A Systematic Approach to Fitting Keratoconus Lenses

Paul Rose, BOpt, BSc, FNZSCLP

Keratoconus is a non-inflammatory ectasia of the axial cornea characterized by corneal distortion and irregular astigmatism. It is usually bilateral and is very rarely truly monocular. It is characterized by corneal thinning, distortion, and irregular astigmatism, so one of the first indications of a keratoconic cornea is some steepening and distortion of the topographical map or keratometer mires. Visually it often presents initially as normal astigmatism, but frequently progresses quickly so that the astigmatism is in excess of three diopters or more and best acuity levels drop below 20/20. At this point, the eye’s shape is not normal and it starts to protrude forward (Fig.1).

Rate of Incidence

The percentage of occurrence of keratoconus in the general population is very difficult to determine. Researchers will quote substantially different amounts because there is a significant variation in how the data is collected and the diagnostic criteria may vary from one country to another. In some countries, until a patient is clinically blind and the condition is very advanced, keratoconus often goes undiagnosed. However, in more developed countries where sophisticated healthcare delivery services and advanced technology are available (e.g. topography), it is diagnosed much earlier.

Keratoconus is more common in hot, dry climates. The gender effect is equal, so males and females are affected at the same rate. It is more common in isolated population demographics. For example, off the southern end of Australia in Tasmania, the incidence is much higher than in the rest of Australia. As soon as a population demographic is identified that is isolated by religion, tradition or geography, more keratoconus tends to be observed.

The estimated occurrence of keratoconus in New Zealand, where the country is relatively geographically isolated, is approximately one in every 330 people. In Cypress the rate of occurrence is one in 3000; and in the United Kingdom, one in 10,000. These numbers were compiled over five years ago, so it is safe to say that in any specific population today there is more keratoconus than anyone ever thought there was. As screening and diagnostic tools, such as corneal topography, continue to gain acceptance and are used in increasing numbers within the eye care community, these patients are being identified more accurately and earlier than ever before.

The Etiology of Keratoconus

It is not understood what causes keratoconus. However, several considerations and associations are indicators. Collagen abnormality is implicated, yet familial association is slight, since keratoconus is only genetically traceable in approximately 30% of all cases. Environmental factors do seem to be involved, and environments that produce high pollen counts with associated high numbers of atopia in the general population have corresponding high numbers of keratoconus. Associated allergies of some kind are found in over 50% of all keratoconus patients. Eye rubbing is also an associated factor, but it is not clearly established which comes first, the eye rubbing or the keratoconus. However, there is now a general consensus amongst keratoconus experts that eye

Figure 1: As keratoconus progresses, the conical protrusion becomes more evident.

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rubbing should be avoided with keratoconic patients.

Keratoconus is also associated with other conditions, such as Down Syndrome, as well as some connective tissue disorders. Vernal keratoconjunctivitis, associated with allergies such as hayfever, asthma, eczema and food allergies, is seen in a significant number of keratoconus patients. To some contact lens fitters, this might indicate these patients may not be particularly successful GP wearers. However, it is possible to overcome these contraindications and achieve successful wear in the majority of cases when fitting the keratoconus patient with GP lenses.

**Categorizing Keratoconus**

Generally speaking, keratoconus can be broken down into three categories; early, moderate, and advanced. Early keratoconus, with average keratometer readings less than 50.00 diopters (6.8mm), is only able to be detected within a small area around the central cornea. One of the first signs of early keratoconus is irregular astigmatism and an inability to improve a patient's visual acuity to normal standards. Moderate keratoconus is categorized by average K readings between 50.00 to 58.00 diopters (6.8 to 5.8 mm). Any cornea with a reading greater than 58.00 diopters (5.8mm) is categorized as being advanced keratoconus.

There are basically three types of keratoconus, nipple, oval and globus cones, although in some cases these overlap. The nipple cone tends to be a small, steeply curved central cone of less than 5mm. It is usually located slightly inferiorly, around the central cornea and within 1 to 1.5 mm of the visual axis/pupil center and so it affects the cornea over a relatively small area.

The oval cone is probably the most common type. Its optical center is usually displaced inferiorly, with a cone diameter of about five to seven millimeters. The oval cone affects a larger area of the cornea than the nipple cone.

