous consensus (100%) was reached that ICRS can help regularize the corneal surface, reduce spherical aberrations, and provide limited refractive correction. Panelists also agreed that implant thickness correlates with the degree of corneal flattening, and that asymmetric ICRS allows customization for asymmetric keratoconus patterns, and 76% agreed that ICRS can be used as a refractive procedure. Furthermore, 76% of panelists agreed that ICRS does not halt disease progression, and complications such as ring migration or extrusion remain concerns. Regarding contraindications, 92% of panelists agreed that ICRS should not be used in the presence of acute hydrops, central corneal opacity or opaque Descemet folds, central corneal thickness below 400 μm, or keratometry readings above 64 diopters (D).

To address these limitations, the CAIRS technique was developed, in which the ring segments are made from human donor stromal tissue, rather than PMMA. Recent developments in the technique have included the use of femtosecond lasers to create both stromal tunnels and custom-shaped allogenic ring segments, thereby enhancing precision and enabling personalized treatments. Techniques such as air-drying (the "jerky" technique), crosslinked CAIRS, and high-fluence CXL of the donor segments (Extracorporeal Optimization of Corneal Allogenic Intrastromal Ring Segments or ECO-CAIRS) have been introduced to stiffen the implants and facilitate insertion.[31]

Regarding CAIRS, 92% of panelists agreed that the technique and current nomogram remain investigational, and 76% agreed that CAIRS offers improved biocompatibility compared with synthetic ICRS. Although 69% agreed that the use of femtosecond lasers may enhance reproducibility and predictability, no consensus was reached on whether CAIRS currently offers better predictability than ICRS.

Regarding phakic IOLs, 92% of panelists agreed that acceptable CDVA or stable keratoconus are minimum prerequisites for implantation. In addition, 69% of panelists considered an anterior chamber depth greater than 3.0 mm and an endothelial cell count above 2500 cells/mm^2 as minimum requirements. Unanimous consensus was reached

(100%) that phakic IOLs offer superior correction for high ametropia compared with ICRS and topography-guided PRK, and 79% agreed that phakic IOLs also provide better refractive predictability. This consensus reflects agreement that phakic intraocular lenses represent an effective surgical option in appropriately selected patients and does not imply exclusion of nonsurgical modalities, such as contact lenses, when these remain effective and tolerated. In the absence of contraindications, phakic IOLs have demonstrated high efficacy in correcting substantial refractive errors, offering benefits such as predictability, precision, and reversibility, provided that accurate ocular biometric data and IOL power selection are ensured.

SLAK, a technique that involves implanting a negative meniscus-shaped donor lenticule into the patient's stroma, was also discussed.[32] This approach enables customization tailored to the individual's corneal architecture. A total of 92% of panelists agreed that SLAK can serve both therapeutic and refractive purposes, and that lenticule customization may improve outcomes. However, no consensus was reached regarding the current use of myopic small incision lenticule extraction (SMILE) lenticules in this context (Table 6).

Group 5: Keratoplasty Approaches

The panel agreed that the most essential factors to indicate keratoplasty in keratoconus are insufficient CDVA and poor tolerance or intolerance to contact lenses. The following diagnostic assessments are appropriate for determining both indication and type of keratoplasty in these patients: CDVA (93% consensus), AS-OCT (87% consensus), contact lens CDVA (87%), biomicroscopy, fundus examination, corneal tomography, and endothelial/confocal microscopy (each at 67% consensus).

Among tissue-replacement keratoplasty techniques, 93% of panelists agreed that DALK was the preferred surgical option for keratoconus, unless specific conditions warranting PK are present. Similarly, 93% of panelists highlighted the presence of a healthy endothelium as the most essential corneal morphological requirement—and a prerequisite—for selecting DALK in cases of corneal ectasia, including keratoconus. Conversely, 78% of panelists considered PK appropriate in cases of endothelial dysfunction, and 71% in the presence of deep stromal scarring involving Descemet membrane. In total, 84% of panelists agreed that DALK is the preferred planned surgical approach, with PK reserved for specific indications such as endothelial dysfunction or deep stromal scarring.

TABLE 6. Group 4 Summary of Recommendations

Invasive Visual Rehabilitation
Main findings:
• Invasive visual rehabilitation techniques might be considered in patients intolerant to contact lenses.
• Regarding PRK in keratoconic eyes:
o If PRK is considered in some keratoconic eyes, it should be associated pre- or perioperatively with CXL
o When combined with CXL, the use of MMC is not accepted by all surgeons
o Transepithelial techniques are more adequate than the standard approach for reducing aberrations
• About ICRS in keratoconic eyes:
o ICRS can be used both as a therapeutic and refractive procedure.
o When CXL and ICRS are both indicated, there is not enough evidence to support the best order of treatment.
o ICRS cannot halt keratoconus progression.
o ICRS improve vision by centering the cone and diminishing coma-like aberrations.
o The flattening effect of the ICRS is thought to be correlated with implant thickness and optical zone of implantation.
o Contraindications include hydrops; opaque apex or Descemet folds; keratometry >64 D; CCT <400 μm.
• Phakic IOLs in keratoconic eyes. Minimum requirements include the following:
o Acceptable spectacle best-corrected vision.
o Stable keratoconus.
o Anterior chamber depth >3 mm from endothelium.
o Minimum endothelial cell count >2500 cells/mm^2.
• Phakic IOLs offer superior correction of higher ametropia and better refractive predictability compared with ICRS and TE-PRK.
• Regarding CAIRS in keratoconic eyes:
o The surgical technique and nomogram are still under investigation.
o CAIRS should have better biocompatibility than ICRS.
o The use of femtosecond laser technology is expected to improve reproducibility and predictability.
• Regarding SLAK in keratoconic eyes:
o SLAK can serve as both a therapeutic and refractive procedure.
o Lenticule customization is anticipated to enhance outcomes.

