# Myelodysplastic syndrome

**Myelodysplastic syndrome (MDS)** is a hematopoetic disease caused by a hematopoetic **stem cell mutation** that results in the formation of a pathological clone of cells of one or more blood lines. We can find pancytopenia in the peripheral blood, on the contrary, the bone marrow is normo- or hypercellular with signs of dysplasia. In about 10 % of cases, the bone marrow can also be hypocellular and difficult to distinguish from aplastic anemia. In the late stage, MDS can progress to acute myeloid leukemia. The incidence is 4.5 patients per 100,000 inhabitants. The incidence of patients with MDS increases in people over 60 years of age.

### Etiology

Based on the action of pathological insults (virus, toxin, radiation, exposure to mutagen), a mutation of the hematopoetic stem cell occurs, as well as a pathological immune response, which leads to the apoptosis of more mature cells from the hematopoetic line.

## Clinical picture

- anemia: anemic syndrome (fatigue, paleness, shortness of breath);
- leukocytopenia: infection;
- thrombocytopenia : bleeding manifestations.

### Laboratory finding

In the peripheral blood, cytopenia (of one or more blood lines) is expressed differently, in the later stages, blastic variants of cells are washed out into the peripheral blood. In the bone marrow, the cells are morphologically and dysplastically changed, there are signs of cellular maturation (blasts) and sideroblasts.

### WHO classification

- **Refractory cytopenia with dysplasia of one line RCUD**

1. ***refractory anemia – RA***
2. ***refractory neutropenia – RN***
3. ***refractory thrombocytopenia – RT***

- **Refractory anemia with annular sideroblasts - RARS**
- **Refractory cytopenia with multilineage dysplasia – RCMD**
- **Refractory anemia with excess blasts 1 - RAEB-1**
- **Refractory anemia with excess blasts 2 - RAEB-2**
- **MDS unclassified – MDS-U**
- **MDS with isolated 5q deletion**

## Treatment

- supportive treatment: transfusion of erythrocytes, platelets, administration of vitamin B, chelating agents;
- substances inhibiting apoptosis: growth factors (erythropoietin, G-SCF);
- immunosuppressive and immunomodulating substances: corticosteroids + cyclosporine A , lenalidomide;
- hypomethylating agents – 5-azacytidine, decitabine;
- chemotherapy:
  - monoterapie – hydroxyurea, etoposid;
  - combined – anthracyclines + cytosine;
- allogeneic hematopoietic cell transplantation.

### Prognosis

The prognosis for patients with MDS is variable. Negative prognostic factors are a high number of circulating blasts (patients with more than 20 % blasts are classified as AML), dysplasia of all three lines, complex karyotype changes and abnormalities of the 7th chromosome. The most common causes of morbidity and mortality in patients with MDS are: progression to AML, severe neutropenia or thrombocytopenia, iron overload, and cardiac disease. Average time of survival is 2 years.

## Summary video

<mediaplayer width="500" height="300">https://www.youtube.com/watch?v=fT_NTuZSbkQ</mediaplayer>

## Sources

## Related Articles

- Bone marrow
- Hematopoesis

## External links

- PGS informace (https://web.archive.org/web/20160331222721/http://zdravi.e15.cz/clanek/postgradualni-medicina/myelodysplasticky-syndrom-458605)
- Myelodysplastický syndrom- pohled internisty (http://www.solen.cz/pdfs/int/2005/02/02.pdf)

## Literature


- ČEŠKA, Richard – TESAŘ, Vladimír. *Interna.* 132. edition. Triton, 2012. ISBN 978-80-7387-629-6.

- NEČAS, Emanuel – ŠULC, Karel – VOKURKA, Martin. *Patologická fyziologie orgánových systémů. Část I.* 1. edition. Karolinum, 2006. 0 pp. ISBN 978-80-246-0615-6.