The last category is the globus cone, which involves about 80 or 90% of the cornea but is fortunately relatively rare occurring in less than 2% of all keratoconus patients.

Statistically, 71% of cones are inferior, with the majority located on the temporal inferior side, and only 11% are found superiorly. Only 18% of cones are found centrally, which causes centration problems because contact lenses tend to position over the steepest part of the cornea. Fortunately, 76% of cones are displaced less than one millimeter from the optical center of the cornea so most displacement is only within one millimeter of the central position.

**Patient Symptoms and Clinical Signs**

Initially, patients tend to be asymptomatic and are often unaware they have keratoconus. The first symptom usually reported is some visual degradation, monococular ghosting/diplopia, flare, and glare or halos around lighted objects particularly at night. Frequently the first reported symptom is problems with night vision while driving. But reports of recent photophobia and excessive use of sunglasses, whereas previously they had no problems with glare or light during waking hours, is also reported.

Early clinical signs include distorted retinoscopy reflex, decreased unaided vision, marked changes in the refractive sphere, cylinder and axis, irregular astigmatism, closely held reading material and a narrow palpebral aperture as the patient squints to improve vision. Later signs will present upon examination as distorted photokeratoscopy rings, inferior steepening in corneal maps (Fig. 2), apical corneal thinning and cone formation.

Advanced keratoconus presents with advanced cone formation topographically, Munson’s sign, Fleischer’s iron ring (Fig. 3) and an increased visibility of corneal nerve fibers (Fig. 4). As the keratoconus advances further, Vogt’s striae (Fig. 4), apical corneal thinning and cone formation (Fig. 5), non-uniform red reflex with ophthalmoscopy, ruptures of Descemet’s membrane (hydrops), and reduced intraocular pressure are also seen.

Adolescent onset keratoconus, which is early onset keratoconus, tends to appear in the early to mid-teens and is normally bilateral, but it often presents in one eye first and is often more advanced in one eye than the other. Very rarely does the keratoconus condition manifest exactly the same in both eyes. The rate of progression varies dramatically over the first five to ten years. Progression of the cone is usually quicker in the first five years, often slowing down after about ten years, although in some cases it can continue to progress for many years thereafter. Early onset keratoconus, which occurs during puberty, typically doesn’t change much after the age of 30. Later onset keratoconus progresses much longer.

**Fitting the Keratoconus Patient**

How does one fit keratoconus effectively and what are the primary fitting objectives? Ninety percent of keratoconus fittings require GP lenses, and a proper fitting process requires a careful assessment and evaluation of fluorescein patterns. Some fitters believe keratoconus lenses can be fitted using topography alone. However, different mapping machines may give different pictures and even different operators may produce different maps. Various scales also give an array of pictures so it is important when comparing maps to make sure the scale is the same in both maps. It is not as simple as just taking a map, sending it to the lab, and saying, “Send me a lens.”

The most useful aspect of topography is indicating where the cone displacement is located, the size of the cone, whether there is any significant astigmatism outside of the cone, and providing some idea about where the contact lens is probably going to sit, as well as defining the initial diagnostic lens selection.
The primary objectives in keratoconus lens fitting are to provide the best possible visual acuity, with a final fit that creates minimal interference to the corneal physiology. It is vital that the cornea continues to function well, without disturbing the epithelium.

The fitter must 1) **Optimize lens comfort** by placing the major bearing surface of the lens on the stronger para-central part of the cornea, not the weaker, thinner cone portion of the cornea. 2) **Achieve slight apical touch** that does not produce any insult to the cone apex. 3) **Maximize tear exchange around the cone** to minimize potential problems with edema, which causes the cone to swell further forward, and promotes a sloughing off of epithelial cells over the cone. 4) **Provide optimum lens movement**, not too much or too little, but enough to exchange tears from behind the lens and wash over the cone apex on blinking. 5) **Position the lens over the pupil**, which is sometimes the biggest challenge, because cones are not typically centered over the pupil. To achieve an optimum fit, it is absolutely essential to look at the lens to cornea fitting relationship centrally, and then separately look at the peripheral lens fit. The singularly most important part of keratoconus fitting is the peripheral fit, but this is the area of keratoconus GP fitting which is often ignored. But, this is what the patient feels. They do not feel the central fit, they only feel the peripheral fit and the edge profile.