AC, anterior chamber; CAIRS, corneal allogenic intrastromal ring segments; CCT, central corneal thickness; CXL, corneal cross-linking; D, diopters; ICRS, intrastromal corneal ring segments; IOL, intraocular lens; MMC, mitomycin C; PRK, photorefractive keratectomy; SLAK, stromal lenticule addition keratoplasty; TE-PRK, transepithelial photorefractive keratectomy.

DALK was favored over PK because of several advantages. Complete consensus (100%) was achieved on its lower risk of graft rejection. Additional benefits included fewer intraocular complications (93%), reduced long-term graft failure (86%), lower incidence of postsurgical glaucoma (80%), and a decreased risk of postoperative cataract formation (73%). In the long term, DALK offers improved graft survival and better preservation of endothelial cell density.

To standardize the nomenclature in DALK techniques, panelists agreed on the following terminology for describing intraoperative outcomes when performing big-bubble DALK (with air or viscoelastic injection):

1. 85% agreed on "DALK with type 1 big-bubble: exposure of the recipient's pre-Descemet/Dua layer";
2. 78% agreed on "DALK with type 2 big-bubble: exposure of the recipient's DM";
3. 71% agreed on "DALK with mixed bubbles or type 3 bubble: formation of both type 1 and type 2 bubbles, either partially or completely."

In cases where big-bubble formation is not achieved or not attempted, 71% of panelists recommended using the term pre-Descemet DALK (pdDALK) to describe manual dissection techniques that preserve the pre-Descemet layer and posterior stroma.

Furthermore, 73% agreed that the ideal surgical end point during DALK is exposure of the recipient's pre-Descemet layer (type 1 equivalent). The preferred approach in routine cases was air big-bubble DALK, although manual layer-by-layer dissection and femtosecond laser–assisted DALK were also deemed appropriate in selected cases. The panel acknowledged femtosecond laser as a significant advance for anterior lamellar keratoplasty (73%) and stromal additive techniques (80%), such as SLAK and CAIRS.

If big-bubble formation is unsuccessful, 80% of panelists recommended proceeding with manual dissection techniques aimed at achieving a pre-Descemet dissection plane. This may include layer-by-layer dissection or manual peeling. When manual DALK is performed initially or after failed big-bubble attempts, 71% agreed on the term "manual DALK with retention of residual posterior stroma." Long-term satisfactory visual acuity can still be achieved without full baring of Descemet membrane or Dua layer, provided the residual stroma is limited and regular. Panelists considered <150 μm acceptable, whereas literature suggests that thinner residual stromal beds (ideally <80 μm) are associated with better visual outcomes.[33,34] Overall, residual bed regularity remains the key factor, with thinner stromal beds potentially offering additional visual benefit.

Conversion to PK during DALK was most commonly indicated by intraoperative perforation of Dua layer or Descemet membrane (78% agreement). However, 67% of panelists agreed that 1 to 2 microperforations do not necessarily require conversion. In type 1 big-bubble DALK, most panelists supported continuing the procedure in the presence of up to 2 small perforations of the pre-Descemet layer. In type 2 big-bubble cases, continuing DALK was acceptable if a single, minor Descemet membrane rupture occurred. Overall, the panel agreed that small perforations do not uniformly necessitate conversion.

Regarding postoperative management, the most commonly used postoperative steroid therapies included prednisolone acetate 1%, dexamethasone 0.1%, or equivalent agents, and 80% of panelists agreed that topical steroids are generally maintained indefinitely at minimal doses after PK to prevent immune-mediated graft rejection. In contrast, only 6.7% of panelists recommend lifelong use of topical steroids after DALK (Table 7). The differentiation of postoperative use of topical steroids between phakic and pseudophakic patients was not addressed.

Group 6: Elective Refractive Surgery

There was strong consensus (92%) that corneal tomography should be considered a mandatory tool during preoperative screening for refractive surgery. It was recognized as essential for evaluating corneal structure and detecting early abnormalities that may compromise surgical outcomes. Adjunctive diagnostic tools were also valued: epithelial thickness mapping was endorsed by 92% of respondents as a valuable complement to tomography. In comparison, corneal biomechanical assessment was supported by 71%, highlighting its emerging role in risk assessment.

Genetic testing did not reach consensus for inclusion in preoperative evaluation, with only 7% of participants considering it essential. Consequently, the panel did not recommend routine genetic screening in candidates for refractive surgery.

In contrast, there was strong agreement that a positive family history of keratoconus should be considered a relevant risk factor, as it may indicate an increased risk of postoperative corneal instability. A total of 85% of respondents reported incorporating family history into their clinical decision-making process. Although family history alone was not deemed sufficient to exclude a patient from surgery, it was recommended as an additional factor to be weighed alongside tomography findings and other screening tools.

