Effect of congenital ptosis correction on corneal topography- A prospective study

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Purpose: The aim of this study was to evaluate the changes in corneal topography, cycloplegic refraction, and best-corrected visual acuity (BCVA) after ptosis correction surgery in patients with congenital ptosis. **Methods:** Our study represents a prospective observational study conducted on 27 eyes of 21 patients with congenital ptosis. All patients underwent complete ophthalmological evaluation, cycloplegic refraction, and baseline Orbscan prior to ptosis surgery. At 6 months postoperative review, the cycloplegic refraction and Orbscan were repeated to evaluate the changes in these parameters. The main outcome measures in our study were Steepest K, Inferior-Superior Asymmetry (I-S Asymmetry), cycloplegic refraction and BCVA. **Results:** A significant decrease in Steepest K postoperatively ($P < 0.001$) was noted. Superior K and Inferior K also decreased, but the decrease in Inferior K was statistically significant ($P = 0.044$). However, change in I-S Asymmetry was not significant. Variation in BCVA, and cycloplegic sphere and cylinder was minimal. Sim K astigmatism, Surface Regularity Index, I-S Asymmetry and Central Corneal Thickness did not show significant variation. **Conclusion:** Ptotic eyelid constantly presses on the cornea causing significant changes in corneal contour and surface remodeling. This pressure when relieved, results in significant flattening and regression of anterior corneal surface to its near normal anatomy. This further resulted in improvement of corneal surface irregularity and symmetry.

Access this article online

Website:
www.ijo.in

DOI:
10.4103/ijo.IJO_2650_20

Quick Response Code:

Key words: Blepharoptosis, congenital ptosis, corneal topography, Frontalis Sling, Orbscan

Congenital ptosis is defined as drooping of the upper eyelid since birth or within 1 year of age. The prevalence of simple congenital ptosis is 1 in 842 of which unilateral is more common.[1,2] Marcus Gunn Jaw Winking Syndrome (MGJWS) accounts for almost 5% of cases of congenital ptosis.

In a normal person, corneal topography of the two eyes are like mirror images. The eyelid pressure causes flattening of the peripheral cornea and steepening of the central cornea of the ptotic eye, leading to higher incidence of corneal astigmatism and loss of symmetry.[3,4]

Corneal topography of ptotic eye also shows more surface irregularity and asymmetry as compared to the normal eye.[4]

Various theories propose that upper eyelids cause a "band like pressure" on the cornea and may cause With the Rule astigmatism due to altered corneal shape.[5] Several studies have recorded the refractive error, astigmatism and corneal topography changes produced by the ptotic eyelid on the cornea.[3-20] Amblyopia has also been reported as a result of upper eyelid ptosis in children.[14,15]

Timing and need for intervention mainly depends on severity of ptosis, chances of developing amblyopia and also the other associated ocular and systemic syndromes.[2] Choice of surgery relies on factors like amount of ptosis and Levator Palpebrae Superioris (LPS) function. We included only patients with severe ptosis in our study for whom the surgery of choice was Frontalis Sling. Patients with MGJWS underwent Frontalis Sling with LPS disinsertion.

In our study, we analyzed the mechanical effects of severe ptosis on corneal curvature. We recorded baseline visual acuity, cycloplegic refraction and corneal topography of our patients and compared them with the postoperative data in order to study the effects of ptosis correction on these parameters.

Methods

Study population and methods

This is a prospective study conducted on 27 eyes of 21 patients who were diagnosed with congenital ptosis. The patients were recruited from the Orbit and Oculoplasty department of a tertiary eye hospital in South India between October 2017 and March 2019. Informed written consent was obtained from all patients who underwent the surgery and in case of minors, informed written consent of parent or guardian was obtained. Institutional Committee approval was considered in accordance with tenets of the Declaration of Helsinki.

The primary outcome measure was change in Steepest K and I-S Asymmetry (Inferior – Superior Asymmetry) postoperatively. Secondary outcome measure was change in BCVA and cycloplegic refraction post ptosis correction.

