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Systematic Red Volchanka

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Abstract

Rheumatoid disease is a disease of unknown etiology (idiopathic) with damage to systems and organs due to an antigen-antibody complex. SQV is an autoimmune disease, and one of the main causes of clinical symptoms is the deposition of antinuclear immune complexes in tissues and basement membranes formed by antibodies with binding parts of cell nuclear elements (DNA, RNA, histones). In addition, cell fragments that die with apoptosis, increased netosis, and reduced autophagy become targets of immune system cells. Immune complexes enter plasmacytoid dendritic cells through FcyRIIa receptors, then nucleic acids of immune complexes activate interferon receptors. In this way, activated interferons (IFN -α) begin to be produced in abundance. These cytokines, in turn, produce autoreactive antibodies by monocytes and Betta cells. The production of monocytes, neutrophils and dendritic cells increases under the influence of type I IFN. All this leads to an increase in the number of immune complexes.

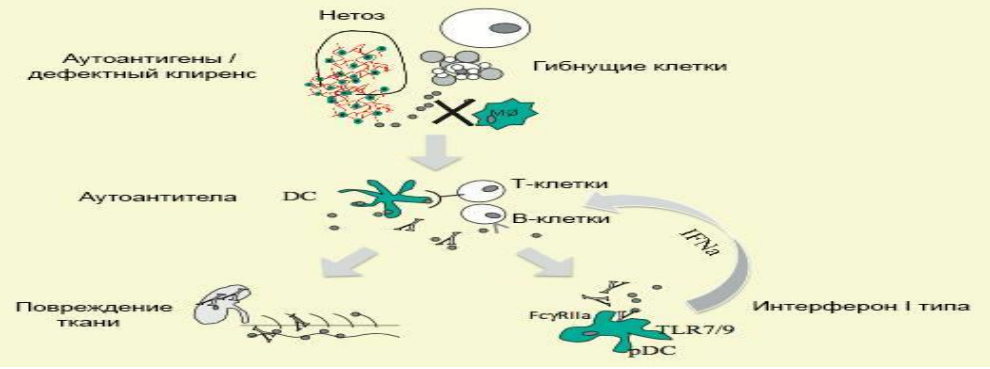
Key words: SQV (Systemic red volchanka *)*

Statistics:

It occurs in children over 1.8 years old, mostly in girls.

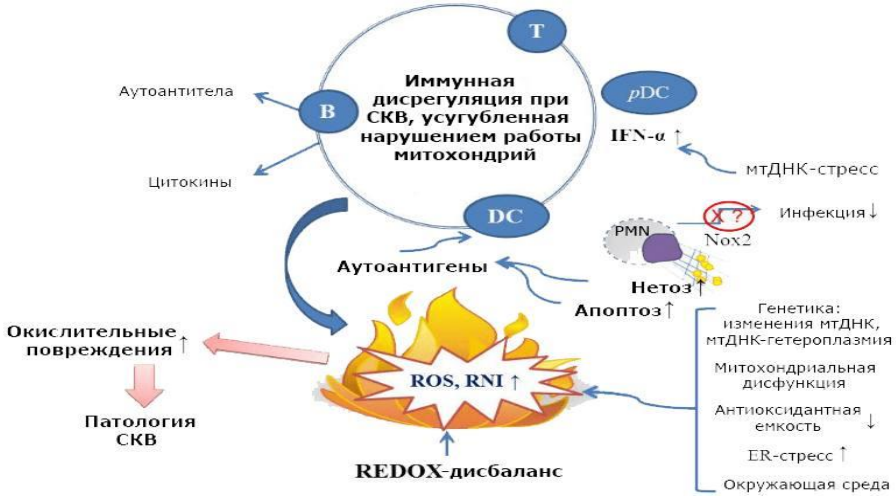
2. The highest incidence occurs at the age of 15-25.

