

# GEORGIAN MEDICAL NEWS

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ЕЖЕМЕСЯЧНЫЙ НАУЧНЫЙ ЖУРНАЛ

Медицинские новости Грузии  
საქართველოს სამედიცინო სიახლენი

## GEORGIAN MEDICAL NEWS

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**GMN: Georgian Medical News** is peer-reviewed, published monthly journal committed to promoting the science and art of medicine and the betterment of public health, published by the GMN Editorial Board since 1994. GMN carries original scientific articles on medicine, biology and pharmacy, which are of experimental, theoretical and practical character; publishes original research, reviews, commentaries, editorials, essays, medical news, and correspondence in English and Russian.

GMN is indexed in MEDLINE, SCOPUS, PubMed and VINITI Russian Academy of Sciences. The full text content is available through EBSCO databases.

**GMN: Медицинские новости Грузии** - ежемесячный рецензируемый научный журнал, издаётся Редакционной коллегией с 1994 года на русском и английском языках в целях поддержки медицинской науки и улучшения здравоохранения. В журнале публикуются оригинальные научные статьи в области медицины, биологии и фармации, статьи обзорного характера, научные сообщения, новости медицины и здравоохранения. Журнал индексируется в MEDLINE, отражён в базе данных SCOPUS, PubMed и ВИНТИ РАН. Полнотекстовые статьи журнала доступны через БД EBSCO.

**GMN: Georgian Medical News** – საქართველოს სამედიცინო სიახლენი – არის ყოველთვიური სამეცნიერო სამედიცინო რეცენზირებადი ჟურნალი, გამოიცემა 1994 წლიდან, წარმოადგენს სარედაქციო კოლეგიისა და აშშ-ის მეცნიერების, განათლების, ინდუსტრიის, ხელოვნებისა და ბუნებისმეტყველების საერთაშორისო აკადემიის ერთობლივ გამოცემას. GMN-ში რუსულ და ინგლისურ ენებზე ქვეყნდება ექსპერიმენტული, თეორიული და პრაქტიკული ხასიათის ორიგინალური სამეცნიერო სტატიები მედიცინის, ბიოლოგიისა და ფარმაციის სფეროში, მიმოხილვითი ხასიათის სტატიები.

ჟურნალი ინდექსირებულია MEDLINE-ის საერთაშორისო სისტემაში, ასახულია SCOPUS-ის, PubMed-ის და ВИНТИ РАН-ის მონაცემთა ბაზებში. სტატიების სრული ტექსტი ხელმისაწვდომია EBSCO-ს მონაცემთა ბაზებიდან.

### WEBSITE

[www.geomednews.com](http://www.geomednews.com)

## К СВЕДЕНИЮ АВТОРОВ!

При направлении статьи в редакцию необходимо соблюдать следующие правила:

1. Статья должна быть представлена в двух экземплярах, на русском или английском языках, напечатанная через **полтора интервала на одной стороне стандартного листа с шириной левого поля в три сантиметра**. Используемый компьютерный шрифт для текста на русском и английском языках - **Times New Roman (Кириллица)**, для текста на грузинском языке следует использовать **AcadNusx**. Размер шрифта - **12**. К рукописи, напечатанной на компьютере, должен быть приложен CD со статьей.

2. Размер статьи должен быть не менее десяти и не более двадцати страниц машинописи, включая указатель литературы и резюме на английском, русском и грузинском языках.

3. В статье должны быть освещены актуальность данного материала, методы и результаты исследования и их обсуждение.

При представлении в печать научных экспериментальных работ авторы должны указывать вид и количество экспериментальных животных, применявшиеся методы обезболивания и усыпления (в ходе острых опытов).

4. К статье должны быть приложены краткое (на полстраницы) резюме на английском, русском и грузинском языках (включающее следующие разделы: цель исследования, материал и методы, результаты и заключение) и список ключевых слов (key words).

5. Таблицы необходимо представлять в печатной форме. Фотокопии не принимаются. **Все цифровые, итоговые и процентные данные в таблицах должны соответствовать таковым в тексте статьи**. Таблицы и графики должны быть озаглавлены.

6. Фотографии должны быть контрастными, фотокопии с рентгенограмм - в позитивном изображении. Рисунки, чертежи и диаграммы следует озаглавить, пронумеровать и вставить в соответствующее место текста **в tiff формате**.

В подписях к микрофотографиям следует указывать степень увеличения через окуляр или объектив и метод окраски или импрегнации срезов.

7. Фамилии отечественных авторов приводятся в оригинальной транскрипции.

