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Pellucid marginal degeneration classification development based on investigation of relationship between functional and refractive changes

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BACKGROUND: One of the problems in the diagnosis and treatment of pellucid marginal degeneration of the cornea is the difficulty of systematizing its manifestations due to the lack of classification. This is due to the low frequency of pellucid marginal degeneration in the structure of primary keratectasia, the main type of which is keratoconus. The developed classifications of keratoconus cannot be fully applied to pellucid marginal degeneration.

AIM: The aim was to develop a classification of pellucid marginal degeneration based on investigation of relationship between functional and refractive changes.

MATERIALS AND METHODS: The study included 42 people (42 eyes) with pellucid marginal degeneration. Keratometry and refractometry were performed, uncorrected and best corrected visual acuity, as well as cylindrical and spherical components of subjective refraction were studied, and retinal visual acuity was determined. The 1st group – 12 patients (12 eyes) with fully corrected induced ametropia (best corrected visual acuity ≥ 0.8), the 2nd group – 17 patients (17 eyes) with partially corrected induced ametropia (<0.8 and ≥ 0.3), the 3rd group – 13 patients (13 eyes) with uncorrected induced ametropia (<0.3).

RESULTS: To develop a clinical classification of pellucid marginal degeneration by stages, we selected: the values of corneal astigmatism, best corrected visual acuity and Index of difference between the values of maximum and minimum keratometry (ΔK), all of which had good separation of obtained data, and their demarcate values in groups.

CONCLUSION: The study showed the presence of relationship between functional and refractive changes indices of eyes with pellucid marginal degeneration. The leading parameters of refractive status, objectively determining the value of best corrected visual acuity, are induced corneal astigmatism and ΔK . The developed classification of pellucid marginal degeneration is easy to use and makes it possible to determine the stage of keratectasia even if there is only induced corneal astigmatism or ΔK values.

Keywords: pellucid marginal degeneration; astigmatism; classification; visual acuity.

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Разработка классификации пеллюцидной маргинальной дегенерации роговицы на основе исследования взаимосвязи функциональных и рефракционных изменений

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Актуальность. Одной из проблем в диагностике и лечении пеллюцидной маргинальной дегенерации роговицы является сложность систематизации её проявлений вследствие отсутствия классификации. Это обусловлено небольшой частотой пеллюцидной маргинальной дегенерации роговицы в структуре первичных кератэктазий, основным видом которых выступает кератоконус. Разработанные классификации кератоконуса не могут быть полноценно применимы к пеллюцидной маргинальной дегенерации роговицы.

Цель — разработать классификацию пеллюцидной маргинальной дегенерации роговицы на основе исследования взаимосвязи функциональных и рефракционных изменений.

Материалы и методы. В исследовании участвовали 42 человека (42 глаза) с пеллюцидной маргинальной дегенерацией роговицы. Проводили кератометрию и рефрактометрию, исследовали некорригируемую и максимально корригируемую остроту зрения, а также цилиндрический и сферический компоненты субъективной рефракции, определяли ретинальную остроту зрения. Первую группу составили 12 пациентов (12 глаз) с полностью корригируемой индуцированной аметропией (максимально корригируемая острота зрения ≥0,8), вторую группу — 17 человек (17 глаз) с частично корригируемой (<0,8 и ≥0,3), третью группу — 13 человек (13 глаз) с некорригируемой индуцированной аметропией (<0,3).

Результаты. Для разработки клинической классификации пеллюцидной маргинальной дегенерации роговицы по стадиям были выбраны показатели: величины роговичного астигматизма, максимально корригируемой остроты зрения и разницы между значениями максимальной и минимальной кератометрии (ΔK), имевшие хорошие разделения полученных данных и их разграничивающие значения в группах.

Заключение. Исследование показало наличие взаимосвязи между функциональными и рефракционными показателями глаз с пеллюцидной маргинальной дегенерацией роговицы. Ведущими параметрами рефракционного статуса, объективно определяющими величину максимально корригируемой остроты зрения, определены индуцированный роговичный астигматизм и ΔK . Разработанная классификация пеллюцидной маргинальной дегенерации роговицы обладает простотой применения и позволяет определить стадию эктазии даже при наличии только значения индуцированного роговичного астигматизма или ΔK .

Ключевые слова: пеллюцидная маргинальная дегенерация роговицы; астигматизм; классификация; острота зрения.

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BACKGROUND

Despite some advances in diagnostics of pellucid marginal corneal degeneration (PMCD), data interpretation is still impossible and remains one of the serious problems because of the lack of a classification of this pathology [1, 2]. This circumstance is probably due to the low frequency of PMCD in the range of primary keratectasia, with keratoconus (KC) as the most common type. However, the need to systematize the clinical and diagnostic manifestations of PMCD for the treatment of this category of patients is quite relevant [1–5]. The literature presents few publications where the authors attempt to classify PMCD into stages according to the magnitude of induced corneal astigmatism (ICA); however, no other signs are considered in relation to this indicator [1, 6].

