INTRODUCTION

Corneal ectasia is a noninflammatory, bilateral, asymmetrical condition, causing progressive corneal steepening and thinning, types of corneal ectasia include Keratoconus (KC), Pellucid Marginal Degeneration (PMD), Keratoglobus, post refractive ectasia and wound ectasia after Penetrating Keratoplasty (PK) ***(Spadea et al., 2012)*.**

KC is the most prevalent form of corneal ectasia and affects all ethnicities, defined as a bilateral, asymmetric degenerative disorder of the eye caused by collagen disorganization in the cornea, the annual incidence of which is 2 per 100,000 with a prevalence of 54.5 per 100,000 (approximately 1 per 2,000) ***(Weed et al., 2008) (Rabinowitz, 1998) (Kennedy et al, 1986).***

Another form of corneal ectasia is PMD (Pellucid Marginal Degeneration); it's an idiopathic, noninflammatory thinning corneal disorder normally involving the cornea inferiorly, between the 4 and 8 o’clock meridians. The zone of thinning is generally 1–2mm wide, and separated 1 or 2mm from the limbus by an area of normal cornea. The area of ectasia is cylindrical, creating an against the rule astigmatism ***(Sridhar et al., 2004)*.**

Keratoglobus is a rare type of corneal ectasia, characterized by generalized thinning and globular protrusion of the cornea. Both congenital and acquired forms have been shown to occur, and may be associated with various other ocular and systemic syndromes including the connective tissue disorders ***(Feder and Kshettry, 2005)*****.**

Refractive surgeries are various elective procedures that modify the refractive status of the eye, procedures that involve altering the cornea are collectively referred to as “keratorefractive surgery”, other refractive surgery procedures include the placement of intraocular lens (IOL) implant either in front of the crystalline lens “phakic IOL” or in place of the crystalline lens “refractive lens exchange”. The first excimer laser tissue ablation keratorefractive surgery performed was Photorefractive Keratectomy (PRK) after being approved by the FDA in 1995; subsequently laser-assisted in situ keratomileusis (LASIK) has become the most commonly performed keratorefractive surgery ***(McLeod et al, 2013).***

Yet, these surgeries may uncommonly be associated with complications; the most serious long-term complication of keratorefractive surgery is the weakening of the cornea and the development of keratoectasia (post LASIK ectasia) which is a corneal disorder characterized by progressive stromal thinning and central or asymmetric inferior corneal steepening. It has been suggested that central ectasia is associated with low residual stromal bed thickness, whereas inferotemporal ectasia is a sign of pre-existing forme fruste keratoconus (FFK) or pellucid marginal degeneration (PMD) ***(Binder, 2007) (Shortt et al, 2006) (Twa et al., 2004***)

Refractive surgery practice allowed us to see more patients with corneal ectatic dystrophies and other topographic abnormalities than would be expected from the incidence of each of these disorders in the general population, thus, meticulous preoperative screening is one of the key factors that contributes to success in refractive surgery ***(Ambrósio and Wilson, 2001)***.

Corneal topography is a valuable diagnostic tool for diagnosing subclinical keratoconus and for tracking the progression of the disease. Evolution in keratoconus detection has resulted in continued refinement of indices such as the (keratometry, Inferior-Superior dioptric asymmetry, skew percentage, astigmatism). Several topographic devices provided additional information through various features, for example, the Orbscan provides data on anterior and posterior elevation and best-fit sphere and a corneal pachymetry map. Later a recently introduced imaging device that provides accurate measurement of corneal power, elevation and pachymetry is the Pentacam; this device uses a rotating Scheimpflug camera. The Scheimpflug system determines net corneal power, elevation maps, anterior chamber depth and corneal wavefront. It is also an excellent method to detect form fruste keratoconus and subclinical keratoconus ***(Swart et al,*** **2007); *(Konstantopoulos et al,*** **2007).**

AIM OF THE WORK

The aim of the study is to determine the prevalence of ectatic corneal conditions among keratorefractive population admitted for refractive surgery at Al-Mashreq Eye Centre, during the period from June 2017 till December 2017, using data from the scheimpflug tomographer imaging system.

