Intraocular pressure, corneal thickness, and corneal hysteresis in Steinert's myotonic dystrophy

Pressão intraocular, espessura corneal e histerese corneal em distrofia miotônica de Steinert

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ABSTRACT

Purpose: Low intraocular pressure (IOP) measured by Goldmann applanation tonometry (GAT) is one of the ocular manifestations of Steinert's myotonic dystrophy. The goal of this study was to evaluate the corneal-compensated IOP as well as corneal properties (central corneal thickness and corneal hysteresis) in patients with myotonic dystrophy.

Methods: A total of 12 eyes of 6 patients with Steinert's myotonic dystrophy (dystrophy group) and 12 eyes of 6 age-, race-, and gender-matched healthy volunteers (control group) were included in the study. GAT, Dynamic Contour Tonometry (DCT-Pascal) and Ocular Response Analyzer (ORA) were used to assess the IOP. Central corneal thickness was obtained by ultrasound pachymetry, and corneal hysteresis was analyzed using the ORA device. In light of the multiplicity of tests performed, the significance level was set at 0.01 rather than 0.05.

Results: The mean (standard deviation [SD]) GAT, DCT, and corneal-compensated ORA IOP in the dystrophy group were 5.4 (1.4) mmHg, 9.7 (1.5) mmHg, and 10.1 (2.6) mmHg, respectively. The mean (SD) GAT, DCT, and corneal-compensated ORA IOP in the control group was 12.6 (2.9) mmHg, 15.5 (2.7) mmHg, and 15.8 (3.4) mmHg, respectively. There were significant differences in IOP values between dystrophy and control groups obtained by GAT (mean, -7.2 mmHg; 99% confidence interval [CI], -10.5 to -3.9 mmHg; P<0.001), DCT (mean, -5.9 mmHg; 99% CI, -8.9 to -2.8 mmHg; P<0.001), and corneal-compensated ORA measurements (mean, -5.7 mmHg; 99% CI, -10.4 to -1.0 mmHg; P=0.003). The mean (SD) central corneal thickness was similar in the dystrophy (542 [31] µm) and control (537 [11] µm) groups (P=0.65). The mean (SD) corneal hysteresis in the dystrophy and control groups were 11.2 (1.5) mmHg and 9.7 (1.2) mmHg, respectively (P=0.04).

Conclusions: Patients with Steinert's myotonic dystrophy showed lower Goldmann and corneal-compensated IOP in comparison with healthy individuals. Since central corneal thickness and corneal hysteresis did not differ significantly between groups, the lower IOP readings documented in this dystrophy seem not to be related to changes in corneal properties.

Keywords: Intraocular pressure; Tonometry, ocular; Myotonic dystrophy; Ocular hypotension; Cornea; Corneal topography

RESUMO

Objetivos: *Pressão intraocular (PIO) baixa medida por meio da tonometria de aplanação de Goldmann (TAG) é uma das manifestações oculares da distrofia miotônica de Steinert. O objetivo deste estudo foi avaliar a pressão intraocular compensada para as propriedades corneais (espessura corneal central e histerese corneal) em pacientes com distrofia miotônica.*

Métodos: *Um total de 12 olhos de 6 pacientes com distrofia miotônica de Steinert (grupo distrofia) e 12 olhos de 6 voluntários sadios (grupo controle) pareados para idade, raça e sexo foram incluídos no estudo. Tonometria de aplanação de Goldmann, tonometria de contorno dinâmico (TCD-Pascal) e analisador de resposta ocular (ORA) foram usados para medir a pressão intraocular. A espessura corneal central foi obtida por meio da paquimetria ultrassônica e a histerese corneal foi analizada usando o aparelho ORA.*