Patients aged between 4 and 25 years with severe ptosis obscuring the visual axis, or Marginal Reflex Distance 1 (MRD1) <+1 were included in the study. Patients less than four years were excluded in view of their inability to co-operate for the corneal topography. Patients with Marcus Gunn Syndrome

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Received: 15-Aug-2020 Revision: 15-Oct-2020
Accepted: 29-Jan-2021 Published: 21-May-2021

Cite this article as: Assadi FA, Narayana S, Yadalla D, Rajagopalan J, Joy A. Effect of congenital ptosis correction on corneal topography- A prospective study. Indian J Ophthalmol 2021;69:1527-30.

were also included in this study. Patients with other causes of ptosis like Mechanical, Neurological and Traumatic Ptosis; Ocular Myasthenia; Double Elevator Palsy; Neurofibromatosis; Blepharophimosis syndrome; corneal abnormalities due to vernal conjunctivitis and dry eye; other causes of decreased visual acuity like Congenital Cataract and Corneal Opacity; previous ptosis surgery; posttrauma to lid/cornea and all patients unable to cooperate for corneal topography were excluded from the study.

Patients with Isolated Congenital Ptosis underwent Frontalis Sling surgery whereas patients with Marcus Gunn Jaw Winking Syndrome underwent LPS disinsertion with Frontalis Sling Surgery. In cases of bilateral congenital ptosis, each eye was considered separately. Both eyes were operated at the same time in order to avoid the need for general anesthesia twice. The surgeries were performed by only two experienced surgeons to get best possible surgical outcome. Frontalis Sling was done using Fox Pentagon technique. Two eyelid crease incisions, two brow incisions and one incision on the forehead in between the brow incisions were marked. Stab incisions were made through all the marks. Silicon sling (Aurolab, India) was passed through the incisions deep to the orbicularis plane, and sling tightened till desired height was achieved. Frost suture was placed at the end of the surgery.

All patients underwent complete ophthalmologic evaluation including Best-corrected visual acuity (BCVA), cycloplegic refraction, anterior segment and fundus evaluation preoperatively. Cycloplegic refraction was done by an experienced refractionist using Snellen's chart. For statistical calculations, visual acuity was expressed in log MAR. MRD1 and LPS function was measured. Corneal topography was recorded using Orbscan 3 (Bausch and Lomb) by a trained technician. Corneal Topography parameters that were included in the study were $SimK_{max}$, $SimK_{min}$ in the central 3 mm of cornea, Simulated K Astigmatism (Sim K Astigmatism), Steepest K, SRI (Surface Regularity Index) at 3 mm and 5 mm zone, S-I Symmetry Index, Central Corneal Thickness (CCT). Steepest K was defined as the steepest point on the anterior surface of cornea using Tangential map. Location of the steepest point with respect to the distance from center of cornea and axis in degrees was recorded preoperatively and also post operatively to compare the difference. Inferior-Superior Asymmetry Index (I-S Asymmetry Index) was defined as difference between Inferior K and Superior K along the central vertical axis using tangential map. I-S Asymmetry <0 indicates steep superior cornea whereas I-S Asymmetry >0 indicates steep inferior cornea. Tangential keratometric map was used for deriving the Steepest K, Superior K, Inferior K and I-S Asymmetry Index. All patients underwent ptosis correction surgery.

At the sixth month review, Orbscan was repeated; BCVA, cycloplegic refraction, and MRD1 were evaluated. The preoperative and postoperative data were then analyzed and compared. Orbscan analysis was done by SN, a cornea surgeon.

Statistical analysis

Data was presented in the form of mean ± standard deviation for approximately normally distributed continuous variables or median and inter-quartile range for skewed data. Categorical variables were presented in frequencies and percentages. In each analysis, parametric or non-parametric tests were employed based on the results of normality test. A Wilcoxon sign rank test was used to compare the postoperative visual acuity and spherical equivalent with baseline measurements because it was not distributed normally, as judged by the Shapiro-Wilk test. The differences of corneal topographic measurements between the baseline and follow-up were tested with paired *t* test. The statistical analyses were performed using STATA, 14.0 (Texas, USA); a two-sided *P* value less than 0.05 was considered to represent statistical significance.

