INTRODUCTION

Keratoconus

Keratoconus is a non-inflammatory disorder, characterized by corneal thinning and anterior protrusion.[1,2] With increasing use of the excimer laser for the correction of myopia, detecting early keratoconus in the absence of slit-lamp findings has gained increasing importance.[3] Because these patients do not achieve high-quality vision with either glasses or contact lenses, they tend to seek out refractive surgery. Recent reports suggest that patients with early keratoconus or keratoconus suspects comprise 2–5% of patients presenting for refractive surgery for myopia.[4,5] Keratorefractive procedures may have unsatisfactory results and cause postoperative complications in these patients.[3, 6] Moreover, apart from excessive ablation of corneal tissue, unidentified subclinical keratoconus is considered to be the main cause of ectasia after LASIK.[7, 8]

I. Epidemiology:

Keratoconus, classically, has its onset at puberty and is progressive until the third to fourth decade of life, when it usually arrests. It may, however, commence later in life and it may progress or arrest at any age. Rarely may it be congenital.[9] The reported incidence of keratoconus varies, with most estimates being between 50 and 230 per 100,000 in the general population (approximately 1per 2,000). The variability in the reported incidence reflects the subjective criteria often used to establish the diagnosis. Keratoconus occurs in all ethnic groups with no male or female predominance.[10]

II. Etiology and Pathogenesis:

A. Associated disorders

Keratoconus has been reported in various clinical settings. It may be an isolated sporadic disorder, or it may be associated with other rare genetic disorders. It may be associated with Down syndrome and Leber's congenital amaurosis, with connective tissue disorders, with hard contact lens wear and eye rubbing, and with a positive family history of the disorder.[11,12] Down syndrome has been reported to have a high association with keratoconus, with reported incidence ranging from 0.5% to 15% (i.e., 10–300 times more common than in the general population).[13,14] Similarly, there is a high incidence of keratoconus in patients with Leber's congenital amaurosis (up to 30% of patients older than 15 years).[2] The frequent occurrence of keratoconus has been attributed to a high incidence of eye rubbing in these two disorders, owing to increased blepharitis in Down syndrome and an oculo-digital sign in Leber's congenital amaurosis. However, a recent study of children in a school for the blind contradicts this theory and suggests that the association with keratoconus might be due to genetic factors rather than eye rubbing.[15] Mechanical trauma has also been implicated in the pathogenesis of keratoconus. Although a number of studies report a high association of eye rubbing with keratoconus, a cause-and-effect relationship is difficult to prove.[1]

Contact lenses are also suggested as a source of mechanical trauma related to keratoconus.[1] Because early in the disease process patients have mild myopic astigmatism with clinically normal-looking corneas and their vision is best corrected with rigid contact lenses, it is extremely difficult to determine which came first, the keratoconus or contact lens wear. It is possible that mechanical trauma induced by eye rubbing and hard contact lens wear act as environmental factors that enhance the progression of the disorder in genetically predisposed individuals. Atopy is often cited as being highly associated with keratoconus. A review of the literature reveals conflicting data in favor of and against this association. [16-18]

B. Biochemical studies

Despite intensive biochemical investigation into the pathogenesis of keratoconus, the underlying biochemical process and its etiologic basis remain poorly understood. Corneal thinning appears to result from loss of structural components in the cornea, but why this occurs is not clear. Early biochemical studies demonstrated that collagen composition in corneas with keratoconus was unaltered.[14] Recent biochemical assays and immune-histological studies of corneas with keratoconus suggest that the loss of corneal stroma after digestion by proteolytic enzymes could be caused by increased levels of proteases and other catabolic enzymes or decreased levels of proteinase inhibitors.[19, 20]

Wilson et al have proposed a role for an interleukein-1(IL-1) system in the cornea in the pathogenesis of keratoconus.[21] It has previously been demonstrated that keratocytes from keratoconus corneas have a fourfold greater number of IL-1 receptors than normal corneas; Wilson et al suggested that the increased expression of the IL-1 receptor sensitizes the keratocytes to IL-1 released from the epithelium or endothelium, causing a loss of keratocytes through apoptosis and a decrease in stromal mass over time. This hypothesis makes sense of the occurrence of keratoconus in relation to eye rubbing, contact lens wear, and atopy, if it is presumed that epithelial micro trauma leads to an increased release of IL-1 from the epithelium.[22]

Wilson et al have also suggested that abnormalities in the processes that regulate apoptosis, besides the IL-1 system, could be the cause of keratoconus, even in the absence of epithelial cell injury.[21]

C. Genetics

Although formal genetic analysis using current methodology has not been reported for keratoconus, review of the published literature provides strong pointers to suggest genetic influences in the pathogenesis of this disorder. This includes at least eight reports of its occurrence in identical twins, the bilaterality of the disorder, and multiple reports of its occurrence in family members in two and three generations.[23-25]

The majority of reported studies suggested an autosomal dominant mode of inheritance with variable expression and included subtle forms of the disorder, such as forme fruste keratoconus or mild irregular astigmatism.[2]

III. Histopathology

Thinning of the corneal stroma, breaks in Bowman's layer, and deposition of iron in the basal layers of the corneal epithelium comprise a triad of the classical histopathologic features found in keratoconus. Depending on the stage of the disease, every layer and tissue of the cornea can, however, become involved in the pathological process. Fine details of these processes are most clearly appreciated by electron microscopy. The epithelium may show degeneration of its basal cells, breaks accompanied by down-growth of epithelium into Bowman's layer, particles within a thickened sub-epithelial basement membrane like layer and between basal epithelial cells, and accumulation of ferritin particles within and between epithelial cells most prominently in the basal layer of the epithelium.[1]

Histopathologic features detected in Bowman's layer may include breaks filled by eruptions of underlying stromal collagen, periodic acid Schiff–positive nodules, and Z-shaped interruptions, possibly due to separation of collagen bundles and reticular scarring. Features noted in the stroma are compaction and loss of arrangement of fibrils in the anterior stroma, decrease in the number of collagen lamellae, normal and degenerating fibroblasts in addition to keratocytes, and fine granular and microfibrillar material associated with the keratocytes.[1]

Descemet's membrane is rarely affected except for breaks seen in acute hydrops. The endothelium is usually normal. However, some abnormalities have been reported, including intracellular dark structures, pleomorphism, and elongation of cells with their long axis toward the cone. Gross histopathologic analysis of corneal buttons undergoing penetrating keratoplasty for keratoconus has revealed the presence of two types of cone morphology: "nipple"- type cones, located centrally, and "oval"-(sagging) type cones, located inferiorly or inferotemporally.[26]

IV. Clinical Features

Keratoconus is a condition in which the cornea assumes a conical shape as a result of non-inflammatory thinning of the corneal stroma. The corneal thinning induces irregular astigmatism, myopia, and protrusion, leading to mild to marked impairment in the quality of vision.[1] It is a progressive disorder ultimately affecting both eyes, although only one eye may be affected initially.[24, 25]

Symptoms are highly variable and, in part, depend on the stage of the progression of the disorder. Early in the disease there may be no symptoms, and keratoconus may be noted by the ophthalmologist simply because the patient cannot be refracted to a clear 20/20 corrected vision. In advanced disease there is significant distortion of vision accompanied by profound visual loss. Patients with keratoconus fortunately never become totally blind from their disease.

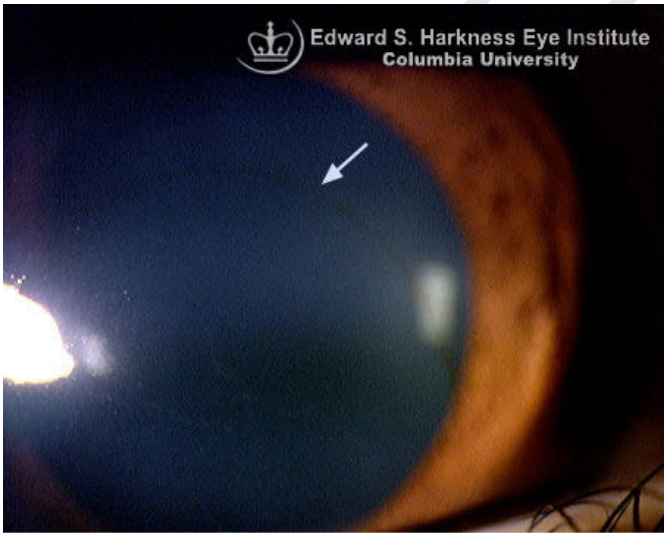
Clinical signs also differ depending on the severity of the disease. In moderate to advanced disease any one or combination of the following signs may be detectable by slit-lamp examination of the cornea: stromal thinning (centrally or para-central, most commonly inferiorly or infero-temporal) conical protrusion; an iron line partially or

completely surrounding the cone (Fleischer's ring); and fine vertical lines in the deep stroma and Descemet's membrane that parallel the axis of the cone and disappear transiently on gentle digital pressure (Vogt's striae). Other accompanying signs might include epithelial nebulae, anterior stromal scars, prominent corneal nerves, and increased intensity of the corneal endothelial reflex and sub epithelial fibrillary lines.[1,24] Munson's sign and Rizzuti's sign are also useful adjunctive external signs associated with keratoconus. Munson's sign is a V-shaped conformation of the lower lid produced by the ectatic cornea in down gaze. Rizzuti's sign is a sharply focused beam of light near the nasal limbus, produced by lateral illumination of the cornea in patients with advanced keratoconus.[24] (Fig.1)

Early in the disease process the cornea may appear normal on slit-lamp biomicroscopy; however, there may be slight distortion or steepening of keratometry mires centrally or inferiorly. In such instances it is useful to dilate the pupil. Retro illumination techniques and scissoring of the retinoscopic reflex or the "Charleux" oil droplet sign are useful clinical signs to confirm the diagnosis in suspicious cases. [24]

Keratoconus, Munson's Sign

Keratoconus, Vogt's striae



Fleischer Ring

Fig.1: Signs of keratoconus: top Munson's sign, middle Vogt's striae and bottom Fleischer Ring [27]

V. Classification

Several classifications of keratoconus based on morphology, disease evolution, ocular signs and index-based systems have been proposed in the literature:

Morphological classification:

Classically, keratoconus has been classified into: [2, 26, 28, 29]

- Nipple—the cone has a diameter≤5mm, round morphology and is located in the central or paracentral cornea, more commonly in the infero-nasal corneal quadrant. Correction with contact lenses is normally relatively easy
- Oval—the cone has a diameter>5mm and a paracentral to peripheral location, more commonly in the infero-temporal corneal quadrant. Contact lens correction is more difficult.
- Keratoglobus—the cone is located throughout 75% of the cornea. Contact lens correction is a difficult challenge, except in very limited cases.