The main pathognomonic sign of PMCD is the keratotopographic crab claw pattern. However, as this pattern can be also present in KC, it cannot be considered specific; thus, other indicators should be taken into account for making a diagnosis, particularly keratopachymetry, to identify the location of the thinning zone (Fig. 1) [1, 5, 7, 8].

Since a group of the world's leading ophthalmologists involved in the study of various issues of diagnostics and treatment of primary keratectasia achieved a global agreement in 2015 (Global Consensus on Keratoconus and Ectatic Diseases of the Cornea), within which PMCD and KC were recognized as clinical manifestations of the same disease, the use of the same classification is perhaps applicable for both types of keratectasia. However, this would not be entirely correct [9]. Thus, for example, one of the signs of the Amsler-Krumeich classification of KC (1998), namely, the presence of Vogt's striae and corneal opacities, which can cause a pronounced deprivation effect, is not registered in PMCD [1, 3-5, 10, 11]. Our studies have also shown that with equal ICA values, the values of uncorrected (UCVA) and best-corrected visual acuity (BCVA) in patients with PMCD were higher than that in KC [2]. This phenomenon occurred because in PMCD, the thinning zone is located on the extreme periphery of the cornea, whereas in KC, it is located in the central sections, which can lead to higher aberrations [1, 3-5].

Many authors have proposed classifications of KC. However, all of them are unlikely to be fully applicable to PMCD, as some consider the keratotopographic pattern aspects, which is very variable in KC, whereas others consider it necessary to use the maximum number of signs as criteria, characteristic precisely for KC [12–16].

The aforementioned absence of corneal opacities and the crab claw keratotopographic pattern characteristic in all cases in PMCD eyes necessitate the use of indicators of functional refractive changes in the affected eyes as the main signs characterizing the degree of pathological changes.

The work aimed to develop a PMCD classification based on the study of the relationship between functional and refractive changes.

MATERIALS AND METHODS

The study involved 42 patients (42 eyes) with PMCD, including 23 men and 19 women. The age of the patients ranged from 28 to 67 (average 48 ± 9.2) years. The main selection criteria for the follow-up group were the presence of a keratotopographic crab claw pattern and peripheral thinning of the cornea.

All patients underwent keratometry and refractometry using an NRK 8000 device (Nikon, Japan) to determine the ICA value and cylindrical and spherical components of objective refraction. The UCVA, BCVA, and cylindrical and spherical components of subjective refraction were examined using a character projector R2047 (CSO, Italy) at a decimal scale on a TAKAGI VT-5 phoropter (Japan). In addition, retinal visual acuity was determined in all patients using a Lambda 100 retinometer (Heine, Germany) on a decimal scale from 0.06 to 0.8.

All patients were distributed into three groups according to the degree of correction of induced ametropia (IA) and the BCVA value. Group 1 included 12 patients



Fig. 1. Pentacam Map Report. Keratotopographic pattern and keratopachymetric map of a patient with pellucid marginal degeneration of the cornea

Рис. 1. Кератотопографический паттерн и кератопахиметрическая карта пациента с пеллюцидной маргинальной дегенерацией роговицы на анализаторе Pentacam (OCULUS, Германия) (12 eyes) with fully correctable IA (BCVA \ge 0.8), group 2 included 17 patients (17 eyes) with partially correctable IA (BCVA <0.8 and \ge 0.3), and group 3 included 13 patients (13 eyes) with uncorrectable IA (BCVA <0.3).



Fig. 2. Uncorrected visual acuity (UCVA) in patients with pellucid marginal degeneration of the cornea

Рис. 2. Значения некорригируемой остроты зрения (НКОЗ) у пациентов с пеллюцидной маргинальной дегенерацией роговицы



Fig. 3. Best corrected visual acuity (BCVA) values in patients with pellucid marginal degeneration of the cornea

Рис. 3. Значения максимально корригируемой остроты зрения (МКОЗ) у пациентов с пеллюцидной маргинальной дегенерацией роговицы



Fig. 5. Cylindrical component of subjective refraction value in patients with pellucid marginal degeneration of the cornea **Рис. 5.** Величина цилиндрического компонента субъективной рефракции (ЦКСР) у пациентов с пеллюцидной маргинальной дегенерацией роговицы

In all eyes, the Pentacam device (OCULUS, Germany) was used to determine the keratotopographic pattern, minimum keratopachymetry (KPM_{min}), maximum keratometry (K_{max}), and minimum keratometry (K_{min}), and the difference between them was calculated (ΔK).

The study did not include patients with retinal visual acuity less than 0.8, cataracts and macular pathology, and corneal opacities of various origins.

Statistical data processing was performed using the IBM SPSS Statistics for Windows, version 20 (IBM Corp., Armonk, NY, USA). The delimiting values of the studied parameters to justify the classification were determined using the ROC analysis.