3. The incidence ratio in women and men is 8:1



In addition, the pathogenesis of SQV involves abnormal oxygen exchange, which increases inflammation, cell death, and the introduction of autoantigens. This occurs mainly as a result of mitochondrial dysfunction. Increased secretion of pro-inflammatory cytokinins, tissue damage, and other processes that characterize SQV result in the generation of large amounts of reactive oxygen

species (ROS) that further damage surrounding tissues and induce the self-destruction of specific neutrophils. Self-destruction - *netosis* is observed .



Classification

Development periods of systemic lupus erythematosus (according to VANasanova's classification)

1. Acute period: High immunologic activity and rapid development of multi-organ damage, including kidney damage, are observed.

2. Acute period: periodic exacerbation of injuries (not as clear as in the acute period), especially in the first year of the disease, the development of kidney damage is observed.

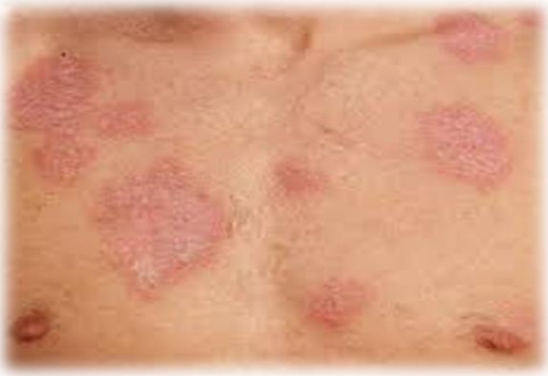
3. Chronic period: one or more symptoms dominate for a long time: skin damage, polyarthritis, hematolytic diseases, Raynaud's phenomenon, light proteinuria.

Clinical and immunological classification : Elderly people (over 50 years old) have a higher incidence of SQV than young people. Joint damage (usually in adults), pulmonary atelectasis , pneumonitis , pulmonary fibrosis, Sjögren's syndrome, peripheral neuropathy are observed.

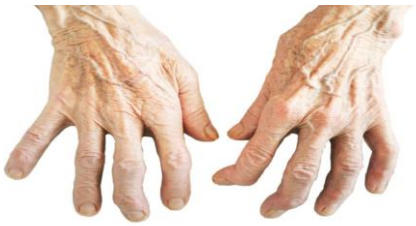
Neonatal SQV classification : Antigen antibody immune complexes can be found in the blood serum of newborns of mothers infected with SQV. Clinical manifestations develop from several weeks to several months after birth. These include erythematous rashes, complete heart block (35%).

Clinic. The clinical picture of systemic lupus erythematosus has a number of specific features and is rapidly progressing in a polysyndromal form , resulting in death in most cases as a result of the rapid development of functional failure of one or another internal organ. The disease often begins with general weakness, a rapid decrease in body weight and a long-term steady rise in temperature , joint syndrome, in some cases, Raynaud's phenomenon, and then is characterized by inflammation of internal organs and systems. syndromes occur . They are mainly associated with inflammation of the skin, joint, muscle , kidney, heart and blood vessels, lung, liver, gastrointestinal and psycho-nervous system and serous membranes and in different degrees and severity. will pass. Among those

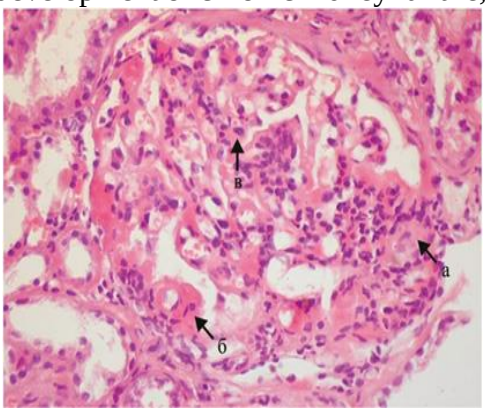
mentioned above, skin changes are often diagnostically typical of systemic lupus erythematosus, and are manifested mainly by the appearance of "butterfly"-shaped erythema on the face, cheeks , and nose. Especially in the chronic course of the disease, focal signs with a discoid character can be observed on the skin. In most cases, there are many lupus lesions on the skin (a hard and dry crust on the red part of the lip, sometimes ending with atrophy or erosion) and trophic changes (dry skin, diffuse hair loss, brittleness and deformation of nails) is determined. Symptoms observed on the skin are often associated with vascular inflammation (vasculitis), mainly edematous erythema, atrophy and telangiectases on the finger pads and palms, and bullous nodular, hemorrhagic and papulenecrotic rashes on other parts of the body, and Can manifest as Raynaud's syndrome. In turn, the presence of enanthema and sores, that is, stomatitis, is determined in the mucous layer, in the oral cavity.