8. При оформлении и направлении статей в журнал МНГ просим авторов соблюдать правила, изложенные в «Единых требованиях к рукописям, представляемым в биомедицинские журналы», принятых Международным комитетом редакторов медицинских журналов - <http://www.spinesurgery.ru/files/publish.pdf> и [http://www.nlm.nih.gov/bsd/uniform\\_requirements.html](http://www.nlm.nih.gov/bsd/uniform_requirements.html) В конце каждой оригинальной статьи приводится библиографический список. В список литературы включаются все материалы, на которые имеются ссылки в тексте. Список составляется в алфавитном порядке и нумеруется. Литературный источник приводится на языке оригинала. В списке литературы сначала приводятся работы, написанные знаками грузинского алфавита, затем кириллицей и латиницей. Ссылки на цитируемые работы в тексте статьи даются в квадратных скобках в виде номера, соответствующего номеру данной работы в списке литературы. Большинство цитированных источников должны быть за последние 5-7 лет.

9. Для получения права на публикацию статья должна иметь от руководителя работы или учреждения визу и сопроводительное отношение, написанные или напечатанные на бланке и заверенные подписью и печатью.

10. В конце статьи должны быть подписи всех авторов, полностью приведены их фамилии, имена и отчества, указаны служебный и домашний номера телефонов и адреса или иные координаты. Количество авторов (соавторов) не должно превышать пяти человек.

11. Редакция оставляет за собой право сокращать и исправлять статьи. Корректур авторам не высылаются, вся работа и сверка проводится по авторскому оригиналу.

12. Недопустимо направление в редакцию работ, представленных к печати в иных издательствах или опубликованных в других изданиях.

**При нарушении указанных правил статьи не рассматриваются.**

## REQUIREMENTS

Please note, materials submitted to the Editorial Office Staff are supposed to meet the following requirements:

1. Articles must be provided with a double copy, in English or Russian languages and typed or computer-printed on a single side of standard typing paper, with the left margin of 3 centimeters width, and 1.5 spacing between the lines, typeface - **Times New Roman (Cyrillic)**, print size - 12 (referring to Georgian and Russian materials). With computer-printed texts please enclose a CD carrying the same file titled with Latin symbols.

2. Size of the article, including index and resume in English, Russian and Georgian languages must be at least 10 pages and not exceed the limit of 20 pages of typed or computer-printed text.

3. Submitted material must include a coverage of a topical subject, research methods, results, and review.

Authors of the scientific-research works must indicate the number of experimental biological species drawn in, list the employed methods of anesthetization and soporific means used during acute tests.

4. Articles must have a short (half page) abstract in English, Russian and Georgian (including the following sections: aim of study, material and methods, results and conclusions) and a list of key words.

5. Tables must be presented in an original typed or computer-printed form, instead of a photocopied version. **Numbers, totals, percentile data on the tables must coincide with those in the texts of the articles.** Tables and graphs must be headed.

6. Photographs are required to be contrasted and must be submitted with doubles. Please number each photograph with a pencil on its back, indicate author's name, title of the article (short version), and mark out its top and bottom parts. Drawings must be accurate, drafts and diagrams drawn in Indian ink (or black ink). Photocopies of the X-ray photographs must be presented in a positive image in **tiff format**.

Accurately numbered subtitles for each illustration must be listed on a separate sheet of paper. In the subtitles for the microphotographs please indicate the ocular and objective lens magnification power, method of coloring or impregnation of the microscopic sections (preparations).

7. Please indicate last names, first and middle initials of the native authors, present names and initials of the foreign authors in the transcription of the original language, enclose in parenthesis corresponding number under which the author is listed in the reference materials.

8. Please follow guidance offered to authors by The International Committee of Medical Journal Editors guidance in its Uniform Requirements for Manuscripts Submitted to Biomedical Journals publication available online at: [http://www.nlm.nih.gov/bsd/uniform\\_requirements.html](http://www.nlm.nih.gov/bsd/uniform_requirements.html)  
[http://www.icmje.org/urm\\_full.pdf](http://www.icmje.org/urm_full.pdf)

In GMN style for each work cited in the text, a bibliographic reference is given, and this is located at the end of the article under the title "References". All references cited in the text must be listed. The list of references should be arranged alphabetically and then numbered. References are numbered in the text [numbers in square brackets] and in the reference list and numbers are repeated throughout the text as needed. The bibliographic description is given in the language of publication (citations in Georgian script are followed by Cyrillic and Latin).

9. To obtain the rights of publication articles must be accompanied by a visa from the project instructor or the establishment, where the work has been performed, and a reference letter, both written or typed on a special signed form, certified by a stamp or a seal.

