

HEREDITARY HYPOFIBRINOGENEMIA IN PRIMARY CARE PEDIATRICS: A CASE REPORT

Gomellya M.V.¹,
Krupskaya T.S.¹,
Belkova T.Yu.²,
Vorzhheva A.N.²,
Mironova A.V.¹

¹ Irkutsk State Medical University
(Krasnogo Vosstaniya, 1, Irkutsk 664003,
Russian Federation)

² Ivano-Matreninskaya City Children's
Clinical Hospital for Outpatient and
Polyclinic Services (Sovetskaya St., 57,
Irkutsk 664047, Russian Federation)

Corresponding author:
Anastasia V. Mironova
e-mail: tav23_97_1997@mail.ru

ABSTRACT

Introduction. Hypofibrinogenemia is a rare coagulation disorder characterized by reduced plasma fibrinogen levels and an increased risk of hemorrhagic complications. Inherited forms, caused by mutations in the FGA, FGB, or FGG genes, have an estimated prevalence of 1:500,000 and often remain undiagnosed due to an asymptomatic course.

Aim. To present a rare clinical case of hereditary hypofibrinogenemia with a paucisymptomatic course in a child, in order to address challenges in diagnosis, long-term follow-up, and determining indications for prophylactic therapy to prevent hemorrhagic complications.

Materials and Methods. We report a clinical case of hereditary hypofibrinogenemia in an 8-year-old child. The diagnosis was suspected incidentally during a routine pediatric examination. The diagnosis was subsequently confirmed by a hematologist based on persistently low fibrinogen levels, a positive family history (maternal hereditary hypofibrinogenemia), and genetic testing. Differential diagnosis with acquired forms of hypofibrinogenemia was performed based on clinical and laboratory evaluation, including complete blood count, coagulogram, aggregatogram, biochemical parameters, and instrumental studies.

Results. The key feature of this case was the mild clinical presentation, which included a neonatal cephalohematoma, a single episode of blood in stool, and rare epistaxis. This aligns with literature data on the highly variable phenotypic expressivity of hypofibrinogenemia. The importance of a detailed family history and genetic testing in diagnosing inherited coagulopathies, even in the absence of overt bleeding, is emphasized.

Conclusion. This case highlights the need for physician vigilance regarding inherited coagulation disorders in patients with abnormal coagulation test results, even in the absence of significant clinical symptoms. The development and implementation of strategies for long-term patient monitoring and the prevention of potential hemostasis-related risks are of paramount importance.

Keywords: hereditary hypofibrinogenemia, coagulopathy, hemostasis, fibrinogen, children

Received: 23.09.2025
Accepted: 29.06.2026
Published: 23.07.2026

For citation: Gomellya M.V., Krupskaya T.S., Belkova T.Yu., Vorzhheva A.N., Mironova A.V. Hereditary hypofibrinogenemia in primary care pediatrics: a case report. *Acta biomedica scientifica*. 2026; 11(3): 117-123. doi: 10.29413/ABS.2026-11.3.14

НАСЛЕДСТВЕННАЯ ГИПОФИБРИНОГЕНЕМИЯ В АМБУЛАТОРНО-ПОЛИКЛИНИЧЕСКОЙ ПРАКТИКЕ ВРАЧА-ПЕДИАТРА (КЛИНИЧЕСКИЙ СЛУЧАЙ)

Гомелля М.В.¹,
Крупская Т.С.¹,
Белькова Т.Ю.²,
Воржева А.Н.²,
Миронова А.В.¹

¹ ФГБОУ ВО «Иркутский
государственный медицинский
университет» Минздрава России
(664003, г. Иркутск, ул. Красного
Восстания, 1, Россия)

² ОГАУЗ «Городская Ивано-
Матренинская детская клиническая
больница» (664009, г. Иркутск,
ул. Советская, 57, Россия)

Автор, ответственный за переписку:
**Миронова Анастасия
Витальевна**
e-mail: tav23_97_1997@mail.ru

РЕЗЮМЕ

Введение. Гипофибриногенемия является достаточно редкой коагулопатией, характеризующейся снижением уровня фибриногена и повышением риска геморрагических осложнений. Наследственные формы, обусловленные мутациями в генах FGA, FGB, FGG, встречаются с частотой 1:500 000 и часто остаются недиагностированными из-за бессимптомного течения.