Unanimous consensus (100%) was reached on the use of phakic IOLs to correct residual ametropia in patients with keratoconus who have undergone both ICRS implantation and/or CXL, provided they demonstrate refractive and tomographic stability. Although no specific timeframe was defined, the panel emphasized that stability should be confirmed before implantation, at the discretion of the treating physician. This consensus reflects agreement that phakic intraocular lenses represent an effective surgical option in appropriately selected patients and does not imply exclusion of nonsurgical modalities, such as contact lenses, when these remain effective and tolerated. Furthermore, 78% of panelists supported the use of toric phakic IOLs in patients with more than 2 D of astigmatism. However, it was noted that visual outcomes may be limited by underlying irregular astigmatism and persistent higher-order aberrations, particularly coma, even after correction of lower-order refractive errors.

A total of 71% of the panel reached a consensus that CXL, when performed independently of any refractive procedure (laser or nonlaser), does not consistently improve

TABLE 7. Group 5 Summary of Recommendations

Keratoplasty Approaches
Main findings:
• The most important factors to consider when indicating keratoplasty surgery for keratoconus patients are as follows:
o Insufficient CDVA.
o Poor tolerance or intolerance to contact lenses: presence of moderate to severe discomfort while using contact lenses, not relieved by lubrication or pauses.
• The main goal of performing keratoplasty for corneal ectasia is to restore or improve CDVA.
• The following examinations are considered appropriate to define the indication and type of keratoplasty surgery: CDVA, AS-OCT, contact lens-CDVA, biomicroscopy of the anterior segment and fundus examination, corneal tomography, and endothelial/confocal microscopy.
• DALK should be the preferred choice of keratoplasty technique in keratoconus (in the presence of a healthy endothelium), unless specific conditions indicating the need for PK are present (significant endothelial alterations and/or the presence of deep stromal scarring involving the DM).
• Femtosecond laser technology represents a significant advance in the surgical management of anterior lamellar keratoplasty and intrastromal additive surgery (SLAK and/or CAIRS).
• When performing DALK, the preferred technique in regular cases is air big-bubble DALK, with the aim of exposing the pre-Descemet layer.
• Regarding standardization of nomenclature in big-bubble DALK, the following terms are used to describe the surgical result achieved:
o DALK with type 1 big-bubble = exposure of recipient pre-Descemet layer.
o DALK with type 2 big-bubble = exposure of recipient Descemet membrane.
o pdDALK, when pre-Descemet layer and some residual posterior stroma is retained.
• The main advantages of DALK over PK are lower rate of graft rejection, reduced intraocular complications, lower rate of long-term graft failure, better preservation of the endothelial cell density, lower risk of postsurgical glaucoma, and a lower incidence of postoperative cataract.
• When a big-bubble cannot be achieved, the next best step is to continue with other dissection techniques (manual DALK with retention of residual posterior stroma) to achieve a pre-Descemet DALK, with the aim of exposing a regular surface of deep (less than 150 μm residual stroma.
• When performing DALK, the primary reason for conversion to PK is intraoperative rupture. DALK can be generally completed in the event of no more than 1 small (type 2 big-bubble) or 2 (type 1 big-bubble) perforations.
• Postoperative therapy with topical steroids should be used in the long term (with minimal dosage and lifelong in selected cases) after PK to prevent rejection episodes. In contrast, routine lifelong use of topical steroids is not recommended after DALK.

CAIRS, corneal allogenic intrastromal ring segments; CDVA, corrected distance visual acuity; CL, contact lenses; DALK, deep anterior lamellar keratoplasty; DM, Descemet membrane; OCT, optical coherence tomography; PD-DALK, pre-Descemet deep anterior lamellar keratoplasty; PK, penetrating keratoplasty; SLAK, stromal lenticule addition keratoplasty.

UDVA. The emphasis was placed on the word *consistently*. Although some patients may experience notable corneal flattening, with improvement in UDVA, the effect is highly variable and not reliably predictable with standard techniques. Customized CXL approaches that aim not only to halt progression but also to improve corneal irregularity may offer such potential; however, as these protocols are not yet widely available in most countries, they were not considered separately in the current analysis.

Conversely, combining CXL with excimer laser ablation, such as in topography-guided or wavefront-guided PRK, was considered a strategy capable of improving UDVA, with 85% of panelists supporting this approach.[19] In patients at elevated risk of ectasia, 71% favored PRK over LASIK, citing PRK's more conservative biomechanical impact on the cornea. Importantly, LASIK is generally contraindicated in keratoconus and ectasia-susceptible eyes because of the very high risk of postoperative ectasia progression. Nevertheless, the minority of responses favoring LASIK reflect variability in global practice rather than a recommendation of a standard of care.

When evaluating the safety of refractive laser procedures in cases of suspected or early keratoconus, 71% of panelists emphasized the need for a multimodal screening approach that integrates refractive error, tomography, corneal biomechanics, and epithelial thickness mapping.

In addition, there was strong consensus (93%) that educating candidates about the risks associated with eye rubbing is essential before refractive surgery, particularly in patients with a potential risk of corneal instability (Table 8).

Group 7: Cataract in Keratoconus

Cataract management in keratoconus has emerged as a growing concern because of advances in corneal stabilization and visual rehabilitation. In the past, cataract surgery was primarily considered in eyes that had previously undergone PK. However, it is now increasingly encountered in untreated corneas or in eyes that have undergone corneal remodeling with CXL, ICRS, or lamellar grafts. In such cases, the challenge lies less in the cataract surgery itself, but in accurately calculating the IOL power, which is complicated by irregular astigmatism and extreme keratometric values often seen in keratoconus.