CORNEAL ANATOMY

The cornea is the transparent, avascular anterior portion of the eye that covers the iris, pupil, and anterior chamber. It comprises the external layer of the eye along with the sclera, with which it is continuous. The transition area between the cornea and sclera is the limbus, a highly vascularized area of pluripotent stem cells ***(Abbey and Yoo, 2010).***

The adult human cornea measures about 11-12mm horizontally and 9-11mm vertically. It is approximately 0.5mm thick at the center and the thickness increases gradually towards the periphery where it is about 0.7mm thick. The curvature of the corneal surface is not constant being greatest at the center and smallest at the periphery giving it a prolated shape creating an aspheric optical system. The radius of curvature is between 7.5mm and 8.0mm at the central 3mm optical zone of the cornea. The refractive power of the cornea is 40-44 diopters and constitutes about two third of the refractive power of the eye ***(Grosvenor, 2007).***

The human cornea consists of 5 recognized layers, 3 cellular (epithelium, stroma, endothelium) and 2 interfaces (Bowman membrane, Descemet membrane) and a sixth layer has recently been discovered called "Dua's Layer". ***(Figure 1)***

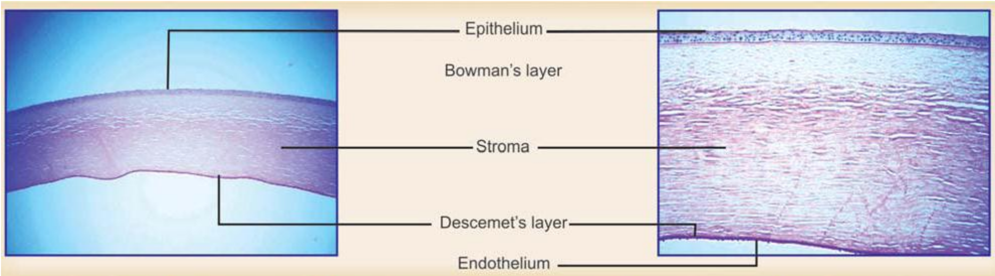

Figure (1): Layers of the normal human cornea ***(Pallikaris et al., 2005)***

1) Epithelium

The epithelial surface of the cornea creates the first barrier to the outside environment and is an integral part of the tear film–cornea interface that is critical to the refractive power of the eye. It is one of the most highly innervated parts of human body. It is a stratified, nonkeratinizing squamous layer that's 4-6 cell layers thick (40μm to 50 μm) accounting for 10 % of the corneal thickness, characterized by extreme uniformity from limbus to limbus ***(Delmonte and Kim, 2011).***

The *most superficial corneal epithelial cells* form a mean of 2 to 3 layers of flat polygonal cells. These cells have extensive apical microvilli and microplicae, which in turn are covered by a fine, closely apposed, charged glycocalyceal layer. This layer's apical membrane projections increase the surface area of contact and adherence between the tear film's mucinous layer and the cell membrane, these surface cells maintain tight junctional complexes between their neighbors, which prohibit tears from entering the intercellular spaces ***(Delmonte and Kim, 2011).***

Below the superficial epithelial cells are one to three layers of *wing cells* ***(Figure 2)***, they are so named for their thin wing like extensions that project laterally from the cell body ***(Abbey and Yoo, 2010)*.**

The deepest cellular layer of the corneal epithelium is the *basal layer*, which comprises a single cell layer of columnar epithelium approximately 20 μm tall. Only the basal cells of the corneal epithelium proliferate. The daughter cells differentiate into wing cells and subsequently into superficial cells, gradually emerging at the corneal surface. The differentiation process requires about 7 to 14 days, after which the superficial cells are desquamated into the tear film ***(Wiley et al., 1991) (Nishida and Saika, 2011)*.**

The basal cells are attached to the underlying basement membrane by a hemidesmosomal system. This strong attachment is what prevents the epithelium from separating from the underlying corneal layers.

The epithelial *basement membrane*, approximately 0.05 μm thick, comprises type IV collagen and laminin secreted by the basal cells; and the major proteoglycan is perlecan ***(DelMonte and Kim, 2011)*.**