Joint syndrome in systemic lupus erythematosus It is considered one of the symptoms that occurs in almost all cases and usually takes the form of polyarthralgia and polyarthritis. They are mostly symmetrical in nature and develop more often in the small, middle and knee joints of the hands and feet. During the active period of the disease, its symptoms are stable and long-lasting. Sometimes the combination of tendinitis and tendovaginitis causes an irreversible flexion contracture in the fingers. In the case of muscle inflammation (myolgia and myositis), the patients are disturbed by pain in the arms and legs and some restriction of movement.



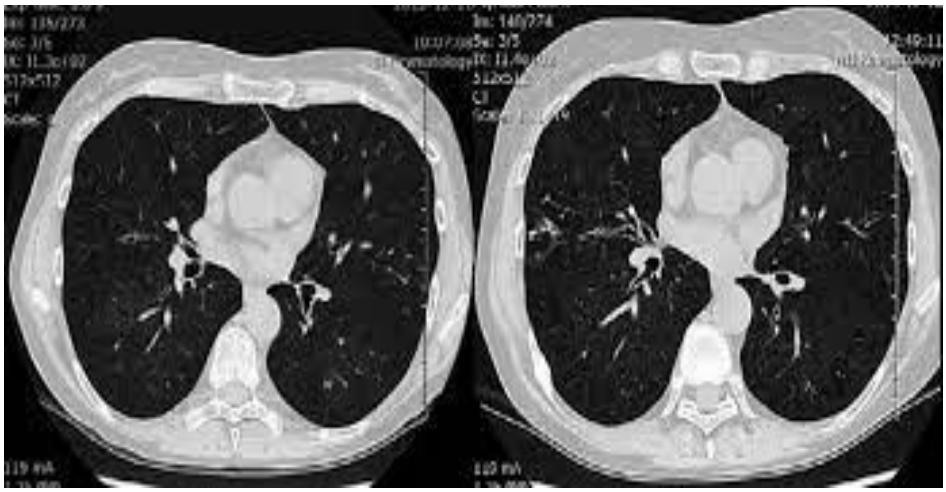
Kidney damage in systemic lupus erythematosus is called *lupus-nephritis* and is characterized by a number of specific features. Early or late clinical manifestations in patients are of great importance for the direct outcome of the disease. Its occurrence in this disease leads to the early development of chronic kidney failure, which in almost all cases ends with the death of the patient.



According to the data, lupus-nephritis is observed in 50-70% of patients. *Proteinuria is the main symptom of kidney damage in* TQV and manifests with nephrotic syndrome. According to the course, its fast and slow developing forms are distinguished. Rapidly developing lupus nephritis manifests itself in the form of rapidly developing glomerulonephritis and soon ends with the development of kidney failure. Slowly developing lupus-nephritis is also observed with nephrotic syndrome (30-40%) or obvious proteinuria accompanied by hematuria.

Also, in some cases, kidney damage in systemic lupus erythematosus can be complicated by nephrotic crisis.

