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Wavefront aberrations in Marfan syndrome over time after refractive surgery

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ABSTRACT

AIM: To study aberrations of the optical system of the eye in patients with ectopia lentis in Marfan syndrome and their changes after surgical treatment.

MATERIAL AND METHODS: The study included 20 patients (38 eyes) with Marfan syndrome with lens subluxation of varying severity, aged 5 to 36 years (23.7±5.2), and who received treatment at the Helmholtz National Medical Research Center of Eye Diseases in 2017 to 2024. Surgical treatment of ectopia lentis involved removal of the subluxated lens followed by implantation of an intraocular lens (IOL) and an intracapsular tension ring. It was performed in 9 patients (16 eyes) from the study group of patients with Marfan syndrome, namely, in 5 girls and 4 boys aged 5 to 15 years (mean: 12.1±3.2 years). The examination of children included autorefractometry, biomicroscopy, ophthalmoscopy, keratometry, tonometry, ultrasound biometry, and aberrometry. Follow-up periods ranged from 3 months to 2 years.

RESULTS: The conducted examinations showed that eyes with keratoconus had a sharp increase in all common (and, above all, internal) aberrations caused by a change in the position of the lens, its tilt (which contributes to the tilt aberration), vertical and horizontal decentration (vertical and horizontal coma and trefoil), a more convex shape due to the impossibility of tensioning the ciliary zonules that flatten the lens in the healthy eye (spherical aberration), a change in the clarity and quality of the lens surface (trefoil).

CONCLUSION: In Marfan syndrome, all common and internal wavefront aberrations of the eye are sharply increased. Vertical (×2,000 times) and horizontal (×1,000 times) tilt, vertical (×7,000 times) and horizontal (×130,000 times) coma, and vertical trefoil (×900 times) demonstrate extreme values. After surgical replacement of the lens and its centration, all listed aberrations are significantly reduced, however, they still remain increased compared to the normal values.

Keywords: myopia; Marfan syndrome; aberrations.

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Аберрации волнового фронта глаза при синдроме Марфана и их динамика после оптико-реконструктивной хирургии

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АННОТАЦИЯ

Цель. Изучение аберраций оптической системы глаза у пациентов с эктопией хрусталика при синдроме Марфана и их изменений после хирургического лечения.

Материал и методы. В исследование включено 20 пациентов (38 глаз) с синдромом Марфана с подвывихом хрусталика II–III степени в возрасте 5–36 лет (23,7±5,2), наблюдавшихся в НМИЦ глазных болезней им. Гельмгольца в 2017–2024 гг. Хирургическое лечение эктопии хрусталика, а именно удаление подвывихнутого хрусталика с имплантацией интраокулярной линзы (ИОЛ) и внутрикапсульного кольца получили 9 пациентов (16 глаз) из общей группы с синдромом Марфана — 5 девочек и 4 мальчика в возрасте 5–15 лет (в среднем 12,1±3,2 года). Обследование детей включало авторефрактометрию, биомикроскопию, офтальмоскопию, кератометрию, тонометрию, ультразвуковую биометрию, аберрометрию. Сроки наблюдения составили от 3 месяцев до 2 лет.

Результаты. Проведённые исследования показали резкое повышение в глазах с подвывихом хрусталика всех общих (прежде всего, внутренних) аберраций, вызванное изменением положения хрусталика, его наклоном (что увеличивает аберрацию тилт), смещением от зрительной оси по вертикали и горизонтали (вертикальные и горизонтальные кома и трефоил), более выпуклой формой ввиду невозможности натяжения цинновых связок, уплощающих линзу в норме (сферическая аберрация), изменением прозрачности и качества поверхности хрусталика (трефоил).

Заключение. При синдроме Марфана все общие и внутренние аберрации волнового фронта глаза резко повышены. Экстремальных значений достигает повышение вертикального (в 2000 раз) и горизонтального (в 1000 раз) тилта, вертикальной (в 7000 раз) и горизонтальной (в 130 000 раз) комы, вертикального трефойла (в 900 раз). После хирургической замены хрусталика, централизации его положения все перечисленные аберрации значительно снижаются, однако, всё же остаются повышенными по сравнению с нормой.

Ключевые слова: миопия; синдром Марфана; аберрации.

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ЭКО•ВЕКТОР

INTRODUCTION

Marfan syndrome is a genetic connective tissue disorder characterized by typical involvement of the eye and the musculoskeletal and cardiovascular systems [1]. The prevalence of the syndrome in the population, according to various sources, ranges from 4 to 6 per 100,000 newborns [2–5]. In Marfan syndrome, weakened zonular fibers may initially manifest as myopia and lens subluxation. These patients are often prone to early cataract development.