10. Articles must be signed by all of the authors at the end, and they must be provided with a list of full names, office and home phone numbers and addresses or other non-office locations where the authors could be reached. The number of the authors (co-authors) must not exceed the limit of 5 people.

11. Editorial Staff reserves the rights to cut down in size and correct the articles. Proof-sheets are not sent out to the authors. The entire editorial and collation work is performed according to the author's original text.

12. Sending in the works that have already been assigned to the press by other Editorial Staffs or have been printed by other publishers is not permissible.

**Articles that Fail to Meet the Aforementioned  
Requirements are not Assigned to be Reviewed.**

## ავტორთა საქურაღებოლ!

რედაქციაში სტატიის წარმოდგენისას საჭიროა დაიცვათ შემდეგი წესები:

1. სტატია უნდა წარმოადგინოთ 2 ცალად, რუსულ ან ინგლისურ ენებზე დაბეჭდილი სტანდარტული ფურცლის 1 გვერდზე, 3 სმ სიგანის მარცხენა ველისა და სტრიქონებს შორის 1,5 ინტერვალის დაცვით. გამოყენებული კომპიუტერული შრიფტი რუსულ და ინგლისურენოვან ტექსტებში - **Times New Roman (Кириллица)**, ხოლო ქართულენოვან ტექსტში საჭიროა გამოვიყენოთ **AcadNusx**. შრიფტის ზომა – 12. სტატიას თან უნდა ახლდეს CD სტატიით.

2. სტატიის მოცულობა არ უნდა შეადგენდეს 10 გვერდზე ნაკლებს და 20 გვერდზე მეტს ლიტერატურის სიის და რეზიუმეების (ინგლისურ, რუსულ და ქართულ ენებზე) ჩათვლით.

3. სტატიაში საჭიროა გაშუქდეს: საკითხის აქტუალობა; კვლევის მიზანი; საკვლევი მასალა და გამოყენებული მეთოდები; მიღებული შედეგები და მათი განსჯა. ექსპერიმენტული ხასიათის სტატიების წარმოდგენისას ავტორებმა უნდა მიუთითონ საექსპერიმენტო ცხოველების სახეობა და რაოდენობა; გაუტკივარებისა და დაძინების მეთოდები (მწვავე ცდების პირობებში).

4. სტატიას თან უნდა ახლდეს რეზიუმე ინგლისურ, რუსულ და ქართულ ენებზე არანაკლებ ნახევარი გვერდის მოცულობისა (სათაურის, ავტორების, დაწესებულების მითითებით და უნდა შეიცავდეს შემდეგ განყოფილებებს: მიზანი, მასალა და მეთოდები, შედეგები და დასკვნები; ტექსტუალური ნაწილი არ უნდა იყოს 15 სტრიქონზე ნაკლები) და საკვანძო სიტყვების ჩამონათვალი (key words).

5. ცხრილები საჭიროა წარმოადგინოთ ნაბეჭდი სახით. ყველა ციფრული, შემაჯამებელი და პროცენტული მონაცემები უნდა შეესაბამებოდეს ტექსტში მოყვანილს.

6. ფოტოსურათები უნდა იყოს კონტრასტული; სურათები, ნახაზები, დიაგრამები - დასათაურებული, დანომრილი და სათანადო ადგილას ჩასმული. რენტგენოგრამების ფოტოასლები წარმოადგინეთ პოზიტიური გამოსახულებით **tiff** ფორმატში. მიკროფოტოსურათების წარწერებში საჭიროა მიუთითოთ ოკულარის ან ობიექტივის საშუალებით გადიდების ხარისხი, ანათალების შედეგის ან იმპრეგნაციის მეთოდი და აღნიშნოთ სურათის ზედა და ქვედა ნაწილები.

7. სამამულო ავტორების გვარები სტატიაში აღინიშნება ინიციალების თანდართვით, უცხოურისა – უცხოური ტრანსკრიპციით.

8. სტატიას თან უნდა ახლდეს ავტორის მიერ გამოყენებული სამამულო და უცხოური შრომების ბიბლიოგრაფიული სია (ბოლო 5-8 წლის სიღრმით). ანბანური წყობით წარმოდგენილ ბიბლიოგრაფიულ სიაში მიუთითეთ ჯერ სამამულო, შემდეგ უცხოელი ავტორები (გვარი, ინიციალები, სტატიის სათაური, ჟურნალის დასახელება, გამოცემის ადგილი, წელი, ჟურნალის №, პირველი და ბოლო გვერდები). მონოგრაფიის შემთხვევაში მიუთითეთ გამოცემის წელი, ადგილი და გვერდების საერთო რაოდენობა. ტექსტში კვადრატულ ფხიხლებში უნდა მიუთითოთ ავტორის შესაბამისი N ლიტერატურის სიის მიხედვით. მიზანშეწონილია, რომ ციტირებული წყაროების უმეტესი ნაწილი იყოს 5-6 წლის სიღრმის.

