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Schenlein-Genoch's Disease In Children: Clinical Characteristics

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Abstract. The article retrospectively analyzes the clinical course of hemorrhagic vasculitis in children and the characteristics of the course in children with associated pathologies. Currently, hemorrhagic vasculitis is one of the most common and frequent pathologies in the pediatric population. Its frequency is very variable and depends on the region, its level of economic development, diagnostic and statistical features. Various manifestations of hemorrhagic vasculitis clinical signs, the severity of the course of the disease, the consequences largely depend on the state of the coagulation and anticoagulation systems of hemostasis, and which organ is damaged, and kidney damage determines the probability of a severe course of the disease.

Key words. hemorrhagic vasculitis, Schönlein-Henoch disease, clinical, congenital pathologies, children

Introduction. Schenlein-Genox's disease is a disease belonging to the group of systemic vasculitis with microcirculatory disturbances as a result of the accumulation of immunoglobulin A (IgA)-containing immune complexes in the blood vessels of the skin, joints, gastrointestinal system, and kidneys. This disease is one of the most common systemic vasculitis in childhood. Worldwide, the incidence among children ranges from 3 to 26.7 per 100,000 children. In the pathogenesis of the disease lies the damage of the intima of small blood vessels in the skin, joints, gastrointestinal system, kidneys with IgA-immunocomplexes. As a result, endothelial dysfunction - a decrease in the synthesis of fibrinolysis activators, activation of the lipid peroxidation system, coagulation-platelet hemostasis processes occurs [1,3,4,6]. Different manifestations of clinical symptoms, severity of the disease, consequences largely depend on the state of the coagulation and anticoagulation systems of hemostasis. The course and clinical features of HV in children depend on which system or organ is damaged. The course and consequences of the disease largely depend on the damage to the kidneys. In children, clinical signs of kidney damage in GVs occur in 26-60% of cases [2,12,15].

Purpose of study. Retrospective analysis of clinical types and degrees of Schenlein-Genoch's disease in children, age- and sex-related characteristics of the course, and the level of concomitant pathologies.

Research materials and methods. The study was conducted at the clinic of the Tashkent Medical Academy. Medical history of children aged 3 to 17 years were copied and recorded on specially designed cards, who were treated in hospital with the diagnosis of hemorrhagic vasculitis (GV) at the Children's Cardiorheumatology Department of the multidisciplinary clinic of the Tashkent Medical Academy in 2012-2022, a total of (416). In the study were divided into groups and analyzed according to age, gender, clinical course, seasonal dynamics, severity level, concomitant diseases.

For a long time, they have been used to diagnose HV diagnostic criteria proposed by the American Society of Rheumatology in 1990 (J.A. Mills et al., 1990). Availability of 2 any of the 4 signs presented in Table 1 makes it possible to diagnose HSP with a sensitivity of 87.1% and a specificity of 87.7% (J.A. Mills et al., 1990).

Table.
Classification New diagnostic criteria for HSP were proposed in 2010

Criterion	Description
Palpable purpura	Slightly raised hemorrhagic skin changes not associated with tombocytopenia
Age <20 years	Age of onset of disease less than 20 years

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Stomach ache	Diffuse abdominal pain, worse after eating or intestinal ischemia (may be intestinal bleeding)
Detection of granulocytes on biopsy	Histological changes revealing granulocytes in the wall of arterioles and venules

New diagnostic criteria for HV were proposed in 2010 (S. Ozen et al., 2010). Making a diagnosis of HV is possible if you have the patient has characteristic skin rashes (purpura, in most cases palpable) with their characteristic localization mainly on the skin lower extremities (mandatory sign) in combination with at least one additional feature; abdominal pain; arthritis or arthralgia; kidney damage (proteinuria and/or hematuria); histological changes in the form of leukoplastic vasculitis with predominant deposition of IgA deposits or glomerulonephritis with deposition of IgA deposits; histological changes in the form of leukoplastic vasculitis with predominant deposition of IgA deposits or glomerulonephritis with deposition of IgA deposits. Sensitivity are 100% and specificity 87.8% of the new diagnostic criteria for HV, respectively (S. Ozen et al., 2010). In most cases, diagnosing HV is not difficult and is based on identifying the characteristic clinical signs of the disease - the presence of skin rashes, damage to the gastrointestinal tract, involvement of joints and kidneys [9]. To date, there are no laboratory methods that can reliably confirm diagnosis of HV. Serum IgA concentrations are elevated in pproximately half of children with HSP and do not correlate with either the severity or activity of the disease [2]. Thus, it is now necessary to highlight laboratory parameters as diagnostic criteria for HV and activity of its manifestations.