From an epidemiological standpoint, 80% of panelists agreed that the incidence of cataract in keratoconus may be influenced by allergic phenotypes, particularly in individuals with a history of corticosteroid use. Among grafted eyes, cataract development was considered more likely to occur earlier after PK than after DALK, as the latter preserves the endothelium and reduces the risk of steroid-induced opacification (87% agreement). Cataract surgery should be delayed until several weeks after suture removal, once corneal stability has been established (73% consensus).

The primary concern in planning cataract surgery in keratoconus is ensuring corneal stability. The panel

TABLE 8. Group 6 Summary of Recommendations

Refractive Elective Surgery
Main findings:
• We recommend that corneal tomography be a mandatory tool for refractive surgery screening. In such cases, corneal biomechanical assessments and epithelial maps are a valuable add-on in addition to tomography.
• Genetic testing is not essential for preoperative screening.
• Family history of keratoconus should be considered in the decision-making process for refractive surgery.
• In patients with keratoconus already treated with ICRS and/or CXL, without progression, phakic IOLs can be indicated to treat residual ametropia. A toric phakic IOLs in such cases might be indicated for astigmatism greater than 2 D.
• CXL (not linked with excimer laser) in eyes with keratoconus cannot consistently provide an improvement in UDVA.
• CXL combined with excimer laser in eyes with keratoconus can provide an improvement in UDVA.
• It is acceptable to perform surface ablation on a patient who would be excluded from LASIK because of a slightly increased risk of ectasia.
• The decision-making process for determining the safety limits of treating a suspicious patient or early keratoconus with laser refractive surgery includes refractive error, tomography, and biomechanics/epithelial mapping.
• We consider information on the prevention of eye rubbing before refractive surgery to be important.

CXL, corneal cross-linking; ICRS, intrastromal corneal ring segments; ICSR, intrastromal corneal ring segments; LASIK, laser-assisted in situ keratomileusis.

unanimously agreed (100%) that cataract surgery should be postponed for at least 3 months after corneal reshaping procedures, such as CXL or ICRS, when these are indicated. These interventions are often required beforehand in cases of documented keratometric progression, defined by 80% of panelists as an increase of ≥1.0 D over 12 months or ≥0.5 D over 6 months. Timing of surgery should also take into account lens opacity (87% agreement), visual acuity (73%), patient-reported symptoms (73%), and overall disease stability (67%).

The timing of cataract surgery in corneal ectasia should be guided by a combination of factors, including lens opacity (87% consensus), visual acuity (73%), patient-reported visual limitations (73%), and disease stability (67%), recognizing that refractive outcomes remain less predictable in these eyes.

In advanced cases, for example in severe keratoconus with loss of corneal transparency and coexisting cataract, combined cataract and keratoplasty ("triple") procedures may be considered (73% consensus). However, the panel advised that such combined surgeries carry reduced refractive accuracy (87% consensus). A total of 80% of panelists recommend that when DALK is the graft technique of choice, a sequential approach (graft first, followed by cataract surgery) is preferable.

As previously emphasized, IOL power calculation remains a significant challenge in keratoconus.[35] This is related to the steep keratometry values that frequently exceed 50 D (93% consensus) and substantial discrepancies between anterior and posterior corneal curvatures (67% consensus), both of which contribute to unpredictable refractive outcomes. To mitigate these challenges, a multi-formula approach is recommended by 87% of panelists, and targeting slight postoperative myopia can help offset refractive surprises (80% agreement). The Barrett True-K formula was identified as the preferred first-line method, ideally used in conjunction with topography-based calculations, online tools, and averaging across multiple formulas to increase reliability. In cases of high corneal irregularity, the SRK/T (Sanders–Retzlaff–Kraff/Theoretical) formula is not considered appropriate. Furthermore, the Haigis formula is not recommended for use in eyes with long axial lengths.

Regarding IOL selection, key technical preoperative factors related to the IOL calculations identified by the panel included anterior maximal keratometry (87%), corneal stability confirmed over 2 examinations spaced 6 to 12 months apart (100%), posterior corneal curvature (73%), and thinnest pachymetry (87%). Given that keratoconus induces irregular astigmatism, the use of toric IOLs must be considered cautiously and only under specific conditions. These include the following:

1. Reproducibility of topographic astigmatism and biometric cylinder measurements (93% consensus);
2. Correlation between topographic and refractive astigmatism, both in magnitude and axis (87% consensus);
3. Favorable topographic pattern (eg, central or inferior cone, mild pellucid marginal degeneration) (67% consensus);
4. Documented access to reliable spectacle correction before cataract development (80% consensus);
5. Availability of posterior corneal astigmatism measurements (67% consensus); and
6. Confirmed stability of ectatic disease (100% consensus).

Only when these criteria are met can toric IOLs be responsibly considered in patients with keratoconus or other ectatic disorders.

Among the currently available IOL options, pinhole IOLs occupy a distinct role because of several advantageous properties: they increase the depth of focus, reduce peripheral higher-order aberrations, and offer greater tolerance to residual refractive error and IOL calculation inaccuracies. These characteristics make pinhole IOLs particularly suitable for eyes with irregular astigmatism.

The selection of other advanced monofocal IOLs should be guided by the IOL's optical design (80% consensus) and the degree of corneal irregularity and asphericity (73% consensus). In these complex cases,

residual refractive error is not uncommon and may require further management.

Spectacles should always be attempted as the first-line correction and may be sufficient when they achieve a CDVA of ≥0.4 (73% agreement) and anisometropia remains tolerable. Hybrid contact lenses or custom-designed RGP or scleral lenses based on corneal topography are a viable next step (80% agreement).