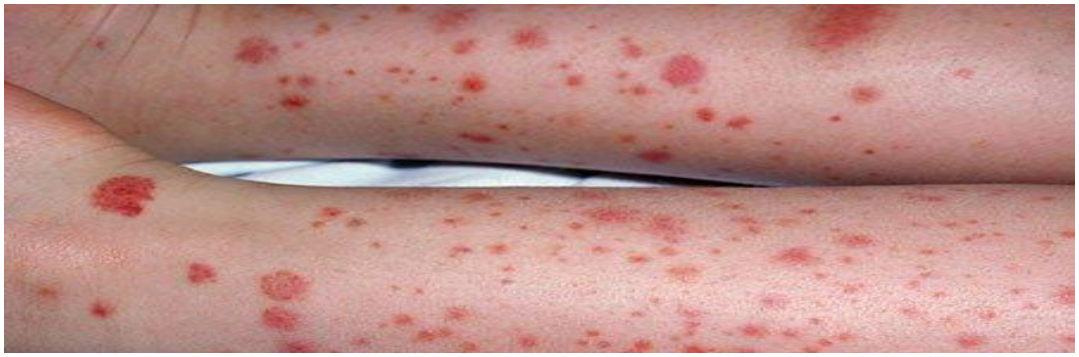
Lungs damage is often observed on the basis of vasculitis, and the patient is disturbed by spitting up blood and a dry cough. Alternatively, during the exacerbation of the disease , there is a possibility of developing *lupus -pneumonitis , which sometimes ends with infiltrates in the lungs.* In such cases, patients have fever, shortness of breath, and cough, and on X-ray (X-ray) there is an elevated position of the diaphragm and disk-like atelectasis in the basal parts of the lungs. Dry and exudative pleurisy may occur in case of inflammation of serous membranes .



Systemic red volchanka ***of the heart*** there is a possibility of inflammation of all three shells, they are mainly adhesive and rarely exudative pericarditis, myocarditis sometimes takes the form of endocarditis ending with mitral insufficiency. In addition, when coronary inflammation develops due to inflammation of most vessels, there is a possibility of myocardial infarction.

Damage ***to the gastrointestinal system*** can be manifested in different ways, and in addition to dilatation of the esophagus and disruption of its motility, ischemia of the stomach and intestines, and in some cases, perforation can be observed. Clinical symptoms of abdominal crisis sometimes appear in patients during the exacerbation of the disease . It is associated with inflammation of the endothelium of the intestines, serous membrane and mesenteric vessels, and the formation of ulcers. In turn, in this disease, inflammation of the liver manifests itself in the form of **lupus-hepatitis with specific changes** , usually the process goes from mild hepatomegaly to sometimes, clinically, in a form reminiscent of severe aggressive hepatitis. .

Changes in ***the nervous system*** are observed in almost all patients with systemic lupus erythematosus. From a clinical point of view, they are characterized by comprehensive symptoms at all stages of the disease, including asthenoneurotic syndrome (weakness , rapid fatigue, adynamia, depression, sleep disturbance, etc.) in the initial period, and polyneuritis, slow reflexes, from paraesthesia, pelvic dysfunction, meningoencephalopolyradiculoneuritis to convulsions, strokes and paralysis. In a number of cases, mental changes are also detected in patients , and they take a special place in the course of the disease. Because in patients with systemic lupus erythematosus, along with emotional sensitivity, euphoria, insomnia, decreased intelligence and ability to remember and think, loss of critical approach to oneself and high assessment of one's own capabilities, symptoms such as hallucinations are also found.



(this picture shows Verlgoff syndrome)

Systemic lupus erythematosus, as mentioned above, can be acute, subacute and chronic depending on its clinical features. In its acute course, the disease begins suddenly, along with damage to many internal organs and tissues, noticeable changes in laboratory parameters are observed for the first time. As a rule, this course of the disease progresses rapidly, and the probability of death of the patient is high, often within the next few years. In turn, the acute course is manifested by the priority of signs of damage to the kidneys and nervous system. The chronic course is relatively mild, the process begins slowly, and in most cases, signs of damage to a single organ (monosyndrome) are observed. But with the passage of certain years, it gradually damages other organs and tissues, and a polysyndromal clinical picture appears.

It should also be mentioned that one of the most important risks associated with SQV is the occurrence of many complications during pregnancy . The majority of patients are young women of childbearing age, so family planning, pregnancy management, and fetal monitoring are of great importance. It has been found that women with systemic lupus erythematosus often experience fetal loss, miscarriage, premature birth, and preeclampsia. In the 1960s, 40% of the fetuses of women infected with this disease died. By the 2000s , such cases had more than halved. Today, researchers estimate this figure to be 10-20%.

