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RESEARCH REPORT

Differences of the Anterior Segment Parameters in Children with Down Syndrome

Lokman Aslan[1], Murat Aslankurt[1], Adnan Aksoy[1], and Yakup Gümüşalan[2]

[1]*Ophthalmology Department and* [2]*Anatomy Department, KSU Faculty of Medicine, Kahramanmaraş, Turkey*

ABSTRACT

Purpose: The study was undertaken to investigate whether anterior segment findings are different in children with Down syndrome (DS) to normal children and to focus on its clinical significance.

Methods: A cross-sectional case control study was conducted in a total of 38 children with DS and 42 healthy children. This is the first report in the literature stating that. All subjects underwent ophthalmologic examination including slit-lamp biomicroscopy, cycloplegic refraction, intraocular pressure measurement and Scheimpflug imaging measurement. Customized software for Pentacam was used to analyze structural indices of anterior segment parameters. The mean anterior segment values of right eyes were compared between the groups.

Results: The mean anterior chamber parameters of patients with DS and controls were measured respectively: Corneal thickness was 502.31 ± 40.5 and 541.8 ± 37.42 mm ($p < 0.001$), corneal volume was 56.63 ± 4.5 and 61.02 ± 4.3 mm^3 ($p < 0.001$), corneal radius curvature was 7.41 ± 0.29 and 7.67 ± 0.34 mm ($p < 0.001$), iridocorneal angle was 39.7 ± 6.2 and $39.5 \pm 6.4^\circ$ ($p = 0.944$), anterior chamber volume was 181.65 ± 27.38 and 185.77 ± 32.53 mm^3 ($p = 0.528$), anterior chamber depth was 3.08 ± 0.24 and 3.02 ± 0.31 mm ($p = 447$), pupil size was 2.95 ± 0.48 and 3.29 ± 0.45 mm ($p < 0.05$).

Conclusion: The majority of the anterior segment parameters were found to be different in children with Down syndrome. While pupil size, corneal thickness, corneal volume and corneal curvature in DS were less than normal, iridocorneal angle, anterior chamber depth and anterior chamber volume were close to controls. The most important parametric differences in children with DS were seen on the cornea.

Keywords: Anterior segment parameters, children, Down syndrome

INTRODUCTION

Ophthalmic findings in individuals with Down syndrome (DS) have been addressed in order to find out if there is any difference from the normal population in some previous studies.[1–5] Anterior segment parameters are important for surgical approach and may be signs of ophthalmic diseases.[3] Increased frequency of keratoconus leading to corneal thinning and biometrical changes have been reported in previous studies in DS.[1,6,7] However, there is no sufficient information on all anterior segment parameters in children with DS.

Anterior segment parameters were measured by using a Scheimpflug imaging system (SI). This device photographs cross-sections of the anterior segment by means of illumination by slit lights at different meridians and acquires three-dimensional images of the anterior segment from the anterior corneal surface to the posterior lens surface within 2 seconds.[8,9] These images ensure the ability to detect subtle changes in the anterior segment.

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Correspondence: Lokman Aslan, Sutcu Imam University, Faculty of Medicine, Department of Ophthalmology, 46050 Kahramanmaraş, Turkey. Tel: +905326069808. E-mail: lokaslan46@yahoo.com

Moreover, accurate measurement can be obtained even in individuals with low cooperation since SI is a non contact method.[9–11] Pentacam has other advantages of automatic shots, short measurement time and a second eye alignment camera.

The study was aimed to determine the parameters of the anterior segment in children with DS and to compare the data with the findings in a group of normal children.

METHODS

The study was approved by the local ethics committee and conducted in accordance with the ethical principles described by the Declaration of Helsinki. Informed consent was obtained from the parents of participants.

The study group included 38 children with Down syndrome randomly selected from rehabilitation centers. The patients had been identified clinically or by chromosomal analyses through their personal health reports, issued by a pediatric psychiatrist and/or neurologist for the rehabilitation centers. The control group consisted of randomly selected 42 age-matched, healthy children among primary school students from the same region. Each subject in the study underwent ocular examination including slit-lamp biomicroscopy, cycloplegic refraction, intraocular pressure (IOP) and Scheimpflug imaging measurements.

