OULUN AMMATTIKORKEAKOULU

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ASSESSMENT AND MANAGEMENT OF KERATOCONUS: CLINICAL GUIDELINE FOR FINNISH OPTOMETRISTS

Research development

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ABSTRACT

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Background: Finnish current care Guidelines do not have any recommendations regarding the diagnosis, treatment, assessment, or management of keratoconus. One international practice pattern exists, nevertheless, it is not designed only for the use of optometrists but also for ophthalmologists. Because the education of optometrists and the professional description and legal rights of optometrist practise in different parts of the world varies the international guideline cannot be straight adapted to the optometrist practise in Finland and a need for a national is justified.

Purpose: The purpose of this study by the Integrative review was to determine and combine appropriate information concerning keratoconus assessment and management, aiming to produce knowledge to be used in the development phase of the study. The purpose of the Research development was to create a clinical guideline concerning keratoconus assessment and management to the use of optometrists in Finland, aiming to select the relevant methods for optometrist practise in Finland. The guidelines should support the utilization of Finnish optometrists as a part of efficient eye health care.

Methods: The primary literature search for the Integrative literature review was conducted by EBSCOhost web source using Academic Search Premier, CINAHL and MEDLINE databases. The search was limited to texts published in English and only texts published between 2013-2023 were accepted. The search resulted in 441 records to undergo further exclusion. PRISMA 2020 Flow Diagram was used to demonstrate the final eighteen articles and one guideline selected and analysed. The results of the literature review were used in the Research Development phase.

Results: The result of this study provides means for Finnish optometrists to identify the clinical signs of keratoconus and tools to assess the severity of the disease. It points out the significance of referral and co-management with ophthalmologists and helps decide the proper management methods provided by optometrists. The personal competence of the practitioner is highlighted. This study provides a baseline proposal for further development of a national guideline.

Conclusions: The competence of optometrists as clinical healthcare professionals is confirmed by this Research Development. Optometrists existing knowledge and capabilities can be utilized more frequently, including in KC assessment and management, providing relief in otherwise overcrowded Eye health care in Finland.

Keywords: clinical guideline, keratoconus, keratoconus assessment, keratoconus management, optometrist

CONTENTS

1 INTRODUCTION

Keratoconus is a progressive eye disorder characterized by the thinning and gradual bulging of the cornea. The altered shape of the cornea can result in distorted vision and finally reduced visual acuity (VA) and visual impairment. While there is no cure for keratoconus, early detection and proper management can halt the progression and improve vision. (Gordon-Shaag et al. 2015; Mas Tur et al. 2017; Santodomingo-Rubido et al. 2022.) Keratoconus is a disease involving both genders and appearing in all ethnicities and it usually develops in the second decade of life. The prevalence of KC is approximately 54 per 100,000 in the general population. (Romero-Jiménez et al. 2010.)

Traditionally keratoconus, as well as many other eye diseases, is a disease mainly assessed, diagnosed and managed by ophthalmologists in Finland and the Finnish eye health care is currently distributed between private sector providers and public eye healthcare (Näe Ry 2020). According to the Ministry of Social Affairs and Health (Sosiaali- ja terveysministeriö 2023 199-200) optometrists' role regarding keratoconus management is currently in contact lens fittings provided by licenced contact lens opticians.

Optical stores, private medical centres and hospitals provide a wide range of eye health care services (Näe Ry 2020) and often optometrists and opticians can be the first contact for the patient with an eye health care professional. Optometrists' education is widely developing towards more clinical knowledge and expertise regarding eye health examinations (European Council of Optometry and Optics 2020; Näe Ry 2020). Therefore, it is justified that the knowledge of optometrists would be considered in the planning and providing of efficient eye health care in Finland.

The increasing amount of aging population will set a demand for sufficient eye health care and the arrangement and organisation of the care should be more closely considered (Näe Ry 2020). With proper education, some of the assessment, management and follow-up of keratoconus could be distributed between optometrists and ophthalmologists in intent to be able to provide more efficient eye health care.

Regarding keratoconus assessment and management one international practice pattern and guideline exists (Garcia-Ferrer et al. 2019). Nevertheless, the practice pattern is not designed only for the use of optometrists but also for ophthalmologists. Because the education of optometrists and the professional description and legal rights of optometrists in different parts of the world vary (European Council of Optometry and Optics 2020), the international guidelines cannot be directly adapted to the optometrist practise in Finland.

Finnish Current Care Guidelines are independent, evidence-based clinical practice guidelines for Finnish health care providers. The purpose of these national guidelines is to cover essential issues concerning Finnish health, medical treatments as well as prevention of diseases. The guidelines are designed as a basis for treatment selection and can be utilized by physicians, dentists, healthcare professionals and citizens nationally. (Current Care Guidelines 2022.)

Current Care Guidelines provide commonly accepted protocols for the management of many eye diseases and conditions but do not have any recommendations regarding the diagnosis, treatment, assessment, management, or follow-up of keratoconus or other ectatic diseases. As the Ethical Council of Optometry (OEN) (2020) states in a statement for OAUS Master's degree students in Clinical Optometry, it would be recommended to create correspondent guidelines to go hand in hand with Nordic and International practice patterns. With reference to these observations regarding the keratoconus assessment and management, a need for the implementation of unified recommendations exists. This development project was later commissioned by the OEN.

The purpose of this study by the literature review is to determine and combine appropriate information concerning KC assessment and management. The aim of the literature review is to produce knowledge of the current status of KC assessment and management to be used in the development phase of the study.

The purpose of the research development phase is to create a clinical guideline concerning KC assessment and management for the use of optometrists in Finland. The aim of the research development phase is to select the relevant methods concerning KC assessment and management for the optometrist practice in Finland. The guidelines should support the utilization of Finnish optometrists as a part of efficient eye health care.

2 THEORETICAL BACKGROUND

A preliminary data search was performed in January 2023 to familiarize with the study subject and gather theoretical background information. A search from PubMed with keywords corneal ectasia, keratoconus AND pathophysiology, keratoconus AND guidelines, keratoconus AND assessment, and keratoconus AND management was conducted including articles not older than 10 years. In addition to background gathering the purpose was to evaluate the number of studies available and requirements for the primary search process. The one existing international guideline (Garcia-Ferrer et al. 2019) was identified from the search and selected for further concept analysis. This guideline was considered as baseline for selecting key elements in the guideline development process.

Some additional searches from PubMed, professional literature from books (Bennet & Henry 2014; Bowling 2016; Seppänen et al. 2022), Finnish laws from online legislation and juridical databases and guideline implementation-related literature were also searched and included in gathering the theoretical background for this thesis. A Swedish clinical guideline for optometrists regarding keratoconus assessment and management (Westerlund & Robertson 2020) was found in an additional web search in September 2023.

2.1 Corneal Histology and Context of Corneal Ectasia

2.1.1 Corneal Histology

Cornea is an avascular tissue with a complex structure of five layers; epithelium with its basement membrane, Bowman's layer, the stroma, Descemet's membrane and endothelium. Cornea plays a protective role and creates about three-quarters of the optical power of the eye. The cornea is a densely innervated avascular structure where nutrients and metabolic products are carried mostly by the aqueous humour and tear film. (Bowling 2016, 168).

The corneal epithelium is on average 50 microns thick and is composed of 4-6 non-keratinized layers of stratified squamous epithelial cells composed of squamous surface cells, wing cells and basal cells. Corneal stem cells are located at the limbus creating a physiological barrier between

the cornea and conjunctiva. (Bowling 2016, 168.) The epithelial basement membrane consists mostly of collagen type IV, laminin, and entactin, and the main proteoglycan is perlecan (Soiberman et al. 2017, 253).

The Bowman's acellular collagen layer is the most superficial layer of the stroma that separates it from the epithelial basement membrane. The Stroma makes up approximately 90% of the corneal thickness with an average thickness of 500 microns. It is formed of regularly arranged collagen fibril layers and an extracellular matrix containing proteoglycans and keratocytes. (Bowling, 2016, 168.) Bowling (2016, 168) mentions the main proteoglycans in the stroma to be chondroitin sulphate and keratan sulphate and Soiberman et al. (2017) adds decorin, biglycan, lumican, keratocan and osteoglycin to the list. The highly ordered collagen network is a critical component in corneal optical clarity. Stroma has no ability to regenerate after trauma resulting in scar tissue formation. (Bowling 2016, 168).

Beneath stroma lies Descemet's membrane, a fine, discrete sheet with regenerative potential functioning as a modified basement membrane for the endothelium (Bowling 2016, 168).
According to Soiberman et al. (2017, 253), the layers related to the pathogenesis of Corneal ectasia and keratoconus (KC) are epithelium and its basement membrane, Bowman's layer, and the stroma.

2.1.2 Corneal Ectasia

Corneal ectasia is a progressive disease of the cornea where steepening and thinning of the corneal structures will occur. Corneal ectasia has many types including keratoconus, pellucid marginal degeneration, keratoglobus, postkeratorefractive ectasia and wound ectasia after penetrating keratoplasty (PK). Corneal ectasia is often associated with reduced uncorrected visual acuity (UCVA), an increase in ocular aberrations, and decreased best-corrected distance visual acuity (BCVDA). Corneal ectasias can cause serious ocular malaise and may lead to surgical intervention. (Garcia-Ferrer et al. 2019, 178.)

Over the last two decades, there has been a revolution in the knowledge related to diagnosing and managing keratoconus and other ectatic corneal diseases. As a result, the ability to identify and diagnose corneal ectasia at a much earlier stage has become straightforward and more reliable

than previously. (Gomes et al. 2015, 359.) The clinical objectives in the treatment and management of corneal ectasia include finding corneal ectasia risk factors and combined conditions and recognising the clinical signs. The main principals considered in corneal ectasia management and assessment are understanding and establishing the appropriate diagnostic methods and non-surgical and surgical treatment options, improving visual capacity and preventing loss of visual function. (Garcia-Ferrer et al. 2019.)

Keratoconus is the most common form of corneal ectasia (Gordon-Shaag et al. 2015). Although there are similarities in the management and assessment of all ectatic corneal disorders (Garcia-Ferrer et al. 2019), the higher amount of keratoconus patients compared to other ectatic diseases justified a decision to limit the theoretical background, the literature review and content of the guideline to concern mainly keratoconus.

2.2 Keratoconus Description, Prevalence, and Risk Factors

Description

Keratoconus (KC) is described as a bilateral and usually asymmetrical corneal disorder leading to progressive stromal thinning, fracture of the anterior limiting membrane, and protruding of the central or paracentral cornea, creating a cone-shaped appearance. Progression of KC often results in myopia progression, irregular astigmatism and finally reduced visual acuity (VA) and visual impairment. (Gordon-Shaag et al. 2015; Mas Tur et al. 2017; Santodomingo-Rubido et al. 2022.) Garcia-Ferrer et al. (2019) also states, that more severe progression of KC can occur due to eye rubbing, family history or younger age of onset and greater progression with more irregular astigmatism, thinning and scarring correlates with more serious visual impairment.

The early stages of KC can be divided into subclinical or form-fruste KC. When topographic signs of KC or suspicious topographic findings are present in one eye without detectable corneal slit-lamp findings and KC is found in the fellow eye, subclinical KC is present. The term form-fruste KC is commonly related to an eye with normal topography and corneal slit-lamp findings and KC found in the fellow eye. However, the adopted purpose of these terms can vary in literature because there are no unified criteria agreed upon. (Santodomingo-Rubido et al. 2022, 8.)

Prevalence

KC often occurs in youth and progresses through the third and fourth decade of life, however, new imaging modalities have proved the ectatic disease can develop already in pre-puberty and progress even after the age of 40 (Gomes et al. 2015).

According to a meta-analysis by Hasheme et al. (2020), the prevalence of KC is approximately 130 per 100,000 population globally although the prevalence was shown to have a wide variation by geographic area. The prevalence seems to be highest in the Middle Eastern countries where Saudi Arabia shows as high as 4790 per 100,000 in a hospital and pediatric study and 3300 per 100,000 in a Lebanese population study which is considered more accurate compared to hospital studies. Population studies from the US show a prevalence of 54.5 per 100,000, from the Far East in Japan 17.3 per 100,000, from Europe Netherlands at 265 per 100,000 and from Finland 30 per 100,000. According to Seppänen et al. (2022), the amount of KC patients in Finland is approximately 2500-5000.

Regardless of the possibility of bias with the different criteria and methods used the in detection and diagnosis of KC in different studies, the prevalence in certain populations and regions seems inevitably higher. (Ferrari & Rama 2020.)

Risk Factors

Crawford et al. (2020) concluded that environmental factors and mechanical factors such as eye rubbing, atopy, floppy eyelid syndrome, contact lens wear, thyroid hormones and pregnancy can be risk factors for KC and eye rubbing shows the most major association with the development of the disease. It seems inevitable that eye rubbing can lead to ocular surface inflammation, the release of stromal matrix-degrading enzymes, thinning of the corneal epithelium, and keratocyte loss which are all associated with KC aetiology. Ferrari & Rama (2020, 368) believe eye rubbing could also be an indirect effect of atopy or ocular allergy-induced itch-promoting KC development or progress. As an addition, the global consensus on ectatic and corneal diseases (Gomes et al. 2015, 364) showed an agreement on the following risk factors for KC; Down syndrome, relatives of affected patients, especially in young patients, ocular allergy, ethnic factors (Asian and Arabian), connective tissue disorders (Marfan syndrome), Ehlers-Danlos syndrome and Leber congenital amaurosis.

Laser refractive surgery (LASIK, SMILE, PRK) induced corneal ectasia risk factors include thinner residual stromal thickness, flap thickness more massive than anticipated or the patient had hidden characters of a subclinical KC preoperatively. Nevertheless, KC can also develop without the appearance of these situations. (Garcia-Ferrer et al. 2019.)