Instrumental laboratory tests. The following changes are important in the diagnosis of systemic lupus erythematosus:

- discovery of LE cells;
- Presence of antinuclear antibodies;
- Detection of antinuclear factor;
- Discovery of antibodies against DNA antigens;
- Presence of rheumatoid factor;
- When the immune system is examined, immune deficiency and changes in immunoglobulin fractions are observed.

However, it should be noted that the negative results of the above tests do not completely rule out systemic lupus erythematosus. *Microscopic examination of tissue biopsy* is of particular importance in all systemic diseases of the connective tissue, including SQV . Finding specific changes in the skin biopsy in the early stages of the disease and analyzing them together with certain clinical signs play an important role in its diagnosis. In this case , skin epidermal atrophy, hyperkeratosis with keratotic barrier, vacuolar dystrophy of the basal layer, destruction of connective tissue material and composition due to fibrinoid swelling of collagen fibers in the dermis, accumulation of basophils, and presence of hemotoxylin bodies are observed. In addition, productive and productive-destructive changes are characteristic of the walls of more small vessels, lymphoid and plasmacytic infiltration of the tissue is determined.

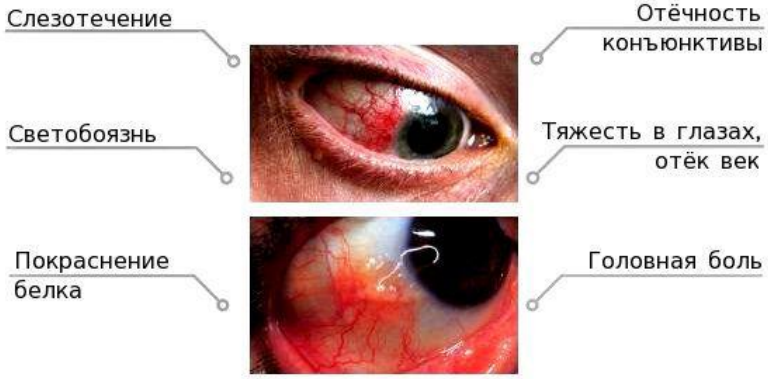
are characteristic of systemic lupus erythematosus , the following indicators are important in determining the severity of the disease:

- Dysproteinemia (especially γ- high globulin indicators);
- Anemia;
- Leukopenia;
- Thrombocytopenia;
- ECHT opening;
- Increased amount of fibrinogen;
- High titer of seromucoid proteins;
- In some cases, the Wesserman test is positive.

In addition to the above, the use of the following criteria recommended by the American Association of Rheumatologists makes it easier to diagnose SQV in a timely manner without complex laboratory and instrumental tests:

- "Butterfly"-shaped rash on the face;
- Discoid foci;
- Photosensitization;
- Ulcers in the mouth and nose;
- Arthritis without deformation;
- Serositis (pleurisy or pericarditis);
- More than 0.5 g of protein in the amount of urine excreted during the day or changes in urine sedimentation;
- Mental changes (psychosis) or convulsions;
- Hemolytic anemia, leukopenia, thrombocytopenia;
- LE- cells, detection of antibodies in DNA, probability of positive Wasserman reaction;
- Detection of antinuclear antibodies in the blood

The presence of at least 4 of the above criteria is the basis for making a diagnosis of systemic lupus erythematosus.



Treatment. Although the problems of SQV are regularly studied, its treatment remains a complex issue. Since the etiological factors are not clear, therapeutic agents are mainly aimed at alleviating the individual symptoms of the disease. Due to the diversity of the course of the disease, some of its forms have a dangerous, rapidly developing, sometimes severe course with long spontaneous remissions, it is difficult to create treatment methods. GKS, cytostatic immunodepressants (azotioprine , cyclophosphamide, chlorambucil) and 4-aminocholine products (plaquenil, delagil) are used in its treatment. Also, mechanical blood purification - plasma exchange, lymphapheresis, immunosorption methods are used. In our country, filtration of blood through activated charcoal - hemosorption is often used. YaQNDs are recommended as an additional tool .