Results

The study included 27 eyes of 21 patients with congenital ptosis, of which 15 (71.4%) were unilateral and 6 (28.6%) were bilateral. The mean age of patients recruited was 11.62 ± 4.4 (range; 5 to 21 years). 12 (57.1%) of the subjects were males and 9 (42.9%) were females. 3 (14.3%) of these patients had MGJWS. LPS function was poor in all except one eye which had fair LPS function [Table 1].

Only 16 patients (21 eyes) came for follow up, hence only these 21 eyes were included for pre and postoperative data comparison.

Median of MRD1 at preop was -2 mm which significantly improved to + 3.5 mm postoperatively ($P < 0.001$). Mean BCVA preoperatively was logMAR 0.05 ± 0.1 which changed to logMAR0.07 ± 0.1 in the postoperative period. No significant change in BCVA was noted. The mean changes in cycloplegic sphere and cylinder were also statistically insignificant postoperatively [Table 2].

We noted a significant decrease in mean Steepest K from a baseline mean value of 45.59 ± 2.1 to 44.03 ± 1.9 postoperatively ($P < 0.001$) indicating flattening at the same point as depicted in Figs. 1 and 2. K-min and K-max decreased but not significantly. Baseline mean Sim K corneal astigmatism was –0.70 which decreased to -0.60 after the ptosis correction [Table 3].

We noted a decrease in Superior K from 44.20 ± 2.1 to 43.66 ± 1.8 indicating flattening postoperatively but the decrease was statistically insignificant. There was a statistical significant decrease in Inferior K from 44.99 ± 2.1 to 44.07 ± 1.6 ($P = 0.044$). This indicates that there was a generalized flattening of the cornea after surgery with predominant inferior flattening [Table 3].

I-S Asymmetry index in both baseline and postop operative scans show values >0 indicating steep inferior corneal

Table 1: Demographic characteristics of the study participants

Variable	Frequency (*n*=21)	Percentage
Age, years		
Mean±SD	11.62±4.4	-
Min to Max	5 to 21	
Gender		
Male	12	57.1
Female	9	42.9
Laterality		
Unilateral	15	71.4
Bilateral	6	28.6
Eye		
Right	14	51.8
Left	13	48.2
MGJWS		
Present	3	14.3
Absent	18	85.7
LPS function*		
Fair	1	3.7
Poor	26	96.3

*No. of eyes=27. MGJWS-Marcus Gunn Jaw Winking Syndrome, LPS - Levator Palpebrae Superioris hemisphere. Though there was a postoperative decrease in I-S Asymmetry index from 0.75 to 0.58, it was statistically insignificant. Other variables like SRI (at 3 mm and 5 mm) and CCT were also compared in our study. No significant variation was noted in these values [Table 3].

Discussion

Upper eyelid pressure on the cornea induces astigmatism and refractive error changes. Ptosis not only causes lower order aberrations but also results in higher order aberrations in the eye.[10] Ugurbas *et al*.[4] studied the various corneal topography patterns in patients with ptosis. It was noted that the mirror like symmetry between the two corneas was lost in eyes with unilateral ptosis. This study also concluded that ptotic eyes had increased incidence of astigmatism, corneal irregularity (SRI, $P < 0.05$) and corneal asymmetry (SAI, $P < 0.05$). In literature, there are several studies that have documented corneal topography changes in patients with lid abnormalities like lid hemangiomas, involuntary eyelid spasms, chalazia, ptosis and ectropion.[21-24] It is thus conclusive that corneal topography to a great extent is influenced by eyelid morphology and position.