Surgical enhancement should be reserved as a last resort (67% consensus), pursued only after adequate time has passed for postcataract stability and neuroadaptation. In this context, piggyback IOL implantation offers a reversible surgical solution when placed after a primary in-the-bag IOL. It may be considered a secondary intervention in select cases (Table 9).

DISCUSSION

Group 1: Definition, Diagnosis, and Staging

The present consensus developed a new staging system for keratoconus. Previous staging systems, such as the Amsler–Krumeich and CLEK (Collaborative Longitudinal Evaluation of Keratoconus) staging classifications, were recognized as limited in their ability to incorporate modern diagnostic technologies, particularly corneal tomography, which are now considered essential for accurate diagnosis and staging.[36–41]

In the first round of voting, panelists agreed that the classification of keratoconus severity should be based on 3 core parameters derived from tomographic imaging:

1. Anterior corneal curvature,
2. Posterior corneal curvature, and
3. Corneal pachymetry at the thinnest point.

During the face-to-face meeting, the Global Consensus panel recognized the need for a practical, universally applicable system to stage the severity of keratoconus. To address this, the panel proposed a new classification based on the grading system described initially by Belin.[27] This system incorporates 4 parameters:

i. Radius of corneal curvature at the thinnest point,
ii. Posterior radius of corneal curvature at the thinnest point,
iii. Pachymetry at the thinnest point, and
iv. CDVA.

The Belin ABCD classification provides a more quantitative description of the disease and is available on both Scheimpflug- and OCT-based platforms, ensuring broad applicability in clinical practice.[42]

In the fourth round of voting, the diagnostic group's proposed classification, which stratifies keratoconus into mild, moderate, and severe categories, was submitted to the full panel (n = 86) and received 79.1% approval. This new framework enables the integration of modern imaging technologies that support earlier and more accurate detection, moving beyond legacy systems that are now limited by their inability to incorporate tomographic data. This proposal is intended to be progressively incorporated, through a simple and applicable classification that is both physician- and patient-friendly. However, given the variety in pachymetric and anterior and posterior curvature characteristics of eyes with keratoconus, we aim to explore the performance of this classification in the future, without ruling out possible refinements. Because the proposed classification is based on the Belin ABCD classification, the strengths and limitations of the above could influence the proposed classification.

Since the publication of the first keratoconus consensus a decade ago, significant advances have been made, most notably, the demonstrated effectiveness of CXL and the emergence of a wide array of therapeutic and refractive interventions. These developments have not only enabled reliable disease stabilization but have also achieved levels of corrected and UDVA previously unattainable in keratoconus. Together, they underscore the critical importance of early diagnosis and timely treatment.

The expert panel aimed to clarify terminology and clinical strategies related to the early diagnosis of keratoconus, particularly in high-risk populations. In this context, the term "suspicious keratoconus" was formally defined. Analogous to the designation of "glaucoma suspect," which permits more rigorous monitoring before a definitive diagnosis, the term "suspicious keratoconus" allows corneal specialists to investigate ambiguous or borderline cases more thoroughly.[38]

The panel agreed that patients classified as having suspicious keratoconus should be monitored closely for potential progression to clinically manifest disease, particularly if they are young or candidates being considered for refractive surgery. Such cases should be evaluated using advanced diagnostic modalities, including Scheimpflug tomography, AS-OCT, corneal biomechanical analysis, and mapping of epithelial thickness.[42]

Although the panel was unable to reach consensus on formal definitions for the terms "subclinical keratoconus" and "forme fruste keratoconus," it was agreed that these terms retain clinical relevance and may continue to be used to describe preclinical stages of the disease. Furthermore, recent literature reviews have helped clarify prevailing usage patterns and contextual distinctions between these terms.[39,43]

The panel also addressed the clinical approach to children with astigmatism, recognizing that keratoconus is often diagnosed in this population only after a decline in CDVA has already occurred. To facilitate earlier detection and intervention, the panel agreed that additional diagnostic testing should be performed in all children aged 6 to 18 years presenting with more than 2.0 D of astigmatism on subjective refraction. Specifically, it was recommended that these patients undergo Placido disk–based topography and/or Scheimpflug tomography to rule out keratoconus.[41]

The expert panel considered it essential to define the criteria for keratoconus progression, particularly given the implications for timing intervention. Panelists agreed that the principal indicators of progression mirror those used for disease classification:

1. Significant and reproducible changes in anterior curvature,

TABLE 9. Group 7 Summary of Recommendations

Cataract in Keratoconus

Main findings:

- Indication for CXL or CAIRS (±CXL) before cataract surgery (±CXL) is appropriate under the following conditions: CXL is indicated when there are changes in keratometry >1 D in 1 yr or >0.5 D in 6 months.
- The appropriate timing for cataract surgery in corneal ectasia is defined by lens opacity, patient demand, decreased visual acuity, and stability of the disease.
- Regarding cataract development after corneal grafting (DALK, PK):
  - It is more frequent after PK than DALK.
  - Surgery is indicated after suture removal.
- Regarding cataract surgery after CXL or CAIRS, surgery should be postponed for at least 3 months post-CXL/CAIRS.
- In preoperative assessment (topography, stability), the following parameters are crucial:
  - Anterior maximal keratometry.
  - Stability verified by 2 exams separated by 6 or 12 months.
  - Posterior curvature.
  - Thinnest pachymetry.
- Selection for an advanced monofocal IOL should be based on the following points:
  - The optical principle of the IOL.
  - Corneal irregularity.
- Regarding IOL selection, the following factors play a role in the use of toric IOLs:
  - Reproducibility of topographic astigmatism measurement.
  - Reproducibility of biometric astigmatism measurement.
  - Correlation between topographic and refractive astigmatism in both amplitude and axis.
  - The topographic characteristics of ectasia (central, inferior, PMD).
  - Access to spectacle correction before cataract.
  - Keratoconus stability.
  - Posterior astigmatism value.
- As for IOL selection and the specific placement of pinhole EDOF IOL:
  - Increased depth of focus.
  - Elimination of peripheral HOA.
  - Indication for irregular astigmatism.
  - Better tolerance to inaccurate IOL calculations and remaining refractive error.
- Regarding the selection of a calculation formula for pseudophakic IOL in keratoconus:
  - We recommend Barret True-K as the first choice; topographic keratometry-based calculations for comparison; the use of online calculators.
  - We do not recommend using Haigis for long eyes or the mean value of all formulas.
  - In cases of high irregularities, the use of SRKT is not considered appropriate.
- Concerning IOL calculation in irregular corneal astigmatism (ectasia, graft), the following are true:
  - For Kmax >50 D, predictability decreases significantly.
  - Discrepancies between anterior and posterior curvatures lead to unpredictable results.
  - Predictability decreases significantly.
  - A multifocal formula is recommended.
  - Slight myopia should be targeted.
  - Discrepancies between anterior and posterior curvatures lead to unpredictable results.
- Pertaining to combined surgery (cataract and keratoplasty):
  - The triple procedure (cataract, graft, IOL) results in poor initial refractive predictability.
  - Sequential procedures are recommended in DALK.
  - Should be discussed in the context of grade 4 (opacity), advanced keratoconus associated with cataract.
- Regarding IOL selection and, when needed, secondary piggyback IOL implantation: this procedure may be performed sequentially after primary in-the-bag IOL implantation.
- Concerning the management of residual refractive error, optical treatment options include the following:
  - Spectacles, which may be sufficient when they provide more than 0.4 CDVA.
  - Hybrid contact lenses, RGP, and scleral lenses, which can be customized based on topography.
- With respect to the surgical management of residual refractive error, for surgical treatment, we recommend the following:
  - Surgery can be considered after 3 months postcataract surgery.
  - Surgery should be the last option after the failure of optical treatments.

CAIRS, corneal allogenic intrastromal ring segments; CDVA, corrected distance visual acuity; CL, contact lenses; CXL, cross-linking; DALK, deep anterior lamellar keratoplasty; DM, Descemet membrane; ICRS, intrastromal corneal ring segments; IOLs, intraocular lens; pdDALK, pre-Descemet DALK; PK, penetrating keratoplasty; PRK, photorefractive keratectomy; OCT, optical coherence tomography; SLAK, stromal lenticule addition keratoplasty; TG, topography guide.

2. Posterior curvature,
3. Thinnest pachymetry, and
4. Worsening of visual acuity.

The role of Kmax in assessing keratoconus progression was critically examined. Although 91.7% of panelists agreed that Kmax has low repeatability and reproducibility, 75% continue to use it in clinical practice, and 67% still regard it as a relevant parameter, reflecting its persistence in the current literature and diagnostic workflows.[40] The panel identified the corneal radius of curvature as a promising parameter for progression assessment. It is recommended that future imaging devices incorporate a broader set of curvature-related metrics automatically to support clinical decision making.

Regarding the role of the corneal epithelium, the panelists agreed that epithelial thickness maps are useful for diagnosis. Nevertheless, they emphasized that quantitative epithelial cell data are currently insufficient to support reliable diagnosis, classification, or progression monitoring.

Group 2: Clinical Treatment and Noninvasive Visual Rehabilitation

Serological studies in keratoconus have reported low serum levels of vitamin D, zinc, copper, and selenium, and an imbalance in oxidant/antioxidant status. However, no clinical treatments or noninvasive rehabilitation strategies have yet been established based on these biochemical findings.

The primary nonsurgical approach to managing irregular astigmatism in keratoconus remains contact lens wear. A variety of lens types reached consensus, except for soft contact lenses, unless they are used as part of a hybrid system.[29] A detailed depiction of this step-by-step process is presented in Figure 2.

Eye rubbing and the management of ocular allergy and surface irritation were recognized as key modifiable factors in slowing disease progression. Importantly, RGP lenses were not found to influence keratoconus progression, although their role in visual rehabilitation remains central.[44] Both eye rubbing and allergic eye diseases were acknowledged as risk factors in the development of keratoconus.

In cases of acute corneal hydrops, a variety of nonsurgical treatment options were endorsed, including topical corticosteroids, IOP reduction, and hypertonic saline.

Overall, the panel emphasized the need for high-quality clinical trials evaluating both clinical efficacy and patient-reported outcomes in the nonsurgical management of ectatic corneal disorders.[45] In addition, registries were seen as valuable tools for generating real-world evidence, informing both natural history of the disease and effectiveness of current and emerging treatments, including everyday patient-reported experiences.[6,46,47]

Group 3: Corneal Cross-linking for Progression

The panel reaffirmed that CXL should be performed in patients with definitive tomographic or topographic evidence of disease progression. In younger patients, where the risk of rapid progression and visual decline is elevated, treatment may be warranted at the time of diagnosis, even in the absence of documented progression.[48] These recommendations are consistent with recent studies underscoring the aggressive course of keratoconus in pediatric populations.[49,50] Importantly, although cessation of eye rubbing was strongly endorsed as a behavioral intervention, the panel emphasized that it should neither delay nor substitute for CXL in individuals at high risk.