A word of caution: It is imperative to use a keratoconus lens design as opposed to a standard spherical GP lens design when fitting the cone patient. This is because a keratoconus lens design will accommodate the transition from the steeper central portion to the mid-peripheral bearing portion of the posterior lens surface and produce a greater edge lift than a normal GP design. These elements are essential for comfort and good tear exchange. The normal, spherical GP lens design does not accommodate the dramatic change in radius of curvature present on the keratoconus cornea. With a regular, spherical GP lens there is not sufficient flattening between the optic zone and the periphery so it becomes difficult to achieve the ideal apical touch while maintaining optimum peripheral edge lift. Ideal apical touch and optimum edge lift facilitate good tear exchange and ideal lens movement, which leads to good comfort and minimal insult to the cornea. Certainly, large diameter, regular, spherical GP lenses can work in some cases for early keratoconus. However, often this is not optimal and causes a challenge in later fittings when the design will have to be changed as the cone progresses. The more advanced the cone, the less likely it is that a normal, spherical GP lens will fit optimally, and therefore this will create more centrally flat and peripherally tight fitting lenses with subsequent corneal compromise. Scar tissue is produced from flat lenses with tight peripheries. Fitting lenses specifically designed for the keratoconus cornea will lessen the probability of scar tissue formation as the cone progresses.

However, the dilemma the fitter is faced with is that a contact lens with large amounts of apical touch (flatter) will often provide better vision than a lens fitting with no touch. If apical staining occurs however, the lens is too flat. Optimum base curve selection provides for a three point, very light apical touch. If, when the patient blinks, tears flush over the cone apex, base curve selection should be adequate and should not normally result in damage to the cone even if central apical touch is present.

**The Five Steps to Keratoconus Fitting**

There are five steps for fitting keratoconus lenses, and the order presented is how fitting should be approached (Table 1). Base curve selection is first, and then evaluate the peripheral fit, followed by diameter, lens location and finally lens movement.

So first, the **base curve**. If using a keratometer, choose the first diagnostic lens by averaging the two meridians, then choose the initial diagnostic lens 0.2mm steeper than this mean “K.” This is useful in early keratoconus, but will prove to be more and more inaccurate as the keratoconus becomes more advanced.
Keratometry or topography is only ever a guide to choosing the first diagnostic lens with keratoconus, and the fitter must always then evaluate this initial lens using fluorescein, and make subsequent changes to facilitate acceptable apical touch based purely on the fluorescein pattern.

Corneal topography is useful, but not absolutely essential. The initial diagnostic lens base curve is the same as the radius of the third ring out on the temporal quadrant. There should be a millimeter section set on the topographer to easily identify the third ring in the horizontal meridian. However, to reinforce the fitting philosophy, the fitter must always place a diagnostic lens on the eye and evaluate this with fluorescein (Table 2).

Very often, during the initial fitting, a lens will settle inferiorly (Fig. 7). To evaluate how the contact lens fits over the central primary position, manipulate the lens position by using the lids. Push the lens up and evaluate the fit in the central primary position (Fig. 8) because it will always be wrong if evaluated in the inferior position. When the lens is observed in the central position and there is no apical touch, it is too steep (Fig. 9). A contact lens showing significant central touch is too flat and can position inferior or superior to the central cornea (Fig. 10, 11). If the fit is not changed, apical staining will occur and if not corrected will eventually lead to scar tissue formation (Fig. 12).

With a light apical touch (what some fitters may consider slightly flat), visual acuity is often better because there is a larger and better visual pathway through the cornea. Also with most keratoconus lens designs, the flatter the base curve, the larger the posterior optical zone. Conversely, the steeper the base curve, the smaller the posterior optical zone. This may also affect the vision as larger posterior optical zones often produce better acuity and less ghosting particularly in poor lights.

Unfortunately, often with the “perfect fit” the vision is not as good as that of a slightly flatter fit so the aim is to try and achieve an acceptable balance while maintaining corneal integrity. The principle of the fitting is to go steeper with the diagnostic lens until cone touch can no longer be seen, and then flatten the base curve selection back until cone touch just begins again (Fig. 13).