The results and their discussion. In the study of the dynamics of children treated with the diagnosis of GV in TTA cardiorheumatology, it was found that there was an increase in the trend of the disease until 2019, but this is not statistically reliable. The decrease in the dynamics of inpatient treatment of children with this diagnosis in 2020-2021 can be attributed to the coronavirus pandemic. These patients are more likely to be treated on an outpatient basis. In addition, from this data, it is not possible to determine the trend of increasing or decreasing GV among children, because children with GV are not treated only in this clinic. Patients from different regions of the republic are treated in the clinic of the Tashkent Pediatric Medical Institute.

According to the results, hemorrhagic vasculitis in children that occurred in 7-14 years of age in 71.7% of cases, the disease occurred in 2.1% of cases in 1-3-year-olds, and in 2.8% of cases in 15-18-year-olds. 51.4±2.4% of 416 children are boys and 48.6±2.4% are girls, that is, the disease is more common in boys. This is consistent with the data from the studied literature [2,3,4]. It was found that there is a seasonality in the increase of the disease, in particular, the disease increases in autumn (September, October, November), spring (March, April, May).

Other researchers also observed a decrease in the occurrence of the disease in the summer months. In particular, Kudryasheva M.A. (2015) found that the disease is recorded 3-4 times less often in the summer months than in other seasons, but more severe forms of the disease occur in the summer season. The duration of inpatient treatment of children with hemorrhagic vasculitis depends on the clinical form of the disease and its severity, as well as accompanying pathologies in the child. According to our results, the average duration of children's inpatient treatment was 10±2.3.

The most obvious clinical and diagnostic sign of hemorrhagic vasculitis in children is a palpable erythematous rash on the skin. [2,4,9,11,15,17]. It is difficult to diagnose the disease before the rash appears in patients, but the disease is confirmed when the rash appears. The rash appears mainly on the legs and hands of the patient after 3-4 days of the disease. From the anamnesis of the patients, acute respiratory tract infections before the manifestation of clinical symptoms: nasopharyngitis 221 (53.1±2.4%), tonsillitis 266 (23.7±2.0%) and fever 99 (63.9±2.3%) clinical signs were observed.

In many studies devoted to the study of hemorrhagic vasculitis in children, the allergic background and frequent acute respiratory infections in children's bodies are noted as factors that trigger the development of the disease [2,3,4,7,10].

The onset of the disease is often accompanied by a nasopharyngeal or intestinal infection, food allergy [17]. The presence of an infectious disease (in most cases, a nasopharyngeal or intestinal infection) before the onset of GV determines the ongoing interest in the role of various infectious agents in the development of

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the disease. According to various reports, an upper respiratory tract infection occurs before the onset of HV in 30-65% of cases [16]. In addition, a number of studies have shown a high prevalence of chronic infections, particularly chronic sinusitis or tonsillitis, in 74% of children with HV [15].

In our study, 403 (96.8±0.8%) children had an allergic background, in particular, when studying the child's life history, it was found that he was sick with various diatheses, worms, frequent infections of the upper respiratory tract. Pathologies of the body (chronic diseases) in children affect the course of any disease. From the results of our study, 252 (60.5%) patients had associated pathologies. According to the results of examination of the sick children, atopic dermatitis (92 22.1±2.0%), anemia (266 23.7±2.0%) and various infections predominated, in particular enterovirus infection - 46 (11.1±1 .5%), gastrointestinal tract infections: 76 (18.2±1.8%), helicobacter infection - 105 (25.2±2.1%), gastroduodenitis - 43 (10.3±1.4%). Acute respiratory viral infections observed in 221 (53.1±2.4%) patients were noted as the causative factor of the disease. According to the literature, the most common form of the disease is the skin and skin-joint form [2,4,5], and in all the patients in our observation, the skin and skin-joint form was the leader, and hemorrhagic rashes on the skin were observed in 100% of patients. At the same time, in 93 patients (22.3±2.0%) a normal (skin) form was detected, in which the rash was observed in a purple type with a hemorrhagic tint. The rash is symmetrically located and has a typical localization, it is detected around the joints, on the buttocks, on the legs, and often rises above the skin level. After the rash, pigmentation appears, the skin becomes dry and flaky. In most 74 (17.7±1.8%) patients, skin-hemorrhagic syndrome and arthritic syndrome coexisted, children's legs, hand joints, rarely knee joints, spine joints were injured. A characteristic feature of HV is that there are no complications in the joints and full recovery, changes can be observed in rare cases. In hemorrhagic vasculitis, arthritis (injury of the joints) often coincides with the appearance of the rash and usually manifests itself in the form of joint pain, hyperemia, and periarticular swelling.