Currently, the Ghent criteria are considered highly valuable for diagnosing Marfan syndrome. According to these, ectopia lentis is classified as a major ophthalmic criterion, whereas increased axial length of the globe, abnormally flat cornea, and hypoplasia of the iris and ciliary muscle are classified as minor criteria. Other developmental anomalies may also occur, including congenital cataract, microphakia and/or spherophakia, lens coloboma, and atrophic retinal tears. The known connective tissue weakness in Marfan syndrome, specifically involving changes in the position, shape, size, and transparency of the lens, combined with a potential increase in axial length, leads to refractive errors and the development of optical system aberrations. Given the inability to achieve correction with glasses or contact lenses, the risk of amblyopia, difficulties with social adaptation, and impaired psychological development in children, growing ophthalmological experience has demonstrated that the most effective modern method for managing ocular manifestations of Marfan syndrome is surgical intervention, specifically removal of the subluxated lens with intraocular lens (IOL) implantation [6–8].

Currently, the removal of a subluxated (dislocated) clear or cataractous lens in Marfan syndrome can be performed using two approaches. The first approach involves removing the lens along with the capsular bag (lensectomy–vitrectomy), followed by IOL implantation with scleral fixation. The second approach consists in removing the subluxated lens while preserving the capsular bag, with simultaneous IOL implantation either into the bag or using a combined fixation technique, sometimes with the additional use of a capsular tension ring or other implants [9–13].

Anatomical shifts occurring in the eyes of patients with Marfan syndrome — such as lens displacement, subluxation, impaired accommodation and disaccommodation processes, and elongation of the anteroposterior axis — predispose to changes in wavefront aberrations. There are few reports in the literature dedicated to aberrometry in Marfan syndrome. For instance, Lteif and Bahar demonstrated that in lens subluxation, the total level of aberrations, particularly tilt and astigmatism as well as higher-order aberrations (RMS HOAs, coma, trefoil), increased three- to fourfold compared to healthy eyes. At the same time, spherical aberration did not show such differences and was even non-significantly lower than in healthy individuals [14–16].

AIM: To study optical system aberrations in the eye in Marfan syndrome patients with ectopia lentis and their changes after surgical treatment.

MATERIAL AND METHODS

The study included 20 patients (38 eyes) with Marfan syndrome and grade 2–3 lens subluxation. The patients were aged 5 to 36 years (mean age: 23.7±5.2 years); they were followed-up at the Helmholtz National Medical Research Center of Eye Diseases between 2017 and 2024. Surgical treatment for ectopia lentis (removal of the subluxated lens followed by implantation of IOL and intracapsular ring) was performed in 9 patients (16 eyes) from the overall group with Marfan syndrome — 5 girls and 4 boys aged 5 to 15 years (mean age: 12.1±3.2 years). The examination of children included biomicroscopy, ophthalmoscopy, tonometry, and methods required for the implanted IOL calculation (autorefractometry and ultrasound biometry). The follow-up period ranged from 3 months to 2 years.

To study the wavefront, all patients underwent examination using the OPD-Scan III aberrometer (Nidek, Japan). Of these, nine patients underwent repeated aberrometry — both before surgery and 3 months after surgery.

Phacoaspiration of the subluxated lens was performed using the Megatron S4 surgical system (Geuder, Germany), with implantation of an intracapsular polymethylmethacrylate ring and a hydrophobic acrylic IOL into the capsular bag. A controlled anterior vitrectomy was performed to center the capsular bag. The postoperative period in all patients was uneventful, with standard anti-inflammatory therapy.

RESULTS

The clinical and anatomical characteristics of the examined patients are presented in Table 1.

Optical system aberrations of the eye in patients with Marfan syndrome are shown in Table 2. The table also includes previously published wavefront parameters in healthy children with various refractions ranging from high hyperopia to high myopia [17].

All total wavefront aberrations in eyes with ectopia lentis were elevated by several orders of magnitude, rather than by 3–4 times. In particular, RMS increased 18-fold, vertical tilt increased 2000-fold, horizontal tilt increased 1000-fold, vertical coma increased 7000-fold, horizontal coma increased 130,000-fold, vertical trefoil

Table 1. Clinical and anatomical characteristics of patients pre-surgery

Таблица 1. Клинико-анатомические характеристики пациентов до операции

	All examined Все обследованные	Eyes treated surgically Из них в дальнейшем оперированные
Number of patients (eyes) Количество пациентов (глаз)	20 (38)	9 (16)
Age, years Возраст, годы	23,7±5,2	12,1±3,1
Spherical equivalent (D) Сферический эквивалент рефракции, дптр	-17,8±3,7	-18,5±1,6
Best corrected visual acuity Максимальная корригированная острота зрения	0,31±0,15	0,24±0,27
intraocular pressure (mmHg) Внутриглазное давление, мм рт. ст.	13,8±3,9	14,3±3,3
Anteroposterior axis, (mm) Переднезадняя ось, мм	24,3±1,7	23,31±1,2