9. სტატიას თან უნდა ახლდეს: ა) დაწესებულების ან სამეცნიერო ხელმძღვანელის წარდგინება, დამოწმებული ხელმოწერითა და ბეჭდით; ბ) დარგის სპეციალისტის დამოწმებული რეცენზია, რომელშიც მითითებული იქნება საკითხის აქტუალობა, მასალის საკმაობა, მეთოდის სანდოობა, შედეგების სამეცნიერო-პრაქტიკული მნიშვნელობა.

10. სტატიის ბოლოს საჭიროა ყველა ავტორის ხელმოწერა, რომელთა რაოდენობა არ უნდა აღემატებოდეს 5-ს.

11. რედაქცია იტოვებს უფლებას შეასწოროს სტატია. ტექსტზე მუშაობა და შეჯერება ხდება საავტორო ორიგინალის მიხედვით.

12. დაუშვებელია რედაქციაში ისეთი სტატიის წარდგენა, რომელიც დასაბეჭდად წარდგენილი იყო სხვა რედაქციაში ან გამოქვეყნებული იყო სხვა გამოცემებში.

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## SYSTEMIC OR LIMITED IS HEMISCLERODERMA OF FACE IN A PERSON WITH UVEITIS? EXPERIENCE OF 10 CASES OF UVEITIS IN HEMISCLERODERMA OF FACE FROM ONE RHEUMATOLOGY CENTER

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### Abstract.

**Introduction:** Linear scleroderma of head and face (LSH) in children is a severe disorder, that results in hemiatrophy of skin, subcutaneous tissue, bones with functional disabilities, neurologic disorders and uveal involvement.

**Aim:** The aim of the research was to establish uveal involvement in children with hemifacial scleroderma.

**Materials and methods:** A retrospective analysis was done in a group of 110 children with hemifacial scleroderma. A comprehensive clinical, laboratory and instrumental examination was performed, including MRI of the brain, EEG, and an ophthalmologist's examination, which included visometry, biomicroscopy, and ophthalmoscopy.

**Results:** 10 cases of uveal involvement were detected (9% of 110 pt). 9 patients had anterior segment inflammation (iridocyclitis), in 2 iridocyclitis was combined with retinal changes (in 1- peripheral focal chorioretinitis, in 1- iridocyclitis and central focal chorioretinitis). In one case, iridocyclitis was combined with optic neuropathy. In 3 children uveitis appeared at the disease debut, in the others 3-10 years later. Uveal inflammation in all cases was on the side of scleroderma skin involvement. In 3 children uveitis was bilateral. Seizures and concomitant foci in white matter of the brain were detected in 2 children with uveitis. 90% of the group had positive antinuclear factor. Persistent decrease in visual acuity developed in 3 patients.

**Conclusion:** Patients with LSH must undergo routine eye examination using basic ophthalmological techniques (visometry, biomicroscopy, ophthalmoscopy) every 6 months and highly necessary in case of relapse of scleroderma. We assume that patients with UI in LSH must be defined as patients with JSS and treated intensively with systemic glucocorticoids, cytostatics and even biologics in case of resistance.

**Key words.** Juvenile hemiscleroderma of face, uveitis, antinuclear antibodies, juvenile systemic sclerosis.

### Introduction.

Juvenile localized scleroderma (JLS) is a fairly common disease in the practice of dermatologists and rheumatologists. The incidence of JLS is 3.4 – 9 cases per 1 million child population annually [1,2]. Linear scleroderma of head and face (LSH) and Parry-Romberg syndrome is one of the most severe JLS types, characterized by deep lesions of the skin and subcutaneous tissues, including bone structures, as well as damage to the nervous system and ocular involvement. The disease leads to gross cosmetic and functional defects of the face, dental apparatus, accompanied by seizures, damage to the cranial nerves (trigeminal, oculomotor), ocular involvement. Uveal inflammation is described in LSH. Besides, signs of

ocular involvement such as damage to globe and adnexa oculi in LSH appear in the form of fibrosclerotic changes in the eyelids, loss of eyelashes (madarosis), damage to the uveal tract, development of dry eye syndrome.