Цель исследования. Представить редкий клинический случай малосимптомного течения наследственной гипофибриногенемии у ребёнка для освещения вопросов диагностики, динамического наблюдения и определения показаний к профилактической терапии для предотвращения геморрагических осложнений.

Материалы и методы. В статье представлен клинический случай наследственной гипофибриногенемии у ребенка 8 лет. Диагноз был заподозрен случайно при плановом обследовании у педиатра, а затем при углубленном обследовании у гематолога установлен на основании стойкого снижения уровня фибриногена, подтвержденного генеалогического анамнеза (наследственная гипофибриногенемия у матери, генетического исследования). Проведен дифференциальный диагноз с приобретенными формами гипофибриногенемии на основании клинко-лабораторного обследования: общего анализа крови, коагулограммы, агрегатограммы, биохимических показателей, инструментальных исследований.

Результаты. Ключевая характеристика данного клинического наблюдения заключается в малосимптомном течении заболевания, проявляющемся неонатальной кефалогематомой, единичным эпизодом крови в стуле и редкими носовыми кровотечениями, что соответствует данным литературы о вариабельности степени тяжести клинической картины при гипофибриногенемии. Важен тщательный сбор семейного анамнеза в диагностике наследственных коагулопатий, несмотря на отсутствие выраженных геморрагических проявлений, а также генетическое исследование.

Заключение. Представленный клинический случай демонстрирует необходимость настороженности врачей в отношении наследственных коагулопатий у пациентов с изменениями коагулограммы даже при отсутствии клинической симптоматики. Особое значение приобретают разработка и реализация подходов к дальнейшему наблюдению за пациентами, а также профилактике потенциальных рисков, связанных с нарушениями системы гемостаза.

Ключевые слова: наследственная гипофибриногенемия, врожденные коагулопатии, гемостаз, фибриноген, дети

Статья поступила: 23.09.2025
Статья принята: 29.06.2026
Статья опубликована: 23.07.2026

Для цитирования: Гомелля М.В., Крупская Т.С., Белькова Т.Ю., Воржева А.Н., Миронова А.В. Наследственная гипофибриногенемия в амбулаторно-поликлинической практике врача-педиатра (клинический случай). *Acta biomedica scientifica*. 2026; 11(3): 117-123. doi: 10.29413/ABS.2026-11.3.14

INTRODUCTION

Hypofibrinogenemia is a coagulation disorder characterized by a reduced plasma fibrinogen concentration [1, 2]. According to the World Federation of Hemophilia and national registries, fibrinogen deficiency accounts for approximately 8.0 % of all rare coagulopathies [3]. The prevalence of this condition is approximately 1 case per 500,000 population [4]. Despite its low prevalence, timely detection of hypofibrinogenemia is critically important for the prevention of potential hemorrhagic complications.

According to the classification of hypofibrinogenemias, hereditary (congenital) and acquired forms are distinguished [1]. Hereditary hypofibrinogenemia often remains unnoticed, but may have clinical consequences, including bleeding and an increased risk of thrombosis [3, 5]. In contrast to acquired forms, which are often caused by liver disease or disseminated intravascular coagulation (DIC), hereditary hypofibrinogenemia is characterized by a persistently low fibrinogen level throughout life and can be confirmed by a positive family history [1].

Historically, hereditary forms of fibrinogen deficiency were first described in the early 20th century, and in 1999, mutations in the genes responsible for its synthesis were identified [6]. Hereditary hypofibrinogenemia, associated with the genes encoding the fibrinogen chains (FGA, FGB, FGG), can lead to hemostatic abnormalities [7, 8]. Fibrinogen plays a key role in the blood coagulation process, being converted to fibrin by thrombin and forming a fibrin clot [1, 3, 9].

In clinical practice, two types are distinguished: hypofibrinogenemia and afibrinogenemia, in which plasma fibrinogen levels are below 1.8 g/L and 0.1 g/L, respectively [10]. In most cases, patients have an asymptomatic course or minimal bleeding from mucous membranes, with bleeding after trauma or surgical interventions usually of the microcirculatory type [3].