It is always advisable to use a corneal anesthetic. Chair time is expensive both for the contact lens professional and the patient so it is preferable to make application of the lens onto the eye as comfortable as possible to facilitate minimal tearing and adaptation. With the use of a corneal anesthetic, it is not necessary to spend ten minutes waiting for the gas permeable lens to settle. Invariably more cone touch will be observed as the lens settles and tearing subsides, and therefore if very light cone touch is seen, it is likely this will increase as the contact lens settles.

The second consideration for a successful keratoconus fit is the periphery and, as stated earlier, it is the most ignored and singularly the most important part of keratoconus fitting. An acceptable fit will present a good fluorescein band at the edge. If not, the lens will be tight, reducing the tear flow behind the lens and overall lens movement. The concept is to spread the paracentral bearing on the cornea as widely as possible while maintaining ideal edge lift (Fig. 14). The ideal edge lift is represented by a fluorescein band at the lens periphery between 0.6 to 0.8 millimeters wide. Evaluate the edge lift in the horizontal meridian within 20 degrees of the 180 degree meridian, never in the vertical meridian. There will often be different values on the nasal and temporal side of the lens. A very narrow, dark ring indicates a tight periphery (an edge lift that is too
Figure 10, 11: A diagnostic contact lens observed in the central position showing significant touch is fit too flat and can position inferiorly (left) or superiorly (right).

Figure 12: If a flat fitting GP contact lens is not changed to allow more tear film beneath the lens, apical staining will occur and scar tissue can form.

Figure 13: An optimum base curve selection for a keratoconus patient shows light apical touch over the cone.

Figure 14: Optimum edge lift—very slight apical touch, central positioning, wide paracentral bearing, and excellent tear exchange.

Figure 15: Peripheral seal edge lift—too tight, no apical touch, central positioning, paracentral to wide bearing, little or no tear exchange, and increased edge lift required.
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Figure 16: Edge lift too high—too flat, too much apical touch, central positioning, paracentral to narrow bearing, excessive lens movement, and decreased edge lift required.

Figure 17: The ideal overall diameter of a keratoconus lens will allow the lens to hang off the top lid, clear the lower limbus and position centrally.

Figure 18: A contact lens that is too large will produce peripheral staining where the secondary curve sits on the cornea.

Figure 19: A contact lens that is too small may result in glare, haloes, ghosting and a general inability to improve visual performance.

Figure 20.

acceptable movement. Excessive edge lift will create excessive movement and decentration of the central, ideal lens position. Again, the peripheral fit is most important in both lens location and movement and is perhaps the single most overlooked factor in lens fitting for keratoconus.

So in summary, these are the fitting goals. Optimize the central fit first with light touch, minimizing the cone contact and clearance, and make sure there is as little tear pooling around the cone as possible. The bigger the optic zone, the more pooling that will get around the base of the cone. Place the main lens bearing on the stronger peripheral cornea. Fluorescein pattern assessment is critical in evaluating the optimum lens to cornea relationship. Choose a design where the peripheral fit can easily be changed. Do not raise a patient's visual expectations too high and don’t always expect to achieve 20/20 visual performance.

It is worthwhile mentioning the evaluation of the correct power when fitting the keratoconus patient and how to approach the over-refraction of a diagnostic lens or the existing patient Rx for the best vision performance.

Keratoconus patients are notoriously difficult to refract, particularly with steep base curves. Often a +1.50D or a –1.50D will yield little or no difference in the acuity subjectively. There is no benefit refracting initially in 0.25D steps. The patient will only fatigue. Always start the refraction with at least a 1.00D increment for all keratoconics. Refine in 0.50D increments and only use 0.25D increments in early keratoconus.

And, of course, the final lens power is the diagnostic lens power and over-refraction taking vertex distance into account if the over-refraction is more than 5 diopters.

Here are some fitting pearls to reduce lens reordering relative to power:

1) Finish the refraction with the lights on. Depending on the pupil size, this influences the entrance pupil for the light passing through the lens and cornea and into the eye and because keratoconus corneas have such a variable shape across the cornea, this can produce a different refraction for different pupil sizes.
2) Do not spend a lot of time trying to get 20/20 vision. Most keratoconics don’t get much better than 20/30 vision. Get the best vision possible with the diagnostic lens and over-refraction. Vision often improves as the lens settles back over the first few weeks.
3) Educate the patient about visual expectations. It’s very unlikely a keratoconic patient will see as well at night as a normal eye.