Resultados: *A pressão intraocular média (desvio-padrão) da TAG, TCD e compensada para a córnea do ORA no grupo distrofia foram 5,4 (1,4) mmHg, 9,7 (1,5) mmHg e 10,1 (2,6) mmHg, respectivamente. A pressão intraocular média (desvio-padrão) da TAG, TCD e compensada para a córnea do ORA no grupo controle foram 12,6 (2,9) mmHg, 15,5 (2,7) mmHg e 15,8 (3,4) mmHg, respectivamente. Houve diferença significativa nos valores da pressão intraocular entre os grupos distrofia e controle obtidas pela TAG (média, -7,2 mmHg; intervalo de confiança (IC) de 99%, -10,5 a -3,9 mmHg; P<0,001), TCD (média, -5,9 mmHg; IC de 99%, -8,9 a -2,8 mmHg; P<0,001) e ORA compensada para córnea (média, -5,7 mmHg; IC de 99%, -10,4 a -1,0 mmHg; P=0,003). A espessura corneal média (desvio-padrão) foi similar nos grupos distrofia (542 [31] µm) e controle (537 [11] µm) (P=0,65). A histerese corneal média (desvio-padrão) nos grupos distrofia e controle foram de 11,2 (1,5) mmHg e 9,7 (1,2) mmHg, respectivamente (P=0,04).*

Conclusão: *Os pacientes com distrofia miotônica de Steinert apresentaram valores menores de pressão intraocular medidas tanto com Goldmann quanto compensadas para a córnea em comparação com indivíduos sadios. Uma vez que os valores da espessura corneal central e histerese corneal não diferiram significantemente entre os grupos, os valores baixos da pressão intraocular encontrados nos pacientes com distrofia miotônica não parecem estar relacionados com as propriedades corneais.*

Descritores: *Pressão intraocular; Tonometria ocular; Distrofia miotônica; Hipotensão ocular; Córnea; Topografia da córnea*

INTRODUCTION

Steinert´s myotonic dystrophy is a multisystemic disease inherited in an autossomal dominant pattern that affects squeletal musculature, cardiac conduction system and endocrine system, and has neurologic and ocular alterations. It is the most common muscular dystrophy with a prevalence of 3-5-cases/100.000 births[1].

Ocular involvement includes cataract, ptosis, retinal abnormalities and ocular hypotony[2-4]. Low intraocular pressure (IOP) is a common finding in myotonic dystrophy patients, but its pathophysiologic mechanism remains unclear. Jung[5] suggested that the hypotony was secondary to a high uveal-scleral outflow. Other authors[6,7] suggested an association of the high uveal-scleral outflow with a reduced production of aqueous humor secondary to ciliary body atrophy. In these studies, Goldmann applanation tonometry (GAT) was used to measure the IOP.

GAT is considered the gold standard method to evaluate IOP, but it is influenced by central corneal thickness (CCT) and curvature as well as axial length of the eye[8,9]. More recently, corneal biomechanical properties have been evaluated using the Ocular Response Analyzer (ORA), which measures the corneal hysteresis (CH) - an

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estimation of the viscous damping properties of the cornea[10,11]. The ORA and the dynamic contour tonometry (DCT) can provide corneal-compensated IOP measurements.

The goal of this study was to evaluate the Goldmann and corneal-compensated IOP, CCT and CH in patients with myotonic dystrophy and healthy individuals and to analyze whether corneal thickness and biomechanical properties were associated with the low IOP found by GAT in this disease.

METHODS

This cross-sectional study was carried out at the Department of Ophthalmology of the Federal University of São Paulo. The study protocol adhered to the tenets of the Declaration of Helsinki and was approved by the Institutional Ethics Committee. Written informed consent was obtained from all subjects.

Participants

A total of 12 eyes of 6 patients followed at the Neurology Department of the Federal University of São Paulo with clinical diagnosis of Steinert's myotonic dystrophy were included in the dystrophy group. The diagnosis was based on family history, typical muscle findings and electromyography, and confirmed by genetic analysis. Twelve eyes of 6 age (within 5 years difference), race, and gender-matched healthy volunteers were included in the control group.

All participants underwent a complete ophthalmological examination including biomicroscopy and fundus examination using a 78-Diopter lens. Patients with any significant ocular disease, history of contact lenses or ocular medication use, previous eye trauma or intraocular surgery were excluded.

