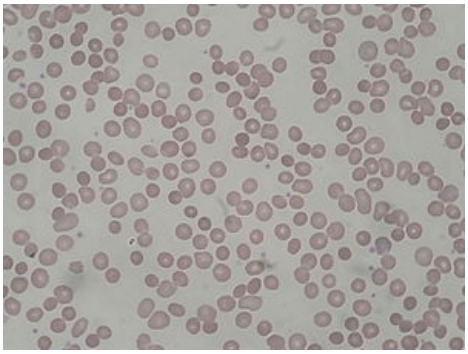
Hereditary spherocytosis

Spherocytosis:

Hereditary spherocytosis is a **genetic disease** (AD, it can also be acquired) conditioned by a **gene defect for structural proteins of the cytoskeleton and membrane of erythrocyte** (spectrin, ankyrin, protein III, protein IV.1). The lipid bilayer is insufficiently anchored to the spectrin layer and parts of it separate when passing through the splenic sinuses. The surface of the erythrocyte decreases relative to its internal volume, which **leads to a change in shape** from biconcave to spherical or ellipsoidal. The deformability of erythrocytes and the resistance to repeated passages through capillaries, especially splenic sinuses (splenectomy has a therapeutic effect) are reduced. It is one of the most common types of hemolytic anemia in Northern Europe.[1]



Hereditary Spherocytosis (smear)

The incidence is reported to be in the range of 1:3000, but is probably higher due to unrecognized mild forms.

Clinical picture

- Anemia (hemolysis in crises, among them only mild anemia);
- splenomegaly;
- increased occurrence of reticulocytes in the blood;
- hemolytic icterus, event. gallstones;
- in the blood smear, small spherocytes (missing central clearing) with slightly changed MCV;
- in liver hemosiderosis and fibrosis;
- bone marrow hyperplasia and osteoporosis.

Links

Related Articles

- Anemia
- Hemolytic anemia corpuscular

Source

- PASTOR, Jan. *Langenbeck's medical web page* [online]. [cit. 12.4.2010]. <http://langenbeck.webs.com>.

=== References ===v

1. POSPÍŠILOVÁ, D. *Anémie u dětí* [online]. The last revision 2007-08-22, [cit. 2011-07-20]. <www.ocol.cz/_data/1188998010_00.ppt>.