The future is indeed bright for new improved keratoconus lens designs as lathing technology improves and lens fitters and fabricators can create complex aspheric and aspheric/spherical designs for specialty GP lens applications.
The vast numbers of fitting curves needed to effectively fit the diverse nature of keratoconus corneas dictates the need for a fitting system. The design system offered by the Rose K™ lens manufactured in Boston® materials automatically compensates all the curves across the entire posterior surface when the fitter adjusts the peripheral edge lift design or the central base curve or diameter, and maintains consistency of fit and performance from lens to lens. This system also predicts the changes that occur in the corneal shape as the condition progresses so the fitter does not need to change to a different design.

For example, in the Rose K™ system, changing the edge lift doesn’t just change the periphery of the lens. The change begins near the central optic of the base curve, widening the paracentral bearing area. This broadens the application of the lens design system and provides the fitter with a simplified, consistent, predictable approach for fitting the keratoconus patient.

When designing the Rose K™ system, the standard lift is set at an arbitrary value of zero. It has no meaning other than it defines how much flatter the secondary curves are in relation to the base curve so it defines the edge lift on a scale that the fitter can use to accurately control the peripheral fit. The system establishes the appropriate edge lift as standard for that specific base curve and diameter. From this point the system compensates the base curve and power based upon what change is made in the periphery. If anything other than a standard lift is ordered, both the base curve and power will be slightly altered by the laboratory to give the same final sagittal depth as the original diagnostic lens, otherwise the lens would fit differently centrally.

It should also be noted that if a fitter orders a different lift, the posterior lens radius changes all the way back to the central posterior optical zone diameter and not just at the edge. A “Standard Increased Lift” represents 1.0 increased lift and a “Standard Decreased Lift” represents 0.5 decreased lift on this scale. The difference in change between the standard increased and standard decreased lift is a clinical observation based on hundreds of fits. It was found that the change needed to make a significant difference to the peripheral fit was almost twice as much for an increased lift as it was for the decreased lift to accommodate the majority of cones.

Historically, Rose K™ lenses were limited to 0.5mm increment changes in edge lift values. However, when Blanchard Contact Lens became the manufacturer for the Rose K™ lenses in North America in 2004, they were able to offer edge lift changes in 0.1mm increments of change. At this time, decreased edge lift values can be fitted from -1.3mm decreased lift values and up to 3.0mm increased values in 0.1mm steps (Fig. 20). This is particularly important as the fitter now has the ability to fine tune edge lift changes in much more detail than was possible in the past. Figure 20 also shows the percentage of usage of the three primary edge lift values as they relate to all lens fittings.

Visual performance and comfort have shown improvement with the new Rose K2 Aberration control lens design. The lens has just finished its introductory launch into the North American market and improvements are promising. The posterior surface has an aspheric base curve incorporated into the lens design. The aspheric curves adjacent to the central posterior optic are manufactured relative to the anterior radius of power. The overall benefit is to increase the posterior optical zone diameter, which improves vision performance particularly at night but also during daylight hours. Trials on over 100 keratoconus eyes have shown significant visual improvement for the majority of wearers as well as improved comfort for the majority of the patients.

The biggest challenge was how to improve vision and comfort without changing the fit. The cause for less than optimum vision performance can be narrowed to three contributing factors: a small optic zone, which is smaller, certainly, than most standard lenses; a multiple number of secondary curves, which often come within the pupil, particularly in poor light creating multiple images and spherical aberration; and, an on-axis aberration in a series of focal points along the visual axis, which basically means that you don’t have the light, like a magnifying glass, coming to one point. The greater the difference between the curves on the front and the back of the lens, the greater the spherical aberration; so this was more noticeable for higher powers than lower powers, where the two surfaces are relatively parallel, and therefore there is a much better chance of getting a single focal point. The steep base curve and higher minus power create a multiple number of focal points along the visual axis as opposed to a single focal point. Aberration increases as base curves steepen and power increases. With an aberration controlled design, the goal is to bring all the focal points onto the retina at a single point regardless of base curve radius or lens power. Incorporating a specific eccentricity value within the posterior optic zone, as well as stretching the optic zone without changing the fit, created the desired results. Specific eccentricity values dependent on five factors are used, with base curve and power being the major factors. Base curve, optic zone, refractive index, lens thickness, and power all influence the choice of eccentricity.