2.3 Keratoconus Aetiology and Pathophysiology

KC is recognized to be a multifactorial disease where genetic, biochemical, biomechanical, and environmental components play a major role in the disease pathophysiology, where no primary pathophysiologic explanation has not been found (Gomes et al. 2015, 364,368). KC has traditionally been considered a noninflammatory disease, but recent studies also report an association with significant alterations in inflammatory mediators demonstrating that KC eyes are often involved with ocular inflammation conditions. (Santodomingo-Rubido et al. 2022.)

KC histopathological changes seem to affect all corneal layers with more noticeable distortion in the central cornea compared to the peripheral cornea. Histopathological changes are largely found in the corneal epithelium, Bowman's layer, and stroma. Endothelial changes due to KC are somewhat debated due to KC-related management methods that can alter the endothelial morphology, e.g. contact lenses. Likewise, Descemet's membrane appears to experience a more limited effect. (Santodomingo-Rubido et al. 2022.)

Biochemical Properties

Sugar & MacSai (2012, 716) investigated KC aetiology and stated that many biochemical properties that alter with KC are linked to corneal collagen diminishment. Additionally, proteolytic enzymes alpha-1-proteinase inhibitor and alpha-2-macroglobulin diminishment were found to have a connection with KC. Also, enzyme activity modifications were found to affect the corneal deterioration as well as elevated activities of acid phosphosphatase, acid esterase, cathepsins B, G and K, and trypsin-2 in the keratoconic epithelium. Other biomechanical properties linked to KC were Matrix metalloproteinases found to cause stromal degradation and conversion of tissue inhibitors of metalloproteinases demonstrated to endorse corneal stromal breakdown. Highly increased SFRP1 regulators that affect apoptosis in the epithelium were found in keratoconic corneas. Keratocyte apoptosis is also present in KC along with increased corneal fibroblast reactivity to oxidative stress.

Associated Genetic Disorders

Because KC is highly associated with collagen diminishment, as mentioned earlier, according to Garcia-Ferrer et al. (2019) specific genetic disorders linked to KC covers a variety of connective tissue diseases with exceptional collagen and hyperelasticity. Such diseases as Ehlers-Danilos syndrome, oculodentodigital dysplasia, osteogenesis imperfecta, ichtyosis, congenital hip dysplasia, nail-patella syndrome, Down syndrome, pseudoxanthoma elasticum, hyper-immunoglobulin E syndrome associated with eczema and atopy have been linked with KC.
It has also been a proven that eye rubbing and diminished mental capacities are linked to other genetic disorders such as Apert syndrome, hyperornithinaemia, Crouzon syndrome, Angelman syndrome, Down syndrome, and Noonan syndrome linked to KC.

Biomechanics

Corneal ectatic diseases and laser refractive surgery cause changes in corneal mechanical and optical quality. It has been demonstrated in several studies that eyes with KC have substantially lower corneal central thickness accompanied by lower corneal hysteresis and corneal resistance factor, terms used to characterize corneal biomechanics. It is believed to be a reaction caused by the distortion of the lamellar matrix in the stroma. (Mas Tur et al. 2017, 775.)

2.4 Keratoconus Assessment

Early detection of KC is an important factor in the process of KC assessment and management. Early detection plays a major role in enhanced outcomes and in selecting the correct intervention methods and the effect will finally reduce the need for corneal transplantation. (Santodomingo-Rubido et al. 2022, 10.)

Patients with symptoms and signs related to corneal ectasia or KC should go through a comprehensive eye examination with relevant aspects. The diagnosis of KC is typically based on distinctive findings on corneal topography and tomography and common patient history. The diagnosis is ideally made before the patient experiences symptoms in a pre-clinical stage, but no convenient and economical patient screening test is available, leading to later diagnosis and more advanced stages of KC. The possibly progressive corneal ectatic eyes, such as subclinical disease

in patients considering keratorefractive surgery, or young people that are more likely to progress, should be identified properly at an early stage. (Garcia-Ferrer et al. 2019, 12.)

2.4.1 Patient History

A thorough patient history is an important aspect of every KC patient assessment aiding in the evaluation of the condition. Patient history should include outbreak and development of the disease, vision history (degree of impairment), ocular history with contact lens history and surgical history, medical history, as well as family history. (Garcia-Ferrer et al. 2019.)

2.4.2 Examination

Visual Function Assessment

Most patients with KC represent features of progressive myopia and astigmatism eventually involving both eyes (Oyeniran & Tauqeer 2021). According to Gomes et al. (2015), subjective refraction should be performed in all KC patients. Unstable refraction with progressively increasing astigmatism is a typical KC sign ultimately resulting in decreased VA where the best spectacle-corrected VA (BSCVA) is less than 20/20. When an unlimited time for optotype reading is permitted during a VA test better results are obtained compared to reading the letters as fast as possible. This indicates that the progression is further than expected and visual function is far worse than suspected. Irregular astigmatism is also detected by scissors-reflex, a light reflex where retinoscopy light appears to create a dark centre instead of being evenly distributed. (Garcia-Ferrer et al. 2019.)

As KC progresses visual distortion and mild blurriness can also occur (Santodomingo-Rubido et al. 2022, 9). Corneal higher-order aberrations can alter the visual capability and retinal image quality considerably in the KC population compared to the normal population with regular corneas. Corneal aberration measurements in the KC population may be helpful in optical aberration correction with custom contact lenses and surgical refractive corrections and impact the improvement of visual performance. (Gobbe & Guillon 2005, 2.) Visual optical quality can be also assessed with a pinhole test to compensate for the effect of refractive error (Bowling 2016, 582). Visual impairment caused by optical aberrations linked to KC can be detected by the pinhole test (Kanclerz 2022).

External Examination

In advanced stages, external examination can deliver clear signs of KC. Munson sign (figure 1), a V-shaped deformation of the lower eyelid during downgaze is recognized widely in the literature and advised to be looked for in an external examination (Garcia-Ferrer et al. 2019; Oyeniran & Tauqeer 2021, 308; Santodomingo-Rubido et al. 2022, 10.) Rizzuti's sign is another hallmark often observed in the advanced stages of KC where a bright reflection of the nasal area of the limbus is detected when light is pointed to the temporal limbal area (Oyeniran & Tauqeer 2021, 308; Santodomingo-Rubido et al. 2022, 10).

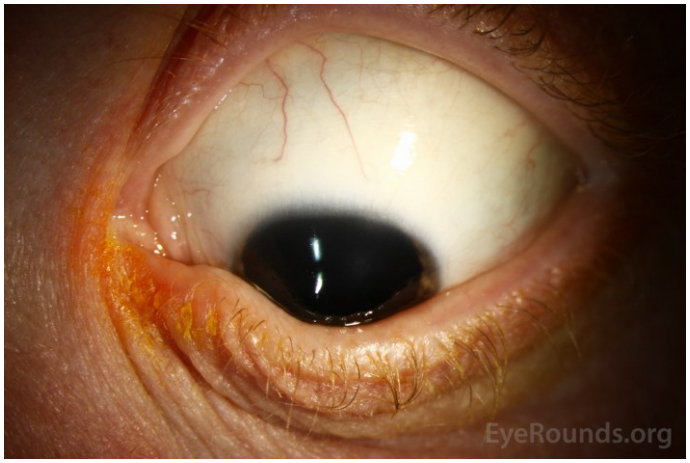

FIGURE 1. Munson sign (Critser 2019).

Corneal Slit-Lamp Biomicroscopy

Biomicroscopy is necessary for the diagnosis of KC where the observer can detect the subtle changes within the cornea (Bennet & Henry 2014.) According to Gomes et al. (2015, 14) and Santodomingi-Rubido et al. (2022, 10) corneal thinning, corneal protrusion, Fleischer's ring and prominent corneal nerve fibres are the hallmark findings of KC in corneal slit-lamp biomicroscopic assessment, with these signs recognized in over 50% of patients with KC. In KC slit-lamp biomicroscopy, corneal thinning is observed with an optic section (figure 2) and is apparent centrally and paracentrally, and the cornea most commonly protrudes at the thinnest point of the cornea creating the cone.

FIGURE 2. A paracentral cone with associated stromal thinning (Vislisel & Karakas 2019).

Around the cone, a yellow-brown ring of iron deposits in the deep epithelium, also known as the Fleischer ring (figure 3), can be observed with or without the assistance of cobalt blue light. Corneal nerve fibres can be recognized entering the stoma from the limbal cornea as slender pale lines. (Nuzbrokh et al. 2020.)

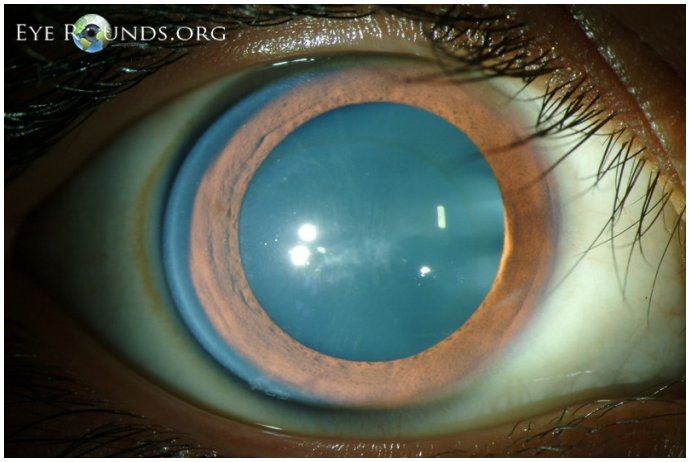

FIGURE 3. Fleischer ring around the base of the cone. Vogt's striae and stromal scarring are also present. (Ward 2019.)

In addition, parallel stress lines formed by corneal lamellae stretching, called Vogt striae, can be visualized in the posterior stroma (figure 4). Vogt striae appear as a series of vertical or oblique lines and are temporarily eliminated by transient pressure applied to the globe through the upper lid. (Bennet & Henry 2014, 524; Nuzbrokh et al. 2020.) Any evidence of apical corneal scarring (figure 3), previous hydrops or previous corneal surgery should be examined (Garcia-Ferrer et al. 2019, 183).

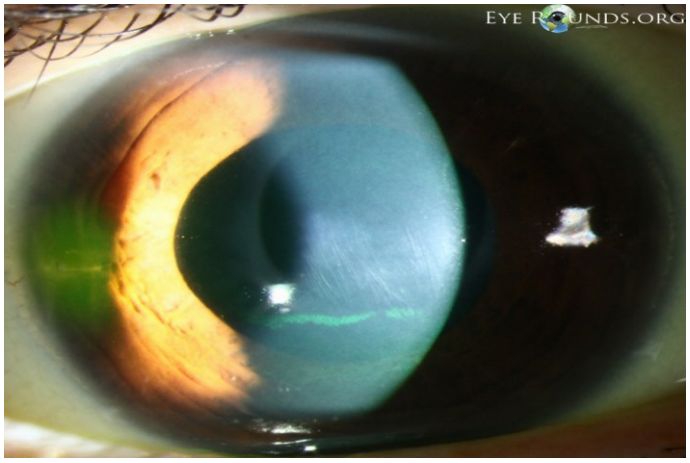

FIGURE 4. Vogt's striae in keratoconus (Kirkpatrick & Venckus, 2019).

IOP and Fundus Examination

Because decreased corneal thickness in KC patients has a reductive effect on Intraocular pressure (IOP) measurement with an applanation tonometer (Goldmann, Perkins etc.), it is advised to operate with alternative devices less reliant on sleek corneal surface. Devices such as Pneumatonometer, rebound tonometer, Mackay-Marg tonometer, dynamic contour tonometer or ocular response analyzer are preferred. (Garcia-Ferrer et al., 2019.)

According to Garcia-Ferrer et al. (2019, 184) during a KC patient or a KC suspect patient assessment, a comprehensive fundus examination should be performed to recognize possible signs of tapetoretinal degenerations associated with KC. Gideon Abou Said et al. (2023, 89) investigated the fundus red reflex and the data it provides as a method for KC screening and diagnosis. In their study they found the red reflex to be a valuable diagnostic evaluation method for KC. In an ophthalmoscopy examination performed in a KC eye, an annular dark shadow in red reflex, also called an oil droplet sign (figure 5), was demonstrated in all their study eyes with KC or KC suspect but in none of the control eyes.

FIGURE 5. Oil droplet red reflex. (Bowling 2016, 214)

Because early detection of KC is essential for KC management and preventing vision loss, oil droplet signs may be used to identify KC and subclinical KC cases especially when other diagnostic devices (e.g. corneal tomography) are not available.

2.4.3 Diagnostic Tests

Keratometry

KC is often related to irregular corneal astigmatism and increased steepening of corneal curvatures. Keratometry can be utilized in the diagnosis and monitoring of KC, however, it has limitations due to measuring only a few paracentral points of the cornea. KC detection can be difficult if a decentred corneal apex is present. (Bennet & Henry 2014, 521.) Corneal keratometry values can be useful in the classification of KC based on the corneal central power magnitude where the increase of corneal curvature and corneal power demonstrates more advanced KC. (Santodomingo-Rubido et al. 2022, 13.) Garcia-Ferrer et al. (2019) also remind us that no keratometric value alone is enough for the KC definition.

Corneal Topography and Tomography

The importance of corneal topography in detecting KC is undeniable and detection, diagnosis and follow-up of the disease has greatly improved. Topographic and tomographic slit scanning and

Scheimpflug imaging devices aid in detecting curvature changes on the anterior and posterior corneal which are often early indicators of KC. (Matalia & Swarup 2013.)