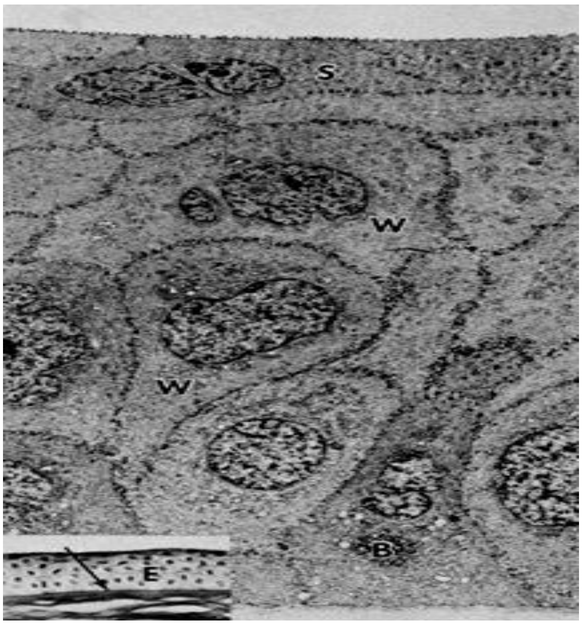

Figure (2): Full-thickness electron micrograph of corneal epithelium, (S) Apical surface cells, (W) wing cells and (B) basal cells. Inset: Epithelium (E) overlies a thin, dense basement membrane **(**arrow**)** ***(Tasman & Jaeger, 2007)***.

2) Bowman's layer

Acellular homogenous zone which is 8-14µm thick immediately subjacent to the basal lamina of the epithelium. Structurally Bowman's layer consists of a meshwork of fine collagen fibrils of uniform size lying in a ground substance. Bundles of stromal lamellae insert into this layer anteriorly. The compact arrangement of collagen confers great strength to this zone. This layer is resistant to trauma, once destroyed it is replaced by scar tissue ***(Seema, 2013).***

3) Stroma

The corneal stroma lies between Bowman's layer and Descemet membrane and comprises approximately 90% of the corneal thickness (500 µm). The cellular components (mainly keratocytes) occupy only 2–3% of the total volume of the corneal stroma, with the remaining portion comprising mostly collagen and proteoglycans. Keratocytes are specialized tissue fibroblasts that reside within the corneal stroma and produce most of the corneal stromal proteins. The role of keratocytes is to maintain the highly ordered extracellular matrix (ECM) of the corneal stroma, mainly: collagens, proteoglycans as well as other proteins. These ECM components, when properly arranged maintain a structurally stable cornea that is optically clear and possesses the desired refractive and biomechanical properties. Collagen constitutes more than 70% of the dry weight of the cornea ***(Figure 3) (Abbey and Yoo, 2010) (Nishida and Saika, 2011) (Hassell and Birk, 2010)***.

Collagen in corneal stroma is mostly type I, smaller amounts of types III, V, and VI also present. The major proteoglycans are decorin, biglycan, keratocan and lumincan. Proteoglycans are distributed among the major collagen fibers. The diameter of collagen fibril is too small (27-35 nm) to allow significant scattering of light. The peripheral stroma is thicker than the central and the collagen fibrils may change direction to run circumferentially as they approach the limbus. ***(Seema, 2013) (Berryhill et al., 2002)*****.**

Transparency of the corneal stroma depends particularly on the degree of spatial order of its collagen fibrils which are narrow in diameter and closely packed in a regular array. The collagen fibrils themselves are weak scatterers since their fibril diameter is less than the wavelength of light, and fibril refractive index is close to that of the ground substance ***(Muller et al., 2001)*.**

The ultrastructure within the organization of the lamella varies based on the depth within the stroma. Deeper layers are more strictly organized than superficial layers, and this difference accounts for the greater ease of surgical dissection in a particular plane as one approaches the posterior depths of the corneal stroma ***(Delmonte and Kim, 2011)*.**