The wide spread use of corneal topography has allowed the detection of new keratoconus patterns affecting the superior, nasal and central cornea [30]. More recently, a new D-shape keratoconus pattern has been described in the literature. This pattern was detected retrospectively in 2 patients who developed post-LASIK ectasia and prospectively in 4 patients who had other corneal abnormalities suggestive of keratoconus.[31]

Classification based on disease evolution:

The first keratoconus classification based on disease evolution was proposed by Amsler [32], who classified the disease in four different severity stages:

1. Fruste or subclinical form; diagnosed by corneal topography; 6/6 visual acuity (VA) achievable with spectacle correction.
2. Early form; mild corneal thinning; corneal scarring absent.
3. Moderate form; corneal scarring and opacities absent; Vogt's striae; Fleischer's ring; <6/6 VA with spectacle correction, but 6/6 VA with contact lens correction; irregular astigmatism between 2.00–8.00D; significant corneal thinning.
4. Severe form; corneal steepening >55.00D; corneal scarring, <6/7.5VA with contact lens correction; severe corneal thinning and Munson's sign.

Treatment of keratoconus:

Keratoconus treatment varies depending on the disease severity. Traditionally, incipient cases are managed with spectacles, mild to moderate cases with contact lenses,

and severe cases can be treated with keratoplasty. Other surgical treatment options include intra-corneal rings segments, corneal cross-linking, intra-ocular lens implants or a combination of these.

1) Spectacles

Spectacles are normally used in early cases of keratoconus only. As the disease progresses, irregular astigmatism develops and adequate visual acuity cannot be achieved with this type of visual correction.[9]

2) Contact lenses

The first to describe the use of contact lenses to manage keratoconus was Adolf Fick in 1888. Since then, contact lens wear has represented the most common and successful treatment option for early to moderate cases of keratoconus.[33]

3) Intracorneal ring segments:

Intra corneal ring segments is a surgical technique originally developed for the treatment of low myopia. The technique consists of the implantation of one or two polymethyl methacrylate segments in the corneal stroma to reshape its abnormal shape in an attempt to improve visual acuity, contact lens tolerance and prevent or, at least, delay the need for corneal graft. Contact lens-intolerant keratoconus patients without central scarring, who have mild or moderate disease, may be candidates for intrastromal ring segment insertion [34-36] The procedure improves visual acuity by flattening the central cornea, reducing astigmatism and centering the cone. The goal of the procedure is to improve contact lens fit and comfort. It may also improve best-spectacle-corrected visual acuity (BSCVA). Ferrara rings (Ferrara Ophthalmics, Belo Horizonte, Brazil) and Intacs (Addition Technology Inc, Des Plaines, IL, USA), commonly used ring segments, are made of rigid polymethyl methacrylate. Ferrara rings have a fixed inner diameter of 5.0 mm and a triangular anterior contour. Intacs have an inner diameter of 6.8 mm, a flat anterior surface, and are available in thicknesses of 0.25–0.45 mm, in 0.05 mm increments.[36,37]

To achieve the desired effect, the ring segments must be inserted at approximately two-thirds depth; therefore, the cornea should be at least 450 μm thick. Stromal channels have traditionally been prepared with a specially designed keratome. Some femtosecond lasers can now be programmed to create stromal channels of the appropriate width and depth.[38]

Colin and Malet reported that contact lens wear was restored in over 80% of cases. The 2-year best-corrected visual acuity (BCVA) improved in 68.3% of eyes ($p < 0.001$), and the manifest refraction spherical equivalent improved from a mean of −6.93 D ± 3.91 (SD) preoperatively to −3.80 ± 2.73 D at 2 years ($p < 0.001$). Measurements of mean keratometry decreased from 50.1 ± 5.6 D to 46.8 ± 4.9 D at 2 years ($p < 0.001$).[36]

Complications: The traditional mechanical technique of tunnel creation can lead to the following complications:[39,40] epithelial defects at the keratotomy site, anterior and posterior perforations during channel creation, extension of the incision toward the central visual axis or toward the limbus, shallow placement of the intra-corneal ring segments,

infectious keratitis, persistent incisional gaping, decentration, stromal thinning, and cornea stromal edema around the incision and channel, extrusion of the implant and migration, undercorrection, overcorrection, intrastromal deposits, and glare. Kanellopoulos et al[41] reported postoperative complications at 35%. Ruckhofer et al [42] reported the frequency of intrastromal deposits after Intacs implantation in myopic eyes. Galvis et al.[43] reported a case of a patient who developed culture-proven bacterial keratitis 4 months after intra-corneal ring segment implantation for keratoconus. The femtosecond laser offers several advantages that could reduce these complications due to more precise location of the channel and its dimensions, depth, diameter, and width. However, some reports have shown complications. Ertan and Kamburoglu [44] observed decentration after Intacs placement by femtosecond laser.

4) Corneal Cross Linking:

Collagen cross-linking (CXL or C3-R) is the most recent addition to the surgical armamentarium and may slow or halt the progression of keratoconus by using a photo-oxidative treatment to increase the rigidity of the corneal stroma.[45-47] At the time of this writing, CXL is not FDA approved for use in the USA. The procedure begins with the removal of the central epithelium to enhance stromal saturation with the topically applied riboflavin (vitamin B_2). The cornea is then irradiated with ultraviolet A (UVA) light at 370 nm for 30 minutes using a special UVA generating device. The irradiation of the riboflavin results in chemical reactions that create covalent bonds, which bridge amino groups of the stromal collagen fibrils.[48]

The principal effects of cross-linking are localized to the anterior 300 μm of the stroma.[49] The biomechanical effect in human corneas has been shown to be a 328.9% increase in corneal rigidity.[50] Morphologically, the formation of cross-links increases intermolecular spacing as the collagen polypeptide chains are pushed apart.[48] Increased resistance to enzymatic degradation shown in porcine eyes following cross-linking may also contribute to biomechanical stability.[51]

The improvement in visual acuity after CXL is the result of decreasing both corneal curvature and astigmatism. Raiskup-Wolf et al reported a 2.68 D reduction in corneal power at 1 year postoperatively.[45] Three years after the treatment, the BCVA improved one line in 58% of 33 eyes and remained stable in 29% of eyes ($p < 0.01$). The astigmatism had diminished by a mean of 1.45 D in 54% of eyes. These topographical changes have the potential to improve contact lens fit. CXL appears to be most beneficial for patients with mild progressive keratoconus. It is less effective in patients with advanced keratoconus and may be less effective in patients with ectasia following LASIK surgery. CXL poses a risk of dose-dependent keratocyte apoptosis affecting the anterior 300 μm of the cornea. UVA irradiance of 0.36 mW/cm^2 has been found to be cytotoxic to the rabbit corneal endothelium, which corresponded to a human corneal stromal thickness of less than 400 μm.[52,53] Therefore, pachymetry should be performed preoperatively to confirm that the corneal stroma is greater than 400 μm thick. Preliminary studies have shown that the lens and retina are not adversely affected.[54]

5) Keratoplasty:

When a stable, comfortable contact lens fit cannot be obtained or fails to provide adequate vision, more invasive surgery such as keratoplasty is recommended. The type of keratoplasty surgery depends greatly on the individual patient's needs and the surgeon's preferred technique. While penetrating keratoplasty has traditionally been the surgery of choice, lamellar surgery is becoming more popular for patients with mild to moderate disease. Recurrent keratoconus in the donor following cornea transplant has been reported;[55, 56] however, recurrent keratoconus is far more likely to be related to incomplete excision of the cone. The iron ring, found at the base of the cone, should be used as a reference when planning graft size.[57]

Postkeratoplasty myopia can be reduced by using the same-sized donor and host corneal buttons.[58-60] Some have even suggested undersizing the donor to further flatten the postoperative corneal contour.[61] Axial length can be an important factor in the refractive error outcome following keratoplasty.[62] Ultrasound axial length measured from the anterior lens capsule to retina reveals a broad range in length from 18.77 to 25.65 mm. Reducing donor size, in a relatively short eye, could result in significant postoperative hyperopia. The flattened corneal contour could complicate contact lens fitting in the anisometropic patient. Same-size donor and host corneal buttons should not be used when the anterior lens-to-retina length is less than 20.19 mm, the mean length for non-keratoconic individuals with emmetropia.[63]

Deep anterior lamellar keratoplasty (DALK) is a recently developed alternative to traditional lamellar keratoplasty as well as penetrating keratoplasty (PK). One advantage of the procedure is that the host endothelium is preserved, thus reducing the risk of rejection. The risk of endophthalmitis is theoretically less because this is largely an extraocular procedure. Intact host Descemet's membrane may afford greater wound stability than occurs in penetrating keratoplasty. In addition, the reduced need for topical steroid may help the DALK patient to heal faster than the PK patient. Visual outcomes are comparable to PK. The main drawbacks are that DALK is technically more challenging and time consuming.[64-66]

The procedure involves a staged dissection of stroma down to the level of Descemet's membrane and transplantation of donor tissue that has had Descemet's membrane removed. Anwar and Teichmann introduced the big bubble technique in which air is injected into the deep stroma after an initial stromal dissection. This acts to safely separate the posterior stroma from Descemet's membrane, resulting in a shorter and safer surgery.[67] Viscoelastic has also been used to separate the host Descemet's membrane. Reducing the intraocular pressure (IOP) with a paracentesis also helps to prevent perforation. Melles et al has described using intracameral air as a reference to help judge the depth of the host stromal dissection.[68]

History of Contact Lenses: [69]

There are many statements in the literature that the original inventor of contact lenses was Leonardo de Vinci in 1508. This idea came from the discovery of some marginal notes in one of his writings that showed the head of an observer that was immersed in a large bowl of water and some notes indicating that vision was possible. While this

demonstration may have had some of the elements of a contact lens, it most certainly cannot be considered a contact lens, even in crude form, as we know it today.

René Descartes proposed another idea in 1636, in which a glass tube filled with liquid is placed in direct contact with the cornea. The protruding end was to be composed of clear glass, shaped to correct vision; however, the idea was impractical, since it would make blinking impossible.

It would appear that the true inventors of contact lenses were three men of different nationalities who came upon the idea at nearly the same time in the year 1888. Fick (Switzerland), Kalt (France) and Mueller (Germany). Lenses by Muller can be viewed in a museum in Munich. Whether this simultaneous discovery was the result of pure coincidence or whether the three men had knowledge of the work of each other is uncertain and probably will remain unknown. Each had as his primary interest the correction of keratoconus and each made lenses from blown glass. Glass lenses were relatively heavy and could not be modified after their original formation. Nevertheless, lenses of glass were made until the late nineteen thirties when they were finally replaced by plastics.

The older contact lenses were scleral lenses; there were two types of scleral lenses: fluid lenses and fluidless lenses. The first were called fluid lenses because before insertion one needed to pour into the concavity of the contact lens a small amount of fluid that was compatible with corneal physiology. These lenses could be worn for only a few hours (3 - 5 hours) each day, and eventually patients would experience a clouding effect due to significant corneal swelling (edema). The fluidless contact lens looked nearly the same as the fluid contact lens except that it had an aperture to allow fluid to pass behind the contact lens. This design did not work effectively because the tears still could not supply the minimum amount of required nourishment, but it did extend the wearing time of the contact lens to about 4 to 6 hours a day.