The choice of CXL protocol remains dynamic. Epi-off CXL remains the preferred approach, particularly in cases of progressive or advanced disease. Although no formal consensus was reached on specific parameters, a trend toward accelerated protocols was observed, with 9 mW/cm^2 for 10 minutes identified as the most commonly used regimen (approximately 40% of respondents). Epi-on CXL is used to a lesser extent, and most panelists do not currently use high-fluence or pulsed light protocols. It is important to note that many modern epi-on protocols demonstrating improved efficacy of the epi-on technique incorporate these very features, suggesting a potential future shift in practice patterns.[18]

Historically, thin corneas—defined by the traditional 400 μm threshold—were considered ineligible for CXL. However, most clinicians now use modified protocols to treat such cases. Despite variability across regions and techniques, most experts now routinely offer CXL to patients with thin corneas, reflecting a more inclusive and adaptable therapeutic approach.[18,51–54]

Finally, there was unanimous agreement that oral riboflavin supplementation and sunlight exposure are not adequate substitutes for CXL. This position is supported by the absence of compelling scientific evidence demonstrating their efficacy in halting disease progression. As such, CXL remains the standard of care for stabilizing keratoconus.

Group 4: Invasive Visual Rehabilitation

Once keratoconus has reached a state of natural stability or has been stabilized through intervention (eg, CXL), clinical focus shifts toward visual rehabilitation. In most cases, satisfactory visual function can be achieved with spectacles and/or contact lenses, the latter of which have seen substantial improvements in design and performance over the past 2 decades.

However, in a subset of patients, spectacle correction remains inadequate, and contact lens intolerance—whether because of biological, occupational, or psychological factors—precludes their use. In such cases, invasive visual rehabilitation techniques should be considered, including those outlined in previous sections (eg, PRK with CXL, ICRS, CAIRS, phakic IOLs, SLAK).

Group 5: Keratoplasty Approaches

The panel reached consensus that keratoplasty should be considered when patients are not fully satisfied with