According to a multicenter study [3], among 750 children with JLS, 24 (3.2%) had significant eye damage. The majority of children with eye damage had LSH (66.7%). Eye lesions had the following nature: ten children (41.7%) had damage to the auxiliary apparatus (eyelids and eyelashes), seven patients (29.2%) had inflammatory diseases of the anterior segment of the eyeball (five anterior uveitis – iridocyclitis, two – episcleritis). Eye damage was often accompanied by neurological symptoms: three patients revealed central nervous system disorders of systemic etiology, four had paralytic strabismus, pseudopapillitis and refractive disorders. Involvement of orbital structures in the process, such as enophthalmos, damage to the eyelids and orbit are also pointed out. The most common eye lesions in LSH include changes of the cornea and retina [4,5]. And the most common neuro-ophthalmological diseases include damage to the optic nerve, oculomotor nerves and pupillary dysfunctions [6-8].

In Parry-Romberg syndrome (PRS), ophthalmological symptoms usually appear due to the involvement of the eyeball, orbital soft tissues and orbital walls in the pathological process. So, the dysfunctions range from slight visual impairment to complete blindness [9]. As tissue atrophy spreads deeper, such patients usually suffer from enophthalmos and extraocular muscle dysfunction. In some cases, paralytic strabismus may develop [10]. The development of acute retinal vasculitis in patients with PRS proves the vascular etiology of the disease. Other ophthalmological clinical manifestations such as uveitis [11], neuroretinitis, glaucoma, papillitis, cataracts, changes in retinal pigment epithelium, iris heterochromia [12-14], associated nasolacrimal duct obstruction and prolonged dacryocystitis in PRS have also been reported [15].

Uveitis is quite rare in JLS, in 1% of cases [3], so we present our own cases of uveal damage in a group of 110 children with LSH.

**Aim:** The aim of the study was to establish the frequency and features of uvea involvement in children with LSH.

### Materials and Methods.

In the Childhood Department of Sechenov University for the period 2000-2023, 110 children with LSH were monitored, among them the uvea involvement (UI) was detected in 10 patients. The diagnosis of LSH was established according to the Preliminary proposed classification of juvenile localized scleroderma in 2004 [16]. The group of patients with LSH included both patients with linear scleroderma of the head and

**Table 1.** Clinical characteristics of patients with UI.

Patient	1	2	3	4	5	6	7	8	9	10
<b>Gender</b> Female\male (f/m)	f	f	f	f	f	m	f	m	m	f
<b>the age of the LSH debut (years)</b>	3	8	3	4	3	6	5	6	5	6
<b>Duration LSH by the time of UI (months)</b>	3	24	120	72	1	8	6	36	72	84
<b>UI is unilateral -* bilateral -**</b>	*	**	*	*	*	**	**	*	*	*
<b>The type of the UI Iridocyclitis (IC)</b>	IC, central chorioretinitis	IC	IC, optic nerve neuropathy	Peripheral chorioretinitis	IC	IC	IC	IC	IC	IC
<b>Relapses of IC</b>	+	+	-	-	+	-	+	-	-	-
<b>Decreased visual acuity in the acute period</b>	+	+	+	-	+	+	+	+	+	+
<b>Decreased visual acuity in the outcome of the disease</b>	+	+	+	-	-	-	-	-	-	-
<b>Antinuclear factor (ANF) positive</b>	+	+	+	+	+	+	+	+	-	+
<b>Seizures</b>	+	+	-	-	-	-	-	-	-	-
<b>Focal changes brain MRI</b>	+	+	-	-	-	-	-	-	-	-

face (94 children) and patients with Parry-Romberg syndrome (16 children).

All patients underwent a comprehensive clinical, laboratory and instrumental examination, including examination by a pediatric rheumatologist, ophthalmological examination (visometry, biomicroscopy, ophthalmoscopy), brain magnetic resonance imaging (MRI), electroencephalogram (EEG.)

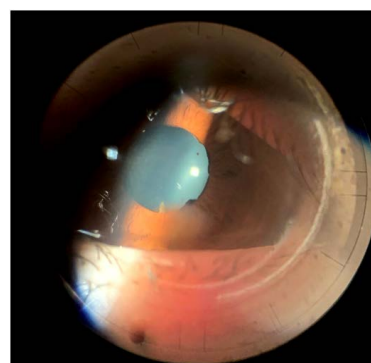
The age of the children with UI ranged from 3 to 15 years, averaging 6.4 years, among them 7 girls and 3 boys, the duration of follow-up in our clinic averaged 5.5 years. The clinical characteristics of the patients with UI are summarized in Table 1.