In the beginning of 1936, some major improvements took place. William Feinbloom tried to improve the design of the scleral contact lens by devising a two-section scleral lens in which the outside scleral section (also known as the "haptic" section) was made out of a new resin (plastic) that could be molded and adjusted. The central optical section was made of glass because clear plastics could not provide the same optical quality as glass. Unfortunately, these contact lenses still had a limited wearing time, and there were probably only a dozen or so optometrists in the U.S. who could fit these contact lenses because it was so difficult to do so successfully.

The next major advancement in scleral contact lenses was made by Fredrick Ridley. In 1938 Ridley observed that airplane cockpit canopies made of polymethylmethacrylate (PMMA) did not cause an allergic reaction when the shattered pieces became embedded in the eyes of fighter pilots. From this observation he conceived the idea of making contact lenses from PMMA.

In the U.S., Kevin Tuohy, an optician practicing in Los Angeles in partnership with two optometrists, Louis Zabner and Solon Braff, was instrumental in starting the modern era of contact lenses. Touhy was a -7.00

Diopter myope (his wife was a -3.00D myope) and had a great interest in both manufacturing and fitting contact lenses. Actually, his invention was made unexpectedly

while producing scleral contact lenses in their office. One day the technician who made the scleral contact lenses showed them a lens he had made for a particular patient who was soon to arrive to receive her lenses. The transition point between the central and scleral portion of the contact lens was so thin that the two pieces fell apart. One of the men suggested trying to fit only the center (corneal) piece. Fortunately it remained on the eye and the patient could also see very well. It was referred to as the "Tuohy Contact lens.

The original Tuohy lens was a simple lens design in which there was only one curve on either the back or front surface with no peripheral curve. At the edge of the lens, there was a bevel that provided a channel for tears to pass underneath. Although it was a great improvement over scleral contact lenses, it still offered far less than the capabilities of today's contact lenses.

George Butterfield, an optometrist in Oregon, filed a patent in 1950. His contact lens had a peripheral curve that flattened away from the cornea. His design also reduced the thickness of the lens.

One important disadvantage of PMMA lenses is that no oxygen is transmitted through the lens to the conjunctiva and cornea, which can cause a number of adverse clinical effects. By the end of the 1970s, and through the 1980s and 1990s, a range of oxygen-permeable but rigid materials were developed to overcome this problem. Chemist Norman Gaylord played a prominent role in the development of these newer, permeable contact lenses. Collectively, these polymers are referred to as "rigid gas permeable" or "RGP" materials or lenses. PMMA and RGP lenses could be correctly referred to as being "hard" or "rigid", the term hard is now used to refer to the original PMMA lenses, which are still occasionally fitted and worn, whereas rigid is a generic term that can be used for all these lens types: hard lenses (PMMA lenses) are a sub-set of rigid lenses. Occasionally, the term "gas permeable" is used to describe RGP lenses, but this is potentially misleading, as soft lenses are also gas permeable in that they allow oxygen to move through the lens to the ocular surface.

Gas Permeable Lens Fitting

A contact lens prescription is different from a spectacle because in addition to the power needed, the CL prescription includes other information required to describe the fit of the lens on the eye. A detailed prescription for all CL parameters should therefore be given to the laboratory to allow for the most complete and accurate fabrication of the lens.[70]

Diameter:

The overall diameter of the lens is specified in millimeters (mm). Most standard RGP fits use lens diameters ranging from 8.80 - 10.0mm. During the past several years there has been a trend in RGP fitting to prescribe larger diameters because they provide more comfort, result in better vision, and are easier for the patient to insert and remove.[70]

Posterior central curve radius (PCCR): mm vs. Diopters

The Posterior Central Curve Radius (also called base curve radius or BCR) can be specified in both millimeters (mm) and diopters (D). Many clinicians prefer to use diopters

because it is easier to relate the BCR to the patient's keratometry reading. For example, it is helpful to determine easily and quickly the bearing relationship (i.e., the relationship between the BCR and central cornea), that is, whether it is flatter than K, steeper than K, and so on.[70]

Optic zone diameter (OZD; given in mm)

The OZD is the dimension on the posterior surface that corresponds to the area of the central lens over which the BCR is lathed. For example, if we state that the OZD is 8.0mm, then we mean that that the BCR is effective over that 8.0mm area. It is the OZD that limits the effective area of vision and therefore must be sufficiently large to cover the pupil in different illuminations.[70]

Power:

The power of the lens is specified in diopters.[70]

Peripheral curves radius (PC; given in mm)

For each RGP lens there are one or more curves in addition to the BCR on the posterior lens surface. These curves are designed to improve the alignment of the lens on the cornea, provide proper movement, and serve as a reservoir of fluid for tear interchange. The "peripheral" curve immediately adjacent to the BCR is called the secondary or intermediate curve and any additional curves are called the peripheral curves. For smaller diameter lenses there may be only two curves on the posterior surface (i.e., bicurve lens); however, for larger lenses (equal or > 9.2mm) there are usually at least three curves (tricurve lens).[70]

Peripheral curve width (PCW; given in mm)

The peripheral curve width corresponds to the area that is covered by the curve. It is specified in mm. It is important to understand that when this width is specified, it corresponds to only one half of the lens. To describe the total width that this curve uses relative to the total lens diameter, it is necessary to multiply by 2.[70] (Fig .2)

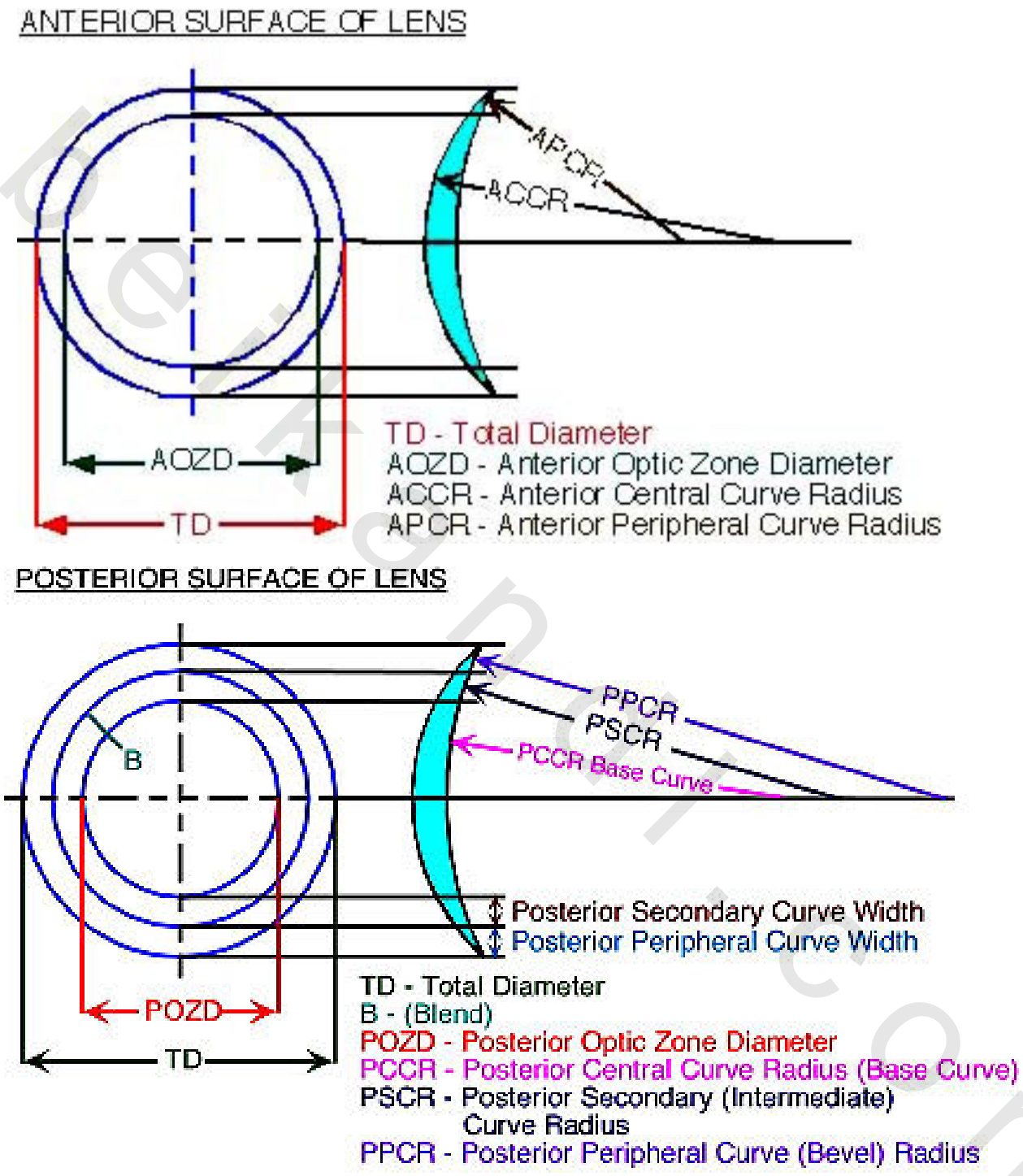


Fig. 2: Rigid contact lens terminology [70]

Goals of Gas Permeable Lens Fitting: [71]

The goal of a contact lens (CL) fitting is to provide good vision, comfort, and normal physiological ocular response while wearing lenses. In addition to these goals it is also necessary to achieve an adequate wearing time. To achieve these goals there are the following prerequisites:

a) Adequate tear interchange:

Tear interchange is the amount of tears "exchanged" (in and out) with each blink. An adequate tear interchange is important for oxygen and carbon dioxide exchange and also for removing trapped debris that can build up under a GP lens. We usually assess the degree of tear exchange by observing the amount of fluorescein at the peripheral tear reservoir and the amount of lens movement. If trapped debris or an immobile lens is observed, tear exchange is likely to be poor. If the fluorescein pattern indicates an area of sealing, tear exchange would be minimal.

There are several factors that affect tear interchange, including overall diameter, base curve, OZD, and peripheral curve and width. Finally, all these lens factors must translate into a lens that moves on the cornea. Without movement there will be very little, if any, tear exchange.

b) Lens centration & lens position before and after the blink

Good lens centration and positioning are required for both good vision and comfort. The lens factors that affect centration and position are diameter, OZD, bearing relationship, and peripheral curves. Patient factors include corneal toricity (both direction and magnitude), lid tension, and palpebral aperture size.

