

History of the Development of the Treatment of Keratoconus

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ABSTRACT

Keratoconus (from Greek meaning horn-shaped cornea) is a degenerative, ectatic disease of the cornea, causing corneal thinning and remodelling into a more conical shape.¹ Symptoms include blurring of vision, and a gradual decline in visual acuity.¹ It is usually bilateral, but can affect each eye at different severities.¹ There are various treatment modalities, both surgical and nonsurgical, both simple and more invasive. Here, we review the history of the development and advances of the wide range of treatment options, since it was first recognized in 1748 up until now.

Keywords: Keratoconus, History of keratoconus, Intrastromal corneal rings, Cross linking, Contact lenses, Corneal transplant.

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INTRODUCTION

Keratoconus (from Greek meaning horn-shaped cornea) is a degenerative, noninflammatory, ectatic disease of the cornea, causing corneal thinning and remodelling into a more conical shape.¹ Symptoms include blurring of vision, and a gradual decline in visual acuity.¹ It is usually bilateral, but can affect each eye at different severities.¹ There are various treatment modalities, both surgical and nonsurgical, both simple and more invasive. Here, we review the history of the development and advances of the wide range of treatment options, since it was first recognized in 1748 up until now.

EARLY HISTORY

Burchard David Mauchart (1696-1751) was an ophthalmologist and a professor of anatomy and surgery at the University of Tübingen, Germany.^{2,3} In 1748, he was the first to write a dissertation of a case of keratoconus and became one of the first to document this corneal disorder in the literature.^{2,3} He named the condition 'staphyloma diaphanum',^{2,3} meaning 'bulging of the cornea'.

In 1854, John Nottingham, a British physician, described the condition in greater depth and detail, and most importantly distinguished it from other forms of corneal ectasia.^{2,4} Nottingham wrote a book called 'Practical

Observations on Conical Cornea: And on the Short Sight, and Other Defects of Vision Connected with it,' where he reported cases of keratoconus, described by him as 'conical cornea,' and listed common signs and symptoms of the disease like diplopia, blurred vision, corneal weakness, and the refractive difficulties of spectacle correction of visual acuity of patients.⁴

In 1851, Hermann von Helmholtz invented the ophthalmoscope,⁵ which revolutionized the methods of assessing and examining patients in the field of ophthalmology.⁶ In 1859, this new invention was utilized by the British surgeon William Bowman, where he was one of the first ophthalmologists to use it to examine the cornea and diagnose keratoconus.⁷ He was successful in assessing and examining keratoconic eyes, by a method which he developed, where he was able to angle the ophthalmoscope mirrors in a way which allowed him to best visualize the conical shape of the cornea,⁸ a method still used to this date in the clinical assessment of keratoconic eyes. Bowman was one of the first to contemplate and consider management and treatment options for this condition, once he understood the nature of the disease, its effects on the cornea and the ultimately the visual impairment as a result.^{7,8} One of the cases he reported was of an 18-year-old female, where she was unable to count figures at 20 cm, and who had some improvement in her visual acuity to the level of counting fingers following his attempt to restore her vision.⁷ He was successful by inserting a fine hook, through the cornea, and stretching the pupil into a vertical slit,⁷ mimicking the pin-hole effect.

It was not until 1869, that the disorder acquired its name as 'keratoconus,' meaning 'horn-shaped cornea,' when the Swiss ophthalmologist Johann Horner (1831-1886) conducted a thesis entitled; 'On the treatment of keratoconus.'⁹⁻¹¹ In his understanding and releasing the nature and pathology of the disease, Horner's aim was to attempt to change the physical shape of the cornea, and make it more a normal-like gradually curved.⁹⁻¹¹ His method was based on chemical cauterization of the cornea^{2,9-11} to reshape its conical curvature. This management approach was also endorsed by the German ophthalmologist Albrecht von Graefe (1828-1870).^{2,9-11} It was based on the application of silver nitrate solution to the cornea and applying a pressure dressing on top.^{2,9-11}

CONTACT LENSES

In 1887, the German ophthalmologist Adolf Gaston Eugen Fick (1852-1937) invented the first successful contact lens, made from heavy blown glass.¹² This new invention transformed and revolutionized the management of keratoconus, when in 1888 contact lenses were first used to compress and flatten the conical cornea into a more curved flatter shape.¹³ The French ophthalmologist Eugène Kalt (1861-1941) was the first to envision hard glass contact lenses use in correcting the shape of the cornea in keratoconic eyes.¹³ He made a glass contact lens scleral shell, constructed from the bottoms of glass test tubes.¹³ His flat-fitting sclera lenses were designed in a way to compress the steep cornea into a more regular shape and thus improve the visual outcome.¹³ In the 1930's, plastic scleral lenses were manufactured when polymethyl methacrylate (PMMA) was developed.¹⁴ The American optometrist William Feinbloom (1904-1985) was the first, in 1936, to introduce plastic, rigid, lighter and more convenient contact lenses than the glass-blown contact lenses thus improving the compliance and the management outcome of keratoconus.^{14,15} The developments of better quality designs and materials of contact lenses further enhanced and improved this method as a management option of keratoconus.

