# Corneal Topography Changes in Cases with Duane Retraction Syndrome in Different Gaze Positions

Duane Retraksiyon Sendromu Olan Vakalarda Farklı Bakış Pozisyonlarında Korneal Topografi Değişiklikleri

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Geliş Tarihi/*Received:* 23.06.2009 Kabul Tarihi/*Accepted:* 01.12.2009

This study was presented as a verbal announcement at 42<sup>nd</sup> National Congress of the Turkish Opthalmology Associaton in 19-23 November 2008, Antalya.

Yazışma Adresi/Correspondence: Barış YENİAD, MD İstanbul University İstanbul Faculty of Medicine, Department of Ophtalmology, İstanbul, TÜRKİYE/TURKEY byeniad@yahoo.com ABSTRACT Objective: To evaluate the corneal topographic changes in patients with Duane retraction syndrome in different gaze positions and to investigate the effect of eyelids on corneal topography. Material and Methods: Thirty-three cases with Duane retraction syndrome and 19 control patients were included in the study. Corneal topographies of the cases with Duane retraction syndrome were first measured during their preferred gaze and then during primary gaze position. Topographies of the cases in control group were measured in their right eye during primary gaze position and when the eye was on adduction. Results: Mean horizontal sim K value of the cases with Duane retraction syndrome with preferred gaze position was 41.4 ± 2.3 D, while vertical sim K value was  $42.7 \pm 1.9$  D, and with their primary gaze position, mean horizontal sim K value was  $40.5 \pm 1.8$  D, and vertical sim K value was  $42.7 \pm 1.8$  D. Statistically significant horizontal flattening was noted in primary gaze position (p= 0.009). No statistically significant difference was found between the measurements of horizontal and vertical sim K values of control patients in different gaze positions. Conclusion: Duane retraction syndrome is a rare congenital ocular motility disorder. Although the pathogenesis is not clear, abnormal innervation of the extraocular muscles is mostly blamed. Most of the patients with Duane retraction syndrome develope a face turn position to achieve binocular vision. In our study, we concluded that eyelid position and the changes in corneal topography might affect the development of face turn position.

Key Words: Duane retraction syndrome; astigmatism; eyelids; corneal topography

ÖZET Amaç: Duane retraksiyon sendromu olan vakalarda farklı bakış pozisyonlarında korneal topografik değişiklikleri değerlendirmek ve korneal topografide göz kapaklarının etkisini arastırmak. Gereç ve Yöntemler: Duane retraksiyon sendromu olan 33 vaka ve 19 kontrol vakası çalışmaya alındı. Duane retraksiyon sendromu olan vakaların kornea topografileri önce kendi tercih ettikleri bakış pozisyonunda, sonra da primer bakış pozisyonunda ölçüldü. Kontrol grubundaki vakaların sağ gözlerinde gözlerinde addüksiyonda ve primer bakış pozisyonda topografik ölçümler yapıldı. Bulgular: Duane retraksiyon sendromu olan vakaların tercih edilen bakış pozisyonunda vertikal sim K değeri 42.7 ± 1.9 D iken, ortalama horizontal sim K değeri  $41.4 \pm 2.3$  D olup, primer bakış pozisyonda ortalama horizontal sim K değeri  $40.5 \pm 1.8$  D ve vertikal sim K değeri 42.7 ± 1.8 D bulundu. Primer bakış pozisyonunda horizontal eksende istatistiksel olarak anlamlı düzleşme saptandı (p= 0.009). Farklı bakış pozisyonlarında, kontrol hastalarının horizontal ve vertikal sim K değerlerinin ölçümleri arasında istatistiksel olarak anlamlı bir fark saptanmadı. Sonuç: Duane retraksiyon sendromu nadir görülen bir konjenital oküler hareket bozukluğudur. Patogenezi tam olarak açıklanamamasına rağmen, sıklıkla ekstraoküler kasların anormal inervasyonu suçlanmaktadır. Duane retraksiyon sendromu bulunan vakaların çoğu binoküler görmeyi sağlamak için baş pozisyonu geliştirirler. Çalışmamız sonucunda göz kapağı pozisyonunun ve korneal topografideki değişikliklerin baş pozisyonu gelişimininde etkili olabileceği düşünülmektedir.