The duration of LSH at the time of UI detection ranged significantly from 3 months to 10 years. The occurrence of UI developed at different stages of the disease, at the onset (in 3 children) and from 3 to 10 years after the debut of the disease. Two children had significant damage to the skin of the eyelids and loss of eyelashes (Figure 1), five children developed enophthalmos. In all the children, UI was noted on the side of the skin lesion, while in three children, it was detected in the second eye-was bilateral. It is noteworthy that the overwhelming number of children with UI (9 out of 10) showed immunological activity of the disease, in the form of an increase in the level of antinuclear factor, while scleroderma-specific antibodies (anti-centromere antibodies) were found in three patients.

In 9 out of 10 patients, UI changes were in the form of iridocyclitis (Figure 2) with posterior synechiae, precipitates on the posterior surface of the cornea were detected in all the patients. One girl with a long-term scleroderma (72 months) had a chorioretinal atrophic lesion in the central and paracentral zone (Figure 3), and in two more children iridocyclitis was combined with peripheral atrophic retinal changes (peripheral



**Figure 1.** Patient with hemiscleroderma of face and chorioretinitis. Hemiatrophy of the right part of the face, eyelid fibrosis, loss of eyebrows and eyelashes.



**Figure 2.** Iridocyclitis, posterior synechiae.



**Figure 3.** Chorioretinal atrophic lesion in the paracentral zone of the retina of the left eye.

chorioretinitis) and optic neuropathy. One patient had ptosis of the affected eye, which disappeared after immunosuppressive systemic therapy. A decrease in visual acuity caused by UI was detected in all the patients with iridocyclitis and central chorioretinitis in the acute period of the disease. We interpret the lesion of the anterior and posterior segments of the uvea as a prognostically unfavorable event, since it can lead to persistent visual impairment. Against the background of the treatment (local and systemic), the signs of iridocyclitis were stopped with a complete improvement of visual functions. And only in three patients, in the outcome of the disease, visual acuity did not recover due to damage to the central parts of the retina and complications of recurrent iridocyclitis. At the same time, 8 out of 10 had concomitant myopia and astigmatism. Iridocyclitis was recurrent in 4 out of 10 patients and worsened with exacerbation of the skin syndrome.

It should be noted that in two children, the onset of iridocyclitis was accompanied by seizures, which required long-term use of anticonvulsants. The same patients revealed focal hyperintensive signals in brain tissue at MRI on the side of iridocyclitis and scalp lesions.

The patients were treated in a specialized rheumatology hospital using immunosuppressive therapy. All the children received glucocorticosteroids orally at a dose of 0.7-1 mg/kg per day for 8-10 weeks, followed by gradual complete withdrawal by the 15<sup>th</sup> -18<sup>th</sup> months of the treatment. One patient received D-penicillamine as a sparing drug, the rest received methotrexate 15 mg/ square meter of body surface per week parenterally; after the withdrawal of glucocorticosteroids two children received 2 drugs - methotrexate and mycophenolate mofetil for 2 years until remission was achieved. Three patients with insufficient effect of cytostatic therapy additionally received Tocilizumab. Of these, only one girl had an indication for initiating therapy with Tocilizumab, since she had a recurrence of iridocyclitis in combination with the progression of skin lesions and seizures. Along with basic therapy, immediately after the detection of iridocyclitis, the children received topical treatment - corticosteroids, nonsteroidal anti-inflammatory drugs, mydriatics

## Discussion.

Ten cases of UI were detected in our group of 110 patients with LSH, which makes 9% of the whole group, and this could be compared with UI in juvenile chronic arthritis. Our data shows a rather high percentage of UI as compared with UI occurrence presented in literature. In a larger investigation, among the whole group of 113 patients with the “en corpe de sabre” subtype, uveitis was in 5 children, which is 4,4% [3].

It is difficult to explain this difference in frequency of uveitis, but we could assume that the long duration of the disease in our group might have influenced UI. Four of our children had less than 12-month duration, in the other cases it ranged from 24-120 months (averaging 68 months). Another cause might have been the immunological activity and high disease activity in our patients: 90% of the group had ANF positive, three patients additionally had anti-centromere antibodies positive.

In the above-mentioned investigation, 50% of the patients were ANF positive, wherein 25% had neurologic disorders. On the other hand, only two patients in our group had concomitant seizures and white matter changes on brain MRI.