Corneal topography maps reveal corneal curvature, and irregular astigmatism with steepening in a non-invasive qualitative and quantitative interpretation. Corneal topographer provides a wide range of maps to be analysed: anterior, sagittal, and tangential curvature maps and corneal thickness maps. In addition, corneal tomography provides further data on the corneal anterior and posterior surfaces (figure 6). (Nuzbrokh et al. 2020.)

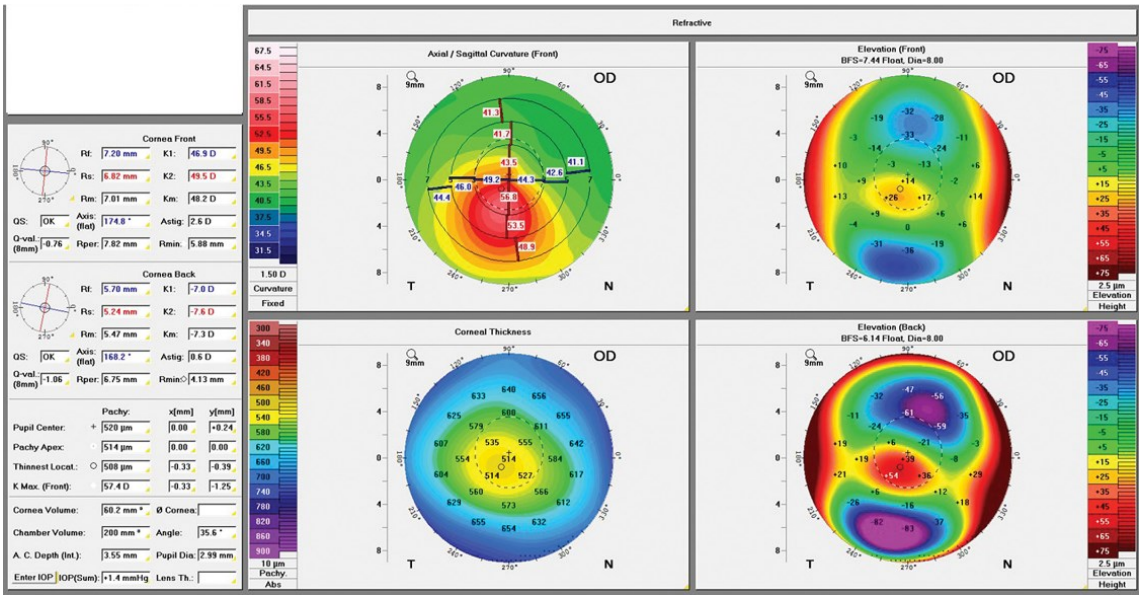

FIGURE 6. Tomography image of inferior corneal steepening and posterior corneal elevation in keratoconus (Fan et al. 2018).

According to Koc et al. (2020), it is not difficult to diagnose KC in the intermediate or advanced stage with corneal topography because the topographic changes are often evident. The early stages of KC or subclinical KC are more complicated since topographic data is limited to the anterior surface of the cornea. The early changes in KC are more sensitively detected by corneal tomography. (Koc et al. 2020.) However Garcia-Ferrer et al. (2019, 185) state ectatic corneas often possess isolated islands of elevation that can be helpful characteristics in KC recognition but also believe that subclinical KC is less obvious on posterior elevation mapping and tomography compared to KC detection. Therefore, anterior and posterior corneal surfaces need to go through a thorough evaluation by topography and tomography when properly assessing and managing KC.

Optical Coherence Tomography (OCT)

Anterior segment optical coherence tomography (AS-OCT) devices are able to deliver two-dimensional cross-section images of the anterior ocular surface including cornea, angle, anterior chamber and anterior lens (figure 7) and are more widely used in KC assessment and KC-related contact lens practice. OCT images create a high-resolution image of the corneal layers and enable a detailed analysis valuable in diagnosis, management and follow-up of anterior segment changes. The advantage of this technology in contact lens design decisions and fit evaluations, especially with scleral lenses, is already evident. (Bennet & Henry 2014, 526.)

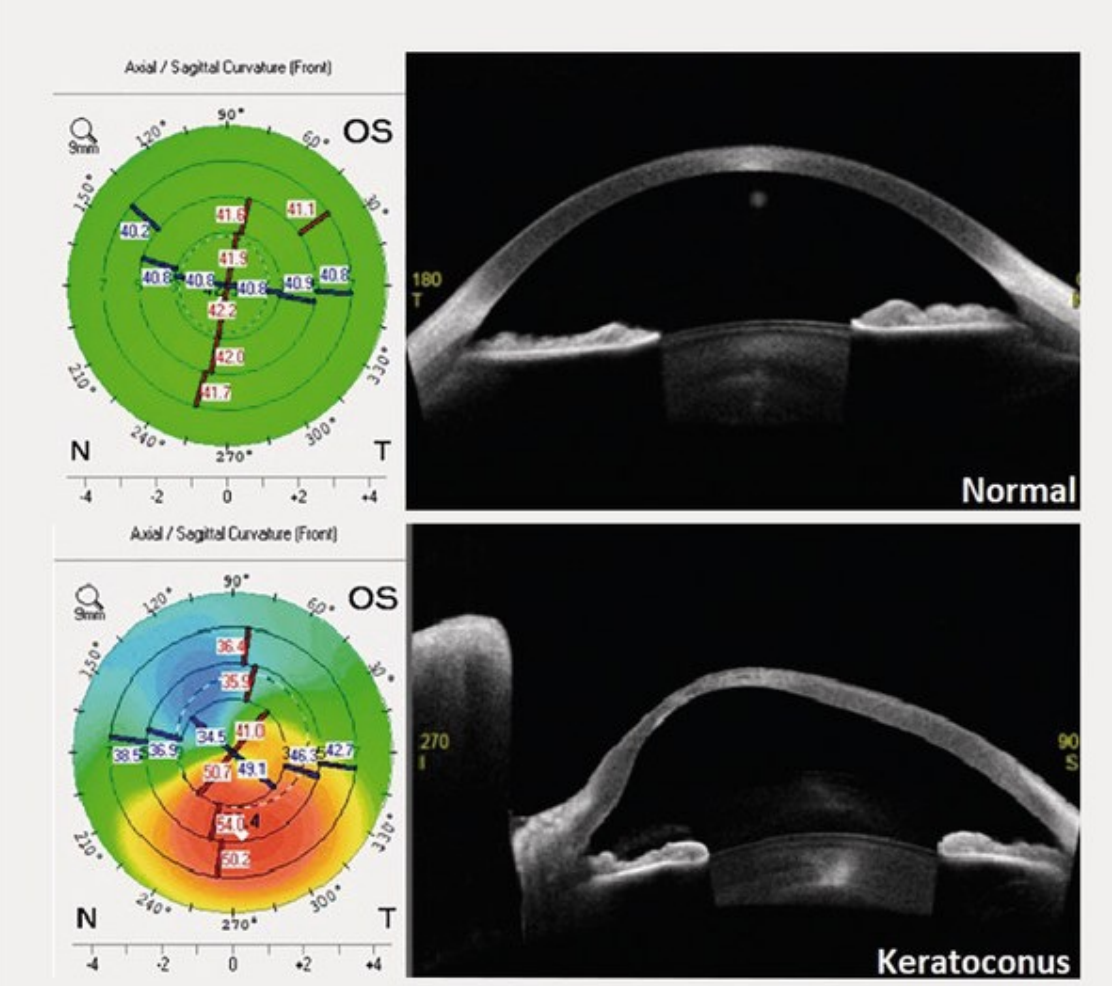

FIGURE 7. Corneal tomography and anterior segment OCT in normal and keratoconus patients (Sideroudi et al. 2023).

2.4.4 Keratoconus Progression, Classification and Grading

According to Gomes et al. (2015), there are various classification systems created for KC, despite no universally accepted classification model or a true or obvious definition of KC progression exists.

However, for documenting KC progression, it has been agreed that at least two of the following changes need to be present: steepening of the anterior corneal surface, steepening of the posterior corneal surface and/or thinning or decrease in corneal thickness. The degree of these changes to clearly demonstrate progression is still under debate. (Andreanos et al. 2017, Gomes et al. 2015; Santodomingo-Rubido et al. 2022.)

Two widely used classification systems, the Amsler-Krumeich system introduced in 1947, and the CLEK study classification system introduced in 1996, are found to be outdated and current information and technological advances in their systems are not utilized. (Gomes et al. 2015, 363, 364, 367.)

In the CLEK (Collaborative Longitudinal Evaluation of Keratoconus) study vision fluctuation, keratometry, biomicroscopic signs, corneal scarring and vision-specific life quality was utilized in the KC classification (Belin & Duncan 2016).

Based on spectacle refraction, central keratometry values, presence or absence of scarring and central corneal thickness, The Amsler-Krumeich system grades KC into 4 stages (Belin & Duncan 2016). The system was further developed and by including corneal scarring and anterior corneal aberrations into consideration, the Alio-Shabayek grading system was developed. The two grading systems are compared in the table 1.

TABLE 1. The Amsler-Krumeich and Alio Shabayek classification systems for grading keratoconus severity (Santodomingo-Rubido et al. 2022).

Amsler – Krumeich	Alio - Shabayek
Grade 1	
Corneal steepening	No scars
Refraction > - 5 D	Coma-like RMS 1.50 to 2.50 um
Mean central K readings < 48 D	Mean central K readings < 48 D
Grade 2	
No scars	No scars
Corneal thickness > 400 um	Corneal thickness > 400 um
Refraction > - 8 D	Coma-like > 2.5 to ≤ 3.50 um
Mean central K readings < 53 D	Mean Central K readings < 53 D
Grade 3	
No scars	No scars
Corneal thickness > 300 um	Corneal thickness > 300 um
Refraction > - 10 D	Coma-like RMS > 3.50 to ≤4.5 um
Mean central K readings < 55 D	Mean central K readings < 55 D
Grade 4	
Central scarring	Corneal scarring
Corneal thickness > 200 um	Corneal thickness > 200 um
Not reliable refraction	Coma-like RMS > 4.5 um
Mean central K readings > 55 D	Mean central K readings > 55 D

Nore: Coma-like RMS (root mean square error) values refer to a 6mm analysis diameter.

Because none of the previously existing classification systems utilized current tomographic data, Belin & Duncan (2016) established a new system called the ABCD Grading system (table 2) where the Anterior radius of curvature (A), Posterior radius of curvature (B for back surface), thinnest Corneal pachymetry (C), Distance best-corrected VA (D) are observed. Also, a modifier (-) for no

scarring, (+) for scarring that allows iris details to be seen and (++) for scarring that blocks iris details. (Belin et al. 2020).

TABLE 2. The New ABCD system for classification of keratoconus (Belin et al. 2020).

ABCD Criteria	A ARC (3mm zone)	B PRC (3mm Zone)	C Thinnest Pach um	D BDVA	Scarring
Stage 0	> 7.25 mm (<46.5 D)	> 5.90 mm (<57.25 D)	> 490 um	= 20/20 (=1.0)	-
Stage 1	> 7.05 mm (<48.0 D)	> 5.70 mm (<59.25 D)	> 450 um	< 20/20 (<1.0)	-,+,++
Stage 2	> 6.53 mm (<53.0 D)	> 5.15 mm (<65.5 D)	> 400 um	< 20/40 (<0.5)	-,+,++
Stage 3	> 6.15 mm (<55.0 D)	> 4.95 mm (<68.5 D)	> 300 um	< 20/100 (<0.2)	-,+,++
Stage 4	< 6.15 mm (>55.0 D)	< 4.95 mm (>68.5 D)	=300 um	<20/400 (<0.05)	-,+,++

Note: ARC: Anterior radius of curvature, PRC: Posterior radius of curvature, BDVA: Best distance visual acuity.

2.5 Keratoconus Management

The main principles in KC management are to prevent visual loss, cut down the signs and symptoms of KC and endure, alleviate or increase visual function based on the patient's demand (Garcia-Ferrer et al., 2019, 12). KC severity and progression define the need for management and treatment of the disease and the correct methods are selected according to the thorough assessment (Santodomingo-Rubido et al. 2022, 14).

2.5.1 Prevention and Early Detection

According to Garcia-Ferrer et al. (2019), early detection and treatment of KC is important for visual function maintenance. KC progression and progression-related visual loss are known to affect life

quality and the economic burden induced by caring for KC patients throughout their life is considered a health problem. Therefore, for any young patient with changing refractive error, KC suspicion should always be ruled out by a careful evaluation and follow-up.

Corneal Cross-Linking (CXL)

Several studies stated, that as a rule, anyone with progressive KC with clear cornea and minimal corneal thickness of 400 microns should undergo CXL (Gănescu 2022; Garcia-Ferrer et al. 2019; Gomes et al. 2015; Nuzbrokh et al. 2020; Santodomingo-Rubido et al. 2022). CXL treatment utilizes ultraviolet A and riboflavin to stabilize corneal tissue, increasing the biomechanical stability and rigidity of the cornea as an effort to prevent KC progression. Additionally, CXL has been found to increase best corrected VA by 1-2 lines and cut down corneal maximum keratometry (Kmax) by 1-2 diopters. (Nuzbrokh et al. 2020.) According to Andreanos et al. (2017, 251) Corneal and total wavefront aberration improvement has also been noted. Long-term follow-ups have also shown continuous corneal flattening for up to several years after the treatment showing an extended CXL effectiveness beyond the first months of surgery.