Diagnostic evaluation of hereditary hypofibrinogenemia is based on plasma fibrinogen levels, other baseline coagulogram parameters, and molecular genetic analysis to identify mutations in the relevant genes [11, 12]. In severe fibrinogen deficiency, prolongation of activated partial thromboplastin time (APTT), prothrombin time (PT), and thrombin time (TT) is observed, which is due to the absence of the necessary substrate for the conversion of fibrinogen to fibrin by thrombin [3].

Diagnosis of the hereditary form requires high accuracy and reliability, which underscores the need for timely detection and differential diagnosis with acquired forms of hypofibrinogenemia [13]. Early diagnosis (particularly of severe hypofibrinogenemia and afibrinogenemia) facilitates preventive measures and the prescription of replacement therapy when necessary to reduce the risk of complications associated with hemostatic abnormalities.

This clinical case of a child with hereditary hypofibrinogenemia illustrates the importance of a timely comprehensive approach to diagnosis and follow-up of patients. The study protocol was approved by the Ethics Committee of the Ivano–Matreninskaya City Children's Clinical Hospital (Protocol No. 10 dated February 10, 2025) and complied with the principles of the Declaration of Helsinki.

AIM

To present a rare clinical case of hereditary hypofibrinogenemia with a paucisymptomatic course in a child, in order to address challenges in diagnosis, long-term follow-up, and determining indications for prophylactic therapy to prevent hemorrhagic complications.

MATERIALS AND METHODS

History

An 8-year-old female patient, accompanied by her mother, presented to a hematologist at the outpatient department of the Ivano–Matreninskaya City Children's Clinical Hospital in February 2025 regarding abnormal coagulogram findings from January 15, 2024: decreased fibrinogen to 1.17 g/L, prolonged APTT to 44 sec, prolonged TT to 29 sec, and decreased PT to 61.0 %. The examination was performed while the patient was in full clinical health and had no complaints.

The child was born from the first pregnancy, 1st delivery. According to the mother, the pregnancy was uncomplicated. Delivery was spontaneous at term (38 weeks). Birth weight was 3850 g; length 54 cm. Apgar score was 7–9. The infant cried immediately and was breastfed immediately after birth. Breastfeeding continued until 9 months of age. Growth and development were age-appropriate. Vaccinations were administered according to the national immunization schedule.

During collection of a detailed bleeding history, a family history was revealed: the mother had been observed for several years to have decreased fibrinogen to an average of 0.87 g/L, prolonged APTT to 44 sec, prolonged TT to 29 sec, prothrombin time to 13.2 sec, and decreased PT to 57.9 %, based on which a hematologist had diagnosed hypofibrinogenemia, and she was under dispensary follow-up. Genetic analysis confirmed the presence of a heterozygous mutation in the FGB G455A gene in the mother. Further family history did not reveal any other cases of significant bleeding or established coagulopathies in close maternal relatives (grandmother, grandfather).

From the medical history, it is known that the child suffered from a cephalohematoma at birth. In 2016, at the second week of their life, blood was found

in their stool, prompting the child to be examined at the Irkutsk Regional Infectious Diseases Clinical Hospital, the Irkutsk Regional Children's Clinical Hospital, and the Ivano-Matreninskaya City Children's Clinical Hospital, with a diagnosis of diarrhea of unspecified etiology. At 8 months old, the patient was diagnosed with hemangioma on the left side of the trunk and underwent surgical treatment. Rare episodes of epistaxis have also been noted. However, previous monitoring of complete blood count and coagulation tests were not performed.

Physical examination

General condition was satisfactory. Well-being was not affected. Position was active. Axillary body temperature was 36.2°C. Blood pressure was 106/68 mmHg. Respiratory rate was 22/min. Heart rate was 85/min. Height was 130 cm; body weight was 33 kg. SDS BMI = 1.65. Skin was of normal color and clear. Visible mucous membranes were of normal color and clear. Subcutaneous adipose tissue was moderately developed. Peripheral lymph nodes were not palpable. Musculoskeletal system showed no deformities, joint mobility was full, and palpation was painless. The chest was of normal shape and size, symmetrical, painless on palpation, with clear pulmonary sound on percussion. Auscultation revealed vesicular breathing, conducted to all areas, no wheezing. The borders of relative and absolute cardiac dullness were within age-related norms. On auscultation, heart sounds were clear, rhythmic, with no murmur. The abdomen was of normal shape and size on inspection, soft and painless on palpation. The liver and spleen were not palpable. Intoxication, anemic, hemorrhagic, lymphoproliferative, hyperplastic, and ossalgic syndromes were absent.