It is known that pathogenesis of scleroderma and ocular involvement is complex and still not clear. There are several hypotheses of the pathogenesis of uvea involvement associated with impaired blood supply, pigmentation disorders, and thinning of subfoveal layers [8]. Generalized microcirculatory vasculopathy in scleroderma is characterized by intimal hyperplasia of small arterioles, followed by luminal stenosis due to tissue hypoxia and chronic tissue ischemia; endothelial dysfunction activates coagulation and blood processes, thus leading to formation of microthrombi. Generalized damage to the microvasculature of obliterating endarteritis type occurs. The globe is an intensively supplied organ, therefore, sclerodermal vasculopathy leads to the uveal tract disorders [17]. Many researchers have described these changes [3,6,8,17]. According to the study of Grennan D [18], pathological choroidal changes including non-perfusion areas were detected in 50% of the patients. In addition, fibro-sclerotic processes occur in the skin and subcutaneous tissues, which affect extraocular structures such as eyelids, eyelashes, even bone structures of the orbit, thus resulting in impaired eyelid closure, tear drainage and other disorders [15,19-21].

Summing it up, high frequency of UI in our study elucidates the necessity of detailed examination of these patients, using modern biomarkers, searching reliable disease predictors. We believe that the patients with LSH and UI should be classified as patients with systemic form of juvenile scleroderma. We regard UI as a manifestation of the systemic form of scleroderma, with possibility of decrease in visual acuity, which requires prolonged systemic administration of glucocorticoids and cytostatics and even biologics.

A similar opinion was expressed by Maria Elisabetta Zannin and co-authors in 2007 [3]. Current classification criteria of juvenile systemic sclerosis (JSS) were designated in 2007, and ocular involvement was not taken into consideration among the possible signs or symptoms of the systemic form of the disease [22]. Nerve involvement was included in the form of seizures, neuroimaging abnormalities, headaches, carpal

tunnel syndrome, though headaches are not so specific for scleroderma as uveitis on the side of skin involvement. There is a pathogenetically proven connection between systemic inflammation of small vessels in JSS and UI in scleroderma. Besides, the majority of our patients with UI had positive ANF, some of them revealed concomitant neurologic disorders. In our practice, we evaluate the children with hemifacial scleroderma and UI as JSS and conduct more intensive immunosuppressive therapy, though UI may result in visual impairment, even blindness. It is reasonable to suppose that UI must be considered among possible JSS criteria in future.

### Conclusion.

According to the results of our retrospective examination, UI was detected in 9% of the children from the group of 110 patients with LSH. Nine patients had damage to only the anterior segment of the eye (iridocyclitis); two patients had damage to posterior segment; one child had peripheral chorioretinitis, and one had a combination of iridocyclitis with central chorioretinitis. UI developed both immediately after the debut of LSH, and 6-10 years after the onset of the disease, while in half of the children the disease had a recurrent character. All the children had UI on the side of the lesion of the scalp and face, which confirms the pathogenetic relationship between scleroderma and eye damage. Decreased visual acuity due to UI in the acute period was observed in all the children with iridocyclitis and central chorioretinitis. The UI of the eyeball can lead to persistent visual impairment, especially with a recurrent course and lesions of the posterior segment of the eye. However, in our group a persistent decrease in vision in the outcome of the disease was noted only in 3 children. And it was due to the presence of central atrophic chorioretinal lesions and complications of recurrent iridocyclitis. Decreased visual acuity can be persistent and lead to significant loss in the absence of adequate treatment, thus arising the problem of early diagnosis and therapy. It is important to undergo routine eye examination using basic ophthalmological techniques (visometry, biomicroscopy, ophthalmoscopy) every 6 months and highly necessary in case of relapse of scleroderma. We assume that patients with UI in LSH must be defined as patients with JSS.

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სისტემური ან შეზღუდულია თუ არა სახის ჰემისკლეროდერმია უვეიტის მქონე ადამიანში? უვეიტის უვეიტის 10 შემთხვევის გამოცდილება სახის ჰემისკის სკლეროდერმიაში ერთი რევმატოლოგიური ცენტრიდან.

რეზიუმე  
შესავალი. ბავშვებში თავისა და სახის ხაზოვანი სკლეროდერმია (LSH) არის მძიმე დაავადება, რომელიც

იწვევს კანის, სუბკუტანეუზის ქსოვილის, ფუნქციური შეზღუდული შესაძლებლობების მქონე ძვლების, ნევროლოგიური დარღვევებისა და uveal ჩართულობის ჰემიატროფიას.

კვლევის მიზანი იყო ჰემიფაციალური სკლეროდერმიით დაავადებულ ბავშვებში uveal ჩართულობის დადგენა.

