

Quick Guide to the Management of Keratoconus

Mazen M. Sinjab

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A Systematic Step-by-Step Approach



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#### Preface

Keratoconus is a common disease, and its prevalence increases day by day due to the huge development in diagnostic and screening tools. Management of keratoconus has also developed; new approaches have raised either to halt the progression of the disease or to rehabilitate the cornea or to achieve both.

It is easy to diagnose the disease, but it is not that easy to classify and grade it. Nevertheless, each treatment modality has its own indications, conditions, contraindications, and complications. All of that put the doctor in many cases on crossroads and make a challenge in choosing the modality(s) that may give the patient the desired optimal result.

There are – of course – general guidelines, but tricky things are so many, hence the aim of this book: that is to clarify and specify those guidelines and to build up a mesh among specific criteria that the doctor should look for.

The way that this book deals with this topic is systematic and academic. First, it mentions the disease and its diagnostic tools with the related clinical interpretation. Second, it goes through treatment modalities in a classified and listed manner rather than an elaborating one. Third, it builds up a mesh in a flow chart manner and suggests a checklist together with a three-step approach. The checklist and the three-step approach are finally applied on nine cases taken as examples and studied following the systematic approach.

The strategy in compiling this book is combining excellence in pictorial quality with a concise but ordered text.

I have aimed the book at all those who need some initial assistance in approaching keratoconus. There are sure to be some errors; as the ophthalmology editor, I take full responsibility for these and look forward to being further educated.

Damascus, Syrian Arab. Rep.

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## Abbreviations

AB	Asymmetric bowtie
AC	Anterior chamber
ACD	Anterior chamber depth
AS	Angle supported
BFS	Best fit sphere
BFTE	Best fit toric ellipsoid
BSCVA	Best spectacle corrected visual acuity
CK	Conductive Keratoplasty
CL	Contact lens
CxL	Corneal cross linking
DALK	Deep anterior lamellar keratoplasty
dpt.	Diopters(s)
FFKC	Forme fruste keratoconus
ICR	Intracorneal ring
ICRs	Intracorneal rings
IOL	Intraocular lens
IOP	Intraocular pressure
IORL	Intraocular refractive lens
IORLs	Intraocular refractive lenses
IS	Inferior steep
IS	Iris supported
IS	Iris supported
I-S	Inferior-superior difference
KC	Keratoconus
K-max	Maximal K-readings
KPD	Keratometric power deviation
MR	Manifest refraction
NSAIDs	Nonsteroidal anti-inflammatory drugs
OCT	Optical coherence tomography
PC	Posterior chamber
PH	Pin hole
PKP	Penetrating keratoplasty
PLK	Pellucid-like keratoconus
PMD	Pellucid marginal degeneration
PMMA	Polymethylmethacrylate
PRK	Photorefractive keratectomy
RGP	Rigid gas permeable
S.E	Spherical equivalent
SB	Symmetric bowtie
S-I	Superior-inferior difference

SRAX	Skewed steepest radial axis index
SS	Superior steep
TG	Topography guided
UBM	Ultrasound biomicroscopy
UCVA	Uncorrected visual acuity
UV	Ultraviolet
UVA	Ultraviolet A

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#### Introduction

Keratoconus (KC) is a fairly common bilateral, noninflammatory, degenerative axial ectatic condition of the cornea in which the cornea assumes an irregular conical shape. It is one of the most common corneal diseases that refractive surgeons encounter.

KC is a complex condition of multifactorial etiology, but the exact etiology is unknown. Both genetic and environmental factors are associated with KC. The role of heredity is not clear because most patients do not have a positive family history. Offspring appear to be affected in only about 10% of cases. An autosomal dominant transmission with incomplete penetrance has been proposed. On the other hand, evidence of genetic etiology includes familial inheritance, discordance between dizygotic twins, and association with other known genetic disorders. Several loci responsible for a familial form of KC have been mapped; however, no mutations in any genes have been identified for any of these loci. There are an increasing data suggesting that the environment might also play a role in the development of the condition; the disease is common in dry, cold climates. Reactive oxygen species (i.e., free radicals) are one of the proposed mechanisms of KC development. They include ultraviolet light, atopy, mechanical eye rubbing, and poorly fitted contact lenses.

KC occurs with increased frequency with systemic and ocular conditions:

1. Systemic disorders:

Down's syndrome, Turner syndrome, Ehlers-Dunlos syndrome, Marfan syndrome, atopy, osteogenesis imperfecta, and mitral valve prolapse.

2. Ocular associations:

Vernal disease, retinitis pigmentosa, blue sclera, aniridia, and ectopia lentis.

The onset of the disease is at around puberty and progresses slowly thereafter, although it may become stationary at any time.

The hallmark of KC is central or paracentral stromal thinning, apical protrusion, and irregular astigmatism. This usually results in significant impairment in both the quantity and quality of vision because of the progressive nature of the disease.

In advanced KC with corneal opacities, corneal grafting can be the only surgical alternative, in spite of its technical, biological, and refractive complications. Therefore, modern managements have been developed either to stop the progression of the disease or to rehabilitate vision or to achieve both.

There are so many good books in the market talking about this disease, but the purpose of this book is to build a step by step systematic approach for the management of the disease. Personally, I find this strategy practical in handling such hot topics. This book will cover the theoretical aspects of the disease; it will concentrate on the practical aspects including important parameters that affect the decision of the proper managements.