Results of laboratory and instrumental studies

Complete blood count (January 14, 2025): hemoglobin – 139.0 g/L, erythrocytes – $5.07 \times 10^{12}/L$, reticulocytes – 1.0 %, hematocrit – 41.8 %, platelets – $272.0 \times 10^9/L$, leukocytes – $8.84 \times 10^9/L$, segmented neutrophils – 56.0 %, monocytes – 5.9 %, lymphocytes – 36.1 %, eosinophils – 2.0 %, and ESR – 2 mm/h.

Biochemical blood test (January 14, 2025): total bilirubin – 10.2 $\mu\text{mol}/L$, indirect bilirubin – 4.7 $\mu\text{mol}/L$, direct bilirubin – 5.5 $\mu\text{mol}/L$, ALT – 16.0 U/L, AST – 22.0 U/L, cholesterol – 3.1 mmol/L, total protein – 62.0 g/L, albumin – 41.0 g/L, urea – 3.2 mmol/L, creatinine – 53.0 $\mu\text{mol}/L$, GFR – 124 mL/min/1.73m², alkaline phosphatase – 125.0 U/L, LDH – 223.0 U/L, glucose – 4.3 mmol/L, CRP – 3 mg/L, RF – 1.2 IU/mL, ASO – 120 IU/mL, ferritin – 52.0 ng/mL, serum iron – 15.0 $\mu\text{mol}/L$, transferrin saturation – 34.0 %, total iron-binding capacity – 73.2 $\mu\text{mol}/L$, vitamin B12 – 250.0 pg/mL, and folic acid – 15 ng/mL.

Coagulogram (three measurements):

January 15, 2024: APTT – 44 sec, PT – 61 %, INR – 1.1, TT – 29 sec, fibrinogen – 1.17 g/L, and D-dimer – 0.2 $\mu\text{g}/\text{mL}$.

February 14, 2025: APTT – 43.3 sec, PT – 61.5 %, INR – 1.12, TT – 22.9 sec, fibrinogen – 1.2 g/L, and D-dimer – 0.1 $\mu\text{g}/\text{mL}$.

February 28, 2025: APTT – 44.5 sec, PT – 59.2 %, INR – 1.1, TT – 23.8 sec, fibrinogen – 1.1 g/L, and D-dimer – 0.2 $\mu\text{g}/\text{mL}$.

Fibrinogen concentration was determined using the Clauss clotting method. Activated APTT, PT, TT, and other coagulogram parameters were measured on an automated analyzer. Platelet aggregation was studied using standard inducers (ADP, epinephrine, collagen, ristocetin, arachidonic acid) on a laser aggregometer.

Platelet aggregation with inducers (January 15, 2024): ADP – 71.4 %, epinephrine – 40.4 %, collagen – 77.1 %, ristocetin – 76.3 %, and arachidonic acid – 75 %.

Dysfibrinogenemia testing (March 20, 2025): ancistron test – 19 sec (normal 16–22 sec).

Antinuclear antibodies (February 14, 2025): less than 1:40.

Anti-dsDNA antibodies (February 14, 2025): 15 IU/mL – negative.

Anti-cardiolipin IgM and IgG antibodies (February 14, 2025): negative.

Lupus anticoagulant (February 14, 2025): not detected.

Urinalysis (January 14, 2025): light yellow, clear, pH 5.0, density 1020, glucose – not detected, ketones – not detected, bilirubin – not detected, squamous epithelium – 1 cell/ μL , leukocytes – 1–2 cells/ μL , erythrocytes – 1 cell/ μL , protein – not detected, bacteria – not detected, mucus – trace, and salts – not detected.