2.5.2 Patient Education

After KC diagnosis verbal guidance to the patient is considered as one of the most important measures in non-surgical KC management (Gomes et al. 2015, 363). Patients with KC have a variety of non-surgical and surgical treatment options available, and the importance of early tomographic evaluation is crucial in determining the state of the disease and establishing a baseline for progression determination and deciding appropriate management options. The benefits and probable risks linked with early CXL should be discussed as well as optical and other surgical management possibilities. (Garcia-Ferrer et al. 2019, 198,199.)

Avoidance of eye rubbing, use of topical antiallergic medications in patients with allergy and use of topical lubricants in case of ocular irritation reduces the stimulus to eye rubbing. Proper patient education and counselling aid the patient in controlling persistent habits of unnatural eye rubbing. (Garcia-Ferrer et al. 2019, 199; Gomes et al. 2015, 363; Santodomingo-Rubido et al. 2022, 14.) Post-penetrating keratoplasty patients should be educated about the indications of rejection, including redness, light sensitivity, vision alterations, and/or pain, and to request for medical attention without hesitation. (Garcia-Ferrer et al. 2019, 198). Follow-up visit intervals are

determined based on the disease progression and/or the management method chosen. (Garcia-Ferrer et al. 2019, 197).

2.5.3 Optical Management

Spectacle Correction

Myopia and high astigmatism related to KC are typically corrected with spectacles as long as satisfactory VA can be obtained. In the mild stages of KC, spectacle correction is considered decent when a VA of 20/40 can be achieved. With KC-associated irregular astigmatism, spectacle correction can often be inadequate and contact lenses could be considered to provide a more satisfactory visual outcome. The type of contact lenses prescribed varies depending on the KC state. (Shetty et al. 2015, 47.)

Contact Lenses

According to Lim & Lim (2020), the development of contact lens designs and characteristics has created a wider range of contact lens options available for patients with keratoconus or other ectatic diesease. The optimized visual outcomes, patient satisfaction and consolation can be aimed with different diameter RGP lenses, scleral lenses, hybrid lenses, or custom soft lenses. Together with successful CXL treatment that halts KC progression development in contact lens designs and comfort might help reduce the number of keratoplasty procedures needed in the future.

During contact lens practice it is vital to consider various factors when fitting a keratoconic patient. The type of the cornea and severity of the disease determine possibilities in lens selection and with the aid of variable fitting tools almost every keratoconic patient can be successfully provided with proper contact lenses. Lens selection can be eased by lens selection systems (table 3). (Bennet & Henry 2014, 555-556.)

TABLE 3. Keratoconus Contact Lens Selection (Bennet & Henry 2014, 556).

Type of Cone	Recommended Designs
Nipple	Small OAD/OZD keratoconic design
	Custom soft lens design
Oval	Intralimbal GP

	Small OAD/OZD keratoconic (unless large oval)
	Custom soft design
	Hybrid
Globus	Intralimbal GP
	Mini-Scleral
	Hybrid or piggyback
Marginal	Same as Globus

Progression	Recommended designs
Mild	Small OAD/OZD keratoconic design
	Custom soft lens
	Hybrid
Moderate	Intralimbal GP
	Small OAD/OZD keratoconic design (if nipple cone)
	Mini-Scleral (if "1" or "2" decenters/poor comfort)
Severe	Intralimbal GP
	Mini-Scleral (if "1" decenters/poor comfort)
	Piggyback
	Hybrid

OAD = overall diameter, OZD= optical zone diameter

Visual outcomes and patient comfort levels with different lens designs were compared in a study performed by Lim & Lim (2020). The study showed that with new design scleral and hybrid lenses improved patient comfort levels were obtained compared to comfort levels with traditional RGP lenses. Nevertheless, better comfort did not correlate with better VA and therefore both types of lenses can be used to provide satisfactory visual outcomes.

In a comprehensive literature review concerning nonsurgical procedures for KC management by Rico-Del-Viejo et al. (2017), it was summarized that with the aid provided by contact lenses the

finest visual rehabilitation for KC patients may be achieved, patients' quality of life enhanced and the need for surgical procedures can be delayed.

Soft Contact Lenses

According to Downie & Lindsay (2015) there is a drawback in standard soft contact lenses regarding KC visual correction is their limited capacity to regulate irregular astigmatism and secondary higher-order aberrations resulting in a poor level of visual quality. Therefore, soft contact lenses are considered mainly as an alternative with early, mild or form-fruste and subclinical KC. Nevertheless, silicone hydrogel materials with higher Dk and custom design soft KC lenses have resulted in improved lens designs and are considered necessary tools for contact lens practitioners (Bennet & Henry 2014, 548). Although, advancements in soft contact lens manufacturing technologies their capacity to provide satisfactory visual outcomes is not widely acknowledged (Downie & Lindsay 2015), even though according to Lim and Lim (2020) with soft lens designs decent vision is achievable in mild-to-moderate KC cases.

Rigid Gas-Permeable Contact Lenses (RGP)

Rigid contact lenses have been the most common refractive correction method for KC both before and after the availability of gas-permeable materials. The rigid material allows a tear lens to be shaped between the irregular corneal surface and the posterior lens surface resulting in corneal astigmatism neutralization and higher-order aberration neutralization to some degree. (Downie & Lindsay 2015.)

RGP lenses are often divided based on their diversity in total diameter or by their landing zones, although the terminology can vary. Examples of lens RGP designs include corneal (7.0-12.0 mm), Corneo-scleral (12.1-15.0 mm), and Scleral lenses (15.1>18.0 mm). (Downie & Lindsay 2015; Van der Worp 2016.)

Corneal RGP Lenses

Corneal RGP lenses are the most often fitted contact lenses for KC patients. Various traditional corneal RGP lens designs are used with KC although, the fitting process may be more challenging compared to normal cornea when the cone apex and its position affect the centration of the lens. (Bennet & Henry 2014, 532.) The selection of RGP lenses is wide and lens type used depends on the severity of KC. With mild-to-moderate KC mono-curve lenses are often used, while bi-curve

lenses are more repeatedly utilized in severe and advanced KC cases. (Lim & Lim 2020.) Also, various specialized KC designs for more advanced KC fit are available. Typically, these lenses range from conventional KC designs to more recently introduced designs with small diameters, steep base curve radius, and either spherical or aspheric peripheral curves. Intralimbal designs are utilized as well. (Bennet & Henry 2014, 533-535.)

There are no ultimately agreed or specific criteria regarding the interpretation of successfully fitted RGP lens. Therefore, it is difficult to create objective comparisons between different lenses and their fit. Traditionally RGP lens fit is evaluated by fluorescein image, an image created by fluorescein-stained tear film underneath the lens, observed by slit lamp biomicroscope by cobalt blue light. The image reveals the fluorescein pattern and the amount of tear film beneath the lens. To be able to visualize fluorescein, there must be at least a 20-micron tear reservoir between the lens and the corneal surface. A thinner lacrimal layer will appear comparably darker. (Downie & Lindsay 2015.) Three major fitting philosophies for corneal RGP lenses are based on fluorescein patterns: apical bearing, apical clearance and three-point touch. (Bennet & Henry 2014, 532, 533; Downie & Lindsay 2015).

In apical bearing, the contact lens primarily lies on the corneal apex creating a fluorescein pattern with an overshadowed area in the centre of the lens. Apical bearing is created with a flat base curve lens with a larger diameter. It was believed that creating pressure on the corneal apex would halt the KC progression and improve visual quality compared to other fitting philosophies. Today this method is controversial and rarely used because of its potential to induce or exacerbate apical corneal scarring. (Bennet & Henry 2014, 532, 533; Downie & Lindsay 2015)

The characteristic of an apical clearance fit is to create a noticeable clearance vaulting over the corneal apex with a small diameter and steep base curve lens design resulting in an apical clearance fluorescein pattern. Some practitioners recommend apical clearance with lens support on the peripheral parts of the cornea to reduce the possibility of lens-induced apical scarring. (Bennet & Henry 2014, 533; Downie & Lindsay 2015.) However, the potential for short-term corneal re-shaping effects, peripheral corneal interruption and lens adhesive are associated with the method (Downie & Lindsay 2015).

Three-point touch is nowadays the most favoured fitting philosophy where the aim is to create a light feather touch at the corneal apex with spreading most of the lens-bearing pressure into at least

two other areas at the mid-periphery. This creates a bull's eye shaped fluorescein pattern with four zones: light apical touch, paracentral clearance, mid-peripheral bearing and peripheral clearance. The advantage considered with a three-point touch is the minimal force towards the corneal apex unlikely to cause epithelial abrasion. (Bennet & Henry 2014, 533,544; Downie & Lindsay 2015.)

To reduce the challenges often experienced in Corneal RGP lens fitting, Ortiz-Toquero et al. (2021) demonstrated a guideline for the management of KC patients with RGP lenses. The guideline aims to provide an evidence-based strategy to successfully and efficiently fit RGP lenses for a KC patient, help with clinical decision making and provide quality KC care. A successful fit with an acceptable visual outcome also requires a comprehensive eye examination to discover whether the patient can be fitted with contact lenses as well as a broad overall knowledge regarding contact lenses and fitting procedures and patient education. The outlining protocol created for successful RGP lens fitting in patients with KC is presented in figure 8.

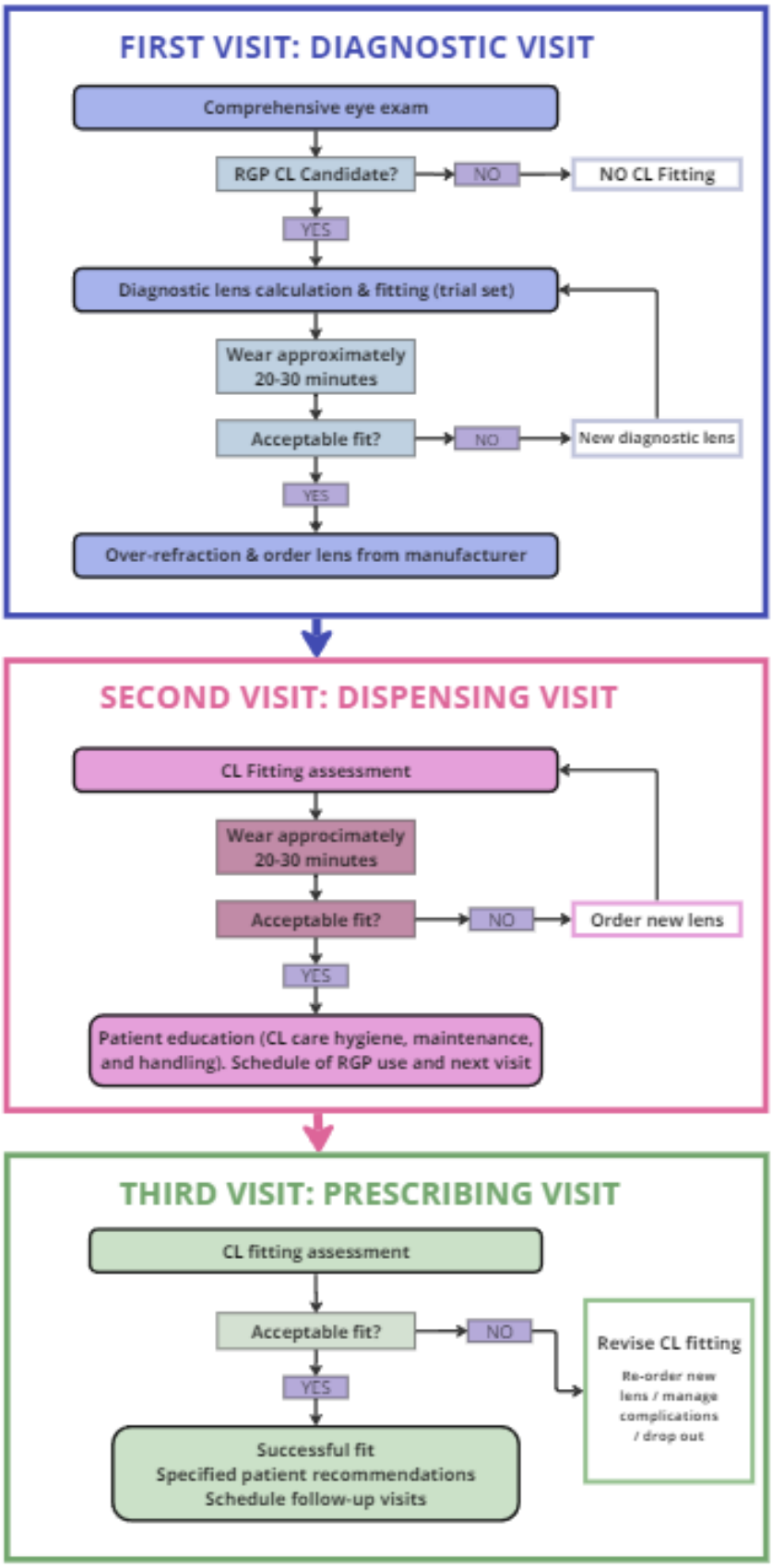

FIGURE 8. Flow chart outlining the protocol for successful rigid gas permeable contact lens fitting in patients with keratoconus (Ortiz-Toquero et al. 2021).

Scleral Contact Lenses

Corneal RGP lens fitting may be demanding in advanced cases of KC as an outcome of lens dislocation, decentration or discomfort. It is thought that large-diameter contact lenses that solely rest outside the cornea are one of the best options for correcting irregular cornea-related vision problems. It seems suggested to avoid any contact between the cornea and lens to maintain true corneal clearance to avoid corneal compromise. The advantage of scleral lenses lies in postponing or even preventing the need for surgical interventions and reducing the risk of corneal scarring. (Lim & Lim 2020; Van der Worp 2016, 1.)