Cyclopentolate 1% was instilled three times with 5-minute intervals for cycloplegic refraction 45 min prior to the examination. All refraction measurements were conducted with autorefractometer (Canon 50RM, Japan) in the cooperative children and with retinoscopy in the uncooperative ones. The spherical equivalent was calculated for assessment of refractive error by applying the following formula: spherical equivalent = spherical refraction + 0.5 × cylindrical refraction. Emmetropia was defined as a refractive error between −1.00 diopter (D) and +1.00 D spherical equivalent. Myopia was defined as less than −1.00 D and hyperopia, more than +1.00 D.

Intraocular pressure (IOP) was measured with a handheld device (Tono Pen) under topical anesthesia in whole participants. Children with over 21 mmHg were considered to have high IOP and referred to the glaucoma section.

Those with clinically detectable corneal pathology such as opacity, scar, degeneration and prior anterior segment surgery were excluded from the study.

Corneal Measurements

Corneal topographic maps were obtained by Pentacam HR (Oculus Inc., Wetzlar, Germany) by the same specially trained person. Measurements were repeated until acceptable quality imaging was obtained. Accurate measurement could be obtained in 38 of 42 targeted children with DS. Obtained quantitative topography values included the central corneal thickness (CCT; μm), anterior corneal radius curvature (CRC; mm), corneal volume (CV; mm^3), irido-corneal angle (ICA; °), pupil size (PS; mm), anterior chamber volume (ACV; mm^3) and anterior chamber depth (ACD; mm) were evaluated. CRC was calculated with the following formula: CRC = 0.03375/K, where K = mean corneal power.

The right eye values of participants underwent statistical analysis with SPSS 16.0. To compare the two groups, the T test was used in normally distributed and the Mann-Whitney U test in non-normally distributed data. Distribution was assessed with Levene's test for equality of variances. Correlation between corneal parameters and clinic results was addressed with Pearson correlation test. Any p values smaller than 0.05 were regarded as significant.

RESULTS

The sex and age distributions were not significantly different between the children with DS and the control group ($p > 0.05$). The DS group included 38 subjects (20 boys, 18 girls), and their mean age was 8.99 ± 2.42 (range 5–13) years. The control group included 42 subjects (20 boys, 22 girls), and the mean age of controls was 8.87 ± 2.34 (range 5–12) years.

Refractive statuses of children with DS were 17 (44.7 %) emmetrop, 12 (31.6 %) hyperopic and 9 (23.7 %) myopic. There was no correlation between refractive status with CCT ($r = 0.293$ $p = 0.146$) and refractive status with CRC ($r = 0.143$, $p = 0.486$).

The mean IOPs of right eyes were 13.83 ± 2.54 in DS and 14.21 ± 2.46 mmHg in controls ($p = 0.453$). There were no pathologically high IOP levels at any time. Fundus examination revealed normal optic nerve, macula and retinal vasculature.

CCT and CV in DS were statistically significantly less than in controls. The range of CCT was 450–500 micron in the DS group and 500–550 microns in controls. Corneal thickness and age were not correlated ($r = -0.301$, $p = 0.135$). The mean CRC in the DS group was also significantly less than in controls, thus corneas of children with DS were steeper than in controls. However, ICA, ACD and ACV values were similar in the two groups. An interesting finding was the mean pupil diameter was smaller in the DS group than in controls, but sufficient pharmacological pupil dilatation was achieved during cycloplegic refraction. Distributions of anterior segment parameters of right eyes in children with DS and in normal controls are shown in Figure 1. The results are further summarized in Table 1.