24-hour proteinuria (January 15, 2025): 0.05 g/day.

Zimnitsky test (January 20, 2025): 6.00–9.00 – specific gravity 1018, 9.00–12.00 – 1022, 12.00–15.00 – 1020, 15.00–18.00 – 1019, 18.00–21.00 – 1017, 21.00–24.00 – 1019, 24.00–03.00 – no urine, 03.00–06.00 – 1010; 24-hour diuresis – 1350 mL, and day/night diuresis ratio – 3:1.

ELISA for helminths (January 21, 2025): anti-Toxocara canis – negative, *Opisthorchis felineus* – negative, *Echinococcus* spp. – negative, *Trichinella spiralis* – negative, and *Ascaris lumbricoides* – negative.

ELISA for TORCH infections (January 21, 2025): anti-CMV IgM, IgG – negative, anti-Toxo IgM, IgG – negative, anti-HSV ½ IgM, IgG – negative, anti-Rubella IgM, IgG – negative, anti-Herpes IgM, and IgG – negative.

ELISA for Epstein-Barr virus (January 21, 2025): anti-Epstein Barr IgM, IgG – negative.

ELISA for hepatitis (January 14, 2025): HBsAg, anti-HBcore – not detected, anti-HCV – not detected, and anti-HAV IgM – not detected.

ELISA for HIV (January 14, 2025): negative.

Mantoux test with 2 TU (May 14, 2024): negative.

ENT (January 14, 2025): nasal breathing free, nasopharyngeal mucosa pink, clear; palatine tonsils of normal size, no plaques, diagnosis – healthy.

Throat and nasopharyngeal swab for flora (January 14, 2025): no growth of pathogenic microflora, single colonies of *Streptococcus viridans* 10^3 CFU/mL.

Dentist (January 14, 2025): superficial caries of tooth 3.5.

Gynecologist (January 15, 2025): healthy.

Electrocardiography (January 15, 2024): sinus rhythm, HR 94–103/min, electrical axis of the heart in vertical position.

Comprehensive ultrasound of the abdominal organs, retroperitoneal space, peripheral lymph nodes, pelvis, and thyroid gland (January 15, 2024): no pathology detected.

Genetic testing of fibrinogen genes *FGB* and *FGG* (Sanger sequencing) (April 15, 2025): a heterozygous mutation *FGB*: -455G>A was identified in the *FGB* gene.

DISCUSSION

The presented case of hereditary hypofibrinogenemia in an 8-year-old child demonstrates the typical features of the medical history (biological, genealogical), clinical, laboratory and instrumental diagnosis of hemostatic disorders, as well as the search for acquired causes that may lead to this condition, and the treatment of this rare inherited coagulation disorder.

The use of a comprehensive range of clinical and laboratory-instrumental studies in our patient made it possible to exclude pathology that could cause acquired secondary hypofibrinogenemia, such as infectious diseases, liver pathology, DIC, drug effects, hemolysis, kidney disease, hematological malignancies, etc.

Based on the data, including the family history (a lifelong persistent hypofibrinogenemia in the child's mother), the paucisymptomatic course of the disease (minimal hemorrhagic manifestations during the 8-year period of the child's life), stable hypofibrinogenemia confirmed by three hemostasis tests, prolonged thrombin time, exclusion of secondary causes of hypofibrinogenemia, and genetic analysis of fibrinogen genes, the patient was diagnosed with hereditary hypofibrinogenemia.

Hereditary hypofibrinogenemia is a well-known condition. In previous reports, there was not always a clear distinction made between hereditary and acquired forms of factor I deficiency [8]. This distinction is crucial for diagnosis and management.

Considering the prevalence of this pathology, it should be noted that the epidemiological characteristics of hypofibrinogenemia have now been adequately studied. Specifically, in a study conducted by Paraboschi E.M. et al. (2017), it was demonstrated that among 342 registered cases of congenital fibrinogen disorders, 58 % were hypofibrinogenemias, 28 % were dysfibrinogenemias, and only 14 % were afibrinogenemias. Additionally, the authors noted significant

variation in clinical manifestations based on the type of mutation [4].

