# Diseases of the white blood component

náhled|vpravo|Leukocytes. Diseases of the white blood component include **quantitative and qualitative abnormalities of white blood cells** (eg granulocyte dysfunction: chemotaxis disorders, phagocytosis disorders, inability to kill phagocytosed bacteria) and **malignancies** (leukemia a lymphoma).

## Bone marrow failure

- **Acquired diseases with involvement of all three hematopoietic lines**:
  - aplastic anemia – pancytopenia with hypocellular bone marrow without the presence of abnormal cells or increased reticulin;[1]
  - paroxysmal nocturnal hemoglobinuria – an acquired clonal hematopoietic disease in which a clone with a deficiency of cell surface antigens inhibiting the terminal complement complex → chronic hemolysis, venous thrombosis and bone marrow failure occurs;[2]
- **Congenital diseases with involvement of all three hematopoietic lines**:
  - Fanconi anemia – a chromosomal instability syndrome characterized by bone marrow failure and an increased incidence of leukemia and carcinomas;
  - dyskeratosis congenita – onychodystrophy, skin reticular changes and leukoplakia of the oral mucosa, bone marrow failure, 10% risk of malignancies;
  - Shwachman-Diamond Syndrome – AR hereditary disease with impaired pancreatic exocrine function, failure to thrive, short stature, metaphyseal bone dysplasia, neutropenia with the risk of bone marrow failure and a higher incidence of myeloid malignancies;
  - amegakaryocyte thrombocytopenia – isolated platelet aplasia manifested by hemorrhagic diathesis, with a delayed risk of developing aplastic anemia and leukemia;[1]
- **Diseases with isolated granulopoiesis**:
  - severe congenital neutropenia (Kostmann's syndrome) – AR hereditary severe neutropenia (<0.5 × 10 9 / l), high block in maturation of granulopoiesis precursors in the bone marrow at the promyelocyte-myelocyt, myelocyte level, recurrent severe bacterial infections in the first days of life skin, umbilical cord and stomatitis infections ( St. aureus, E. coli, Pseudomonas aeruginosa ), treatment with recombinant granulopoiesis growth factors (G-CSF), event. bone marrow transplantation;
  - cyclic neutropenia – (AD hereditary or sporadic) cyclically recurring disorder of stem cell division in the bone marrow, manifestation at the age of about 10 years, neutropenia with fever and ulcers of the oral mucosa occurs at intervals of about 20 days, ev. abscesses, osteomyelitis and sepsis, treatment of antibiotic infections, good prognosis;[3]

### Functional disorders of granulocytes

- chronic granulomatosis – a rare genetically linked functional disorder of granulocytes - defect in the formation of oxygen radicals of phagocytes → impairment of the ability to kill phagocytosed catalase-positive bacteria ( St. aureus, E. coli, Klebsiella, Proteus, Salmonella ) and fungi → chronically recurrent infections from pneumonia , abscesses in the nodes, skin and liver, inflammation of the bone marrow, gums and mucous membranes of the oral cavity ( St. aureus, Burgholderia cepacia and Aspergillus spp. ), higher incidence of inflammatory diseases from rheumatic diseases, treatment of antibiotic infections, bone marrow transplantation.[3]