In children with hemorrhagic vasculitis, the occurrence of concomitant pathologies was observed in 252 (60.5±2.3%) children. 183 (43.9±1.4%) patients had a skin-joint form, 45.3% of them (98 patients) had associated pathologies. In particular, 13.0% of child patients had recurrence of arthritis symptoms. According to the studied literature, changes in the gastrointestinal tract are detected in HVs in 80% of patients [14] The main clinical symptoms are abdominal pain (88%), bleeding (75%), diarrhea (30%) and vomiting (25%). According to the results of our research, this pathology was detected in 62.0% of children. Complaints of these patients were manifested in the form of abdominal pain, diarrhea, vomiting, loss of appetite.

The characteristics of pain in patients range from weak epigastric pain to strong flank pain, and often the clinical picture is similar to acute abdominal pain. In 20-50% of cases, the gastrointestinal clinic should begin before the appearance of skin rashes. In 16-30% of children, abdominal pain is aggravated by bleeding from the gastrointestinal tract caused by thrombosis and necrosis of the vessels of the intestinal mucosa, and in rare cases by intussusception and perforation of the intestine [13].

Skin-abdominal form was found in 38 patients (9.1±1.4%). It was noted that this type of hemorrhagic vasculitis was twice as common in children with concomitant pathology. 25% of these children had Helicobacter infection, 18% had worm infestations, and 10% had gastroduodenitis. Timely detection and treatment of accompanying pathologies in this type of GV led to long-term follow-up of patients in inpatient conditions, in particular, prolongation of the duration of treatment.

Clinically significant damage to the kidney in the form of nephritis develops in an average of 40% of children with GV in the first 4-6 weeks from the onset of the disease, but according to the data presented by different authors, this indicator fluctuates from 20 to 80%. [15]. Often microhematuria, proteinuria sometimes, nephrotic syndrome with massive proteinuria may develop. In some cases, renal arterial hypertension develops against the background of nephritic syndrome syndrome [12]. Morphological changes in the kidneys are often manifested in the form of mesangioproliferative glomerulonephritis [11].

In the course of hemorrhagic vasculitis in children, kidney damage worsens the course and prognosis of the disease. Manifestations of renal syndrome in patients can range from mild proteinuria and microscopic hematuria to acute kidney injury. From the results of the study, it was noted that kidney damage

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in patients was observed in almost half of the children with adjacent pathologies, and its occurrence rate was 30.4%.

The mixed form of the disease (skin+joint+abdominal+kidney) was recorded in 7.4±1.2% of patients, and the duration of inpatient treatment was prolonged in these patients, and it was found that 47% of children were treated in the inpatient hospital for more than 20 days. When patients were analyzed according to the level of activity of the disease, 43% of children with concomitant pathologies had high activity of the disease, while 18% of patients without concomitant pathologies had high activity of the disease.

Conclusion.

Almost all (100%) of patients treated for hemorrhagic vasculitis have signs of skin damage, with palpable hemorrhagic rashes on the skin. In children with GV with concomitant pathology, it was noted that the duration of inpatient treatment was 2.5-3 times higher than in children without concomitant pathology. It was found that the skin abdominal shape, as well as the kidney damage syndrome in HV, were 2 times higher in those with adjacent pathologies. In children, allergic diseases and secondary infections in the body, worm infestations were noted as predisposing factors to the development of hemorrhagic vasculitis. In particular, 35.0% of children had symptoms such as atopic dermatitis and allergy.

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January 2025

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