Scleral lenses have become more popular in the past decades with an increased number of contact lens manufacturers and practitioners fitting scleral lenses. A better design in lens manufacturing due to a new understanding towards the corneoscleral junction and anterior scleral shape as well as improved production processes make lenses more easily manufactured with lower costs. New improved lens materials combined have achieved improved ocular health, increased lens wearing hours, and more uncomplicated lens fit. (Van der Worp, 2016.)

Scleral lenses can be further divided into subcategories by identifying their lens resting point on the ocular surface. Corneoscleral lens design allows lens bearing partly on the cornea and partly on the sclera. The lens that rests completely on the sclera is called a scleral lens. The scleral lens group can be further distinguished by the size of the lens, where a lens with a 6mm larger diameter compared to the horizontal visible iris diameter (HVID) is classified as mini-scleral and a lens with more than 6mm larger diameter compared to HVID is classified a large scleral lens. (Van der Worp 2016, 2-3.)

According to Van der Worp (2016), although there is a difference in the size between scleral lens types, they all endorse good apical clearance compared to corneal contact lenses minimizing the mechanical stress focused on the cornea and thereby scleral lenses are considered an extensive advantage. The most major distinction apart from the landing zone between different scleral lenses is the amount of clearance (tear reservoir) that can be produced between the lens's posterior surface and anterior corneal surface.

When selecting a scleral lens, a thorough consideration concerning oxygen transmissibility is critical. The tear reservoir together with scleral lens thickness and material properties affects the final oxygen transmissibility. Only high oxygen-permeable materials are recommended in addition

to a maximum central lens thickness of 250 microns and not greater clearance than 200 microns to avoid hypoxia-induced corneal oedema. (Downie & Lindsay 2015.)

Piggyback

In a piggyback contact lens system, a soft lens, commonly a disposable silicone hydrogel lens, is placed underneath a rigid lens. Piggyback can be practised with both corneal and corneo-scleral lenses and is generally adopted when a rigid lens alone could lead to corneal insult. A soft lens protects the corneal surface from any excessive lens bearing minimizing the potential complications and accomplishing better comfort. A high positive power (> +4.00 D) soft lens can improve rigid lens centration on a keratoconic cornea with an inferiorly placed cone. The soft lens can also be utilized to correct residual astigmatism with a toric soft lens design. To avoid hypoxia to the cornea it is advised to pay attention to both rigid lens and soft lens oxygen transmissibility (Dk/t) that should be greater than 60 Barrer. (Bennet & Henry 2014, 551,552; Downie & Lindsay 2015.)

Hybrid Contact Lenses

Hybrid contact lenses attempt to combine the preferred visual performance properties of a corneal RGP lens with the comfort and stability of a soft lens by incorporating a rigid lens center with a soft periphery. In addition, this one-lens system contributes certain advantages over a piggyback system such as ease of care and handling. (Bennet & Henry 2014, 553.)

2.5.4 Surgical Management

When a patient is no longer satisfied with nonsurgical KC treatment methods surgical methods should be considered. In a patient with contact lens intolerance, significant corneal scarring, alarmingly thin cornea or the KC is regarded as severe and at probable risk of acute hydrops, surgery may be indicated. In addition to CXL, the most frequently used surgical methods include anterior lamellar keratoplasty (ALK) or more specified descemetic deep ALK (dDALK) and penetrating keratoplasty (PK). (Gomes et al. 2015, 368; Santodomingo-Rubido et al. 2022, 17,18.)

Surgically inserted intra-corneal ring segments (ICRS), phakic and pseudophakic intra-ocular lenses (IOL) and topography-guided photorefractive keratectomy (PRK) can be used as a surgical refractive treatment with a non-progressive KC but are considered less commonly compared to

non-surgical treatments or keratoplasty. (Gomes et al. 2015, 368; Santodomingo-Rubido et al. 2022, 17,18).

2.6 Keratoconus Follow-up

With KC patients the follow-up visit periods are planned based on the treatment decision and the severity and/or development of the disease. The measurement of VA, external examination, slit lamp biomicroscopy, and evaluation of corneal structure by topography and tomography should be included in a medical follow-up. (Garcia-Ferrer et al. 2019, 197.)

The recommended medical follow-up intervals vary from 3-6 months up to one year, however, after the increased availability of CXL procedure the 3–6-month interval is recommended to recognize progression and the need for CXL. Contact lens follow-ups should be performed whenever an unsatisfactory vision or lens instability is present. (Garcia-Ferrer et al. 2019, 197.)

After surgical procedures, slit lamp biomicroscopy and corneal thickness measurement should be performed. In addition, corneal pachymetry and corneal tomography may be useful. After long-term post-op use of corticosteroids, regular IOP check-ups are mandatory to exclude corticosteroid-induced IOP elevation. Early signs of optic nerve damage connected to elevated IOP can be detected by utilizing pupil dilation and fundus examination, visual field testing and stereo disc photography or retinal nerve fibre layer OCT measurements. (Garcia-Ferrer et al. 2019, 197,198.)

2.7 Care Provider, Setting and Referral

According to Garcia-Ferrer et al. (2019, 29), KC evaluations may be provided by optometrists or ophthalmologists. KC diagnosis and management demands comprehensive ophthalmic, medical and surgical knowledge and it is considered necessary to closely monitor these patients. Optometrists should instantly refer patients to an ophthalmologist specialised with KC management if visual loss, loss of functional vision, acute hydrops, evolvement of the disease, or onset at young age is present.

2.8 Creating Guidelines

A definition of Clinical Practice Guidelines by the Institute of Medicine is defined as follows: “Clinical Practice Guidelines are statements that include recommendations intended to optimize patient care that are informed by a systematic review of evidence and an assessment of the benefits and harms of alternative care options” (Institute of Medicine 2011).

Clinical guideline implementation recommendations follow a series of steps considered relative to creating trustworthy and evidence-based knowledge. Typically, evidence synthesis is an essential part of the guideline development process consisting of the identification of the purpose, objectives, and scope of the review, defining literature search methods, executing data elicitation and critical appraisal, integrating findings, and completing the quality assessment. The selected working group translates the gathered evidence and creates recommendations. Systematic or non-systematic methods for evidence synthesis may be used. (Lunny et al. 2021.)

The Current Care Guidelines are Finnish national guidelines created to cover essential issues concerning Finnish health, and medical treatment and to halt disease development. The process of creating Current Care guidelines follows a development process where the Current Care Board selects a topic based on a proposal made by a specialist association. A competent professional information specialist organizes a systematic literature search, and the received evidence-based data is produced as a guideline in Current Care working groups in collaboration with Current Care editors. The guideline is circulated to specific interest panels for their review before its completion. (Current Care Guidelines 2022.)

In the capacity of this thesis, a narrower version of guideline creation was utilized. A literature review-based research development project was selected to create a keratoconus guideline for the Finnish Ethical Board of Optometry (OEN). The OEN holds a right to create changes to the content of this thesis to accomplish the guidelines for the use of Finnish optometrists.

2.8.1 Legislation, Regulations, Guides, and Recommendations

The Act on Health Care Professionals (No. 559/1994) § 2 and §15 states that optometrists and opticians are licenced health care professionals and have the goal of maintaining and promoting

health and preventing illnesses. Optometrists and opticians also have the obligation to take part in further professional training to maintain the knowledge and skills required to practice professional activity.

Regarding the optician practice the Health Care Professionals Decree (564/1994 § 16) states that opticians may not independently prescribe spectacles under the following circumstances:

1) to a child under the age of eight years;
2) to a person who has previously had an eye surgery involving the eyeball;
3) to a person suffering from an eye disease; or
4) to a person whose visual acuity cannot be normalised with spectacles.

The Decree also states that contact lenses may be prescribed and fitted by an optometrist or a licenced optician if separate training has been completed and if there is nothing that makes the use of contact lenses unsuitable (Health Care Professionals Decree No. 564/1994 § 16).

The Decree of the Ministry of Social Affairs and Health on prescribing medicine (2.12.2010/1088 § 6) states that opticians can prescribe from pharmacies those in advance-defined medicines that are needed in practice activities. As an addition an optician or optometrist lack the right to prescribe any medications to patients.

The medical products allowed for optometrist or optician to prescribe and use in practice were updated in January 2020 and the approved medications now include oxybuprocaine hydrochloride, fluorescein, fluorescein and oxybuprocaine combination products, tropicamide, cyclopentolate, and phenylephrine (The Decree of the Ministry of Social Affairs and Health on Prescribing Medicine 2.12.2010/1088, Annex 2). For an optician or optometrist to be able to prescribe medication a separate training needs to be completed (The Decree of the Ministry of Social Affairs and Health on Prescribing Medicine 2.12.2010/1088, § 8). Nowadays optometrists graduating with a Bachelor of Health Care (Optometry) degree automatically obtain diagnostic privileges (Näe ry 2019).

According to the Act on the Status and Rights of Patients (785/1992), optometrists as a health care professional are obliged to provide high-class health care and medical care to the patients and the professional status demands adequate information provided to the patient of relevant findings and proper guidance to seek further medical care if indicated.

The Ethical Council of Optometry in Finland has gathered a recommended good practice for eye examination and contact lens fitting protocol for optometrists and opticians. The recommended protocol covers patient history, vision examination, contact lens fitting and health assessment of the eye and has been conducted to ensure proper compliance with health care laws and regulations and ethical principles. The scope and content of the eye examination is determined by the education, experience, and professional consideration of the examiner. (Ethical Council of Optometry 2019.)

In Finland, speciality contact lenses are provided as a medical aid to a patient with diagnosed corneal disease (e.g. corneal ectasia, keratoconus, central corneal scarring, irregular corneal surface due to eye surgery etc.) causing unsatisfactory vision correction with spectacles. Correctly selected medical aid promotes supports and improves patients' everyday performance and prevents from performance collapsing. An ophthalmologist statement of significant vision improvement compared to spectacles is needed for approval. However, speciality contact lenses are not granted if satisfactory visual quality can be attained with traditional soft daily or monthly contact lenses or if corrected VA is 20/25 or better with reasonable spectacles. (Sosiaali- ja terveysministeriö 2023, 199.)

Whenever possible, it is recommended to incorporate the speciality contact lens practice of medical aid to a specialist optometrist with appropriate skills, experience, and knowledge in advanced contact lens fitting. Contact lens care and insert-related products are included in the initial fitting procedure, and the patient is responsible for following care product acquisition. Contact lenses are renewed by following manufacturer instructions. (Sosiaali- ja terveysministeriö 2023, 199-200.)

These Finnish laws, regulations and instructions set the framework for optometrists' practice in Finland and need to be taken into account when implementing new guidelines.

3 THE PURPOSE, OBJECTIVES AND TASKS OF THE THESIS

3.1 Purpose and Objective

The purpose of this study by the literature review is to determine and synthesize appropriate information concerning KC assessment and management. The aim of the literature review is to produce knowledge of the current status of KC assessment and management to be used in the development phase of the study.

The purpose of the research development phase is to create a clinical guideline concerning KC assessment and management for the use of optometrists in Finland. The aim of the research development phase is to select the relevant methods concerning KC assessment and management for the optometrist practice in Finland. The guidelines should support the utilization of Finnish optometrists as a part of efficient eye health care.

The study objective of the review was to raise awareness of KC as a disease and to determine KC-related assessment and management procedures concerning optometry practice within the boundaries set by Finnish laws, regulations, and recommendations.

3.2 Statement of the Research Question

Based on the literature review purpose and the study objective the following study question was created:

1. How KC patients should be assessed and managed?

Based on the research development purpose and the study objective the following study question was created:

2. What should be included in an assessment and management of KC patients performed by optometrists in Finland to be a part of efficient eye health care in Finland?

4 IMPLEMENTATION OF THE THESIS

The implementation of the thesis included an information search process, defining the key concepts for the KC assessment and management, defining the literature search and selection for the literature review and literature review data analysis followed by the research development part. The thesis report was written between March 2023 and October 2023.

4.1 Literature Review

A Literature review is a review and summary of what is known and unknown regarding the research topic along with discovering the current research place within the existing knowledge. The purpose of a literature review is to recognize where the research stands in the larger educational conversation, help the researcher to accompany the conversation by providing background information, informing methodology, revealing innovation, minimizing duplicative research, and assuring that professional standards are met. An understanding of the current literature is important for all phases of a research study. (Maggio et al., 2016.) A Literature review tries to provide an analysis of today's literature covering a wide scope of material with integrity magnitude based on analyses of literature (Grant & Booth, 2009). Literature reviews can be systematic, integrative or narrative (R. Ferrari, 2015).

4.1.1 Integrative Literature Review as Research Method

The integrative review aims to assess, judge, and arrange the literature so that new theoretical structures and aspects may develop. An overview of the knowledge foundation to critically review and broaden the theoretical base of the topic can be considered the main purpose of integrative review. When a study has a broader research question and reviewing every single relevant article is not possible integrative review seeks to combine context and understanding from different areas of research and results in the advancement of knowledge and theoretical frameworks. Even though integrative review can be administered in several ways, the study is required to follow accepted formalities for reporting article selection and integrative transparency. (Snyder 2019, 334-336.)

The integrative review supports variable methods including experimental and non-experimental studies, resulting in a comprehensive way to incorporate data from theoretical and empirical literature. The integrative literature review process consists of six phases. In the first phase, the research question is determined. Determination of the research question is important because it determines the studies included, factors selected for the identification and information gathered in the selected studies. In the second phase, the literature is searched and selected. Databases for the search are carefully selected, and the inclusion and exclusion criteria must be clearly demonstrated. Data collection as the third phase is carefully operated by following the selected criteria limitations created in the previous phases to collect all the relevant data and to decrease the risk of errors. In the fourth phase, the selected data should be critically analysed by confirming the validity of the methods and results. Discussion of the results consists of the data analysis interpretation, comparing the data with the theoretical references and synthesis of the results. As the sixth and last phase of the study is to present the literature review clearly and completely by conducting the review within the methodological accuracy standards. (Souza et al. 2010.)