So what are the results? Independent trials at Blanchard Contact Lens showed overall visual acuity improvement in 84% of the patients as well as a 68% improvement in comfort. Visual acuity improved 72% by one line or more, 28% remained the same or improved by less than one line with 95% of patients enrolled in the studies preferring the new Rose K2 lens over their existing Rose K1 lenses.
1. According to the article, which is the singularly most important part of keratoconus fitting?
   a. Peripheral lens fit  b. Base curve choice  c. Overall lens diameter  d. Material choice
2. Which of the following clinical signs is not typically found in the keratoconic cornea?
   a. Corneal thinning  b. Irregular astigmatism  c. Corneal steepening  d. Microcysts
3. Which of the following statements is true regarding most keratoconus GP lens designs?
   a. The flatter the base curve, the larger the posterior optical zone  
   b. The flatter the base curve, the smaller the posterior optical zone  
   c. The steeper the base curve, the larger the diameter  
   d. The flatter the base curve, the larger the diameter
4. Which of the following is not considered a classification for the keratoconic corneal shape?
5. Which of the following is most likely to produce apical scarring for the keratoconus patient?
   a. Steeply fitting lenses  b. Large lens diameters  c. Flat fitting lenses  d. Small lens diameters
6. Which of the following is not true regarding the incidence of keratoconus in the population?
   a. It is more common in males than females  
   b. It is more common in hot, dry climates  
   c. It is more common in isolated population demographics  
   d. The rate of incidence is most likely greater than originally thought
7. Which keratoconus shape is the least common, with an incidence of approximately 2%?
8. As a general rule of thumb, when increasing the diameter by 0.3 to 0.4mm, how much should you change the base curve to produce the same central fitting pattern?
   a. Flatten the base curve by 0.3 to 0.4mm  
   b. Flatten the base curve by 0.1mm  
   c. Steepen the base curve by 0.3 to 0.4mm  
   d. Steepen the base curve by 0.1mm
9. Which is a critical tool for evaluating the lens to cornea relationship of a keratoconus GP lens?
10. What is a good criteria to follow in assessing the base curve choice of a keratoconus GP lens to assure that there will be no damage to the apex of the cone?
   a. Tears flush over the cone's apex during a blink  
   b. The GP lens remains inferiorly positioned during the blink  
   c. The patient can see well with the lens  
   d. The lens is comfortable for the patient during all waking hours
11. Given the guidelines in the article, which of the following would be recommended for the initial base curve selection for a GP keratoconus lens, based on the keratometry values 49.00D (6.88mm) / 53.00D (6.36mm)?
   a. 6.8mm  b. 6.4mm  c. 6.3mm  d. 6.0mm
12. Keratoconus is traced within families in approximately how many of the cases?
   a. 10%  b. 30%  c. 50%  d. 70%
13. Allergies of some kind are found in approximately how many keratoconus patients?
   a. Under 10%  b. 20%  c. 30%  d. Over 50%
14. Which single element of GP lens design controls the movement of the lens more than any other?
   a. Base curve  b. Overall diameter  c. Edge lift  d. Center thickness
15. According to the article, which of the following lens parameters should be considered first when fitting the keratoconus patient?
   a. Peripheral fit  b. Base curve  c. Diameter  d. Lens movement
16. Most cones are found in which location of the cornea?
17. Which of the following keratometric value ranges fits into the category of “moderate” keratoconus?
   a. 42.00D to 48.00D  b. 47.00D to 50.00D  c. 50.00D to 58.00D  d. 56.00D to 62.00D
18. Where should the main weight of the GP lens be supported in the keratoconic fitting?
   a. Peripheral cornea  b. Apex of the cornea  c. Sclera  d. Within the optic zone
19. Given the guidelines in the article, which of the following would be recommended for the initial base curve selection for a GP keratoconus lens, based on the keratometry values 51.00D (6.6mm) / 54.00D (6.2mm)?
   a. 6.6mm  b. 6.4mm  c. 6.2mm  d. 6.0mm
20. What is often the first symptom the patient notices of keratoconus?

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