Dysproteinemia

Dysproteinemia is any change in the protein content of the blood. Plasma protein concentrations depend on the ratio between their synthesis and degradation or excretion. When one or more globulin fractions increase, the albumin-globulin ratio - the so-called A / G coefficient - changes (standard 1.5–2).

General causes of dysproteinemias

- **hereditary** (a genetic disorder of protein synthesis or release into the circulation),
- **losses of certain protein fractions:**
    - *kidney* - proteinuria (see nephrotic syndrome),
    - *skin* - severe burns, extensive inflammation,
    - *intestine* - exudative enteropathy,
- **proteosynthesis disorders:**
    - *liver* - a disorder of liver proteosynthesis in cirrhosis, hepatitis, etc .; especially the synthesis of transport proteins (albumin, prealbumin, transferrin, etc.) decreases, in case of a more severe disorder the insufficient synthesis of coagulation factors is manifested by bleeding,
    - *malnutrition with insufficient protein intake in the diet* - protein-caloric or pure protein malnutrition (up to the image of kwashiorkor),
- **relative changes in proteinemia during dehydration, fluid transfer to 3rd space or fluid retention** (clinically known phenomenon of decreased proteinemia after infusion into a dehydrated patient),
- **inflammatory reaction** - synthesis of immunoglobulins, acute phase proteins, attenuation of transport protein synthesis.

Dysproteinemia involving multiple fractions or proteins

Hyperproteinemia

An increase in all serum proteins or alternatively an increase in only some of the proteins, which leads to an increase in the protein concentration as a whole. As for the multiplication of all proteins, then it is actually a relative increase in concentration due to water loss and thickening of the internal environment. An increase in oncotic pressure results in the transfer of water from the interstitium to the plasma. Hyperproteinemia (and paraproteinemia) can lead to hyperviscosity syndrome (see below).

Hypoproteinemia

Reduction of total serum proteins, mainly albumin. It is caused by loss (in the urine, burns), impaired production or a decrease in the synthesis (inflammation) of liver proteins. It leads in a more severe case to a decrease in plasma oncotic pressure and to edema.

Acute inflammation

The so-called acute phase reaction is typical, when in electrophoresis we record a decrease in albumin with a simultaneous increase of all globulin fractions.

Chronic inflammation

An increase in the γ-globulin fraction due to increased IgG (or IgM) synthesis is typical. It is actually a polyclonal hyperimmunoglobulinemia in which the entire immune system is stimulated. It can occur in chronic infections, allergies, autoimmune diseases, malignancies. Elevated Ig levels often make up more than half of plasma proteins.

Dysproteinemia involving a single fraction or protein

Albumin changes

Analbuminemia and hypolbuminemia

Rarely due to an inherited anomaly. Generalized or localized edema. Oncotic pressure is reduced. Although albumin normally makes up about 60% of serum protein, it can be completely absent without serious disorders. Compensatingly increased globulin content. However, the total protein content is below the norm.

Albumin, as a negative acute phase protein, decreases significantly in inflammatory reactions or tumors. Hypalbuminemia may also be a manifestation of malnutrition, liver disorder or increased renal or intestinal loss (see above).

Changes in α1-globulins

A1-antitrypsin deficiency

Hereditary disease caused by a disorder in the release of synthesized protein from liver cells into the blood. It is usually asymptomatic, manifesting itself only when the plasma concentration falls below 10% of the norm, again only in some patients. Insufficient inhibition of neutrophil elastase leads to destruction of the lung lining and the development of juvenile emphysema. Accumulation of protein in hepatocytes can also lead to the development of cirrhosis. The inheritance of all risk genotypes is autosomal recessive. In heterozygotes, serum protein content is also reduced, but without clinical manifestations.

Changes in α2-globulins

Haptoglobin deficiency

Rare hereditary disorder with impaired synthesis - clinically asymptomatic. A more common (and diagnostically significant) is a transient decrease in hemolysis due to haptoglobin consumption after its binding to free globin.

Ceruloplasmin deficiency

Wilson's disease

Ceruloplasmin is the most important serum copper-binding protein. The absence causes the copper to flush rapidly from the blood into the tissues and excreted by the kidneys. Consequently, enteral resorption of copper increases, thus maintaining the predominance of uptake over losses. Copper is stored mainly in the brain in the basal ganglia (neurological form, when the symptoms of Parkinson's disease appear) and in the liver (hepatic form, liver cirrhosis occurs). Furthermore, copper is deposited in the cells of the proximal tubules of the kidneys (transport disorder, renal glycosuria, aminoaciduria, phosphate diabetes). In erythrocytes, copper causes a disorder of individual enzymes (enzymopenic and hemolytic anemia). The deposition of copper salts in the cornea (Kayser-Fleischer corneal ring) is diagnostically important. Penicillamine treatment - a chelating agent with high affinity for copper (renal excretion). Autosomal recessive inheritance. In heterozygotes, the serum content is reduced by about 20%. The disease is one of the most common hereditary anomalies.

Pozn.: externí odkaz: Wilsonova choroba

B-globulin changes

Transferrin deficiency

Genetically determined synthesis disorder leads to impaired serum iron transport, ie insufficient iron supply to hematopoietic tissue. The result is iron deficiency anemia and hemosiderosis. Autosomal dominant inheritance. A transient decrease in transferrin occurs in inflammation, malnutrition, and hepatic proteosynthesis. Increased transferrin is a response to iron deficiency in the body.

Changes in γ-globulins

A-γ-globulinemia

Inherited form

X-linked. Serum is almost completely absent in γ-globulins, levels below 100mg / 100ml (standard is 1600mg / 100ml). Plasma cells are missing in the bone marrow, in the spleen, in the nodes. Apparently a disorder of stem cell maturation in plasma cells (affected by nucleotide metabolism). Manifestation in children after loss of protection by breast milk antibodies. Low resistance to bacterial infections (pneumonia, sepsis). Treatment i.m. injections of γ-globulins.

Acquired form

Rare. It manifests itself in adult men and women. Level of γ-globulins below 500mg / ml. Plasma cells are missing in the nodes. The spleen and liver are enlarged. Manifestations: Often recurrent infections, susceptibility to autoimmune diseases.

Dys-γ-globulinemia

Some types of Ig are missing, others occur in normal or elevated concentrations. IgG or IgA is most often absent and IgM is increased (often the number of plasma cells synthesizing IgM is increased). Manifestations: Susceptibility to infections, thrombocytopenia, haemolytic anemia of the autoimmune type.

Hyperimmunoglobulinemia

Increased levels of γ-globulins due to increased synthesis. Each plasma cell clone produces only one Ig and according to the presence of the given protein types we distinguish:

- **monoclonal hyperimmunoglobulinemia** (ie paraproteinemia)
- **polyclonal hyperimmunoglobulinemia**.

Links

related articles

- paraproteinemia
- plasma protein
- X-linked agammaglobulinemia
- Serum protein electrophoresis
- Wilson's disease