PARAPROTEINEMIC HEMOBLASTOSIS

Paraproteinemic hemoblastosis is a group of diseases characterized by monoclonal proliferation of the cells of B-lymphoid line, secreting immunoglobulins, to which, first of all, belong multiple myeloma and Waldenstroem’s macroglobulinemia.

Multiple myeloma is characterized by hyperparaproteinemia; hyperviscose syndrome (with a level of monoclonal protein Ig G or Ig A over 50 g/l), bleeding, retinopathy, neurological symptoms (paresthesia, sleepiness, headache, dizziness), paraproteinemic coma, osteoporosis and osteolysis, hypercalciemia, peripheral neuropathy, renal functions disorders - myelomic nephropathy (accumulation of myelomic protein and amyloid in the renal tubules, Bence-Jones proteinuria, hypercalciemia).

Hyperviscous syndrome is pronounced in Waldenstroem’s macroglobulinemia, due to the high content of immunoglobulin M, as well as to cryoglobulinemia.
### PLASMAPHERESIS IN PATIENTS WITH APLASTIC ANEMIA

*(IN CASE OF SENSIBILIZATION AND REFRAC TORINESS TO TRANSFUSION)*

Aplastic anemia (ApAn) is a disease characterized by deep pancytopenia and insufficiency of medullar hematopoiesis conditioned by development of aplasia of medulla.

The given nosology calls for frequent and multiple hemotransfusions that bring about sensibilization. In its turn, allosensibilization causes refractoriness to the transfused components of blood.

#### INDICATIONS

- Inefficacy of transfusions of thromboconcentrate in hemorrhagic syndrome

#### CONTRAINDICATIONS

- Reduction of the level of antileucocytic and antithrombocytary antibodies

#### CRITERIA OF EFFICIENCY

- Increase of thrombocytes and erythrocytes after transfusion of components of blood after 1, 6, 15, 24 hours
- Reduction or absence of hemorrhagic syndrome

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**PLASMAPHERESIS IN THE TREATMENT OF PORPHYRIA**

Plasmapheresis (PA) is used for integral treatment of porphyria in the period of exacerbation, basically when fighting such its forms as acute intermittent porphyria and porphyria cutanea.

<table>
<thead>
<tr>
<th>INDICATIONS</th>
<th>CONTRAINDICATIONS</th>
<th>CRITERIA OF EFFICIENCY</th>
</tr>
</thead>
<tbody>
<tr>
<td>➢ Peripheral polyneuropathy</td>
<td>➢ Increased level of porphyrins</td>
<td>✓ Improvement of clinical indicators: disappearance of peripheral polyneuropathy, central nervous system functions disorders, raised to actinism</td>
</tr>
<tr>
<td></td>
<td></td>
<td>✓ Normalization of laboratory indicators of porphyrin exchange</td>
</tr>
</tbody>
</table>
**PLASMAPHERESIS IN HEMATOLOGY**

### PLASMAPHERESIS IN ANTIPHOSPHOLIPID SYNDROME

Plasmapheresis (PA) is used for integral treatment of patients with antiphospholipid syndrome (APhS) in the period of exacerbation.

<table>
<thead>
<tr>
<th>INDICATIONS</th>
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<th>CRITERIA OF EFFICIENCY</th>
</tr>
</thead>
<tbody>
<tr>
<td>➢ Thrombophilia</td>
<td></td>
<td>✔ Improvement of clinical indicators of the main disease</td>
</tr>
<tr>
<td>➢ Lupus anticoagulant (LA), antiphospholipid antibodies (APhA)</td>
<td></td>
<td>✔ Normalization of laboratory indicators of phospholipid-dependant tests [VA, AFA, CIK, immunoglobulins (basically G and M)]</td>
</tr>
<tr>
<td></td>
<td></td>
<td>✔ Normalization of the changed indicators of the coagulogram</td>
</tr>
</tbody>
</table>

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**PLASMAPHERESIS IN HEMATOLOGICAL THROMBOPHILIAS**

Hematogenous thrombophilias manifest themselves by the hypercoagulation syndrome in case of availability of determined defects of cellular and plasmatic elements of blood that bring about thrombosis.