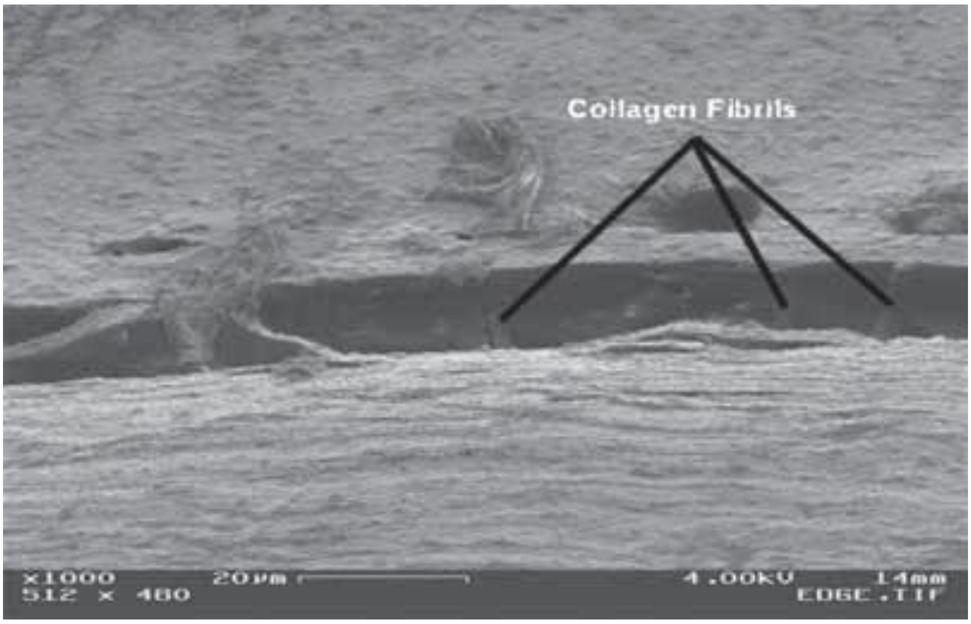

Figure (3): Stromal collagen fibrils (Scanning Electron Microscope) ***(Seema, 2013)***

4) Descemet's Membrane

Descemet's membrane is the basement membrane of the corneal endothelium. It is synthesized by endothelial cells and assembled at the basal surface of the cell layer. This membrane is primarily composed of Type IV and VIII collagen arranged in a hexagonal pattern, and it does not regenerate after injury ***(Takahashi et al., 1990)*.**

When incised, it tends to curl up in the anterior chamber and easily separates from the corneal stroma and the endothelium ***(Richard and Micheal, 1998)***. ***(Figure 4)***

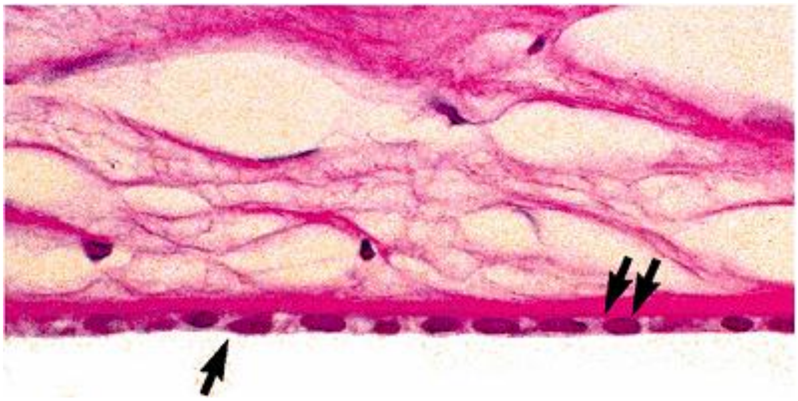

Figure (4): A thin monolayer of corneal endothelial cells (arrow) is adjacent to Descemet's membrane (double arrows). These cells are in direct contact with the aqueous humor of the anterior chamber (H&E, ×132) *(**Gordon and Thomas, 2007**)*.

5) Endothelium

A single layer that covers the posterior surface of Descemet's membrane in a well-arranged mosaic pattern

(Figure 5).These cells are uniformly 5 μm in thickness and 20 μm in width and are polygonal in shape. A major function of the endothelium is the maintenance of corneal transparency through the regulation of stromal hydration. The endothelial cells provide a "leaky barrier" to aqueous humor and contain numerous specialized ion transport systems to transfer excess water out of the cornea. These two functions of the endothelium facilitate its regulation of the amount of water within the corneal stroma ***(Freegard, 1997)(Abbey and Yoo, 2010)***.