Further developments in sciences and the field of medicines expanded the understanding of the disease and its pathophysiology, which has opened up new avenues in the development and advances in the management options of keratoconus.

Surgical Management

Eduard Zirm (1863-1944) was the first ophthalmologist to conduct a successful human corneal transplant in 1905.¹⁶ However, in 1936, the Spanish, American ophthalmologist; Ramón Castroviejo Briones (1904-1987) was the first to perform a successful corneal transplant in an advanced case of keratoconus achieving significant improvement in visual acuity.^{17,18} Briones technique and method became a worldwide standard for corneal transplantation as an option for managing corneal pathologies including keratoconus. He improved and developed the technique of the corneal transplant operation for keratoconus.¹⁹ The invention of the operating microscopic allowed surgeons to magnify their view of the cornea, and the advances of the operating instruments and materials meant that surgeons were able to perform better and more successful corneal transplants and hence better postoperative surgical and visual outcome.

In 1936, the Japanese ophthalmologist Tsutomu Sato made incisions in the cornea, a form of refractive surgery,

attempting to treat keratoconus.²⁰ This principle was further developed by the Russian ophthalmologist, Svyatoslav Fyodorov in 1974 for the management of keratoconus where multiple radial incisions in the cornea were made.²¹ However, this method is not currently used for keratoconus as it involves thinning the already weak cornea.^{22,23}

More advanced and much newer techniques of corneal transplantations continue to develop. The mini asymmetric radial keratotomy was developed by Italian surgeon Marco Abbondanza in 1994, and further improved in 2005.^{24,25} Corneal microincisions are made to produce a controlled scarring of the cornea, and the structural reinforcement of the scars changes the corneal curvature, shape and thickness.^{24,25} Deep anterior lamellar keratoplasty (DALK) is another new surgical technique developed in recent years, where the corneal epithelium and stroma are transplanted, while the patient's own endothelium layer is left unreplaced, giving the benefit of reduced risk of rejection and additional structural integrity.^{26,27}

Cross-Linking

The corneal cross-linking is a new management option for keratoconus, first developed in Germany in 2000.²⁸ It consists of the application of riboflavin solution to the eye, saturating the cornea, which is then activated by the illumination with ultraviolet A light. This method allows the formation of strong new bonds between the corneal collagen strands, improving the shape and the mechanical strength of the cornea.²⁸⁻³⁰ Since its introduction, cross-linking has seen huge advances and developments in the technique and application of this treatment modality has produced significant and successful surgical outcome.²⁸⁻³⁰

Intrastromal Corneal Ring Segments

Intrastromal corneal ring segments are a new and recently developed surgical alternative to corneal transplant where it involves the insertion of intrastromal corneal ring segments.³¹ The segments are made of PMMA and are inserted in the corneal stroma, in a circular arc. The principal of this technique is that the ring segments flatten the curvature of the cornea and reshapes it to a more naturally curved cornea.^{31,32} Two trade names of the intrastromal corneal ring segments are available; Intacs and Ferrara rings.³¹⁻³³ Originally, Intacs were first approved in 1999 for myopia; however their application in the management of keratoconus was finally approved in 2004 by the Food and Drug Administration (FDA) in the United States.³¹⁻³³ Postoperative results with this treatment technique have so far shown promising good outcomes.³¹⁻³³

CONCLUSION

Keratoconus is a degenerative, ectatic, noninflammatory corneal disease, causing thinning, weakness and remodelling of the cornea into a more cone-like shape, thus gradually causing a decline in visual acuity.¹ It was first recognized in 1748,^{2,3} and understood in greater depth in 1854.⁴ Over the years, revolutionary advances in the diagnostic and research methods improved our understanding of the pathophysiology of the disease and ultimately this led to the continuing developments of the management options since it was first recognized until now. From early on, until the present time, the management principal is to reshape and remodel the keratoconic eyes into a more normal, naturally curved cornea. Starting from crude, early surgical interventions in the nineteenth century, the introduction of contact lenses, the conduction of corneal transplantation and more recently, the advances in corneal cross-linking and intrastromal corneal ring segments.

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