In our study, we noted that the mean I-S Asymmetry index was >0 both pre and postoperatively, indicating a steeper inferior cornea. In contrary, Karabulut[20] *et al.* noted a steeper superior cornea in both pre and postoperative topographic scans. This contrasting result is attributable to the difference in inclusion criteria of both these studies. Our study recruited patients with only severe ptosis whereas Karabulut *et al.*[20] enrolled patients with only mild ptosis. The steepest point of the anterior corneal surface is located under the ptotic eyelid margin.[3] Hence, due to the difference in eyelid margin position, severe ptosis results in steeper inferior cornea while in mild ptosis, the superior cornea would be steeper. We also studied changes in I-S Asymmetry occurring postoperatively. Though there was a decrease, it was statistically insignificant, which was comparable to the results of previous studies.[3,20]

Surgical ptosis repair resulted in a significant decrease of Steepest K ($P < 0.001$) indicating flattening. Two previous studies by Savino *et al.*[3] and Karabulut *et al.*[20] also reported a similar decrease in Apical Keratometry front (Steepest K) in the postoperative period. Our study though reported a decrease in Sim K astigmatism, the decrease was statistically insignificant. Other authors also reported no significant change in Sim K Astigmatism.[3,20]

Furthermore, for the first time we also studied the changes in Superior K and Inferior K independently. Both these parameters decreased after surgery but the decrease in Inferior K was statistically significant. Therefore, we infer that surgery resulted in global flattening of the cornea, but more in the inferior corneal hemisphere. The surgical repositioning of the upper lid effects the lid/cornea interaction by elevating the lid that had been constantly pressing on the cornea, thus altering corneal contour. Other corneal topography metrics like $SimK_{min}$, $SimK_{max}$, CCT, SRI at 3 mm and 5 mm were also compared but we did not note any significant variations. The postoperative changes in BCVA, cycloplegic sphere and cylinder was minimal in our study similar to the results in previous studies.[13,20]

In our study, objective documentation of corneal topography changes has been done using Orbscan reports. Corneal surface changes were analyzed using data from Topography which is more sensitive and definitive. There is paucity in literature regarding corneal topography changes post ptosis surgery, especially in severe congenital ptosis; hence, this study contributes to it.

Table 2: Postoperative comparison of visual acuity and cycloplegic refraction

	n	Mean±SD	Median	IQR	*P*[a]
MRD1					
Baseline	27	-1.44±1.2	-2.0	-2.0-1.0	<0.001
Postop	21	3.36±0.8	3.5	3.0-4.0	
logMAR UCVA					
Baseline	27	0.27±0.3	0.18	0-0.48	0.886
Postop	27	0.26±0.3	0.18	0-0.48	
logMAR BCVA					
Baseline	27	0.05±0.1	0	0-0	0.083
Postop	21	0.07±0.1	0	0-0.18	
Sphere					
Baseline	26	-0.28±1.0	0	0-0	0.099
Postop	20	0.06±0.8	0	0-0.5	
Cylinder					
Baseline	26	-0.15±0.7	0	0-0	0.149
Postop	19	-0.05±0.9	0	0-0	
Spherical equivalent					
Baseline	26	-0.36±1.1	0	-0.75-0.375	0.073
Postop	20	0.04±0.9	0	-0.375-0.5	

[a]Wilcoxon sign rank test, IQR - Inter-quartile range, MRD1 - Marginal Reflex Distance1, UCVA - Uncorrected visual acuity, BCVA - Best-corrected visual acuity