Anahtar Kelimeler: Duane retraksiyon sendromu; astigmatizm; göz kapakları; kornea topografisi

doi:10.5336/medsci.2009-14148

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Turkiye Klinikleri J Med Sci 2011;31(3):641-5

Turkiye Klinikleri J Med Sci 2011;31(3) 641

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he factors that affect corneal topography have not been fully discovered yet. It is believed that corneal topography and, in turn, astigmatism are related to factors such as age, ethnicity and genetics.<sup>1,2</sup> On the other hand, there is limited information available regarding the changes displayed by corneal topography in relation to external factors such as eyelids and extraocular muscles. Various studies have demonstrated that the pressure created by the lids on the cornea can cause topographic changes.<sup>3,4</sup> Gresvenor concluded that astigmatism observed in young adults was caused by the band-like pressure of the upper tarsal plate, while astigmatism observed in the older population was caused by the slackening of upper tarsal plate due to age.5

Extraocular muscles can affect corneal topography especially in cases who undergo strabismus surgery. Corneal topography has been shown to become flatter where the muscle is recessed due to decreased muscle tension and steeper where the muscle is resected due to higher muscle tension.<sup>6-8</sup>

Duane retraction syndrome is a congenital syndrome characterized by globe retraction caused by the co-contraction of the medial and lateral rectus muscles and the narrowing of the palpebral fissure. However, the pathogenesis has not yet been fully explained. The cases usually develop a particular head position in order to maintain their binocular vision.<sup>9,10</sup> Duane retraction syndrome is classified into three types; Type 1 is marked limitation of abduction, explicable by maximum innervation of the lateral rectus muscle only when the affected eye is adducted; type 2 is limitation of adduction; and type 3 is limitation of both adduction and abduction. In this study, changes in corneal topography with different gaze positions, which we considered to be effective in the development of face turn position in cases with type 1 Duane retraction syndrome have been analyzed.

# MATERIAL AND METHODS

Thirty-three cases with Type I Duane retraction syndrome followed by the Orthoptics Department in Istanbul University, Istanbul Faculty of Medicine were included in the study. Informed consent was obtained from all patients. The study protocol followed the guidelines of the Declaration of Helsinki. Of 33 cases, 12 were male and 21 were females. The mean age of the cases was found to be  $14.7 \pm 5$  (range: 6-23) years. All cases were subjected to detailed ophthalmologic and orthoptic examination prior to topographic measurement. Average visual acuity of the cases was determined as  $0.8 \pm 0.2$  (0.3-1.0) based on the Snellen scale. During orthoptic examination, all cases demonstrated narrowing of the palpebral fissure, globe retraction on adduction, and limitation of abduction of the affected eye. All cases were esotropic and had a face turn to affected side. None of the cases had undergone strabismus surgery. A total of 19 cases with no ocular or systemic diseases were included in the study as the control group. Mean age of the controls was  $23.8 \pm 6$  (range: 17-29) years.

Pentacam Scheimpflug (Oculus, Germany) imaging was used for the topographical measurements. Topographies of the cases with Duane retraction syndrome were first measured during their preferred gaze and then during their primary gaze position. Topographies of the control group were measured in their right eye during their primary gaze position and when the eye was on adduction. During topography measurements, horizontal and vertical sim K values and cylindrical measurement were taken into account. Measurements of the cases with different gaze positions were compared.

Statistical analysis was performed with the Student's t-test to compare the measurements in different gaze positions. A p value less than 0.05 was considered as statistically significant result.

### RESULTS

The mean horizontal sim K value of the cases with Duane retraction syndrome during preferred gaze position was  $41.4 \pm 2.3$  D, while the vertical sim K value was  $42.7 \pm 1.9$  D, and during the primary gaze position, the mean horizontal sim K value was  $40.5 \pm 1.8$  D, and the vertical sim K value was  $42.7 \pm 1.8$  D. When the results obtained during two different gaze positions were compared, a significant difference was noted in horizontal sim K values (p=

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0.009). When the head was brought to primary gaze position, a flattening in the corneal topography on the horizontal plane was noted compared to the preferred gaze position (Figures 1a,b, 2a,b). When vertical sim K values were compared in both gaze positions, no significant difference was observed (p> 0.05). While the mean cylindrical value measured in the preferred gaze position of the cases was

-1.26 D, it was measured as -1.78 D in the primary gaze position (Table 1).

The mean horizontal sim K value of the cases in the control group in the primary gaze position was  $40.7 \pm 0.9$  D, while the vertical sim K value was  $41.6 \pm 0.9$  D, and the mean horizontal sim K value was  $40.8 \pm 0.8$  D on adduction, while the vertical sim K value was  $41.6 \pm 0.9$  D. No significant diffe-

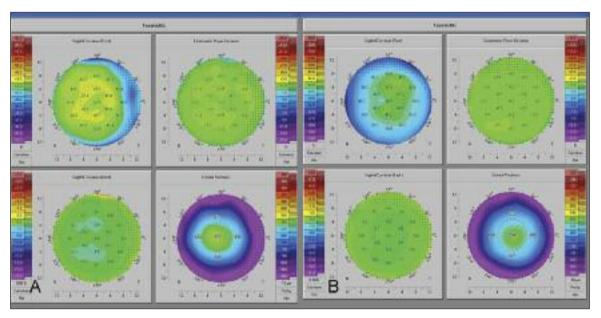


FIGURE 1a: Topographic image of the patient with Duane retraction syndrome in preferred gaze position, 1b: Corneal flattening in primary gaze position.