This study was carried out as an integrative literature review due to the vast scope of the study subject. This literature review-based research development had two phases, where the first phase was the concept analysis of existing KC guidelines followed by an integrative literature review based on the guideline concept analysis. In the second phase, the collected data was organized, thoroughly evaluated, and analysed. The literature review aimed to follow the integrative literature review process presented by Souza et al. (2010). The analysis phase of the study was blended with the research development phase of the study which consisted of bringing all the research data together to produce a guideline for keratoconus patient assessment and management to be used by optometrists.

The Preferred Reporting Items for Systematic Reviews and Meta-analyses (PRISMA 2020) Flow Diagram, borrowed from the systematic review process, was used in this review to demonstrate the different phases of the literature search. The flowchart maps out the number of records identified, included, and excluded and the reasons for exclusions. (Page et al. 2021.)

4.2 Data Search Process and Selection

4.2.1 Literature Search

The primary literature search process was conducted in January 2023 in cooperation with an information specialist from Oulu University Library. According to the subject of interest, the most suitable and comprehensive databases were selected with aid and instructions from the information specialist.

The search was performed by EBSCOhost web source by using Academic Search Premiere, CINAHL and MEDLINE databases. Different appropriate search terms and search operators were considered and finally, the search was performed with the terms `Keratoconus N3 (assessment OR diagnostic*)´, `Keratoconus N3 management´, `Keratoconus AND (guideline* OR "practice pattern*")´ recommended by the information specialist. The search was limited to articles written in English language and since KC knowledge and assessment technologies have improved rapidly during the past decades, texts not older than 10 years were accepted. The article type could not be chosen.

4.2.2 Literature Selection and Evaluation

From the total number of six hundred-ten search results (n=610), one hundred and sixty-nine duplicates (n=169) were removed automatically by a filter applied by the search engine. From the remaining (n=441) texts, their titles, abstracts, and relevance to the study were assessed by one author and further exclusion was made resulting in twenty-four (n=24) articles for eligibility assessment. Articles that did not have full-text access from the EBSCOhost web source were searched again from other databases and all the remaining 24 texts were attained in full text and viewed. Four articles were excluded due to insufficient methodology description, although one exception was made in the exclusion process. One literature review following the literature review process with comprehensive and relevant content and published in a peer-reviewed journal was still evaluated and included regardless of insufficient methodology description. In addition, one editorial text was excluded at this stage. Literature selection resulted in eighteen (n=18) articles and one guideline (n=1) included in the review. All the selected articles were confirmed to be peer-reviewed. The selected articles were saved to the EBSCOhost web server in separate folders to

keep the records in order and saved. From the folder, the selected articles could be easily found when needed for further inspection and analysis. The selection process of included articles is presented in figure 9.

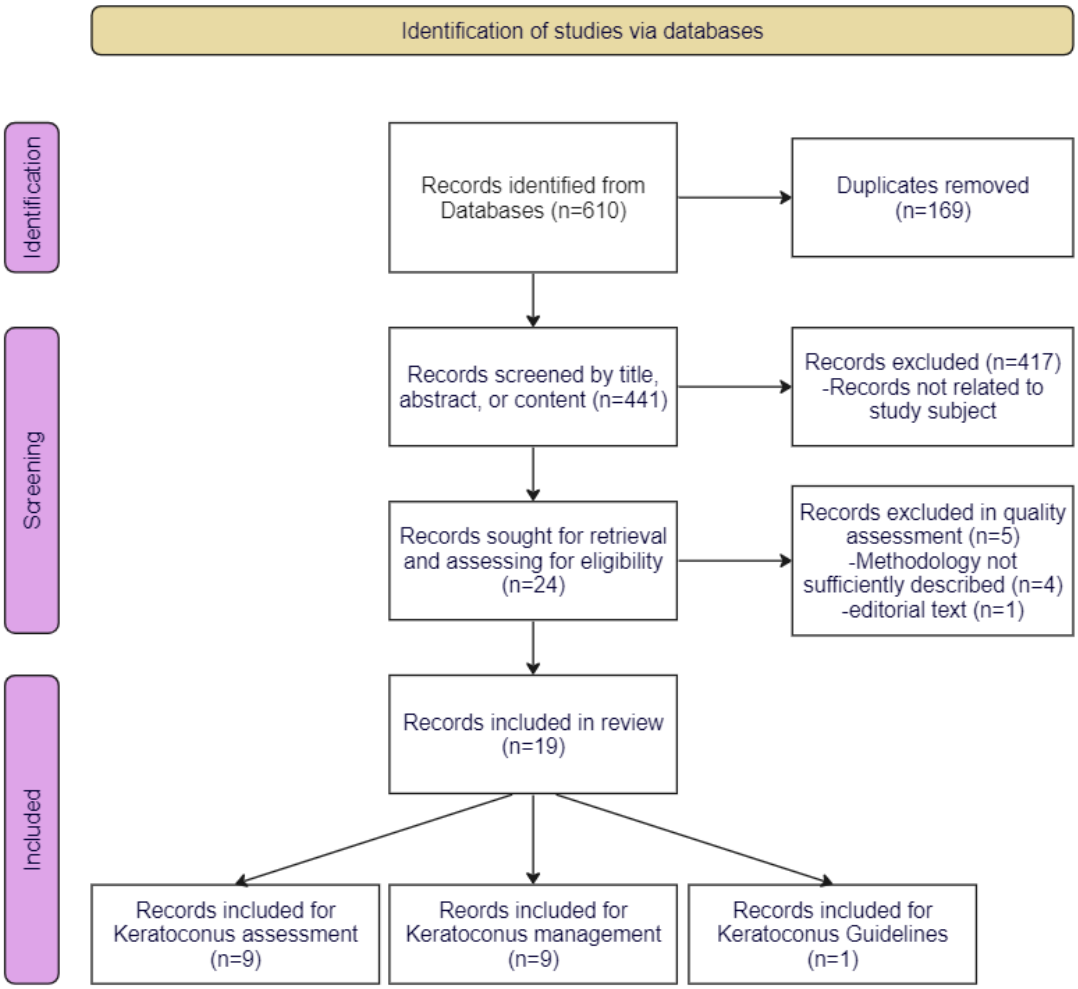

FIGURE 9. PRISMA 2020 (Page et al. 2021) *flow chart of the included article selection.*

Keratoconus guideline evaluation

The clinical guideline detected was selected as a baseline guide concerning KC assessment and management. The guideline was thoroughly read, and the content was analysed. Based on the analysis of the content, key concepts for KC assessment and management were selected. The determination of key concepts is more thoroughly presented in chapter 5.

Literature evaluation

The rest of the texts selected for the literature review for further analysis were read and analysed. Similarities linked to key concepts were searched. Elements repeatedly appearing in the material

were soon identified and selected as relevant sub-elements in KC assessment and management. The determination of sub-elements is more thoroughly presented in chapter 5.

4.2.3 Clinical Guideline as Research Development Phase

The guideline for the use of Finnish optometrists was developed to create a systematic approach to follow while assessing and managing KC patients. The guideline has its foundation built upon the concept analysis of the one international guideline resulted in key concepts, evidence-based literature found in the literature review and the sub-elements recognized from the literature analysis. Key concepts and sub elements are more thoroughly introduced in chapter 5. National recommendations, Finnish laws and regulations concerning optometry practice were taken into consideration when creating the content for the guideline.

5 RESULTS

In this chapter, the literature selection and evaluation results are presented. The results of the literature review are utilized in the guideline development process.

5.1 Determination of the Key Concepts of Keratoconus Assessment and Management

Currently, there are no corneal ectasia or KC-related practice guidelines or recommendations for care in Finland for the use of optometrists or ophthalmologists. After the literature search, one international practice pattern concerning KC was found (table 4).

TABLE 4. Keratoconus guideline identified in the literature search

Guideline	Author	Publisher	Year of Publication
Corneal Ectasia Preferred Practice Pattern®	Garcia-Ferrer et al. (Corneal/External Disease Preferred Practice Pattern panel members consisting of nine MDs and one methodologist)	American Academy of Ophthalmology	2019

Optometry practice in the US is considered to represent the highest competency in the field of optometry in the world (Efron, 2022), and therefore it is justified to assume that Corneal Ectasia Preferred Practice Pattern presents the current view of KC assessment and management. The guideline was read and analysed and as a result, key concepts for KC assessment and management were identified (figure 10) and given due consideration selected as a baseline for components for this guideline. The content of the key concepts is more thoroughly introduced in the theoretical background.

A Swedish Clinical Guideline for Optometrists concerning KC assessment and management (Westerlund & Robertson, 2020) was found in an additional data search at the end of August 2023. After thorough consideration, this guideline was excluded from key concept analysis due to a lack of methodology although the guideline content had numerous similarities compared to the previously introduced Corneal Ectasia Preferred Practice Pattern (Garcia-Ferrer et al. 2019) published by the American Academy of Ophthalmology.

One published article concerning KC global consensus (Gomes et al., 2015) was also identified but not selected for the KC key concept analysis because it was not published as a guideline. Nevertheless, the article was included in the literature review.

5.2 Literature review analysis: Determining Sub-elements of Keratoconus Assessment and Management

Literature search and selection resulted in one guideline and eighteen records for a further literature review. The selected articles and their main content or results are presented in table 5.

TABLE 5. Articles selected for the literature review.

Author, Year, Country	Article/Publication	Journal	Main content or results
Koc et al. / 2020 / Turkey	Topometric and Tomographic Evaluation of Subclinical Keratoconus	Ophthalmic Epidemiology, Vol. 27(4)	A retrospective cohort study of topometric and tomographic imaging methods in KC assessment resulting in earlier diagnosis.
Gideon Abou Said et al. / 2023 / Israel	Revisiting the oil droplet sign in keratoconus: Utility for early keratoconus diagnosis and screening	Ophthalmic and Physiological Optics, Vol. 43(1)	Prospective study. Recognising oil droplet sign as a method for KC recognition and diagnosis.

Castro-Luna & Pérez-Rueda / 2020 / Spain	A predictive model for early diagnosis of Keratoconus	BMC Ophthalmology, Vol. 20(1)	Cross-sectional study discussing topographic, pachymetric and aberrometric characteristics in patients in KC assessment and diagnosis.
Davey et al. / 2013 / USA	Diagnostic accuracy of keratoconus using anterior segment optical coherence tomography	Optometry Reports, Vol. 3(1)	Identifying keratoconic eyes from healthy eyes with anterior segment OCT. OCT alone gives 80-90% accuracy in moderate to advanced KC but a lot less for early KC.
Kreps et al. / 2021 / Belgium	Diagnostic patterns in Keratoconus	Contact Lens and Anterior Eye, Vol. 44(3)	Retrospective patient chart review investigates current patterns of diagnosis and referral in KC, revealing KC stage 2 or higher at the time of diagnosis emphasizing the importance of early diagnosis.
Matalia & Swarup / 2013 / India	Imaging modalities in keratoconus	Indian Journal of Ophthalmology, Vol. 61(8)	A review of Topography and tomography in KC diagnosis.
Santodomingo-Rubido et al. / 2022 / published in the UK	Keratoconus: An updated review	Contact Lens and Anterior Eye	A review of KC epidemiology, pathology, diagnosis and management.
Belin & Duncan / 2016 / US	Keratoconus: The ABCD Grading system	Klinische Monatsblatter Fur	Normative data re-analysing study to create

		Augenheilkunde, Vol. 233(6)	a KC classification/staging system to reflect the anatomical changes seen in KC.
Martínez-Abad & Piñero / 2017 / Spain	New perspectives on the detection and progression of keratoconus	Journal of Cataract and Refractive Surgery, 43(9)	A review presenting corneal topography as the primary diagnostic tool for KC. Pachymetry, aberrometry and tomography make the diagnosis more accurate and enable earlier diagnoses.
Gănescu / 2022 / Romania	Current approaches in the management of patients with keratoconus	Medicine and Pharmacy Reports, Vol. 95(4)	Review of KC diagnostic and management methods.
Ortiz-Toquero & Martin / 2017 / UK and Spain	Current optometric practices and attitudes in keratoconus patient management	Contact Lens and Anterior Eye, Vol. 40(4)	Compares optometric practices and attitudes in the UK and Spain.
Shetty et al. / 2015 / India	Current review and a simplified “five-point management algorithm” for keratoconus	Indian Journal of Ophthalmology, Vol. 63(1),	A review of KC, KC assessment and management by a five-point algorithm was developed.
Song et al. / 2022 / Global	Diagnosis and Management of Keratoconus- A Narrative Review of Clinicians’ Perspectives	Children (Basel, Switzerland), Vol. 9(12).	Review of current practices, attitudes and beliefs in KC assessment and management in adults and children. Attempts to specify the

			differences on a global scale.
Gomes et al. / 2015 / Global	Global consensus on keratoconus and ectatic diseases	Cornea, Vol. 34(4)	Project to create global consensus on KC diagnosis and management. Focuses on definition, concepts, clinical management, and surgical treatments.
Andreanos et al. / 2017 / Greece	Keratoconus treatment algorithm	Ophthalmology and Therapy, Vol. 6(2)	Review that summarizes KC treatment possibilities and. Presenting a treatment algorithm.
Rico-Del-Viejo et al. / 2017 / Spain	Nonsurgical Procedures for Keratoconus Management	Journal of Ophthalmology, 2017	Review discussing different contact lens management options for KC.
Hodge et al. / 2015 / Australia	Therapeutic treatment of keratoconus: A survey of local optometric practice criteria	Clinical and Experimental Optometry, Vol. 98(4)	Survey concerning optometrists' practice patterns and referral criteria regarding KC patients within Australia.
Mohammadpour et al. / 2018 / Iran	Updates on Management for Keratoconus	Journal of Current Ophthalmology, Vol. 30(2)	Review of KC management options.