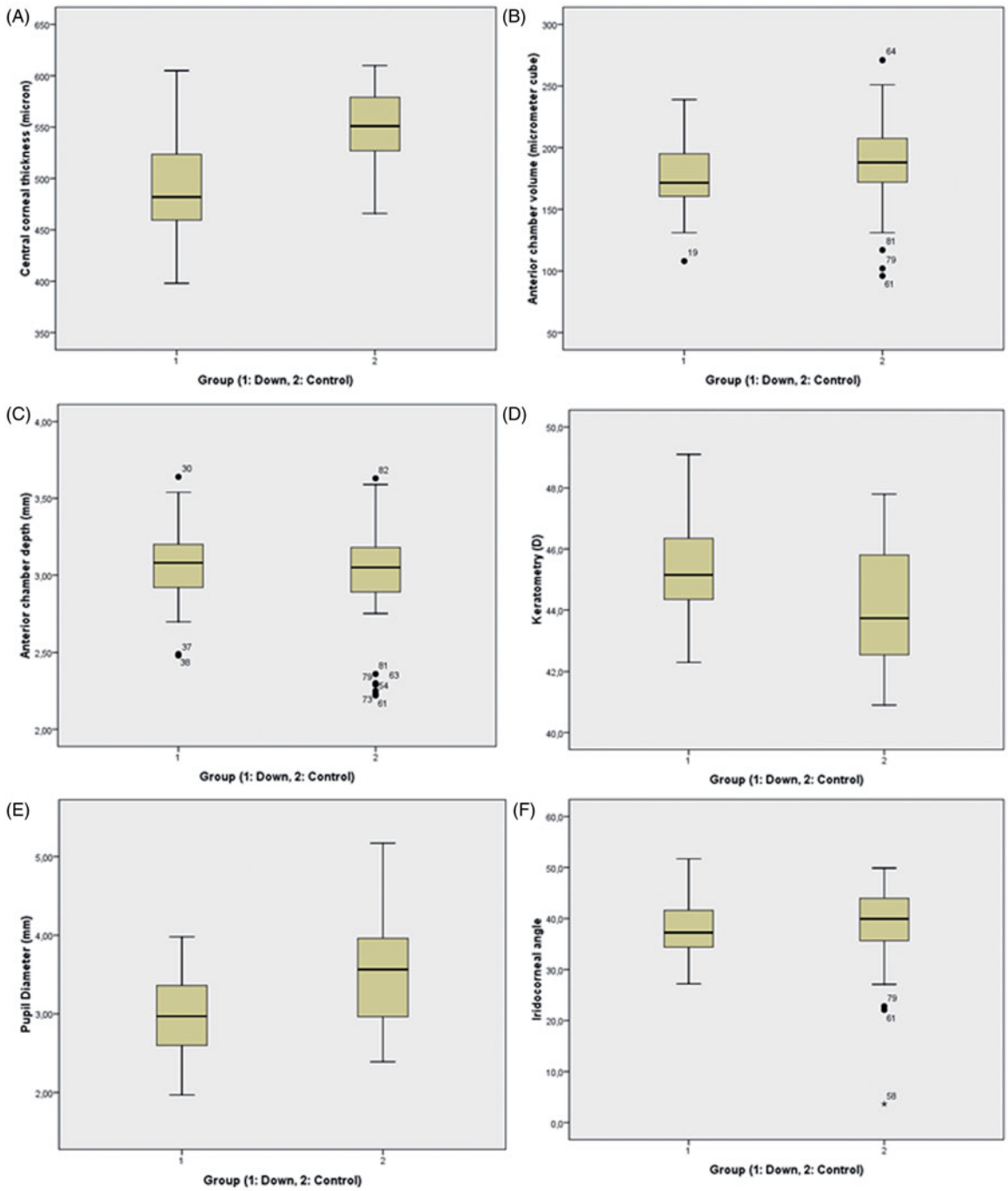

FIGURE 1. Box plots of Pentacam measurements in children with DS and controls: (A) central corneal thickness, (B) anterior chamber volume, (C) anterior chamber depth, (D) corneal power, (E) pupil size, and (F) iridocorneal angle.

DISCUSSION

DS is the most common chromosomal abnormality with an incidence of about 1/5000 live births.[4,5] A number of structural differences in children with DS have been reported in previous studies, and the most prominent clinical finding is refractive errors in children with DS.[3–5] Keratoconus leading to corneal thinning is also common in this population.[6] In this study, CCT in children with DS was found to be thinner than in controls similar to other previous studies in which the findings were generated from only DS individuals.[6,7] This finding could be the initial clinical evidence of congenital or progressive degenerative corneal disease such as kerataconus. The method we describe here is easily applicable and provides technical advantage for the early diagnosis of this disease.[10,11] Early diagnosis of keratoconus is important since some novel preventive treatment methods such as corneal crosslinking may be implemented for halting its progression.[6,7] Additionally, SI is a non-contact, easily applicable and manageable

TABLE 1. Anterior segment parameters in children with Down syndrome and controls.