მასალები და მეთოდები რეტროსპექტიული ანალიზი გაკეთდა ჰემიფაციალური სკლეროდერმიით დაავადებული 110 ბავშვის ჯგუფში. ჩატარდა ყოვლისმომცველი კლინიკური, ლაბორატორიული და ინსტრუმენტული გამოკვლევა, მათ შორის ტვინის MRI, EEG და ოფთალმოლოგის გამოკვლევა, რომელიც მოიცავდა ვიზომეტრიას, ბიომიკროსკოპიას და ოფთალმოსკოპიას

შედეგები გამოვლინდა uveal involvement-ის 10 შემთხვევა (110 pt-ის 9%). 9 პაციენტს ჰქონდა წინა სეგმენტის ანთეზა (ირიდოციკლიტი), 2 - ში ირიდოციკლიტი შერწყმული იყო ბადურის ცვლილებებთან (1 - პერიფერიული ფოკალური ქორიორეტინიტის დროს, 1-ირიდოციკლიტის დროს და ცენტრალური ფოკალური ქორიორეტინიტის დროს). ერთ შემთხვევაში ირიდოციკლიტი შერწყმული იყო ოპტიკურ ნეიროპათიასთან. 3 ბავშვებში uveitis გამოჩნდა დაავადების debute, დანარჩენი 3-10 წლის შემდეგ. Uveal ანთეზა ყველა შემთხვევაში იყო მხარეს scleroderma კანის ჩართულობა. 3 ბავშვებში უვეიტი ორმხრივი იყო. კრუნჩხვები და კომპომიტანტური კერები თავის ტვინის თეთრ ნივთიერებაში გამოვლინდა უვეიტის მქონე 2 ბავშვში. ჯგუფის 90% - ს ჰქონდა anf დადებითი. მხედველობის სიმახვილის მუდმივი შემცირება განვითარდა 3 პაციენტში.

დასკვნა. Lsh-ს მქონე პაციენტებმა უნდა გაიარონ თვალის რუტინული გამოკვლევა ძირითადი ოფთალმოლოგიური ტექნიკის გამოყენებით (ვიზომეტრია, ბიომიკროსკოპია, ოფთალმოსკოპია) ყოველ 6 თვეში ერთხელ და ძალიან საჭირო სკლეროდერმიის რეციდივის შემთხვევაში ჩვენ ვვარაუდობთ, რომ LSH-ში UI-ს მქონე პაციენტები უნდა განისაზღვროს, როგორც jss-ს მქონე პაციენტები.

ყალბი, ვიტისის, ანტიბირთვული ანტისხეულების არასრულწლოვანთა ჰემისლეროდერმია, სკლეროზის მქონე არასრულწლოვანთა სისტემა

**Системной или ограниченной является гемисклеродермия лица с увеитом? Наблюдения 10 детей из одного ревматологического центра.**

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#### **Резюме**

Вступление. Линейная склеродермия головы и лица (ЛСГ) у детей - это тяжелое заболевание, которое приводит к гемиатрофии кожи, подкожной клетчатки, костей с функциональными нарушениями, неврологическими расстройствами и поражением увеальной оболочки.

Целью исследования было установить поражение сосудистой оболочки глаза у детей с гемисклеродермией лица.

Материалы и методы. Проведен ретроспективный анализ в группе из 110 детей с ЛСГ. Всем пациентам проведено комплексное клиническое, лабораторное и инструментальное обследование, включая МРТ головного мозга, ЭЭГ, а также осмотр офтальмолога, который включал визометрию, биомикроскопию и офтальмоскопию

Результаты. Было выявлено 10 случаев поражения сосудистой оболочки глаза (9% из 110 случаев). У 9 пациентов было воспаление переднего сегмента (иридоциклит), у 2 иридоциклит сочетался с изменениями сетчатки (у 1 - периферический очаговый хориоретинит, у 1 - иридоциклит и центральноочаговый хориоретинит). В одном случае иридоциклит сочетался с нейропатией зрительного нерва. У 3 детей увеит развился в дебюте заболевания, у остальных - через 3-10 лет. Воспаление сосудистой оболочки во всех случаях отмечено на стороне поражения в рамках склеродермии. У 3 детей увеит был двусторонним. Эпилептические припадки и очаговые изменения в белом веществе головного мозга были обнаружены у 2 детей с увеитом. У 90% детей группы был положительный антинуклеарный фактор. Стойкое снижение остроты зрения развилось у 3 пациентов.

Вывод. Пациенты с ЛСГ должны проходить плановое обследование глаз с использованием основных офтальмологических методов (визометрия, биомикроскопия, офтальмоскопия) каждые 6 месяцев, и в случае рецидива склеродермии. Мы предполагаем, что пациенты с вовлечением сосудистой оболочки глаза при ЛСГ должны трактоваться как пациенты с системной формой ювенильной склеродермией.