One technique for GP contact lens fitting is using the superior eyelid attachment method, which will allow a 1 to 2 mm movement. If the centering GP contact lens can be enhanced with the help of the superior eyelid attachment, a larger, flatter GP contact lens (a diameter of 9.0 mm) is usually required. If the GP contact lens centers well without superior eyelid influence, a smaller GP contact lens (a diameter of 8.8 mm) is indicated. Centration of the GP contact lens should be within the central corneal diameter, completely covering the pupil, but avoiding the limbus area, leaving it clear and without coming into contact with the edge of the GP contact lens.

c) Lens movement:

Some lens movement is necessary for good tear exchange and normal physiological response. Although some movement is absolutely necessary to provide an exchange of tears, excessive movement can adversely affect comfort. Movement is described by noting the rate of movement, degree, and direction. Several factors affect movement, including:

- **Center thickness**: The greater the center thickness, the greater the movement due to increased lens mass plus moving the center of gravity more anterior, which makes the lens less stable on the cornea.
- **Corneal toricity**: For with-the-rule, the CL is usually stable; however, as little as 1.00D of against-the-rule corneal astigmatism can result in an unstable lens and decenter laterally.
- **Overall lens diameter**: The larger the CL, usually the less the overall movement after the blink.
- **Minus or plus lens**: Minus lenses move less than plus lenses because of thinner lens centers and thicker edges, which tend to hold the lens by the upper lid.
- **Bearing relationship**: Steeper fits generally move less on the cornea.
- **Lid-lens relationship**: lenses that ride under the upper lid (e.g., lid attachment) move less compared to lenses that are in the aperture (intrapalpebral).

Lens/cornea bearing relationship

Definition

The Lens/Cornea Bearing describes the physical relationship between the central base curve radius of the lens and the area of the central cornea, which is covered by the base curve (optic zone). This is usually described by either the relationship of base curve to corneal curvature (e.g., steeper or flatter than K) or by the observation of the lens on the eye when seen with fluorescein (e.g., pooling, touch, and so on). With most fitting strategies the clinician will try to achieve a uniform or equally distributed lens/cornea bearing relationship. If the lens has excessive bearing (e.g., touch, pressure, flatter than K), the lens may irritate the cornea mechanically and also cause discomfort. Yet if the lens has too much central standoff (e.g., clearance, pooling, steeper than K), bubbles may become trapped under the lens, the lens may not move well on the cornea, or excessive lens flexure may occur, all of which can reduce vision and comfort. Also, a lens that is too steep (i.e., apical clearance) may reduce tear exchange and result in a poor physiological response.[71]

Peripheral edge clearance:

The edge lift is described as the distance between the edge of lens and the cornea. In general the greater the lift, the larger the tear reservoir, which means there will be more oxygen available to the cornea with each blink (i.e., better tear exchange). However, the greater the lift, the more the lid-lens contact, which might cause the patient some discomfort during the blink. Conversely too little edge lift will result in very little lens movement and a reduced pump. The clinician relies on experience to determine just how much edge clearance would be optimal. Such ability to assess edge clearance will come from experience with fluorescein pattern observations and fitting RGP lenses.[71]

Lens fit evaluation

Gross evaluation

The gross evaluation involves an inspection of the lens on the eye by using either unaided vision or the slit lamp with low magnification.

During this inspection the clinician notes the position of the lens before and after the blink, the degree of movement, and the general comfort of the patient with the lens. Usually this inspection is made after the lens has had time to "settle" on the eye (approximately 10 - 15 minutes for a patient who has not worn GP lenses). Often fluorescein is instilled before the gross examination so that the fluorescein pattern observations can smoothly follow the gross inspection. Even if the fit evaluated under white light is not acceptable, the clinician should also observe the fit with fluorescein to help determine why the lens fit is not acceptable. In the gross inspection we look for the following: [72]

- Lens position after the blink (lid attachment or intrapalpebral)
- Lens centration after the blink (center, temporal, nasal, inferior)
- Lens centration in the interblink period
- Lens movement (including total excursion) of the lens after the blink

Complications of Contact Lens Use

Risk factors for contact lens induced complications:

Apart from reduction of oxygen supply to the cornea, contact lens acts as a barrier to the elimination of waste products from the anterior corneal surface particularly carbon dioxide and cellular debris. In case of RGP wear, the accumulated cellular debris and exfoliated cells are flushed out more effectively but it can get trapped underneath a soft lens leading to toxic reactions. This can be explained by the larger diameter of the soft lenses and the lower rate of tear exchange.

The pathophysiology of contact lens associated corneal complications is related to factors like hypoxia, mechanical insult, toxicity, hypersensitivity and infection.[73]

1) Corneal epithelium complications

Corneal abrasions:

Corneal abrasions can be due to a foreign body trapped under the contact lens, due to lens damage, or a tear or traumatic lens insertion or during removal. Management of corneal abrasion includes conservative treatment like simple lubrication, patching with antibiotic coverage, removal of debris, replacement of damaged lens.[73]

Punctate epithelial erosions:

Due to mechanical causes like flat peripheral fit producing central punctuate staining or a steep fit in keratoconus causing apical staining. Another cause for punctate epithelial erosions is toxic mediated.[73]

Hypoxic effects:

Hypoxia impairs the epithelial cell metabolism, increases fragility and reduces epithelial adhesion to basement membrane. If mild, hypoxia produces epithelial edema and transient blurring of vision and if severe, it may cause cell death and desquamation.[73]

i) Epithelial microcysts:

Are seen in chronic hypoxia. They are transparent epithelial inclusions of degenerated epithelium which forms in the basal layers of epithelium and with time move towards the anterior surface. They are best visualized with the slit lamp by marginal retro illumination when they appear as tiny translucent irregular dots approximately 10-15 µ. If the hypoxia is reversed there is rebound effect, there is a temporary increase in the number of microcysts which is related to the re-oxygenation of the corneal surface causing recovery of the metabolism and clearance of extracellular debris.[74, 75]

ii) Limbal hyperemia:

The avascularity of cornea is due to the equilibrium maintained between the angiogenic and antiangiogenic molecules.[76] The stimulus and response characteristics of angiogenesis are not fully understood. Chronic increased limbal vessel injection and proliferation has been a risk factor for corneal vascularization.[77] There is considerable substantial evidence linking contact lens induced corneal neovascularization with hypoxia. Metabolic disturbances due to hypoxia cause corneal cells to generate a number of mediators with potentially angiogenic properties. Longer wearing times appears to be associated with increased prevalence of corneal vascularization. In some situations the contact lens factors like inadequate lens fit that causes the lens to directly impinge upon the ocular surface or cause poor ocular surface wetting causes corneal vascularization.[78] (Fig. 3)

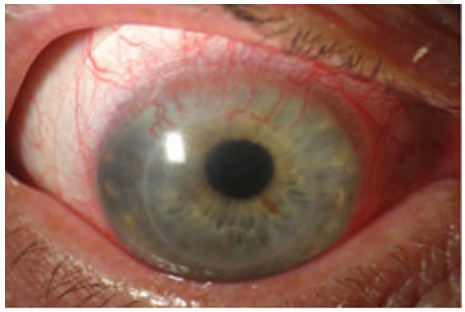


Fig. 3: Corneal neovascularization due to lens wear.[79]

iii) Mucin balls:

They are spherical structures that range in size and clarity. They are thought to be composed primarily of mucin, tear proteins and small amounts of lipid. Mucin balls are described as a new phenomenon accompanying silicon hydrogel lens wear. They have also been observed with conventional hydrogels and GP lenses. The predominant hypothesis

about their formation is that the mechanical interaction between the lens and the ocular surface results in breakup of tear film and tear film debris are formed as spherical bodies as the lens moves. Typically mucin balls does not move along with lens during a blink but remain attached or embedded in the underlying epithelium. This condition usually does not compromise vision or comfort.[73]

2) Stromal Complications

Stromal edema:

Stromal edema due to the chronic hypoxia can have a significant effect on corneal structure and long term function.[75] It manifests as striae or folds in the descement's membrane. Stromal edema induced by contact lenses is evenly distributed across the cornea but there is significantly less swelling in the extreme periphery than in the center.[75, 80]

Stromal thinning:

There are different studies showing conflicting results on the effect of lens wear on stromal thickness. Maintenance of stromal structure is by the keratocytes which comprises up to 10% of the volume of stroma. Lens induced changes to the stroma may be caused by the loss of keratocytes. Jalbert and Stapleton [80] and Efferon et al [81] reported a reduction in keratocyte density with extended wear of soft lenses. Patel et al [82] found no differences in keratocyte density between long term daily hydrogel wearers and non-lens wearers. The mechanisms suggested to explain the keratocyte loss include hypoxia mediated cell death and/ or the pressure induced effects of lens wear.[81, 83]

Refractive error:

Hypoxia induced corneal edema leads to an increase in stromal thickness and this can be associated with changes in corneal curvature or myopic shift.[84-86] Pressure induced changes in corneal shape and central flattening produces an orthokeratolgy like effect and causes changes in refractive error.[85]

3) Endothelial Complications

Endothelial blebs:

Endothelial blebs appear as small dark non reflective areas scattered over the endothelial mosaic. They have little clinical significance but are thought to indicate corneal stress form an acid shift under hypoxic conditions.[87] As intracorneal PH decreases, endothelial cells become edematous causing bulging of the posterior endothelial cell membrane. Endothelial blebs increase in size and number immediately after exposure to a hypoxic stimulus and disappear rapidly following removal.[88]

Endothelial polymegathism

The corneal endothelium is morphologically and physiologically abnormal in long term users of PMMA and soft lenses. Wearers have a greater variation in cell size (polymegathism) and an increased frequency of non-hexagonal cells (pleomorphism) than do non wearers. An association has been reported between significant increases in endothelial polymegathism and pleomorphism and a reduction in endothelial cell density in

long term lens wearers but it is not confirmed whether endothelial polymegathism and pleomorphism are precursors to the reduction in cell density.[89]

4) Infectious keratitis:

Microbial keratitis is the most serious complication of contact lens use, and contact lenses are a major risk factor for corneal ulcers. Corneal infections are more often associated with the use of soft contact lenses than rigid gas-permeable lenses. However, use of RGP lenses for orthokeratology has been associated with bacterial infections and Acanthamoeba keratitis.