The selected data was divided into KC assessment, KC management and clinical guidelines, but some of the selected articles discussed comprehensive of all of the assessment and management topics. Based on the literature KC assessment was divided into patient history, visual function assessment, visual assessment of different signs and anatomical alterations and diagnostic tests. Based on the literature review the KC management was revealed to consist of four main entities;

KC prevention and early detection, optical management, surgical management, follow up and referral.

Patient history

Patient history including the onset and course of the disease, vision history, ocular history including contact lens and surgical history, medical history and family history are all named and separately divided in the guideline by Garcia- Ferrer et al. (2019) as important part of the patient history. The review articles presenting KC aetiology (Gomes et al. 2015; Romero-Jiménez et al. 2010; Santodomingo-Rubido et al. 2022) also discuss the related biochemical, biomechanical environmental factors and linked conditions but do not separately refer to the importance of taking a thorough patient history separately, although are all highly relevant from the aspect of the patient history.

Visual function, external examination, and corneal slit lamp biomicroscopy

According to Garcia-Ferrer et al. (2019), Gomes et al. (2015) and Santodomingo-Rubido et al. (2022), the visual function assessment should be performed on every KC and KC suspect patient. The level of astigmatism and the status of BCVA reveal important aspects and indicate the later need for management. The visual assessment by different slit lamp tests findings and external findings were discussed in several different texts (Garcia-Ferrer et al. 2019; Oyeniran & Tauqeer 2021; Romero-Jiménez et al. 2010; Santodomingo-Rubido et al. 2022; Shetty et al. 2015) and the most often appeared clinical signs were Scissors reflex, Munson's sign, Rizutti's sign, corneal thinning, corneal protrusion, Fleischer's ring, prominent corneal nerves, Vogt's striae and corneal scarring.

According to Garcia-Ferrer et al. (2019) during a KC patient or a KC suspect patient assessment, a comprehensive fundus examination should be performed along with IOP measurements with equipment less reliant on the sleek corneal surface.

Diagnostic tests

The studies of Koc et al. (2020), Matalia & Swarup (2013), Martinez-Abad & Piñero (2017) and Castro-Luna & Pérez-Rueda (2020) all discuss the meaning of topography and tomography in the diagnosis process of KC stating topography being the primary diagnostic tool in KC. Koc et al. and Castro-Luna & Pérez-Rueda both end up with the same conclusion of the importance of the topographic and tomographic measurements in the early diagnosis process. In addition, Castro-

Luna & Pérez-Rueda also mention the central corneal thickness combined with vertical asymmetry as one of the leading signs in KC when measured with topography. Gideon Abou Said et al. (2023) presented the oil droplet sign as a method for KC detection when no topography or tomography was available. Kreps et al. (2021) also highlight the importance of early detection in their study where the unfortunate fact of KC detection rate after stage 2 progression or later was revealed resulting in a greater amount of visual impairment and higher economic burden with more management procedures needed in the future. Davey et al. (2013) introduced anterior segment OCT to the variety of diagnostic imaging methods emphasizing the importance of combined imaging in the early diagnosis process.

Most of the review articles agreed with the statement of topography as the primary diagnostic method for KC (Andreanos et al. 2017; Gănescu 2022; Garcia-Ferrer et al. 2019; Gomes et al. 2015; Oyeniran & Tauqeer 2021; Romero-Jiménez et al. 2010; Santodomingo-Rubido et al. 2022; Song et al. 2022), although there is no true universally accepted consensus of the consistent diagnostic criteria or progression definition criteria for KC (Gomes et al. 2015, Song et al. 2022). However, Andreanos et al. (2017), Gomes et al. (2015) and Santodomingo-Rubido et al. (2022) presented an agreed protocol for KC progression definition where at least two for the following must be present: steepening of the anterior corneal surface, steepening of the posterior corneal surface and/or thinning or changes in the pachymetry determine a difference. These changes are required to document progression, although, the amplitude of the changes is not currently agreed.

Variable grading systems for KC have been presented over the years and one of the latest developments has been a system presented by Belin & Duncan (2016), that combines the anatomical changes seen in KC with anterior and posterior keratometry values, central corneal thickness and presence or absence of corneal scarring.

Prevention, early detection and halting the progression of KC

The main principles in KC management are to prevent visual loss, cut down the signs and symptoms of KC and endure, alleviate or improve visual function based on the patient's demand. KC severity and progression define the need for management and treatment of the disease and the correct methods are selected according to the thorough assessment. (Garcia-Ferrer et al. 2019; Santodomingo-Rubido et al. 2022.)

Early detection is mentioned by Garcia-Ferrer et al. (2019) and Kreps et al. (2021) as an important aspect of KC management. KC progression and progression-related visual loss are known to affect life quality and the economic burden induced by caring for KC patients throughout their life is considered a health problem. Therefore, for any young patient with changing refractive error, KC suspicion should always be ruled out by a careful evaluation and follow-up.

CXL is considered as the primary method for halting the KC progression. Several of the selected studies (Andreanos et al. 2017; Gănescu 2022; Garcia-Ferrer et al. 2019; Gomes et al. 2015; Mohammadpour et al. 2018; Santodomingo-Rubido et al. 2022; Shetty et al. 2015; Song et al. 2022) state, that as a rule, anyone with progressive KC with clear cornea and minimal corneal thickness of 400 microns should undergo CXL. The type of CXL procedure is chosen individually based on the severity and state of the disease.

After KC diagnosis verbal guidance to the patient is considered as one of the most important measures in non-surgical KC management according to the studies by Andreanos et al. (2017), Garcia-Ferrer et al. (2019), Gomes et al. (2015) and Santodomingo-Rubido et al. (2022). They all state that patients with KC have a variety of non-surgical and surgical treatment options available and need to be informed of the possibilities. The benefits and potential risks associated with early CXL should be discussed as well as optical and other surgical management possibilities. To halt the disease progression, it is advised to avoid eye rubbing, use topical antiallergic medications in patients with allergies and use topical lubricants in case of ocular irritation to reduce the stimulus to eye rubbing. Proper patient education and counselling aid the patient in controlling persistent habits of unnatural eye rubbing.

Optical management

Optical management of KC can be mainly divided into spectacles and contact lenses although most of the studies (Andreanos et al. 2017; Gănescu 2022; Garcia-Ferrer et al. 2019; Gomes et al. 2015; Hodge et al. 2015; Mohammadpour et al. 2018; Ortiz-Toquero & Martin 2017; Rico-Del-Viejo et al. 2017; Shetty et al. 2015; Song et al. 2022) state that contact lenses are considered as the main method for KC vision correction. Spectacles can be used as long as the visual performance is considered adequate and satisfactory VA can be achieved. The vast diversity of contact lens options available (soft lenses, corneal RGP lenses, scleral lenses, piggyback lenses, hybrid lenses) are presented in these several studies and the type of contact lenses prescribed varies depending on the KC state. Rico-Del-Viejo et al. (2017) discuss separately soft and different RGP contact

lenses and point out that soft lenses can be used in low levels of astigmatism, but due to the incomplete ability to correct corneal irregularities, RGP lenses are more frequently used. Scleral lenses can provide ocular surface protection, vision improvement, comfort and in advanced irregularities improved centration.

Clinicians' perspectives in contact lens fittings are discussed in the studies by Ortiz-Toquero & Martin (2017), Hodge et al. (2015) and Song et al. (2022) where they acknowledged that RGP and speciality lenses are more difficult to fit compared to regular soft contact lenses (CLs). Successful RGP fittings require special training, a considerable amount of experience and an understanding of the KC visual system and irregularities. Song et al (2022) found that referrals from optometrists to optometrists specialized in contact lens are often made due to RGP fitting difficulties and lack of training. In the survey conducted by Hodge et al. (2015) concerning optometrists practice patterns and referral criteria regarding KC patients within Australia, it was revealed that soft CLs are prescribed daily by 35,4% of the respondents as RGP lenses were prescribed daily only by 9,2% revealing difference in prescribing patterns. The most often experienced difficulties in RGP fittings were lack of experience in fitting procedures and the time required for quality fitting. Also, low market demand was considered a barrier to practice. Topometric devices could provide aid in the CL fittings as the study revealed that RGP fitting rates increased in a practice setting with a topographer available.

Surgical management

Surgical KC management is discussed by Andreanos et al. (2017), Gănescu (2022), Garcia-Ferrer et al. (2019), Gomes et al. (2015), Mohammadpour et al. (2018), Santodomingo-Rubido et al. (2022), Shetty et al. (2015) and Song et al. (2022) and in these studies, it is noted as the management method of KC when a patient is no longer satisfied with nonsurgical KC treatment methods. In a patient with contact lens intolerance, significant corneal scarring, alarmingly thin cornea or the KC is regarded as dangerous and at a potential risk of acute hydrops, surgery may be indicated. In addition to CXL, the most frequently used surgical methods include anterior lamellar keratoplasty (ALK) or more specified descemetic deep ALK (dDALK) and penetrating keratoplasty (PK), although the demand for keratoplasties has decreased after CXL has become more frequent.

Surgically inserted Intra-corneal ring segments (ICRS), phakic and pseudophakic intra-ocular lenses (IOL) and topography-guided photorefractive keratectomy (PRK) can be used as a surgical

refractive treatment with a non-progressive KC but are considered less commonly compared to non-surgical treatments or keratoplasty (Gomes et al. 2015; Santodomingo-Rubido et al. 2022).

Follow-up and referral

Follow-up of KC patients was less frequently discussed in the review articles. According to Garcia-Ferrer et al. (2019), the follow-up visit periods are planned based on the treatment decision and the severity and/or development of the disease. The measurement of VA, external examination, slit lamp biomicroscopy, and evaluation of corneal structure by topography and tomography should be included in a medical follow-up. The recommended medical follow-up intervals vary from 3-6 months up to one year, however, after the availability of CXL the 3–6-month interval is recommended to recognize progression and the need for CXL. Contact lens follow-ups should be performed whenever an unsatisfactory vision or lens instability is present.

Garcia-Ferrer et al. (2019) also point out that after surgical procedures, slit lamp biomicroscopy and corneal thickness measurement should be performed. In addition, corneal tomography may be a useful tool in follow up assessment. After long-term post-op use of corticosteroids, regular IOP check-ups are mandatory to exclude corticosteroid induced IOP elevation. Early signs of optic nerve damage connected to elevated IOP can be detected by utilizing pupil dilation and fundus examination, visual field testing and stereo disc photography or retinal nerve fibre layer OCT measurements.

Referral of KC patients was discussed with Ortiz-Toquero & Martin (2017), Hodge et al. (2015), Garcia-Ferrer et al. (2019) and Song et al. (2022) revealing different referral patterns between practitioners. In the UK and in Spain according to Ortiz-Toquero & Martin (2017), approximately 50% of optometrists made a referral to an ophthalmologist at the initial diagnosis. According to Hodge et al. (2015), referrals from optometrists to ophthalmologists in Australia at the initial diagnosis were made only by 7,4%, but 34,4% were referred when disease progression was detected and 69,2% when possible surgery was considered as a management possibility. Nevertheless, according to Garcia Ferrer et al. (2019), a conclusion is that KC evaluations made by optometrists or by other specialists than ophthalmologists, vision care providers should closely monitor patients and instantly refer them to an ophthalmologist specialized in KC management if visual loss, loss of functional vision, acute hydrops, progression of the disease or onset at a young age is present.

Sub-element establishment

The repeatedly appearing topics from the literature analysis were selected as sub-elements of KC assessment and management and are presented in figure 10 along with the key concept presentation.