In a study conducted by Yakovleva E.V. et al. (2022), data from 47 patients with hereditary fibrinogen disorders were analyzed. The study found that 38 % of the patients exhibited an asymptomatic course [10]. The authors emphasize that, even in the absence of clinical symptoms, these patients remain at a high risk of hemorrhagic complications during invasive procedures [10].

Of particular interest are the findings of Neerman-Arbez M. et al. (2016) that hereditary hypofibrinogenemia often presents in childhood and is frequently detected incidentally during routine examinations, as presented in our clinical case. In some instances, a family history may not be available due to the variable nature of the mutations [11].

The severity of hemorrhagic manifestations in patients with hereditary hypofibrinogenemia directly correlates with the degree of decrease in plasma fibrinogen levels [8]. Patients with severe hypofibrinogenemia (0.2–0.3 g/L) may experience spontaneous bleeding, whereas those with milder forms (1.0–1.5 g/L) usually only bleed or develop hematomas after trauma or invasive procedures [3, 8]. Our patient has a paucisymptomatic course, with a mean fibrinogen level of 1.1–1.2 g/L, comparable to those of her mother, who also has a mean fibrinogen level of 0.87–0.97 g/L.

Despite the typical laboratory signs, genetic testing is a method used to diagnose hereditary hypofibrinogenemia [6, 11, 12].

Long-term follow-up is recommended for the child, including monitoring of the coagulogram profile and other diagnostic tests, due to their hereditary nature and the child's female sex. If necessary, consideration may be given to replacement therapy, particularly in cases involving surgery or other invasive procedures [13].

CONCLUSION

All physicians should be aware that hereditary hypofibrinogenemia may present with a frequently latent or paucisymptomatic course, and may not be accompanied by bleeding for many years, or may present with rare bleeding, usually of the microcirculatory type. It is also important to identify this pathology in patients with a positive family history, as early diagnosis and adequate follow-up can help to implement preventive measures and reduce the risk of hemorrhagic complications in such cases.

Funding

All authors declared no financial support for the preparation of this manuscript.

Conflict of interest

The authors declare no conflict of interest.