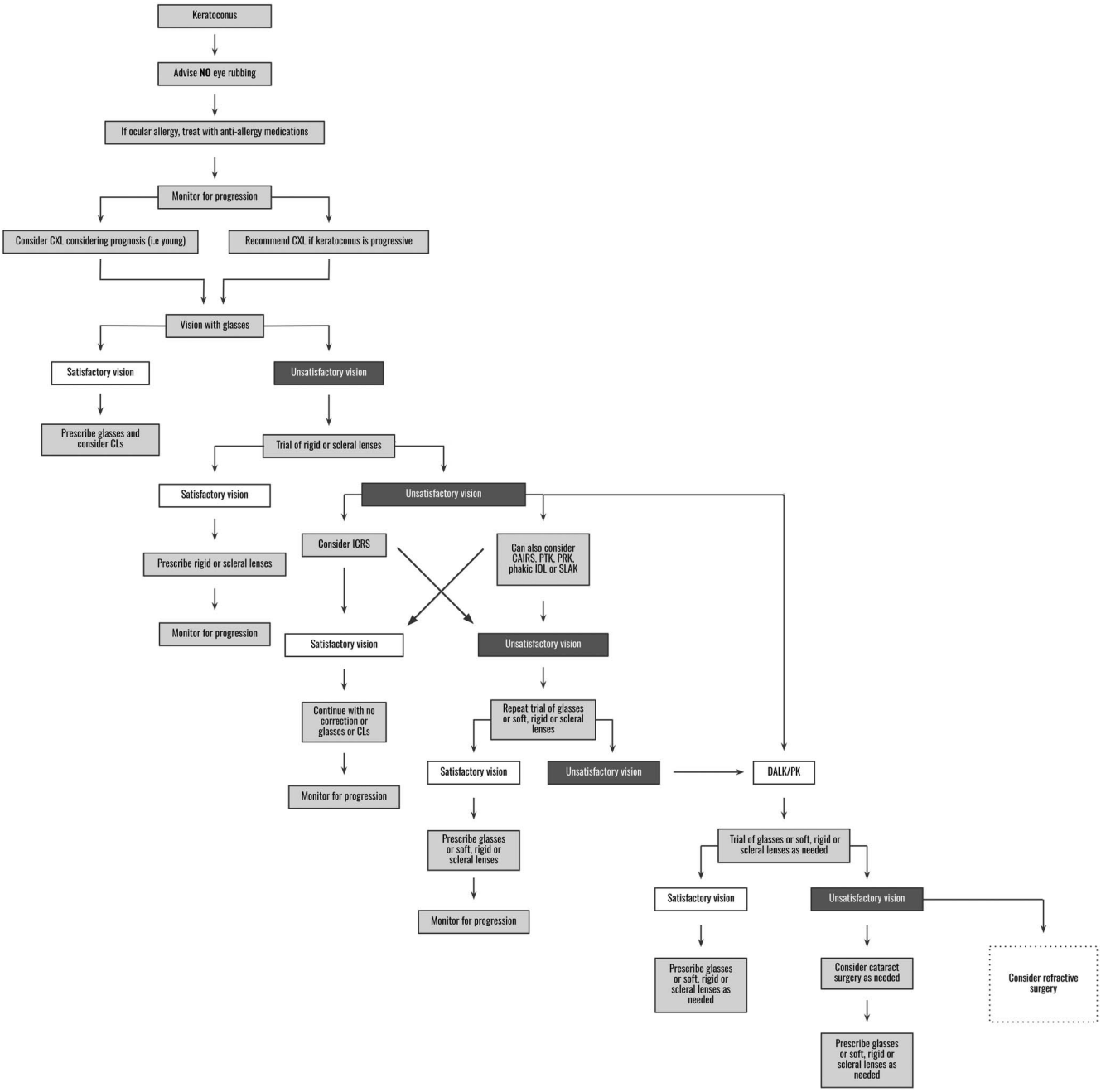

FIGURE 2. Updated keratoconus management flowchart. CAIRS, CL, contact lens; CXL, corneal cross-linking; DALK, deep anterior lamellar keratoplasty; ICRS, intrastromal corneal ring segments; IOL, intraocular lens; PK, penetrating keratoplasty; PTK, phototherapeutic keratectomy; PRK, photorefractive keratectomy; SLAK, stromal lenticule addition keratoplasty.

nonsurgical treatments, particularly when visual function remains compromised. Two primary indications for surgery were identified:

1. Insufficient or unsatisfactory CDVA and
2. Poor tolerance or intolerance to contact lenses.

The preferred definition of contact lens intolerance in this context is moderate to severe discomfort during lens wear that is not alleviated by lubrication or wearing breaks.

The experts further agreed that the primary objective of keratoplasty in keratoconus and other ectatic corneal disorders is to restore or improve CDVA, rather than to achieve complete refractive independence.

Group 6: Elective Refractive Surgery

The panel strongly endorsed the integration of corneal tomography into preoperative screening protocols for

refractive surgery, emphasizing its essential role in mapping corneal curvature and pachymetry to detect early or subclinical forms of ectasia. Although tomography was considered mandatory, a multimodal screening approach was recommended to further reduce the risk of postoperative ectasia. This should include assessment of corneal biomechanics, mapping of epithelial thickness, and evaluation of the family history of keratoconus.

In contrast, genetic testing was not recommended for routine screening, as no consensus was reached on its clinical value in this setting. Improving UDVA in keratoconus remains a challenge. CXL alone does not consistently improve UDVA, although it stabilizes the disease. The addition of excimer laser ablation, such as in combined PRK-CXL protocols, can improve UDVA. Following adjuvant procedures (eg, CXL or ICRS), residual refractive error may persist. In such cases, phakic IOLs, particularly toric lenses for patients with significant astigmatism, were identified as effective options. However, patients should be carefully counseled regarding the variability of refractive outcomes and potential limitations in visual quality, primarily because of residual higher-order aberrations.

The suitability of refractive surgery procedures in patients at increased risk of ectasia remains a matter of controversy. A majority of panelists favored PRK over LASIK in such cases, recognizing its lower biomechanical impact, and the lower risk of causing an iatrogenic ectasia compared with LASIK. Nevertheless, the decision should be based on a comprehensive evaluation, including refractive error, corneal tomography, and epithelial mapping, and biomechanics.

For patients with moderate keratoconus, a combined surgical approach is often necessary to balance visual rehabilitation and ectasia risk. In these cases, techniques such as PRK combined with CXL, ICRS implantation, and phakic IOLs (toric or nontoric) may be integrated to optimize outcomes.

Group 7: Cataract in Keratoconus

Cataract surgery in patients with keratoconus presents unique challenges, particularly regarding IOL selection and predictability of postoperative refractive outcomes. The timing of surgery in eyes with ectatic disease should be guided by multiple factors, including lens opacity, vision impairment, patient expectations, and, critically, the stability of corneal imaging.

IOL power calculation remains a major difficulty in these patients. To minimize error, a multi-formula approach is recommended, with a target of slight myopia to reduce postoperative refractive surprises. The Barrett True-K formula is considered the preferred first-line method, whereas the SRK/T formula may be appropriate in eyes with marked irregularity.

IOL selection should be based on a comprehensive evaluation of the following:

1. Reproducibility of topographic astigmatism,
2. Agreement between biometric and refractive cylinder (magnitude and axis),
3. Topographic cone location and pattern (eg, central, inferior, or pellucid marginal degeneration), and
4. Stability of the ectatic process.

Patients must be counseled preoperatively that precise refractive predictability cannot be guaranteed, even with best practices. Residual ametropia is common and may require further management. In such cases:

1. Spectacles should be the first-line solution and may be adequate when they achieve ≥0.4 CDVA with tolerable anisometropia.
2. Semi-rigid or custom-designed RGP or scleral lenses based on corneal topography may provide enhanced correction.
3. Surgical enhancement should be reserved for cases where optical correction fails, and only after ensuring postoperative stability and neuroadaptation.

CONCLUSIONS

This updated Global Consensus used a modified Delphi methodology to refine guidance on the definition, diagnosis, and management of keratoconus and other ectatic corneal disorders. It offers comprehensive insights from a representative panel of experts in corneal ectatic diseases, producing consensus statements that reflect the best collective judgment currently available, which aligns with current international practices and clinical strategies. The results reflect the group position and do not necessarily represent the individual opinion of each author. The overarching aim is to offer guidance for clinical practice, research priorities, and educational frameworks worldwide. However, consensus does not equate to definitive scientific truth; rather, it represents a structured convergence of expert opinion based on current knowledge, clinical experience, and available evidence. As new data emerge, these positions should be revisited, challenged, and refined to ensure that clinical decision making remains dynamic, evidence-informed, and responsive to evolving scientific understanding.

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