Early diagnosis and appropriate treatment of infections are necessary to reduce loss of vision and, in the most severe cases, loss of the eye. Small ulcers, less than 1 mm, are often treated with intensive hourly topical fourth-generation fluoroquinolones. Larger ulcers, or those unresponsive to this treatment, are cultured and treated with intensive, every 30 minutes, fortified tobramycin and hourly cefazolin/vancomycin. Topical steroids are avoided in the initial management and, in some cases, added cautiously after the organism is identified and the infection is responding to treatment.[90]

5) Giant Papillary Conjunctivitis and Other Allergies:

Giant Papillary Conjunctivitis (GPC) was first reported in the 1970s and was described in detail by Allansmith and colleagues[91] The problem occurs most frequently in patients wearing soft contact lenses, but also occurs in patients wearing RGP lenses, hybrid lenses, ocular prostheses, and scleral lenses.[92, 93] With RGP lenses, it takes longer for GPC to develop, and the papillary reaction tends to be more localized on the superior tarsus. The variety of foreign bodies associated with a similar reaction supports the etiology as a response to the coating on a foreign body, rather than the specific nature of the foreign body. GPC is usually bilateral, but can be asymmetric, especially in patients who wear different types of lenses and/or replace them at different intervals in their eyes.[91]

GPC refers to symptoms associated with a papillary reaction of the superior tarsus. Eversion of the upper lid is necessary to make the diagnosis and should be a part of the routine examination of patients who wear contacts. Symptoms include decreased lens tolerance, increased mucus, and itching. Papillae on the superior tarsus that are greater than 0.3 mm are considered pathologic, unless they are located along the superior border of the tarsal plate, where they can occur in normal people or be associated with seasonal allergies. Allansmith described four stages of GPC. Stage 1 is preclinical, in which symptoms of mucus and itching precede signs. In stage 2, mild GPC, there are more symptoms and a mild papillary reaction of the tarsus, which can be better seen with fluorescein dye. In stage 3, moderate GPC, there may be excessive lens movement, the papillae are elevated, and the apices of the papillae may be white due to scarring, and stain with fluorescein. In stage 4, severe GPC, patients are unable to wear contacts, papillae are larger (>1 mm) and may appear similar to those in vernal conjunctivitis. Histology of the conjunctiva in GPC is also similar to vernal conjunctivitis. Numerous mediators of inflammation are elevated in the tears of patients with GPC. In severe GPC, ptosis can develop. Instead of this staging system, it is common to rate the papillae 1–4 plus. Symptoms and signs may not correlate, as some patients are very symptomatic with mild findings and others are relatively free of symptoms with more findings. Symptoms are more important than signs, since they determine contact lens tolerance.[94]

Standard treatment to reduce the immune response includes the use of mast cell stabilizers. Chronic use of combination mast cell stabilizer and antihistamine drops are helpful to prevent recurrent mucus production and itching. These drops should be instilled before and after contact lens use. It is safest to avoid use of topical steroids, but in severe cases low- or mid-strength steroids can be used to control symptoms when patients discontinue contact lenses, prior to refitting. Necessary treatment depends on the severity of GPC at the time of diagnosis. The goal is to enable patients to wear contact lenses without recurrent symptoms. The papillae often persist, but can decrease slowly over a period of years. With proper management, over 90% of patients with GPC are able to wear contact lenses successfully.(95)

Seasonal and perennial ocular allergies also reduce contact lens tolerance. Approximately 20% of the population has hay fever. Seasonal increase of symptoms associated with contact lens use is significantly higher in patients with than in those without allergies.(96) Contact lenses have been described as an 'antigen depot.(97) Contact lenses are inert plastics. The allergic response is to the coating on lenses, and not the lenses themselves. Contact lens use should be discontinued temporarily when allergies are moderate to severe until they improve with treatment and/or there is a change in seasons.(98)

Treatment of ocular allergies can enhance contact lens tolerance in patients with mild to moderate symptoms. Lubrication with tears can help to increase contact lens tolerance. In addition, topical mast cell stabilizer and or antihistamine eye drops can be used before lens insertion and after lens removal, but not while lenses are being worn, in patients who have mild symptoms. Systemic antihistamine used to control allergic rhinitis can increase dry eyes. If adequate control of allergies is not possible with these measures, the addition of topical 0.05% ciclosporin for treatment of dry eyes and off-label treatment of allergies can be helpful, especially in patients with perennial allergies. Optimal management of ocular allergies is especially important in keratoconus patients who require RGP lenses for visual rehabilitation. Contact lens tolerance can decrease during the allergy season in these patients to the point of developing corneal abrasions. Use of lubricating gels and ointments at bedtime, in addition to the other treatment, can help to prevent this complication. More frequent replacement of RGP lenses, every 4 to 12 months, may also be necessary.(90)

6) Ptosis:

In addition to GPC, there are other contact lens-related causes of ptosis. A retained, lost RGP lens can cause unilateral ptosis with or without inflammation.(99) A mass may be present on lid eversion. Patients who wear RGP lenses can develop ptosis with disinsertion of the levator aponeurosis, probably due to pulling and stretching of the lids during lens removal. Unilateral ptosis that is reversible can develop in patients who wear RGP lenses in one eye, such as for the correction of keratoconus, and does not require a neurological work-up for acquired ptosis.(100)

7) Dry Eye

Similar to allergies, contact lens use can both aggravate and cause dry eyes. Dry eye is a frequent complaint in people wearing contacts, occurring in up to 75%, and the most common reason for discontinuing lens use.(101) The International Dry Eye Workshop has

defined dry eye as a 'disease of the tears and ocular surface that results in symptoms of discomfort and is accompanied by increased osmolality of the tear film and inflammation of the ocular surface.[102] Contact lens solutions also may affect dry eye symptoms.[101]

There is evidence that dehydration of the lens surface by evaporation is more important than water loss from the contact lens itself.[103] Evaporation of the tear film over a contact lens is higher than without a contact, and is independent of the lens water content or material.[104,105] Evaporation of the tear film leads to a thinner, unstable tear film with decreased break-up time.[102] In addition, evaporation results in increased tear osmolality.[106]

Contact lenses often increase dry eye symptoms. Patients may be asymptomatic without contacts and very symptomatic with them. Symptoms that increase later in the day, with longer duration of contact lens use, suggest the possibility of dry eyes. Worse symptoms in dry environments, including indoor heat in the winter and air conditioning in the summer, as well as activities involving prolonged use of the eyes, such as reading or computer work, also point to dry eyes. Lenses become tighter with increased hours of wear and can cause symptoms of dryness and burning. Difficulty in removing lenses also points to tight lenses. Fitting flatter lenses can increase contact lens tolerance in many patients with dry eye symptoms.

Diagnosis and treatment of dry eyes can enhance contact lens tolerance in many patients. Blepharitis and meibomian gland disease cause evaporative dry eye which is aggravated by contact lens use. Treatment with warm compresses, artificial tears, antibiotic ointment at bedtime, topical ciclosporin 0.05% and, if needed, oral doxycycline can be very effective. It is important to recognize that blepharitis is a chronic condition that requires long-term treatment to be controlled. Aqueous deficient dry eye is also common. Treatment includes artificial tears, preferably unit-dose, preservative-free tears if used more than four times daily, topical ciclosporin 0.05% twice daily (before and after lens use), and punctal plugs. Lubricating drops can be used with lenses, and it is more effective to use them regularly in dry eye patients, rather than to wait until symptoms develop. Both lack of effect and benefit have been reported for topical ciclosporin 0.05% treatment of dry eye in contact lens wearers.[107,108] Beneficial effect or oral omega-6 fatty acids for contact lens-associated dry eye has been reported.[109] Many systemic medications can worsen dry eyes, including over-the-counter antihistamines. Oral contraceptives sometimes also can be associated with the development of dry eyes and contact lens intolerance. A careful history may determine if recent changes in systemic medications are factors which could be potentially modified, in collaboration with the patient's other physicians. Patients with mild to moderate dry eye can be successful wearing contact lenses, but contact lenses are contraindicated in patients with severe dry eye.[102]

Use of Contact Lenses for Management of Keratoconus

The Ohio State University (OSU) contact lens clinic has developed what is known internally as the OSU algorithm when instructing residents. Keratoconic patients are treated for keratoconus with contact lenses, contact lenses, and more contact lenses. This simple algorithm means that as a rule 75% of patients can be maintained safely in contact lenses for their condition. Most patients, however, will require re-fitting as their keratoconus progresses. Of those patients who elect to undergo penetrating keratoplasty for keratoconus, most return to contact lenses after successful keratoplasty to achieve binocular balance or their best corrected visual acuity. Smiddy et al have shown that approximately 70% of patients who presented for surgical consideration for penetrating keratoplasty for keratoconus at a university setting can be maintained successfully in contact lenses for their condition. Smiddy and Lass also observed that 60% of patients who elected to have a penetrating keratoplasty resumed contact lens wear for a variety of reasons.[110-112]

Fitting keratoconus has always been a challenge, even for the most experienced of contact lens fitters. The criteria for a successful fit in a keratoconic cornea are really no different than those for a standard GP lens. The lens should be comfortable, wearing time should be all waking hours (if possible), vision needs to be acceptable, as does the post wear biomicroscopy, and the lens needs to stay on the eye.[113]

Contact lens patient selection is the first and most critical element of prescribing contact lenses. With the wide variety of lenses available, almost every motivated patient can wear contact lenses. It is the responsibility of the practitioner to identify the appropriate candidate, determine the best lens options for them, fit the lens to allow maximum vision correction with minimal physiologic change and set realistic expectations with the patient. Primarily, patient selection involves avoiding potentially problematic patients and patients whose expectations do not "fit the product.[114]

The ideal contact lens candidate is an intelligent, motivated individual who is willing and able to make the commitment of both time and finances to properly wear and care for his or her contact lenses. The strongest contraindication is lack of motivation.[115]

Fitting Concepts in Keratoconus:

1) Apical clearance:

With this technique, the lens support or bearing is directed away from the apex and on to the paracentral cornea, with clearance (vaulting) of the apex of the cornea. Practitioners in South America and the USA tend to favor this philosophy.[116] Apical clearance is believed to minimize the chance of long-term apical scarring.[117] (Fig. 4)

Preservation of a clear cornea is important because scarring can further reduce vision. When compared to the other two methods, apical bearing and three-point-touch, apical clearance also contributes to minimal corneal oedema and punctate staining due to the clearance of the contact lens relative to the cone apex. The feasibility of this philosophy was justified by the multi-centred Collaborative Longitudinal Evaluation of Keratoconus (CLEK) Pilot Study.[118]

The feasibility of treating early keratoconic patients was established by the evaluation of changes in corneal curvature, best corrected visual acuity, corneal health, wearing times and any sign of apical corneal scarring. Although wearing time and visual acuity were good, the result showed that corneal moulding was demonstrated by unequal steepening of the flat and steep corneal curvatures after wearing the steep fitting lens for 12 months. Transient compromise such as corneal oedema, punctuate staining, epithelial contact lens imprint and corneal scarring were also present.[118] Another possible physical insult to the cornea is due to tightening of the peripheral portion of the lens, causing a sealing-off of the tear exchange behind the optical zone of the contact lens.[117] Thus, poor circulation of tears will result due to intermediate touch and excessive lid-induced lens flexure.[119]

Fitting a keratoconic eye with a relatively steeper back optic zone radius (BOZR) has been shown to compromise acuity, although the amount is very small.[120] The steep BOZR is associated with a smaller lens diameter, which also reduces the back optic zone diameter (BOZD). Monocular diplopia and flare are common complaints, especially at night when the pupil is more dilated. This is because such a small steep lens tends to be centred on the cone itself, which is often not near the visual axis, allowing the edge of the BOZD to come into view. [116]