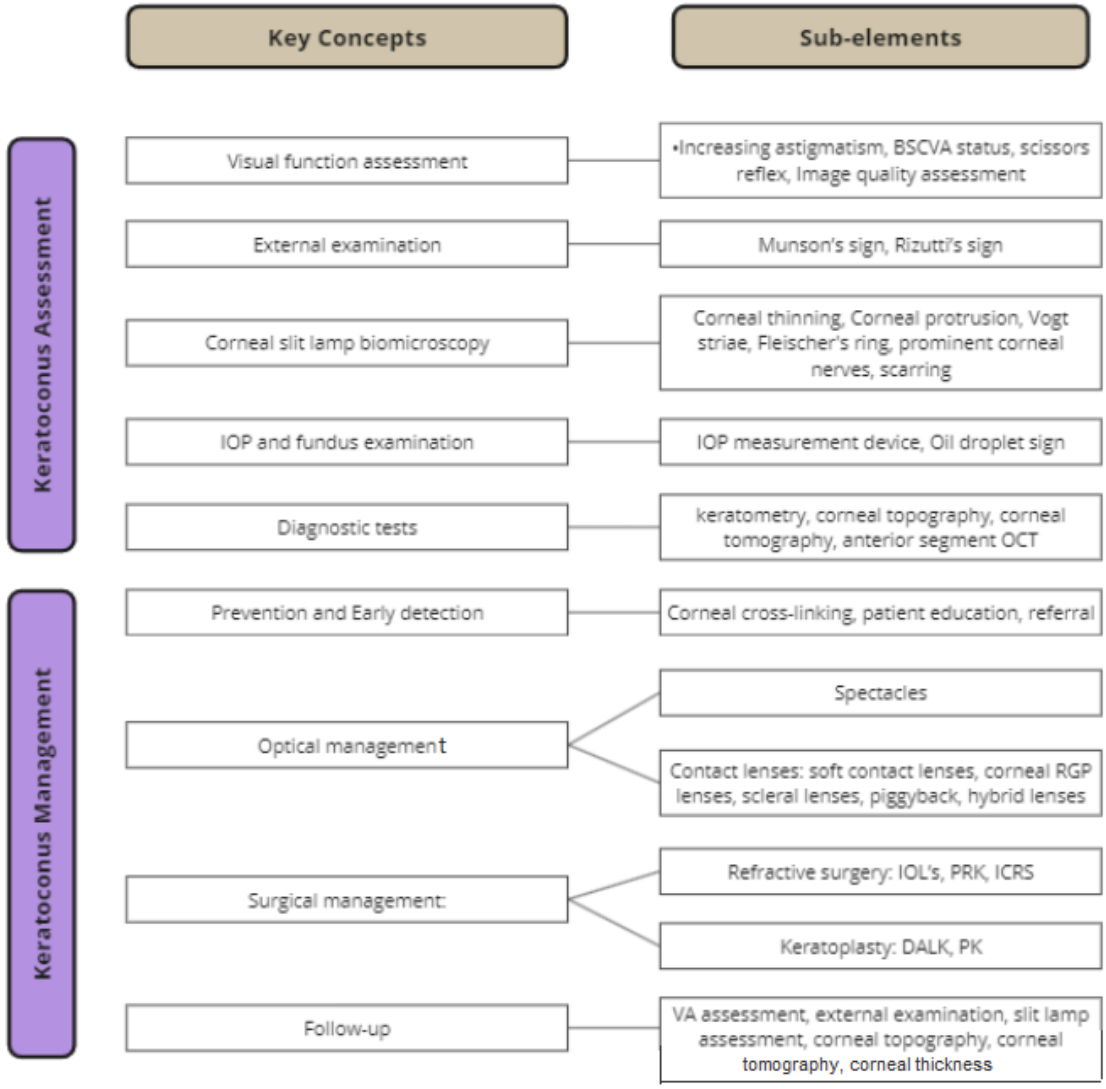

FIGURE 10. Key concepts and sub-elements of keratoconus assessment and management identified by the literature review.

The content of the sub-elements is more thoroughly introduced in the theoretical background.

5.3 Establishing Keratoconus Assessment and Management Guideline

Optometry practice facilities in Finland vary depending on the site of the practice. Hospital-based optometry practice equipment might considerably differ from equipment available in optical store-based practice. In this guideline, all the procedures or diagnostic tests recommended should be performed whenever possible. All the chosen and recommended procedures and tests are explained and justified in the theoretical background and literature analysis. All the recommended procedures should be performed by the guidance of the recommended good practice for eye examination and contact lens fitting protocol for optometrists and opticians gathered by The Ethical Council of Optometry in Finland. The basic and necessary optometry tests are assumed to be done routinely according to the good practice for eye examination and therefore this guideline focuses on pointing out additional tests or where to pay attention during the tests or procedures. All tests and techniques presented in this guideline are assumed as a part of bachelor education and no additional or detailed instructions are given.

A thorough medical and patient history is an important phase of patient care and all KC assessment and management, and necessary test selection should be done following information available from the patient and patient history. The scope and content of the assessment shall correlate to optometrists' personal level of education and professional experience and knowledge. Due to the legislative restrictions of optometrists' practice concerning patients with ocular disease or suspected ocular disease, all action should be in close cooperation with ophthalmologists.

Surgical evaluation and management of KC as well as disease follow up are considered a part of ophthalmologist expertise and have been excluded from this guideline, although patient education regarding surgical possibilities is still considered a component of optometry practice. Monitoring disease progression is considered possible by optometrists only with an approval from the ophthalmologist when close cooperation is available. The recommendations for optometry practice guidelines concerning KC assessment and management are presented in the following chapter.

5.3.1 Assessment and Early Detection

Early detection of KC is crucial for managing the condition effectively. Optometrists should be vigilant in recognizing early signs and symptoms of KCs during routine eye examinations, such as corneal thinning, irregular astigmatism, and changes in VA.

Following procedures or detecting of following signs are recommended:

- Visual Function Assessment:
    - Best spectacle-corrected VA by subjective refraction
    - Astigmatism evaluation
    - Pinhole VA to determine optical quality
    - Retinoscopy to detect scissors reflex
- External examination:
    - Munson's sign
    - Rizutti's sign
- Anterior segment and corneal slit lamp biomicroscopy, signs to look for:
    - corneal thinning
    - corneal protrusion
    - Fleischer's ring
    - prominent corneal nerves
    - Vogt striae
    - scarring
- IOP and posterior segment examination:
    - IOP measurements with applanation tonometer are not recommended. A rebound tonometer such as iCare is preferred.
    - Oil droplet sign in red reflex can be used to identify KC if no topography or tomography devices are available.
- Diagnostic tests should be performed when available, and interpretation in co-operation with an ophthalmologist when needed:
    - keratometry
    - corneal topography
    - corneal tomography
    - anterior segment OCT

5.3.2 Referral to ophthalmologist and collaborative care

If KC is suspected or confirmed, referring the patient to an ophthalmologist or a corneal specialist is essential. These specialist conducts further diagnostic test, including corneal topography, tomography and pachymetry (if not tested previously) to determine the severity and progression of the disease and to select appropriate management.

Optometrists should work closely with ophthalmologists and other eye care professionals to provide comprehensive care for KC patients. KC management demands comprehensive ophthalmic, medical and surgical knowledge. KC evaluations made by optometrists or by other specialists than an ophthalmologist, vision care providers should closely monitor patients and instantly refer them to an ophthalmologist with specialized in KC management if the following signs appear:

- Visual loss
- Loss of functional vision
- Acute hydrops
- Progression of the disease: steepening of the anterior corneal surface, steepening of the posterior corneal surface and/or thinning or changes in the pachymetry
- Onset at a young age

Collaborative management may involve monitoring disease progression and addressing associated conditions.

5.3.3 Optical management

If unsatisfactory VA is not achieved by spectacle correction, speciality contact lenses are provided as a medical aid to a patient with diagnosed KC by an ophthalmologist statement. Contact lenses, particularly rigid gas permeable (RGP) lenses or scleral lenses, are often the primary method for correcting vision in KC patients. Optometrists specializing in KC should have expertise in fitting and managing speciality contact lenses to optimize VA and patient comfort. Contact lens selection is done based on the stage of the disease and corneal evaluation.

Contact lens fitting procedures for KC patients should follow the recommended good practice protocol created by the Ethical Council of Optometry in Finland and the fitting guides provided by contact lens manufacturers. Approved diagnostic medications can be utilized in contact lens fitting.

Specialty contact lens selection for keratoconic cornea include:

- Soft contact lenses
- Corneal RGP lenses
- Scleral lenses
- Piggyback lenses
- Hybrid lenses

Contact lens follow-ups should be performed according to the good contact lens protocol and the interval determined by the selected contact lens type, material, and state of the disease. Often follow-up intervals are recommended yearly or more frequently if indicated.

5.3.4 Patient education and support

KC can be a life-long condition, and patients may require ongoing care and support. Optometrists should educate patients about the nature of the disease, its potential progression, and available treatment options. They should also provide guidance on proper contact lens care, regular follow-up appointments, and lifestyle modifications to minimize disease progression.

Patient education should include:

- Disease explanation
- Explanation concerning signs of progression
- Knowledge of treatment options:
    - optical: contact lenses and spectacles (optical surgery: ICRS or IOLs)
    - surgical: CXL, keratoplasty
- Verbal guidance to avoid eye-rubbing
- Verbal guidance of the importance of allergy medication if allergy is present
- Follow-ups: Annually, or every 3-6 months to look for progression

6 DISCUSSION

Currently there are no corneal ectasia or KC related practice guidelines or recommendations for care in Finland for the use of optometrists nor ophthalmologists. Based on the literature review results during this research development it became obvious that optometrists have a role in KC assessment and management and therefore guidelines regarding KC assessment and management are necessary. By following these guidelines optometrists are able to provide proper eye health care concerning KC and equal quality in KC assessment and management can be ensured for all patients.

The literature search determined KC as a multifactorial corneal disorder leading to progressive stromal thinning, fracture of the anterior limiting membrane, and protruding of the central or paracentral cornea, creating a cone-shaped appearance. Progression of KC often results in myopia progression, irregular astigmatism and finally reduced visual acuity (VA) and visual impairment.

According to the observations made during this research early diagnosis is essential to halt the KC disease progression and to minimise visual impairment. Although diagnostic tests and devices have improved the rate of KC detection, the disease is still often diagnosed at an unfortunately late stage (Kreps et al. 2021). Optometrists' ability to recognize the early signs and symptoms of KC during an eye assessment as well as a referral of the patient to an ophthalmologist is crucial in the early diagnosis process. Because optometrists are most often the first contact a person has with eye health care in Finland, optometrists need to be aware of the signs and symptoms and participate more in the assessment of KC.

Diagnostic tests like topography, tomography and pachymetry provide aid in the early detection, state of progression and in determining the severity of the disease. The appropriate methods for KC management are selected according to severity and progressiveness. Due to the lifelong status of the condition optometrists can provide ongoing care and support to the patients as well as informing the patient of signs of disease progression, and management possibilities. Patient education is critical to minimize disease progression. Verbal guidance to avoid eye rubbing and applying allergy medication if allergy is present is highly suggested.

Optometrists are the main providers of the optical management of the KC patients. Particularly rigid gas permeable (RGP) lenses or scleral lenses are often the primary method for correcting vision in KC. Optometrists providing optical KC management should be specialized in KC and in addition should have expertise in fitting and managing specialty contact lenses. After this project and by relying on the author's experience it seems evident that more education regarding speciality contact lenses is needed amongst Finnish optometrists. Because of the broad subject of this research, speciality contact lenses were only briefly introduced in this thesis even though the importance and significance of contact lenses in KC management is undisputed. Although this research provides basic knowledge on the topic a more thorough education is suggested to be planned separately.

A survey concerning the state of optometry and optician profession in Europe conducted by the European Council of Optometry and Optics (2020) states that most optometrists in Finland have the capability to practise in ocular diagnostic services that include investigation, examination, and evaluation of the eye and associated systemic factors to detect, diagnose and manage disease and to use diagnostic drugs. The survey also noted that the educational recognition of optometrists' skills was more advanced compared to the legislative recognition. The legislative restrictions are outdated.

Due to the aging population, the role of optometrists as eye health care professionals need to transform towards more clinical expertise. Optometrists are no longer only dispensing opticians and need to take more part in providing efficient eye health care to fill in the gap of an inadequate number of ophthalmologists in Finland. The increased state of optometrists' clinical knowledge and the ability to recognise abnormalities need to be utilized in many areas of eye health care, including KC assessment and management.

6.1 Reliability and Ethicality

This integrative review was organized and written by only one author. More than one author could have raised the reliability in the article selection process as well as in the review and analysis process.

To gain greater reliability, the search process was implemented in cooperation with an information specialist from the Oulu University of Applied Sciences. Also, the key terms for the literature search

were selected with guidance from the information specialist to ensure the relevance based on the research question. To ensure all relevant data to be found the search results were not limited to full access texts and all relevant texts not provided by the EBSCOhost web source were sought from alternative sources.

The thesis was implemented to the author's best capability by detailed documentation of the steps followed throughout the literature review process. As a limitation, however, due to lack of previous experience in research, the article selection and evaluation criteria phase was not so accurately documented at the time of data selection and therefore could not be reported better during the writing process.

The clinical guidelines implementation process should follow a carefully selected protocol. A systematic literature review process and expert panellists' evaluation rounds before completion and publishing could have increased the reliability of the thesis. Unfortunately, this was not possible in the scope of this thesis.

The thesis was conducted by following the Oulu University of Applied Sciences thesis protocols. The guidelines on research integrity by the Finnish National Board on Research Integrity (TENK) were followed throughout the thesis writing process. No separate Institutional Review Board (IRB) approval or research permit was required for this literature review-based research development because no patient data was used. The Rectors' Conference of Universities of Applied Sciences Arene thesis recommendations were honoured.

Licences for the use of adopted pictures and tables were required and requested from the owners of the copyrights through web-based licence retrieval systems or by email. Some of the emails remained unanswered by the time of writing this report.

7 CONCLUSIONS

Determining the evidence-based practice patterns in KC assessment and management as well as resolving the elements to be included in KC patient assessment and management guidelines implemented by optometrists in Finland were the main purposes of this thesis.

The literature review pointed out the current practice patterns concerning KC assessment and management and by further analysis of the results and current legislation, the guideline for the use of optometrists in Finland was designed.

The importance of early diagnosis and patient education cannot be overlooked when attempting to halt the KC disease progression and visual impairment. Optometrists' skills and clinical expertise can be utilized more frequently in eye health care to minimize disease progression and visual impairment. Comprehensive care for KC patients should be provided by both optometrists and ophthalmologists working closely together. Optical management of KC relies mostly on optometrists' competence in contact lenses and advanced contact lens fittings.

The increasing amount of aging population will set a demand for sufficient eye healthcare and the arrangement and organisation of the care should be more closely considered. The research development points out the possibilities in easing the economic burden and the lack of eye health care accessibility by taking optometrists into account when planning and providing efficient eye health care in Finland.

This research development provides a baseline structure for the guidelines concerning KC assessment and management by optometrists in Finland. The formal guideline creation procedure requires expert panellists' rounds before the national guideline can be fully implemented in practice. Also, a further pilot use of the guideline in optometry practice is recommended to investigate the informative effectiveness, practicality, and usability of the guideline.

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