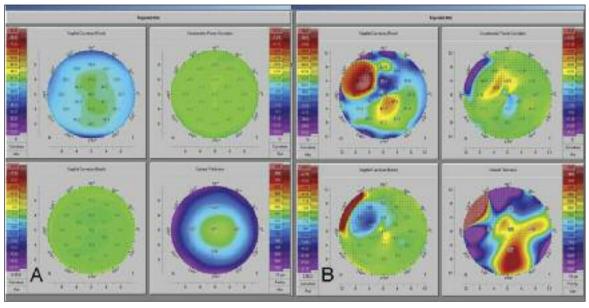


FIGURE 2a: Topographic image of the patient with Duane retraction syndrome in preferred gaze position, 2b: Note the lid pressure on cornea and flattening effect in primary gaze position.

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<b>TABLE 1:</b> Mean horizontal and vertical sim K values with cylindrical values in different gaze positions of patients in both
groups. Note the flattening of the cornea on primary gaze position in Duane retraction syndrome patients

	Duane retraction syndrome group No:33 eyes		Control group No: 19 eyes	
	Preferred gaze	Primary gaze	Primary gaze	Adduction
Horizontal sim K value	41.4±2.3 D	40.5±1.8 D p=0.009	40.7 ±0.9 D	40.8±0.8 D p=0.07
Vertikal sim K value	42.7±1.9 D	42.7±1.8 D p>0.05	41.6±0.9 D	41.6±0.9D p>0.05
Cylindrical value	-1.26 D	-1.78 D p>0.05	-0.80 D	-0.93 D p>0.05

rence was observed between vertical sim K values (p> 0.05) and horizontal sim K values (p= 0.07). In the control group, the mean cylindrical value in the primary gaze position was -0.80 D, while it was -0.93 D on adduction. No statistically significant difference was observed between these two values (p> 0.05).

# DISCUSSION

Duane retraction syndrome is a syndrome in which innervational and central nervous system abnomalies play a role. It is believed that innervational abnomalies of the lateral rectus muscle and paradoxical innervation are responsible for the clinically observed contraction. In cases with Duane retraction syndrome, a face turn position is a common clinical symptom. Patients develop a face turn position in order to achieve binocular single vision and even bifoveal fixation. In this syndrome, abnormal muscle function caused by abnormal innervation of the extraocular muscles changes the palpebral fissure size in different gaze positions and also affects face turn.

Various studies have demonstrated that eyelids affect corneal astigmatism. Shaw et al. indicated that corneal topography can change in adults, downward gaze positions, while Read et al. noted a significant relationship between the change in horizontal lid position and corneal spherocylindrical value. <sup>11,12</sup> In another study where palpebral aperture was measured by changing the lid position, astigmatism up to 2 D was noted by narrowing of the palpebral fissure. <sup>3</sup> In their study, Collins et al. informed that corneal topographical changes were observed mostly with downward and abduction gaze positions and that the topographical changes created by the lids

on the cornea were observed mostly on the vertical meridian due to the position of the lids.<sup>13</sup>

On the other hand, the effects of the extraocular muscles on corneal topography were evaluated in the patients with strabismus surgery. Kwitko et al. noted significant corneal flattening of the upper and upper temporal region in rabbit eyes when they recessed the upper rectus.8 Bagheri et al. noted flattening and astigmatism changes after recession of multiple horizontal rectus muscles in the patients with nystagmus.<sup>6</sup> They indicated that the reason for this effect was a decrase in the mechanical advantage of the horizontal rectus muscles, compared with the vertical rectus muscles, in exerting tension on the anterior globe. In addition, they informed that resection of extraocular muscles had a smaller impact on corneal topography.6

In cases with Duane retraction syndrome, corneal topography is not affected by only a single factor. Since lid position changes depending on the gaze position and extraocular muscle tension is higher with certain gaze positions, many factors can influence topography. Since we have determined significant differences, particularly in the horizontal sim K value, we think that muscle co-contraction might be responsible for these changes. The cylindrical refraction change may occur due to narrowing of the palpebral aperture. Eyelids may have a molding effect on the cornea. The fact that an increase in cylindrical values is observed as a result of a change in corneal topography might be influential in developing a face turn position to decrease the effect of cylindrical refractive error and improve binocularity. Shortcoming of our study is that face turn of the cases was not always to the saOphthalmology Yeniad et al

me degree, and thus, we have obtained very different sim K measurements in certain cases. Duane retraction syndrome has three subtypes, each subtype has different limitation of extraocular movement. Type 1 is the most frequent type (70-80%). Therefore we included only patients with type 1 in this study. Studies with a larger population and other subtypes are needed in order to clearly demonstrate the relationship between corneal topography and face turn position.

## CONCLUSION

The patients with Duane retraction syndrome commonly have a face turn to achieve binocular single vision. In our study, we conclude that changes in corneal topography and lid position may have an effect on the preferred gaze position in patients with Duane retraction syndrome. Studies with larger case numbers may clearly demonstrate this effect.

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