Many practitioners believe that apical clearance is difficult to achieve. In the CLEK Pilot Study, all 30 randomly assigned eyes with early keratoconus were successfully fitted using an apical clearance lens. At a later stage of the disease, an apical clearance fitting relation may become more difficult to obtain.[118] The steepness of curvature of the corneal surface is exceedingly difficult to vault without generating a contact lens with edges that are too steep to gracefully glide over the relatively normal, much flatter peripheral cornea. Cessation of lens movement and lens adherence can result from peripheral curves that are too steep. In addition, an attempt to achieve apical clearance in more advanced cones will result in an optical zone with excessive clearance around the cone area.[117]

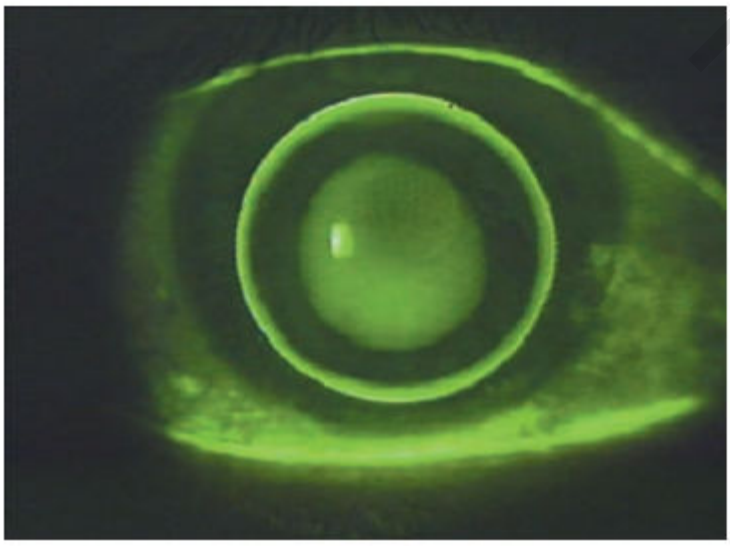


Fig.4: shows apical clearance method of fitting [121]

2) Apical bearing

This technique is characterised by primary lens support and bearing on the apex of the cornea. It was used more widely in the early days of corneal lens fitting [117] and is now mainly being used in Scandinavian and German-speaking countries.[116] this method often has superior visual performance and offers improved comfort. Better visual acuity is achieved, possibly due to the fact that a flat lens better masks irregular astigmatism.[122] It is also the easiest to achieve among the three methods. (Fig. 5)

It was once thought that a larger flat rigid lens had the ability to retard the progression of keratoconus by reshaping the cornea.[117, 122] The theory behind this was similar to orthokeratology. It has not been proven because there is no difference in the rate of progression between lens wearing and non-lens wearing groups as shown by Woodward. The effect of corneal flattening is present but only temporarily and no effect is found on the thinning of the cornea. Hence, the patient's vision may be enhanced for some time after lens removal due to the cornea remaining relatively flattened.[116,119]

Apical scarring is the complication of major concern in an apical bearing lens. Many keratoconus patients eventually show corneal scarring. Although contact lens wearing does not necessarily cause this scarring, it is likely to exacerbate it. [116] the hypothesis that acceleration of scarring can be caused by flat fitting lenses was addressed by the study by Korb [123] but the sample size was very small, having only seven keratoconus subjects (14 eyes). This experiment had strict selection criteria and was designed to eliminate the factor that apical scarring could be a natural progression of the disease and not due to the lens-cornea relationship created by a large flat lens. In the experiment, one eye of each patient was fitted with apical clearance and the contralateral eye with apical bearing. Four of seven eyes assigned to the large flat technique developed permanent apical scarring within one year, whereas none of the eyes fitted with the clearance method showed scarring. More data from a CLEK screening study also suggested increased risk of scarring with apical touch fitting. [124] Observations were made on 741 keratoconic patients. Seventy-five percent of the eyes were fitted with apical touch, 24 per cent were fitted with apical clearance and one per cent in piggyback lens systems. Twenty-five percent of 233 eyes fitted steep were scarred, compared to 46 per cent of 732 eyes fitted flat. However, the magnitude of central touch was not quantified and, therefore, the flat fitting and the three-point-touch cannot be differentiated. In addition, the causal relationship cannot be ascertained with cross sectional data, due to the dependence of fitting technique and disease severity.[124]

The pitfall that one faces when dealing with corneal scarring is that often no attempt is made to address the depth of the scar. It is reasonable to say that when present, lens-induced corneal scars are situated anterior in the stroma. A scar that is located deep in the stromal layer is more likely to be an integral part of the disease. If this factor is taken into account, the actual prevalence of corneal scarring with flat-fitting lenses may be less than the apparent figure. A milder and more common form of corneal insult due to flat-fitting lenses is abrasion. (Fig. 6) this is characterized by epithelial breakdown of the central cornea, which could lead to lens intolerance. Abrasion is capable of progressing into corneal scarring if harsh apical bearing is present for a prolonged period of time due to a poorly fitting lens. [119]

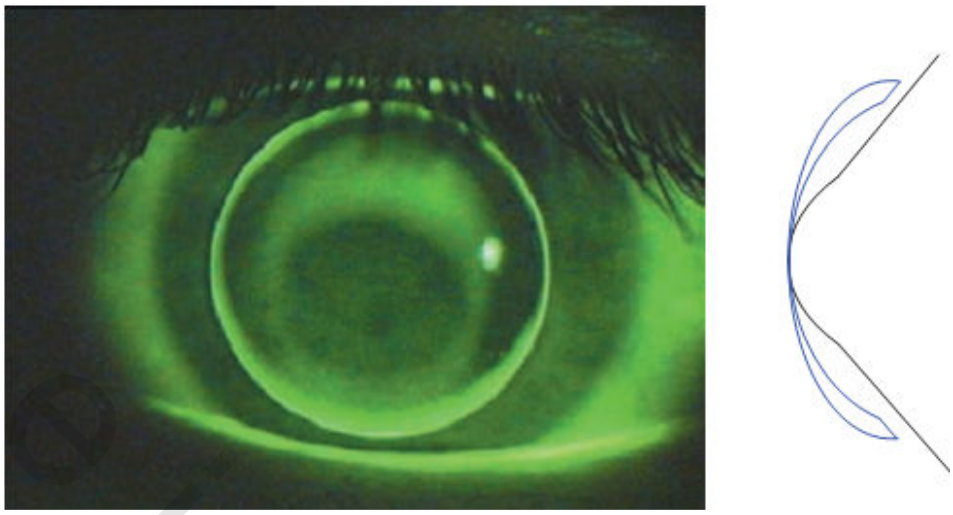


Fig.5: shows apical bearing method of fitting [121]

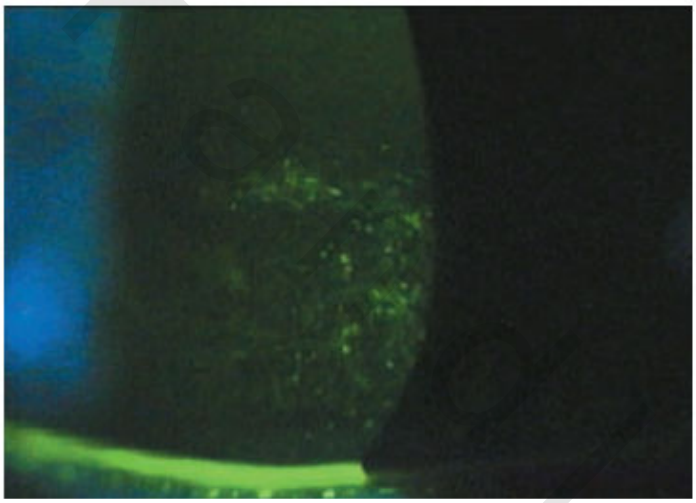


Fig.6: corneal abrasions [121]

3) Divided support or three point touch

This is a delicate balance between the two techniques already described. This method seeks to lightly touch the apex with peripheral alignment. Lens support and bearing is shared between the apex and the relatively normal paracentral cornea. Hence, the weight of the lens is distributed over a large area of the cornea. Woodward [116] defined this as an apical contact area of two to three millimetres, an intermediate clearance zone and a mid-peripheral contact annulus with conventional edge clearance at the periphery. (Fig 7) This delicate balance between a steep and a flat-fitting method was designed to reduce the shortcomings of the two other techniques. A three-point-touch lens-cornea relationship is easier to achieve than apical clearance, especially in more advanced cones. When compared with apical bearing, the chance of corneal scarring is minimized by the reduced apical touch. In addition, this type of RGP lens fitting has been found to be successful for long-term comfort and increased wearing time. There is no published evidence that any cases of three-point-touch cause scarring.[125]

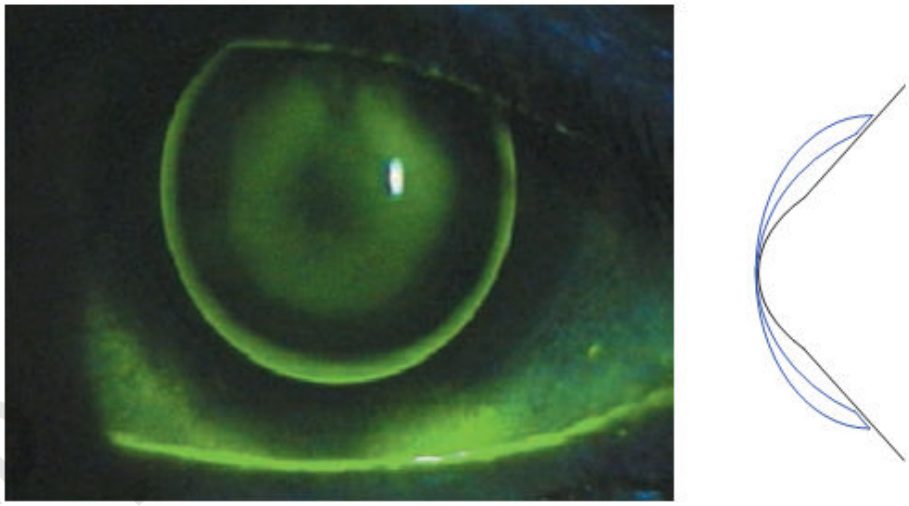


Fig.7: Three point touch [121]

Keratoconus lens options:

There are two parameters in selecting an initial lens in fitting a keratoconus contact lens. The fitter first must decide on what material will be used and then which lens design in that material will achieve a best fit. Available lens materials are: Polymethylmethacrylate (PMMA), rigid gas-permeable, hydrogel and hybrid.[126]

PMMA is a rigid acrylic plastic with no oxygen permeability. This is the original contact lens material used in all contact lenses from the 1940s until the advent of gas-permeable materials. This lens still has a small percentage of usage in keratoconic contact lenses. The material is very rigid, which lessens the chance for flexure or warpage and has the best wetable surface because of its rigid surface. It is inexpensive and easy to manufacture, allowing some fitters to modify lenses in their office to achieve a custom fit.[126]