Procedures

IOP was recorded with Goldmann applanation tonometry (GAT; Haag-Streit, Bern, Switzerland), DCT (PASCAL®, Ziemer Ophthalmic System, Port, Switzerland) and ORA (Reichert Inc., Depew, USA) in random order for each participant. For the ORA device, the corneal-compensated IOP reading, which is a pressure measurement that compensates the biomechanical properties of the cornea, was evaluated. Central corneal thickness was obtained by ultrasound pachymetry (Corneo gage plus, Sonogage, Cleveland, Ohio) and CH was evaluated using the ORA. Three good quality measurement of each device were taken and the mean measurements were used for the analysis. For the DCT measurements, readings with a quality score ≤3 were excluded. Readings from the ORA required consistent and smooth raw signal morphology (clean, sharp, well-defined raw signal peaks, with repeatable characteristics in multiple measurements).

Statistical analysis

Corneal parameters and IOP values were compared using paired t-test. In light of the multiplicity of tests performed, the significance level was set at 0.01 rather than 0.05, and, accordingly, 99% rather than 95% confidence interval (CI) was provided.

RESULTS

The mean (range) age was 41.2 (31 to 53) years in the dystrophy group and 44.5 (27 to 55) years in the control group. There were 3 males and 3 females in each group.

Anterior biomicroscopy and fundus examination were normal in 5 out of 6 patients in the study group. One patient presented a mild nuclear cataract that did not require any further intervention and did not interfere with IOP measurements.

The mean (standard deviation [SD]) GAT, DCT, and corneal-compensated ORA IOP in the dystrophy group were 5.4 (1.4) mmHg, 9.7 (1.5) mmHg, and 10.1 (2.6) mmHg, respectively. The mean (SD) GAT, DCT, and corneal-compensated ORA IOP in the control group was 12.6 (2.9) mmHg, 15.5 (2.7) mmHg, and 15.8 (3.4) mmHg, respectively. The differences in IOP values between dystrophy and control groups for GAT (mean, -7.2 mmHg; 99% CI, -10.5 to -3.9 mmHg; P<0.001), DCT (mean: -5.9 mmHg; 99% CI, -8.9 to -2.8 mmHg; P<0.001), and corneal-compensated ORA IOP measurements (mean, -5.7 mmHg; 99% CI, -10.4 to -1.0 mmHg; P=0.003) were statistically significant.

The mean (SD) CCT was similar between dystrophy (542 [31] µm) and control groups (537 [11] µm) (P=0.65). The mean (SD) corneal hysteresis in the dystrophy and control groups were 11.2 (1.5) mmHg and 9.7 (1.2) mmHg, respectively (P=0.04).

DISCUSSION

In this study, the IOP was significantly lower in patients with myotonic dystrophy compared with healthy subjects when measured by GAT as well as by instruments that compensate for corneal properties (ORA and DCT). This finding is in agreement with several other studies that reported low IOP using GAT[2-6,12,13]. One study, conducted by Rosa et al.[13], also found lower IOP in dystrophy patients using ORA. No publication using DCT to measure IOP in myotonic dystrophy was found.

No significant difference in CCT values between patients with and without myotonic dystrophy was found in this study. Kesler et al.[12], also found similar CCT in patients with myotonic dystrophy and healthy subjects. In contrast, Rosa et al.[13], found thicker cornea in myotonic dystrophy patients in comparison with healthy individuals, but the difference of 20 µm between the groups was not clinically important.

The difference in corneal hysteresis was not statistically significant in the dystrophy group compared with control group. This finding is consistent with those recently published by Rosa et al.[13].

A limitation of this study includes the small sample size and, therefore, it should be replicated with larger sample. In addition, other IOP-related factors such as ciliary body structural and functional integrity could also be evaluated in future studies.

CONCLUSION

In summary, patients with Steinert's myotonic dystrophy had lower Goldmann and corneal-compensated IOP than healthy individuals. Since corneal parameters (CCT and CH) did not differ significantly between groups, the lower IOP readings documented in these patients seem not to be related to changes in corneal properties. Other factors not evaluated in this study could be implicated in the lower IOP observed in this disease.

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