RESULTS

Results of the data analysis are presented in diagrams (Figs. 2–12), demonstrating the difference in the intergroup distribution of the studied values and quantities. Good data distribution, except for BCVA, was noted for ICA, cylindrical component of objective refraction,





Рис. 4. Величина индуцированного роговичного астигматизма (ИРА) у пациентов с пеллюцидной маргинальной дегенерацией роговицы



Fig. 6. Spherical component of subjective refraction value in patients with pellucid marginal degeneration of the cornea **Рис. 6.** Величина сферического компонента субъективной рефракции (СКСР) у пациентов с пеллюцидной маргинальной дегенерацией роговицы

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Fig. 7. Cylindrical component of objective refraction in patients with pellucid marginal degeneration of the cornea

Рис. 7. Величина цилиндрического компонента объективной рефракции (ЦКОР) у пациентов с пеллюцидной маргинальной дегенерацией роговицы



Fig. 9. Maximum keratometry values (K_{max}) in patients with pellucid marginal degeneration of the cornea

Рис. 9. Значения максимальной кератометрии (K_{max}) у пациентов с пеллюцидной маргинальной дегенерацией роговицы



Fig. 11. Difference between values of maximum and minimum keratometry values (ΔK) in patients with pellucid marginal degeneration of the cornea

Рис. 11. Разница между значениями максимальной и минимальной кератометрии (*ΔK*) у пациентов с пеллюцидной маргинальной дегенерацией роговицы



Fig. 8. Spherical component of objective refraction in patients with pellucid marginal degeneration of the cornea

Рис. 8. Величина сферического компонента объективной рефракции (СКОР) у пациентов с пеллюцидной маргинальной дегенерацией роговицы



Fig. 10. Minimum keratometry values (K_{min}) in patients with pellucid marginal degeneration of the cornea

Рис. 10. Значения минимальной кератометрии (*K*_{min}) у пациентов с пеллюцидной маргинальной дегенерацией роговицы



Fig. 12. Minimum keratopachymetry values ($K\Pi M_{min}$) in patients with pellucid marginal degeneration of the cornea

Рис. 12. Величина минимальной кератопахиметрии (КПМ_{min}) у пациентов с пеллюцидной маргинальной дегенерацией роговицы

Table. Classification of pellucid marginal degeneration of the cornea
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Таблица. Классификация пеллюцидной маргинальной дегенераци роговицы

Stage	Induced corneal astigmatism, diopters	Best-corrected visual acuity	Δ <i>K</i> , diopters
I (corrected induced ametropia)	<5.0	≥0.8	<5.0
II (partially corrected induced ametropia)	≥5.0 – ≤10.0	<0.8 – ≥0.3	≥5.0 – ≤14.0
III (uncorrected induced ametropia)	<10.0	<0.3	<14.0

cylindrical component of subjective refraction, and ΔK , where poor distribution was noted for UCVA, $K_{\rm max}$, $K_{\rm min}$, and KPM_{min}. The values of the spherical components of objective and subjective refractions did not show intergroup differences.

To develop a clinical classification of PMCD, the ICA, BCVA, $\Delta \textit{K},$ and their delimiting values in groups were selected (Table 1).

DISCUSSION

First of all, the results of this study confirmed the relationship between the functional condition and refractive status of the PMCD eyes. The main pathological refractive index of corneal ectasia (ICA) determines the visual acuity and process severity; as a result, it was accepted as fundamental for the classification of PMCD. Although the cylindrical components of objective and subjective refractions also had a good distribution into groups, in our opinion, their use as criteria for dividing stages can be difficult in the presence of concomitant pathology of the optical media of the eye, such as cataracts, or in a pseudophakic eye.

When evaluating visual acuity parameters, the value of retinal visual acuity with BCVA <0.8 can be considered reasonable not only for studying the functional status but also for predicting and evaluating visual functions during implantation of intrastromal corneal segments. In addition, this method helps determine the required parameter in the initial cataract, when the BCVA can be sufficiently reduced. Taking into account the above fact and the older age of patients with PMCD, unlike those with KC, the probability of concomitant pathology not only of the optical media but also of the retina is higher in these patients; therefore, the use of BCVA in all cases can be difficult [2, 17].

The ICA, as an objective indicator, can be considered the main parameter for the PMCD distribution by stages. However, in some cases, the keratometer cannot determine high-grade ICA; as a result, ectasia can be attributed to stage III, or keratotopography can be performed to study ΔK and clarify the corneal condition.

In our opinion, the proposed classification will enable to not only systematize the functional and refractive manifestations of PMCD but also develop recommendations in the future for the targeted treatment of patients with various stages of this pathology.

CONCLUSIONS

1. The study results showed the relationship between the functional and refractive parameters of PMCD eyes.

2. ICA and ΔK can be considered major parameters of the refractive status, which determine objectively the BCVA value.

3. The developed classification of PMCD is easy to use and helps in determining the ectasia stage even if only the ICA or ΔK values are available.

ADDITIONAL INFORMATION

Author contributions. All authors confirm that their authorship complies with the ICMJE criteria. All authors have made a significant contribution to the development of the concept, research, and preparation of the article, read and approved the final version before шеы publication.

Author contributions. I.V. Vasilyeva collected and processed the material, created the concept and design of the study, performed statistical processing, and prepared the text. S.V. Kostenev created the research concept and design, edited the text, and approved the manuscript for publication. A.V. Vasiliev edited the text and approved the manuscript for publication.

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