Rigid gas-permeable (RGP) contact lens materials have been available since the mid-1970s. The gas permeability of the material, which can be defined by its DK or diffusion constant, increases the oxygen available to the cornea and is an added benefit. The oxygen available to the cornea, however, primarily is provided by oxygen dissolved in tears. The added benefit of gas permeability of the material has improved comfort and tolerance of these lenses for the keratoconic patient, particularly during the adaptation period. The negative factors that must be tolerated are difficulty in manufacturing these materials and surface spoilage, particularly silicone acrylate containing materials. Gas-permeable materials that are used, are characteristically fluorosilicone acrylate containing materials.[126]

Piggyback systems

Soft contact lenses have been suggested for mild keratoconus to act as a bandage and decrease irregular astigmatism so spectacle lenses may be prescribed. The concept of piggyback fitting using a soft contact lens as a carrier for an RGP lens to provide improve centration and stable optics has been a well-accepted concept for some patients. The use of a high DK silicone hydrogel lens as a carrier and a high DK RGP lens for the refracting surface has increased its success. Because fluorescein is not used, one needs to evaluate movement of the RGP lens and the soft lens during the trial fitting session. [127] (Fig. 8)

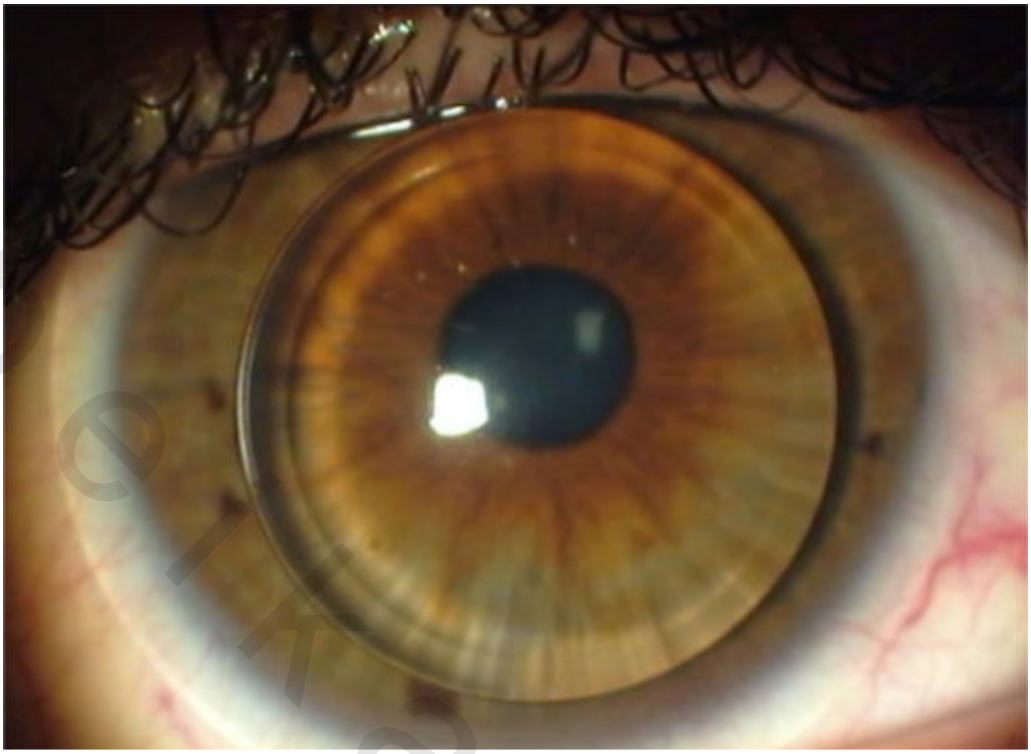


Fig. 8: shows piggyback contact lens fitting. [128]

Hybrid lenses:

These lenses have a rigid lens in the center and a soft skirt in the periphery. The most recent introduction in hybrid lenses is the SynergEyes lens (SynergEyes, Inc, Carlsbad, CA). (Fig.9) The Synergeyes lens has a central rigid gas permeable portion bonded to the nonionic hydrophilic skirt material of 27% water. Indications include RGP intolerance or inability to obtain an optimal RGP fitting, [129] poor lens centration and reduced wearing time with the RGP lenses.[130]

The overall diameter of the lens is 14.5 mm. These lenses are fitted with no or minimal apical touch in the central cornea. The vault can be between 100-600 microns. The soft skirt should exhibit 0.25 mm blink induced movements. The RGP and soft skirt can be changed individually with different lenses. The lenses can be fitted on cones of any severity. The complications noted with these lenses are GPC, tear in the soft skirt and the junction, infection and discomfort.[129]

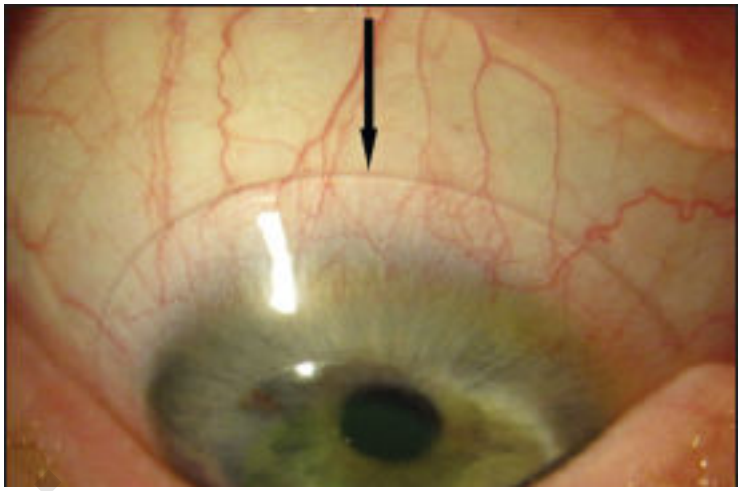


Fig.9: Hybrid contact lens on the eye. [131]

Keratoconic designs: spherical, multi-curve spherical, aspheric base curve

The most commonly used lens design is a single spherical base curve in rigid gas permeable material. The fitter has an infinite variety of base curves, because the RGP materials can be lathe cut. Edge designs also can be varied based on the fitter's experience and skill. The concept of fitting a bicurve lens design for keratoconus was developed by Soper. The concept of this design uses a steep optical portion of 6mm, with a second flatter curve of 2 mm width as a carrier to better approximate the topography of a steep cone with a flat peripheral cornea. Multi-curve lenses have their greatest success with nipple cones. There have been several improvements in the original concept in the past several years. McGuire developed a system using a central steep radius of curvature with four additional progressively flatter spherical base curves. (Fig.10) Another multi-curve design by Siviglia, the NiCone system, uses five base curves.[132]

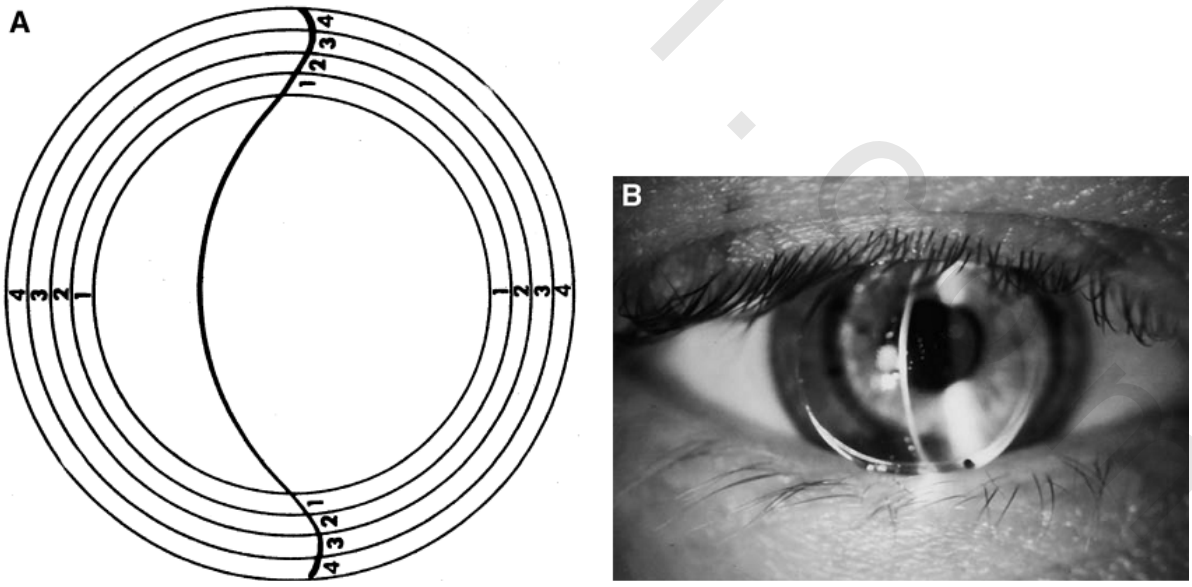


Fig.10: (A) Diagram of posterior curves in McGuire system. (B) McGuire lens.[133]

The most recent multi-curve lens design is the Rose K system. This design has up to six different curves across the back surface of the lens and a decreasing optic zone as the base curve steepens to try to align the back surface of the lens as accurately as possible with the unusual shape of the keratoconic cornea. The lens design also predicts changes that occur in the corneal shape as the condition progresses, so fitters typically don't need to change to a different design. However, patients and fitters were reporting some problems with all GP keratoconus designs that incorporate a small back optic zone and multiple secondary curves. Patients would report ghosting and poorer peripheral vision as compared with conventional-design lenses, which typically have only one curve over the majority of the back surface of the lens. Patients found these visual effects more noticeable in poor light, where the pupil was dilated, and in more advanced keratoconus.[134]

Aspheric lenses:

Rose K2 lenses are example of aspheric lenses for keratoconus. The Rose K2 lens was created to address two critical areas of performance for the keratoconic patient, spherical aberration and small optical zones. The base curve, or back of the lens, of the Rose K2 lens has an aspheric (non-spherical) optical zone unlike the spherical optical zone found on the original Rose K lens. This aspheric optical zone controls spherical aberration found on all contact lenses in higher minus powers, typically present with keratoconus lenses. The incorporation of aspheric optics into the lens design improves vision performance and enhances wearing comfort. The aspheric optical zone is larger than that of the original Rose K reducing glare, haloes and flare, common for many keratoconic patients in dim illumination (night time). Clinical studies indicated a 96% patient preference in visual performance with the Rose K2 lenses when compared with their Rose K lenses and 91% of the patients reported improved comfort.[135]

Corneal Topography

There is an extensive history of methods for measuring the corneal curvature. The earliest and still dominant clinical method is keratometry, which measures corneal slope at two positions in each principal meridian, and then converts the slopes to curvature. From an optical view point, the keratometer has much in common with current instruments that have been developed for measuring curvature over a large corneal area. The principal difference is that a keratometer measurement is restricted to a small central corneal area and is not representative of the entire corneal contour. The only portions of the cornea actually used for a keratometer measurement are two small areas in a given meridian approximately 3 to 3.5 mm apart. (136)