Corticosteroids . GKS are used as first-line drugs in the acute and subacute periods of SQV, which are manifested by painful visceral manifestations . However, the complications that arise in their use, together with the correct diagnosis , require reasonable prescribing with a clear understanding of the nature of biserial changes. Damage to the CNS and kidneys is an absolute indication for prescribing GKS .

Cytostatic immunodepressants . SQV is often used azothioprine, cyclophosphane (cyclophosphamide) and chlorbutin (chlorambucil, leukeran). Unlike GKS, although sufficient studies have been conducted to evaluate the effects of these drugs, there is no consensus on their effectiveness. Instructions for prescribing cytotoxins in the complex treatment of patients with SQV :

- ✓ Lupus- nephritis;
- ✓ High activity of the disease and resistance to GKS or manifestation of side effects in the early stages of treatment with them (especially in adolescents, hypercortisolism, which occurs even in small doses of prednisolone);
- ✓ The maintenance dose of prednisolone is higher than 15-20 mg per day, when it is necessary to reduce it.

In the treatment of SQV, when drugs from different groups are used together, the following scheme is used. Azathioprine and cyclophosphamide should be taken at an average dose of 2.0-2.5 mg (per 1 kg of the patient's body weight per day), chlorbutin at a dose of 0.2-0.4 mg (per kg per day) along with low (25 mg) and medium (40 mg) doses of prednisolone. is used. In recent years, several cytostatics, including azathioprine + cyclophosphamide (1 mg per 1 kg of body weight per day) have been used . In this combination , the development of lupus-nephritis can be slowed down.

are recommended only intravenously (1000 mg once a month for the first six months $1m^3$, then 1000 mg every 3 months for 1.5 years).$1m^3$

Plasmapheresis, hemosorption. In case of failure of commonly accepted methods in the treatment of SQV, the use of plasmapheresis and hemosorption is based on the removal of biologically active substances from the blood, in particular, inflammatory mediators, circulating immune complexes, cryoprecipitins, various antibodies. Such treatment is considered to help reduce the mononuclear system, which stimulates endogenous phagocytosis, which leads to a decrease in the level of organ damage. Plasmapheresis and hemosorption methods should be added to the treatment complex of patients who are resistant to treatment and have severe systemic lupus erythematosus. Under the influence of treatments (course of treatment 3-8 times), the general condition of the patient improves significantly, signs of disease activity decrease, including preservation of kidney function in nephritis, disappearance of strongly manifested skin changes, and a clear acceleration of healing of trophic ulcers on the legs. observed. Plasmapheresis and hemasorption are carried out in parallel with treatment with corticosteroids and cytostatics. ***Immune system modulating drugs*** - levamisole, frentizol - are not widely used in systemic lupus erythematosus. There is some information that their use together with GKS and cytostatics has been effective in some cases when the usual methods do not help or when secondary infection is added. ***Aminoquinoline products and non-steroidal anti-inflammatory drugs*** are prescribed for the treatment of patients with systemic lupus erythematosus without visceral changes and to maintain remission during the period of reducing the dose of GKS and cytostatics. Aminoquinoline drugs usually use Delagil (0.25-0.5 g per day) or Plaquenil (0.2-0.4 g). YaQNDs are prescribed as an additional drug in persistent arthritis, bursitis, polymyalgia. GKS are prescribed in high doses (60-100 mg per day) in the treatment of systemic lupus erythematosus with damage to the central nervous system.

If psychoneurological changes increase when increasing the dose of GKS, it should be reduced. Intravenous cyclophosphamide in pulse-therapy doses is the most effective of cytostatics. In acute psychosis, neuroleptics, tranquilizers, antidepressants should be used together with prednisone to eliminate it. In recent years, the effectiveness of anticoagulants in the treatment of chorea has been proven. In cases of severe damage to the central nervous system, pulse therapy and plasmapheresis are performed. In cerebrovasculitis, which is the first symptom of coma, administration of a large amount of intravenous methylprednisolone (500 mg daily for 4 days) has a positive effect. These treatments reduce mortality from MNS damage from SQV and provide effective benefits.

of GKS and cytostatics in various schemes and together is the basis of treatment of nephritis caused by systemic lupus erythematosus. In cases of marked nephrotic syndrome, high hypertension and kidney failure, and rapidly developing glomerulonephritis in the early stages of the disease, one of the following schemes can be used.