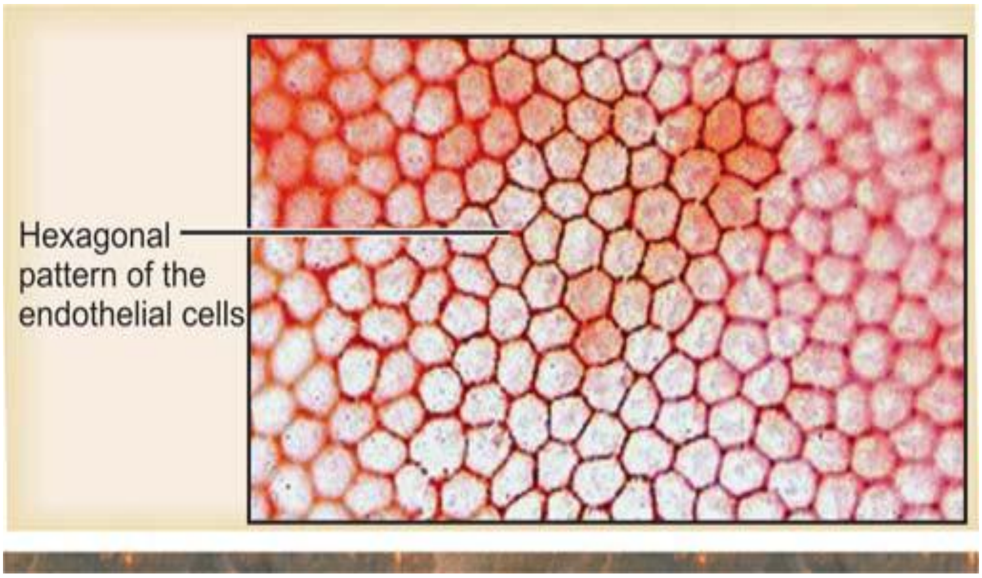

Figure (5): Histology of the corneal endothelial cells (Alizarin red stain) ***(Sharma et al., 2006)***

6- Dua's Layer:

It is described as a novel, well-defined, acellular, strong layer in the pre-Descemet's cornea. Dua's layer is 10.15 +/- 3.6µm thick. It is made up of 5-8 thin compact lamellae of collagen (predominantly type I collagen, also type IV and type VI) transverse, longitudinal and oblique. These strands of collagen extend between posterior stroma and Descemet's membrane. Recognition of the Dua's layer has a considerable impact on posterior corneal surgery and the understanding of corneal biomechanics and posterior corneal pathology such as acute hydrops, Descematocele and pre-Descemet's dystrophies ***(Dua et al., 2013)(Seema , 2013) (Figure 6)***

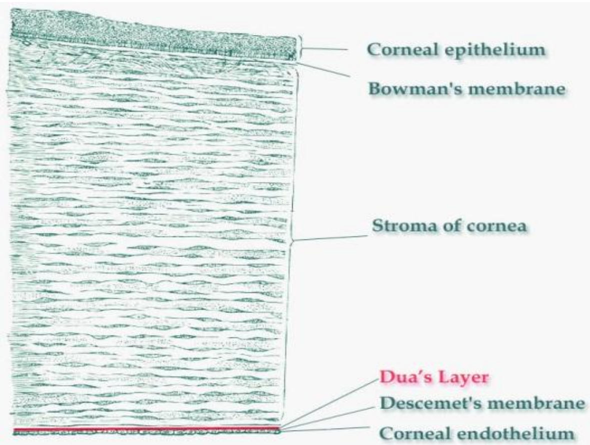

Figure (6): Dua's layer ***(Dua et al., 2013)***

CORNEAL ECTASIA

Corneal ectasia is a non-inflammatory condition, the hall mark of which is progressive corneal steepening and thinning**.** The most common corneal ectatic disorder and a leading indicator for corneal transplantation in developed countries is keratoconus (KC). Other Types of corneal ectasia include: pellucid marginal degeneration (PMD), Keratoglobus (KG), photorefractive ectasia and wound ectasia after Penetrating Keratoplasty (PK). Corneal ectasias are associated with decreased uncorrected visual acuity (UCVA), an increase in ocular aberrations, and often a loss of best-corrected distance visual acuity (BCVA). Corneal ectasia can result in significant ocular morbidity and may require surgical intervention.

The etiology of corneal ectasia can include genetic factors, chromosomal and enzyme abnormalities and mechanical factors. Post refractive ectasias can occur after LASIK and PRK ***(Feder et al., 2013) (Ghosheh et al., 2008)***.

- **Keratoconus:**

KC is a bilateral, progressive ectatic disease where the cornea becomes cone shaped due to significant thinning of the corneal stroma. Visual impairment develops from myopia and irregular astigmatism. KC usually starts at puberty and progresses until it stabilizes in the third or fourth decade. An inverse correlation has been noticed between age and KC severity, the earlier the onset of KC, the more severe the clinical phenotypes. KC appears in all ethnicities and has no gender preference. The annual incidence of KC is 2 per 100,000 with a prevalence of 54.5 per 100,000 (approximately 1 per 2,000).

Clinically, the central or paracentral corneal stroma undergoes progressive thinning and loss of structural integrity that leads to bulging of the cornea, which gives the cornea its typical cone shape appearance in KC ***(Figure 7) (Tambe et al., 2015) (Ertan and Muftuoglu, 2008) (Rabinowitz, 1998)*****.**

Figure (7): Side profile of keratoconic cornea ***(Abbey and Yoo, 2010)***