REFERENCES

1. Voytsekhovskiy VV. Clinical observation of a patient with congenital dysfibrinogenemia. *Amur Medical Journal*. 2022; 1: 68–72. (In Russ.). [Войцеховский В.В. Клиническое наблюдение пациентки с врожденной дисфибриногенемией. *Амурский медицинский журнал*. 2022; 1: 68–72.]. doi: 10.24412/2311-5068_2022_1_68
2. Sturov VG, Chuprova AV, Antonov AR, Anmut SYa. Hereditary dysfibrinogenemias: current state of the problem of clinical and laboratory diagnostics and targeted therapy. *Hematology and Transfusiology*. 2005; 5: 35–40. (In Russ.). [Стуров В.Г., Чупрова А.В., Антонов А.Р., Анмут С.Я. Наследственные дисфибриногенемии: современное состояние проблемы клинико-лабораторной диагностики и направленной терапии. *Гематология и трансфузиология*. 2005; 5: 35–40.].
3. Rukavitsyn OA, Ignatiev SV, Rummyantsev AG. *Hemostasis System: Theoretical Foundations and Clinical Practice: National Guidance*. Moscow: GEOTAR-Media; 2024. (In Russ.). [Рукавицын О.А., Игнатьев С.В., Румянцев А.Г. Система гемостаза. Теоретические основы и клиническая практика: Национальное руководство. М.: ГЭОТАР-Медиа; 2024.]. URL: <https://www.rosmedlib.ru/book/ISBN9785970484975.html>
4. Paraboschi EM, Duga S, Asselta R. Fibrinogen as a Pleiotropic Protein Causing Human Diseases: The Mutational Burden of A α , B β , and γ Chains. *International Journal of Molecular Sciences*. 2017; 18(12): 2711. doi: 10.3390/ijms18122711
5. Piva JP, Garcia PCR, da Rocha TS. Hypofibrinogenemia and Pediatric Sepsis: Another Piece of the Puzzle! *Pediatr Crit Care Med*. 2018; 19(5): 491–492. doi: 10.1097/PCC.0000000000001514
6. Neerman-Arbez M, Honsberger A, Antonarakis SE, Morris MA. Deletion of the fibrinogen alpha-chain gene (FGA) causes congenital afibrinogenemia. *J Clin Invest*. 1999; 103(2): 215–218. doi: 10.1172/JCI5471
7. Fish RJ, Neerman-Arbez M. Fibrinogen gene regulation. *Thrombosis and Haemostasis*. 2012; 108(3): 419–426. doi: 10.1160/TH12-04-0273
8. Barrkagan ZS, Momot AP. *Diagnosis and controlled therapy of hemostasis disorders*. 3rd ed. Moscow: Newdiamed; 2008; 292 p. (In Russ.). [Баркаган З.С., Момот А.П. Диагностика и контролируемая терапия нарушений гемостаза. 3-е изд. М.: Ньюдиамед; 2008; 292 с.].
9. Vorob'ev AI. *Guidelines on hematology*. 4th ed. Moscow: Newdiamed; 2007. 1275 p. (In Russ.). [Воробьева А.И. Руководство по гематологии. 4-е изд. М.: Ньюдиамед; 2007; 1275 с.].
10. Yakovleva EV, Konyashina NI, Gorgidze LA, Lavrova PS. Hereditary hypofibrinogenemia: bleeding or thrombosis? *Hematology and Transfusiology*. 2022; 1: 54. (In Russ.). [Яковлева Е.В., Коняшина Н.И., Горгидзе Л.А., Лаврова П.С. Наследственная гипофибриногенемия: кровотечения или тромбозы? *Гематология и трансфузиология*. 2022; 1: 54.]. doi: 10.35754/0234-5730-2022-67-2-193-201
11. Neerman-Arbez M, de Moerloose P, Casini A. Laboratory and Genetic Investigation of Mutations Accounting for Congenital Fibrinogen Disorders. *Seminars in Thrombosis and Hemostasis*. 2016; 42(4): 356–365. doi: 10.1055/s-0036-1571340
12. Kuznik BI, Sturov VG, Levshin NYu, Maksimova OG, Kudlay DA, et al. *Hemorrhagic and thrombotic diseases and syndromes in children and adolescents: pathogenesis, clinic, diagnosis, therapy and prevention*. 2nd ed., revised and additional. Novosibirsk: Nauka; 2018; 524 p. (In Russ.). [Кузник Б.И., Стуров В.Г., Левшин Н.Ю., Максимова О.Г., Кудлай Д.А. и др. Геморрагические и тромботические заболевания и синдромы у детей и подростков: патогенез, клиника, диагностика, терапия и профилактика. 2-е изд., перераб. и доп. Новосибирск: Наука; 2018; 524 с.].
13. Crighton GL, Huisman EJ. Pediatric Fibrinogen PART II-Overview of Indications for Fibrinogen Use in Critically Ill Children. *Front Pediatr*. 2021; 9: 647680. doi: 10.3389/fped.2021.647680

Information about authors

Marina V. Gomellya – Dr. Sc. (Med.), professor of the Department of Children's Diseases and Children's Infections, Irkutsk State Medical University; e-mail: marina_gomellya@mail.ru, <https://orcid.org/0000-0002-0441-4548>

Tamara S. Krupskaya – Cand. Sc. (Med.), head of the Department of Children's Diseases and Children's Infections, Irkutsk State Medical University; e-mail: krupskaya-tamara@mail.ru, <https://orcid.org/0000-0002-0285-3627>

Tatyana Yu. Belkova – Cand. Sc. (Med.), deputy chief physician for Medical Affairs, Ivano-Matreninskaya City Children's Clinical Hospital; e-mail: belkova@imdkb.ru, <https://orcid.org/0000-0002-1656-9387>

Albina N. Vorzheva – deputy chief physician, Ivano-Matreninskaya City Children's Clinical Hospital for Outpatient and Polyclinic Services; e-mail: albina001@gmail.com, <https://orcid.org/0009-0000-5802-6789>

Anastasia V. Mironova – pediatrician, assistant of the Department of Children's Diseases and Children's Infections, Irkutsk State Medical University; e-mail: tav23_97_1997@mail.ru, <https://orcid.org/0009-0002-7774-4836>