- Pulse therapy together with methylprednisolone + cyclophosphane 3-6 times per month, in the interval - prednisolone 40 mg per day with a gradual reduction of the dose to 20-30 mg per day

in the 6th month, in the remaining 6 months - 5-10 mg per day in maintenance doses 2- 3 years sometimes for a lifetime. Usually, in addition to corticosteroids and cytostatics, aminoquinoline drugs (1-2 tablets Plaquenil or Delagil per day), hypotensive, diuretics, angioprotectors, disaggregatants are prescribed and taken for 6-12 months (the course of treatment is repeated if necessary);

❖ Prednisolone 50-60 mg per day + cyclophosphamide 100-150 mg per day for 2 months + heparin 5000 units 4 times a day for 3-4 weeks and curantil 600-700 mg per day. Then the daily dose of prednisolone is reduced to 40-30mg. Cyclophosphamide up to 100-50 mg and the treatment for another 2-3 months, after which maintenance therapy is prescribed at the dose indicated above (see point 1).

❖ These two schemes of treatment are carried out on the basis of plasmapheresis or hemosorption (once every 2-3 weeks , 6-8 treatments in total), together with hypotensive and diuretic agents. Plasma ultrafiltration when there are a lot of tumors, and 1-2 courses of hemodialysis when the symptoms of kidney failure increase are advisable.

In nephrotic syndrome, it is advisable to choose one of the following three schemes:

- Prednisolone 50-60 mg per day for 6-8 weeks, then the dose is reduced to 30 mg for 6 months and then to 15 mg in 6 months;
- In one day, prednisolone 40-50 mg + cyclophosphamide or azathioprine 100-150 mg for 8-12 weeks, then the dose of prednisolone is the same as above, and cytostatics are prescribed at 50-150 mg per day for 6-12 months.
- Pulse therapy with methylprednisolone and cyclophosphamide or intermittent scheme - pulse therapy with methylprednisolone - hemosorption or plasmapheresis - pulse therapy with cyclophosphamide followed by prednisolone 40 mg per day for 4-6 weeks and then in maintenance doses for 6-12 months.

In active nephritis with pronounced urinary syndrome (proteinuria 2 g per day, erythrocyturia 20-30 in the visual field, but blood pressure and kidney function have not changed significantly), treatment schemes can be as follows:

✓ Prednisolone 50-60 mg every 4-6 weeks + aminoquinoline drugs + symptomatic agents;

✓ Prednisolone 50 mg + cyclophosphamide 100 mg per day for 8-10 weeks, then dose reduction and maintenance therapy is done as indicated above;

✓ Pulse therapy methylprednisolone and cyclophosphamide (methylprednisolone three-day course 1000 mg every day and 1000 mg cyclophosphamide - one day) together, then prednisolone 40 mg for 6-8 weeks, then reducing the dose to 20 mg per day for 6 months. Maintenance therapy according to the above principles during the following months.

when patients develop chronic kidney failure and the level of creatinine in the blood is 1.0 mmol/l or higher.

Prevention. There is no specific *primary prevention of the disease* . However, risk groups are identified in the primary prevention of SQV . Examination of relatives of patients, if even one of its symptoms (persistent leukopenia, increased ECHT, hypergammaglobulinemia, etc.) is detected in the blood test, then the preventive measures in such persons should also be SQV. Patients with isolated lesions of the skin (discoid volchanka) deserve special attention. They cannot receive treatment in spas, gold preparations , ultraviolet rays.

Secondary prevention of SQV should be aimed at preventing disease exacerbation and development. When the patient eats, it is necessary to follow a diet with a small amount of salt and sugar. They should also avoid surgery, vaccinations, extreme heat and cold as much as possible . Light gymnastics and walking are very important. SQV timely prevention and treatment leads to a significant improvement of the consequences of this disease.

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