There have been several attempts to use a keratometer to measure peripheral corneal curvatures, but it is generally recognized that the keratometer was designed to measure only the radius of the central zone and that peripheral corneal measurements are approximations because the area measured is large and includes many different radii. The accuracy can be improved with special techniques, but a standard keratometer nevertheless has poor accuracy for measuring the entire corneal contour. (136)

Videokeratographs

These instruments are based on a Placido disc design and function on the basis of determining an array of discrete radii of curvature for many small areas, based on object-to-image relationships. Currently, there are several videokeratographs available in the commercial market that has many common features. They differ, however, in terms of target design, data processing, and display.(137, 138)

Videokeratography is based on the original concept of photokeratoscopy, developed by Gullstrand in the early part of the 20th century and modified numerous times thereafter. Videokeratography is essentially the same optical system as photokeratoscopy,(139) but differs in that a video camera is substituted for the photographic camera. videokeratography provides reasonable accuracy and repeatability in measuring the corneal topography with a clinical-type instrument.(140)

Basically, the videokeratograph operates by an optical principle that is the same as that of the keratometer. A luminous object (target of rings) is placed in front of patient's cornea, and the image size produced in the corneal reflection is measured. Using the basic optical laws of a convex mirror, the known object and image sizes are sufficient to calculate the radius of curvature of the reflecting surface. In measuring the central corneal curvature with a keratometer, there are only two objects represented, one to measure each of the principal meridians. In contrast, for the videokeratograph, a multiple-object target is used, consisting of a series of concentric rings. The distance from the center of the rings to each point on every ring represents an object distance. Alternatively, the distances between each pair of rings may be used.(141)

The image that is attributed to the corneal surface is actually produced by the tear layer covering that surface. This occurs because the light from the videokeratograph target will have the greatest reflection where there is the largest index of refraction change in the ocular surfaces. However, the normal pre-corneal tear layer is so thin (5-10 μ) that it generally follows the contour of the cornea exactly unless the videokeratograph image is

distorted by corneal areas of drying or unless there is excessive tearing. These areas can be identified on the videokeratogram image of rings as discontinuities or losses in parts of the ring pattern. When an acceptable image is obtained, the computer is directed to process the image, which involves scanning the position of each ring and calculating the radius value that corresponds to each point on the target rings. Various displays of the radii or power data are available, which provide information about the corneal shape and regularity.(136)

Keratometer Simulation (Sim K) : This display gives values that are meant to be the equivalent of a keratometer reading for each of the two principal meridians. It is produced simply by taking the radius value from the target ring that corresponds to the corneal positions where the reflection takes place from the keratometer mires. In some instruments, the readings are also provided for each hemimeridian, which is then used as an expression of corneal asymmetry. Generally, it has been found that there is good correspondence between the readings taken with keratometer and those obtained from the videokeratograph at the keratometer position.(136)

The Corneal Map: By far is the most common and important display in the videokeratography is the corneal map.(142,143) the corneal map allows the operator to visualize the overall characteristics of the corneal contour and to detect various corneal anomalies. A corneal map provides a method for displaying the entire database of several thousand corneal radius measurements in a single display. It allows the operator to immediately determine the radius of curvature and position for any corneal point. It also allows the determination of corneal irregularity and the detection of conditions such as keratoconus.(144) It allows the determination of the type, magnitude, and regularity of corneal toricity, and it allows direct comparisons of corneal changes resulting over time due to contact lens wear or surgical intervention.(145)

The corneal map is not a direct representation of the corneal shape, but rather, an interpretation of corneal shape based on radius values. The corneal map is generated by first plotting the thousands of corneal radius values on a polar graph of the corneal positions. Radius values that are the same or similar are then connected together to form zones of equal radii in the manner of a topographic map. The zones are then colored in spectral order so that the red end (warm colors) corresponds to higher corneal powers and the blue end (cooler colors) corresponds to lower corneal powers. (136)

Color scale options

Topography maps can be viewed with one of two color scaling options-an absolute scale (also called a standard scale), and a normalized scale (also called a color map, auto scale, or autosize scale). (146)

Absolute scale:

Always assigns the same color a predetermined dioptric range. Absolute scales are instrument specific, and the range of curvature values may be vastly different from system to system. Because the scales are consistent each time the absolute map is employed, a direct and rapid comparison can be made between the color maps of one eye and another or

between the same eye on two separate occasions. This comparison avoids confusion and allows visual familiarity for the user. [146]

Using only the absolute scale would have distinct disadvantages. In systems with a large range of curvatures, the scales have large intervals, which may mask clinically significant irregularities. Systems with smaller ranges may not have this disadvantage, but they may suffer from scale saturation. (The smaller-range absolute scales are typically chosen by the manufacturer to simulate the range of a keratometer). If a cornea is unusually steep, such as in keratoconus, the majority of the map may appear as an ‘‘island of red’’ with no interval definition, because most curvatures are steeper than the 52-D top interval of its scale.[146]

The normalized scale:

Automatically adjusts and sub-divides the map into multiple equal dioptric intervals based on the range of dioptric values found for that cornea. The color intervals may vary in range and dioptric values between different eyes or occasions for a given eye; therefore, normalized maps should not be compared visually at a glance without referring to the associated color scale. A normalized display allows more detail, because the color intervals can be much smaller than the corresponding absolute maps. In fact, the normalized scale can produce a misleading map, because it can take a normal cornea and exaggerate its shape to look abnormal with multiple color changes from one region of the cornea to the next.[146]

Curvature map options

Curvature map options vary in their application of a mathematical function to the raw data. The two most informative maps used in contact lens practice are the tangential radius of curvature representation (also referred to as instantaneous, local, or true maps) and the axial representation (also referred to as sagittal, color, or default maps). The axial representation values are defined as the distances along a radial plane measured perpendicularly from a surface tangent at each point along the plane to a central (optical) axis. The axial map is based on a spherically based algorithm that closely mimics keratometer measurements so it has problems mapping abnormal corneas and shows greater error in the corneal periphery.[147,148]

The tangential representation produces true curvatures based on a standard definition of the local curvature at a given point along a curve. The surface curvature is measured along radial planes as in the axial representation; however, the radii of tangent circles at each point along the plane define the curvatures. Although the tangential and axial representations offer qualitatively similar views of corneal shape, tangential maps provide smaller and more centrally located patterns than do the axial maps. Differences between the two representations become significant for corneal points further from the reference optical axis or in the mid-to-far corneal periphery.[148]

Tangential maps depict a more accurate corneal shape better correlated to slit- lamp observations of corneal pathology and RGP fluorescein patterns. They are useful in RGP optical zone selection if the optical zone is targeted to vaulting a chosen area of the cornea. Lastly, tangential maps provide more precise definition to detect irregular astigmatism

before RGP lens fitting, to monitor for contact lens–induced warpage, and to track disease progression.[146]

Corneal Topography and Contact Lenses:

The importance of using CVK in rigid contact lens fitting is that it gives information on the peripheral corneal shape in relation to the central cornea. Pre-fitting analysis of the corneal topography should consist of shape identification and proactive decision-making on the lens design and desired fitting relationship. The practitioner should identify the steepest area of the cornea and assume that the RGP lens will migrate along this ‘‘path of least resistance’’. Once the steepest portion of the corneal surface has been identified, the preliminary RGP lens design may change based on the location of the steep meridian. If the steepest hemimeridian is inferotemporal, one should assume that the lens will decenter in the same position. The fitter may then choose to design the lens flatter, larger, or with a minus-carrier lenticular edge to attempt to achieve a more superior riding fit or lid attachment.[146]

Keratoconus Rigid Gas Permeable Fitting

Data obtained from CVK are invaluable when initiating an RGP diagnostic lens trial fitting in a patient who has keratoconus. Axial and tangential maps differ significantly in apical position and curvature in keratoconus owing to the algorithms applied. The tangential apex is always located closer to the center of the map than the axial apex. Additionally, the tangential apical radius is almost always steeper than the corresponding axial apex curvature. Differences in the axial and tangential apical curvatures are dependent on cone position and disease severity, with larger differences occurring with increased cone displacement and greater peak apical curvature.[146]

RGP Fitting Software:

RGP lens fitting has already been incorporated successfully into contemporary CVK technology.[149-151] Many topographers have their own software program to design and simulate the fluorescein pattern of a rigid lens on a selected eye. Studies have shown that the success rate of such software programs is as high as 93% and matches diagnostic fitting success rates of experienced RGP lens practitioners.[150]

These contact lens modules use corneal eccentricity or height data during the initial base curve selection to calculate suggested posterior lens curves based on a preselected tear layer thickness. Some systems use classical concepts of sagittal depth fitting techniques, wherein the software calculates a posterior lens curve beneath an optic zone until a desired tear layer clearance is achieved between the front surface of the cornea and the back surface of the contact lens. These software programs are vastly different from their first-generation counter parts, which simply used simulated keratometry and applied manufacturer’s RGP fitting nomograms using corneal curvature and toricity data.[151,152] This keratometric-based fitting approach did not provide any more sophistication over the nomograms practitioners had available without using CVK.[153] The ultimate advantage of using RGP fitting soft-ware programs based within CVK systems is an increase in the practitioner’s efficiency. Their use does not necessarily make one a better contact lens fitter. The practitioner must still use clinical data to alter a suggested default lens during a trial lens fitting on the computer screen. All of the systems allow the alteration of

suggested lens parameters with a re-evaluation of the predicted fluorescein pattern to fine-tune the fit. Appropriate fluorescence is determined by the relative depth of the tear film under the contact lens and is helpful for patient education and rapid screening of various lens designs and parameters. This simulated fluorescein pattern should accurately depict the on-eye fluorescein pattern. Nevertheless, parameters such as lid tension may influence the final lens position and may subsequently alter the actual on-eye fluorescein pattern and lens location. Currently, there is no method that can predict blink-induced movement.[153]

The use of CVK-based fitting eliminates the need for reusing and sterilizing diagnostic trial lenses. The elimination of trial lenses is a significant advantage because of the potential risk of trasnsmission of infection. Additionally, chair time can decrease to less than 50% of the time required in a diagnostic lens fitting. The decreased chair time can translate to increased practice efficiency without sacrificing accuracy for experienced RGP fitters and to enhanced first-time fitting success rates for novice fitters.[149]

Although the contact lens modules have been tested on normal eyes with good results, the tear layer and resultant lens power calculations have not been designed for use on an irregular corneal surface.[149]

Although some manufacturers promote their topography software for fitting irregular corneas, no peer-reviewed literature supports the success of such programs for this purpose. CVK is most beneficial as a starting point for the selection of lens design and for obtaining preliminary base curve data for trial lens selection. An appropriate diagnostic lens should be placed on the eye, with the resultant fluorescein pattern viewed and adjusted accordingly. Power should then be determined with a spherocylindrical over refraction over the best fit trial lens available.[146]