## Quantitative changes in white blood count series

| Parameter | Increased | Reduced[4] |
|---|---|---|
| **Neutrophil Count** | ▪ acute bacterial infections,<br>▪ acute a chronic myeloid leukemia, myelopproliferation, generalized malignancies,<br>▪ stress states - pain, cold, heat (so-called distributional leukocytosis with the transfer of leukocytes from the marginal pool to the circulating pool),<br>▪ tissue necrosis (myocardial infarction),<br>▪ vasculitis, decompensation of diabetes with acidosis,<br>drugs (G-CSF and GM-CSF - colony stimulating factors of granulocytes, or granulocytes and macrophages, lithium, corticoids, adrenaline),<br>▪ leukemoid reactions (over 30 000 segmented and younger granulocytes) in sepsis, endocarditis, miliary tuberculosis and tumor metastases | ▪ viral infections,<br>▪ aplastic anemia, agranulocytosis,<br>▪ X-rays, immunosupression, medicine (antibiotics, chemotherapeutics, thyrostatics, analgesics, psychotropic drugs),<br>▪ lymphatic and monocytic leukemias<br>**CAVE!** When evaluating neutropenias, there is a significant decrease in their absolute number. |
| **Number of lymphocytes** | ▪ chronic infections, tuberculosis, infectious mononucleosis, virosis,<br>▪ chronic lymphocytic leukemia, Hodgkin's disease,<br>▪ hypocorticalism,<br>▪ ulcerative colitis, idiopatic thrombocytopenic purpura | ▪ AIDS and related diseases,<br>▪ bone marrow damage after chemo- and radiotherapy, steroid treatment, aplastic anemia,<br>▪ hypercoticalism, neurological diseases (multiple sclerosis) |
| **Monocyte count** | ▪ viral, protozoal and parasitic infections,<br>▪ granulomatous diseases(sarcoidosis, Crohn's disease),<br>▪ tumours (malignant lymphomas, monocyte leukemia) | ▪ aplastic anemia,<br>▪ chronic lymphatic leukemia,<br>▪ glucocorticoid therapy |
| **Number of eosinophils** | ▪ allergic diseases, bronchial asthma , drug allergies,<br>▪ parasitic infections (toxocarosis , trichinosis and intestinal helminthiasis),<br>▪ collagenosis , angioneurotic edema ,<br>▪ Hodgkin's disease a other generalized malignancies,<br>▪ skin diseases ( urticaria , pemphigus) | |
| **Number of basophils** | ▪ chronic myeloid leukemia,<br>▪ hypothyroidism,<br>▪ mastocytoma, event. systemic mastocytosis | |

## Leukemias a lymphomas

- Acute lymphoblastic leukemia – the most common cancer in children under 15 years of age; heterogeneous diseases; uncontrolled proliferation of lymphocyte precursors;
- Acute myeloid leukemia – a heterogeneous disease; uncontrolled proliferation of hematopoietic precursors;
- Leukemia in Down's syndrome 1-2% of children with Down syndrome develop acute leukemia, especially in the first five years of life; acute megakaryocyte leukemia; 10% of children with Down syndrome undergo a so-called transient myeloproliferative reaction after birth, which usually resolves spontaneously by 3 months of age;
- Myeloproliferative diseases: chronic myeloid leukemia – rare in children.
- Myelodysplastic syndrome – a heterogeneous group of acquired clonal stem cell diseases with ineffective hematopoiesis and a different tendency to transform into leukemia.
- Juvenile myelomonocytic leukemia – excessive proliferation of monocytes and granulocytes in early childhood, bears signs of myelodysplastic and myeloproliferative diseases;
- Malignant lymphomas:
  - Hodgkin lymphoma (HL):
    - classic HL - lymphatic tissue tumor formed from mononuclear Hodgkin cells and multinucleated Reed-Sternberg cells;
    - HL with lymphocyte predominance - B-cell tumor characterized by nodular proliferation of isolated large tumor cells;
  - non-Hodgkin's lymphoma - a heterogeneous group of tumors of the lymphatic system; as a result of genetic aberrations affecting lymphocyte proliferation, differentiation and apoptosis; in children they are highly malignant in contrast to adults.[5]

## Diagram hematopoiesis

900px|Schéma hematopoézy.

## References

## Related Articles

- Leukemia: Acute myeloid leukemia • Acute lymphatic leukemia • Chronic myeloid leukemia • Chroni lymphatic leukemia
- Malignant lymphoma: Hodgkin lymphoma • Non-Hodgkin's lymphoma
- Histiocytosis
- Disease of red blood component: Anemia • Polyglobulia
- Physiological and pathophysiological notes on pediatric hematology (pediatrics) • Neutropenia in children • White blood cell pathology (pediatrics)

## Reference


1.
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3.
4.
5.


Kategorie:Pediatrie Kategorie:Hematologie