Groups	*N* Right eyes	Mean ± SD	Minimum	Maximum	Δ (Control-DS)	*p*-value[a]
Corneal thickness						
Down syndrome	38	502.31 ± 40.5	430	584		
Control group	42	541.8 ± 37.42	496	603	39.49	$p < 0.001$
Corneal volume						
Down syndrome	38	56.63 ± 4.5	47.8	69.3		
Control group	42	61.02 ± 4.3	57.0	70.7	4.39	$p < 0.001$
Iridocorneal angle						
Down syndrome	38	39.7 ± 6.2	29.2	51.7		
Control group	42	39.5 ± 6.4	22.8	49.4	−0.3	$p = 0.944$
Radius curvature						
Down syndrome	38	7.41 ± 0.29	6.87	7.89		
Control group	42	7.67 ± 0.34	7.11	8.25	0.26	$p < 0.001$
Pupil size						
Down syndrome	38	2.95 ± 0.48	2.26	3.78		
Control group	42	3.29 ± 0.45	2.39	3.97	0.34	$p < 0.05$
Ant chamber volume						
Down syndrome	38	181.65 ± 27.38	141	239		
Control group	42	185.77 ± 32.53	102	251	4.9	$p = 0.528$
Ant chamber depth						
Down syndrome	38	3.08 ± 0.24	2.48	3.64		
Control group	42	3.02 ± 0.31	2.25	3.63	−0.06	$p = 0.447$

[a]The unpaired student's *t*-test.

method particularly in children with low cooperation. Related to a reduction in the corneal thickness, the mean corneal volume in children with DS was found to be decreased. The measurements of CCT and CV play an important diagnostic role, because they may affect the accuracy of the Goldman applanation tonometer.[13,14] The abnormality of the corneal thickness and CV causes impairment of corneal rigidity. Hence the accuracy of IOP measurements using application may be in doubt. In the present study, we found that intraocular pressure in DS was within normal limits. In previous studies, while glaucoma was reported as low as 1.9% in children with DS,[15] it was 11.5% in an adult DS group with an average age of 35 years. Yokohama reported that glaucomatous optic disc may be seen when the mean IOP is 12.2 mm/Hg. They implied that even though glaucoma was low in children with DS, they should be monitored in terms of glaucoma in their future life.

The corneal radius curvature is related to corneal steepness and there is a negative correlation between corneal steepness and radius curvature. Many authors have reported parallel to our findings that central corneal steepness is increased in DS.[1,5,6,12] Also, decreased corneal radius diameter means more refractive power as well as high corneal steepness and may be a symptom of refractive errors and corneal diseases such as keratoconus. Although the corneal thickness, volume and diameter are important parameters for contact lens applications and keratorefractive procedures, these applications are predominantly not preferred in children with DS.[7,17]

Individuals with DS are more likely to be candidates for cataract surgery since it is more common in DS than in the normal population.[3,4,17–19] Therefore, anterior segment parameters are important for these interventions. Anterior chamber angle, depth and volume in children with DS were found to be close to controls, but the mean pupil size was detected to be significantly smaller in the present study. Small pupil diameter is an important parameter affecting the success of ophthalmic surgery. However, sufficient pharmacological pupil dilatation was achieved during cycloplegic refraction. Furthermore, attenuated responses to sympathoexcitation and accommodation weakness in individuals with Down syndrome have been shown in other studies.[20–22] These autonomic dysfunctions and smaller pupil size may be concerned with a part of the same clinical condition. Moreover, small pupil diameter plays an important role in reducing the spherical aberrations since it prevents more refracted rays on the peripheral cornea passing to the retina. Although it provides an advantage for better visual acuity due to the pinhole effect,[23] the children with DS in our series were found to have more visual impairment due to refractive errors and corneal changes than in the normal population.

As a result, in our study the most common anterior segment differences in children with DS were found on the cornea. These differences have clinical importance in the accurate interpretation of IOP measurements, for the planning of keratorefractive procedures and early detection of degenerative diseases. On the other hand, pupil diameters in children with DS measured in the mesopic condition without pharmacologic dilation were found to be smaller than in the normal control group for the first time in the literature.

DECLARATION OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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