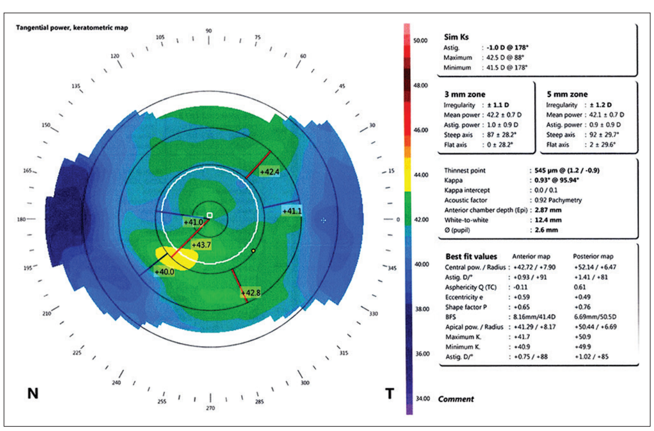

Figure 1: Preoperative Orbscan Image

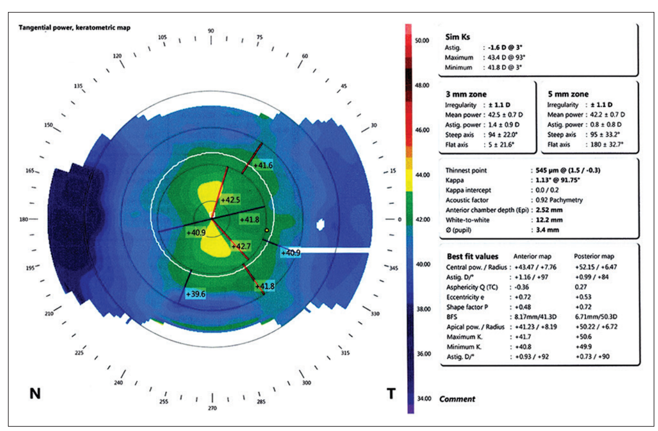

Figure 2: Postoperative Orbscan Image

Table 3: Comparison of Corneal Topographic measurement at baseline and postsurgery

	n	Mean±SD	Min-Max	P^b
K min				
Baseline	27	43.70±1.8	40.8-47.8	0.762
Postop	21	43.37±1.5	41.0-45.8	
K max				
Baseline	27	44.62±1.9	41.8-49.0	0.501
Postop	21	44.29±1.5	41.5-47.4	
Sim K Astigmatism*				
Baseline	27	-0.70-0.60	-1.1-0.5	0.651[a]
Postop	21		-1.1-0.4	
Steepest K				
Baseline	27	45.59±2.1	41.94-49.72	<0.001
Postop	21	44.03±1.9	41.05-47.09	
SRI 3mm				
Baseline	27	1.37 (0.6)	0.20-3.00	0.223
Postop	21	1.24 (0.5)	0.60-2.70	
SRI 5mm				
Baseline	27	1.45 (0.5)	0.60-2.70	0.276
Postop	21	1.35 (0.4)	0.70-2.40	
Superior K				
Baseline	27	44.20±2.1	40.36-48.40	0.488
Postop	21	43.66±1.8	40.07-47.08	
Inferior K				
Baseline	27	44.99±2.1	41.85-49.72	0.044
Postop	21	44.07±1.6	41.61-46.91	
CCT				
Baseline	27	577.96±44.8	474-654	0.145
Postop	21	580.67±42.3	488-653	
I-S Asymmetry*				
Baseline	27	0.75	-0.20-1.20	0.664[a]
Postop	21	0.58	-0.21-1.20	

[a]Wilcoxon sign rank test, [b]Paired *t*-test. *Variables (astigmatism and I-S) were presented with median and inter-quartile range, SRI - Surface Regularity Index, CCT - Central corneal thickness, I-S Asymmetry - Inferior Superior Asymmetry

Our study has potential limitations. This is a type of prospective observational study which is not randomized. Orbscan has its own limitations such as poor repeatability as compared to Pentacam. Also, the sample size is relatively small, the main reason being that most children with pupil obscuring ptosis were operated at an early age of less than four years as they had a chance of developing amblyopia. These children were excluded from the study as they could not co-operate for Orbscan. Few children were lost to follow up after surgery. Another reason for the small sample size is the exclusion of children with mild to moderate ptosis as these children had good vision and were on regular follow up. Surgery in these children was indicated only for cosmetic reasons on a later date.

Conclusion

In conclusion, our study suggests that the ptotic eyelid constantly presses on the cornea causing significant changes in corneal contour and surface remodeling. This pressure when relieved resulted in significant flattening and regression of anterior corneal surface to its near normal anatomy. This further resulted in improvement of corneal surface irregularity and symmetry.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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