Table 2. Aberrations of the optical system of the eye in patients with Marfan syndrome (38 eyes)

Таблица 2. Аберрации оптической системы глаз с синдромом Марфана (38 глаз)

	Aberrations Аберрации			Common aberrations: normal values Общие аберрации в норме
	Common Общие	Corneal Роговичные	Internal Внутренние	
Общий уровень аберраций (RMS HOAs)		3,3±0,6		0,179±0,1
Vertical tilt Вертикальный тилт	17,1±2,7	0,05±0,01	16,9±0,1	0,008±0,03
Horizontal tilt Горизонтальный тилт	35,6±3,9	-0,28±0,04	36±0,1	0,031±0,29
Vertical coma Вертикальная кома	-10,2±1,7	0,03±0,02	-8,5±0,1	0,0014±0,02
Horizontal coma Горизонтальная кома	-19,5±2,1	-0,15±0,1	-17,9±0,1	-0,00015
Vertical trefoil Вертикальный трефойл	24,4±0,1	-0,06±0,1	26,3±0,1	0,027±0,05
Horizontal trefoil Горизонтальный трефойл	8,02±1,1	-0,08±0,01	7,2±0,19	0,024±0,26
Spherical aberration Сферическая аберрация	5,9±0,6	0,05±0,01	4,7±0,1	-0,02±0,14

increased 900-fold, horizontal trefoil increased 300-fold, and spherical aberration increased 300-fold (Table 2).

The increase in aberrations is primarily due to the eye internal optics, while corneal aberrations remain nearly unchanged.

Following lens surgery, total and internal aberrations decrease sharply: RMS by a factor of 3.7, vertical tilt 20-fold, horizontal tilt 320-fold, vertical coma 97-fold, horizontal coma 255-fold, vertical trefoil 62-fold, horizontal trefoil 187-fold, and spherical aberration 90-fold. These changes are attributed to the reduction in internal aberrations, whereas corneal aberrations show a slight increase, which is expected due to the corneal incisions (Table 3).

Thus, the study demonstrated a sharp increase in all total aberrations, primarily internal ones, due to changes in lens position, its tilt (which increases tilt aberration), displacement from the visual axis both vertically and horizontally (resulting in vertical and horizontal coma), a more convex shape caused by the inability of the zonular fibers to exert tension and flatten the lens as they normally do (spherical aberration), and alterations in lens transparency and surface quality following lens replacement (trefoil). After removal of the subluxated lens and implantation of a properly centered IOL, all these aberrations are significantly reduced. However, they remain elevated compared with normal values.

Table 3. Wavefront aberrations in 9 patient before and after the lens surgery

Таблица 3. Аберрации волнового фронта у 9 пациентов до и после операции на хрусталике

Aberrations Аберрации	Pre-surgery До операции			Post-surgery 3 months После операции через 3 месяца		
	Common Общие	Corneal Роговичные	Internal Внутренние	Common Общие	Corneal Роговичные	Internal Внутренние
Общий уровень аберраций (RMS HOAs)		4,5±0,6			1,2±0,2	
Vertical tilt Вертикальный тилт	15,1±2,7	0,05±0,01	14,8±0,1	-0,74±0,1	0,2±0,1	-2,9±0,1
Horizontal tilt Горизонтальный тилт	37,8±3,7	-0,28±0,04	33,7±0,1	-0,11±0,1	-0,15±0,1	0,24±0,1
Vertical coma Вертикальная кома	-11,7±1,9	0,03±0,02	-9,95±0,1	-0,12±0,1	0,18±0,1	-0,3±0,1
Horizontal coma Горизонтальная кома	-17,9±2,7	-0,15±0,1	-18,6±0,1	0,07±0,1	-0,3±0,1	0,4±0,1
Vertical trefoil Вертикальный трефойл	25,1±0,1	-0,06±0,1	24,9±0,1	0,4±0,1	-0,19±0,1	0,58±0,1
Horizontal trefoil Горизонтальный трефойл	7,5±1,1	-0,08±0,01	7,02 ±0,19	-0,04±0,1	-0,001±0,1	-0,07±0,1
Spherical aberration Сферическая аберрация	6,3±0,6	0,05±0,01	5,7±0,1	-0,07±0,1	0,17±0,1	-0,24±0,1

Wavefront analysis of the eye in Marfan syndrome has significant clinical implications for patient monitoring, surgical indication assessment, procedure selection, and assessment of surgical efficacy.

CONCLUSIONS

1. In Marfan syndrome, all total and internal wavefront aberrations of the eye are markedly increased.
2. Extreme increases are observed in vertical tilt (by a factor of 2000), horizontal tilt (1000 times), vertical coma (7000 times), horizontal coma (130,000 times), and vertical trefoil (900 times).
3. After surgical lens replacement and proper centration, all these aberrations are significantly reduced; however, they remain elevated compared with normal values.

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