This occurs under deficit of AT III, anomaly of the system of protein C and protein S, with mutant V Leiden factor, mutations of the gene of methylenetetrahydrofolate-reductase (MTHFR) and hyperhomocysteinemia, mutant prothrombin G202210A, mutations of integrins, presence of lupus anticoagulant - autoantibodies to phospholipids, syndrome of sharply raised aggregation of thrombocytes - the "sticky thrombocytes" syndrome, substantial increase of the level or multidimensionality of the Willebrand factor, activity of the factor VIII, high level of the factor XIII of the coagulation system of blood.

**INDICATIONS**

- Thrombosis
- Modification of the factors of coagulogram: deficit of AT III, anomalies of the system of protein C and protein S, mutant Leiden factor, mutation of the gene of methylenetetrahydrofolate-reductase (MTHFR), substantial increase of the level or multidimensionality of the Willebrand factor and activity of VIII factor, high level of the XIII factor of the coagulation system of blood
- Hyperhomocysteinemia, mutation of integrins
- Syndrome of sharply raised aggregation of thrombocytes - a syndrome of "sticky thrombocytes"

**CONTRAINDICATIONS**

- Improvement of clinical indicators of the main disease
- Normalization of the modified indicators of the coagulogram

**CRITERIA OF EFFICIENCY**

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### PLASMAPHERESIS

**IN INCOMPATIBILITY OF THE RECIPIENT WITH THE DONOR AS TO ANTIGENS OF THE AVO SYSTEM BEFORE ALLOGRAFTING OF BONE MARROW**

The presence of incompatibility between donor and recipient as to antigens of the AVO system does not constitute a contraindication for performing allogenic transplantation of the marrow.

According to international classification, there are 2 types of incompatibility as to antigens of the AVO system. By big incompatibility is meant the presence in the recipient’s blood of natural antibodies to erythrocortary antigens of the donor, arising in such situation the risk of hemolytic complications. For avoiding such sort of complications, it is necessary to remove a maximal quantity of erythrocytes of the donor from the transplantat and to perform exchange plasmapheresis in the recipient in order to remove antibodies. As small incompatibility as to antigens of the AVO system is identified the presence in the donor’s blood of isohemagglutinins to erythrocortary antigens of the recipient, resulting in such cases enough to remove the plasma from the transplantat.

<table>
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</thead>
<tbody>
<tr>
<td>Presence of incompatibility between donor and recipient as to antigens of the AVO system</td>
<td>✔ Reduction of the level of isohemagglutinins in the blood of the recipient below dilution in relation of 1: 16 (being more favorable its reduction to 1: 4 - 1: 2)</td>
<td></td>
</tr>
</tbody>
</table>

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PLASMAPHERESIS IN HEMATOLOGY

PLASMAPHERESIS AND LEUCOCYTAPHERESIS IN THE TREATMENT OF HYPERLEUCOCYTOSIS

Hyperleucocytosis is an urgent condition in hematological patients, associated with severe vitally-dangerous complications and so requiring emergency, special medical action.

By hyperleucocytosis is meant an increase of the level of leucocytes in peripheral blood over 30 x 10^9/l. An increase of the amount of leucocytes above 100 x 10^9/l (in cases of acute leucosis) is considered extraordinary.

Under hyperleucocytosis, in the vessels of the cerebrum, kidneys and heart are formed clumps of leucoaggregates and thrombs from leucosis cells. Clinically this reveals itself in the form of encephalopathy, respiratory distress syndrome, intracranial and pulmonary hemorrhages, hyperuricemic syndrome, which bring about renal failure. As a consequence of a high level of proliferation of tumoral cells, metabolism and cellular breakdown, severe metabolic disorders may develop, including hyperkaliemia, hypocalciemia and hyperphosphatemia.

INDICATIONS

Indications for performing leucocytapheresis in patients with acute myeloblastic leucosis:
- When the quantity of leucocytes in the patient’s blood is above 100 x 10^9/l, even without signs of leucostasis
- On the background of the therapy with hydroxyurea, when the content of leucocytes in the blood is over 100 x 10^9/l
- When presenting clinical signs of leucostasis
- In case of a very quick progression of the disease: duplication of the number of leucocytes from 50 x 10^9/l for a day
- Inefficiency of the treatment with hydroxyurea during 1 day, that is, drop of the level of leucocytes by less than 30% from the initial level
- Impossibility of taking hydroxyurea

Indications for performing LCA in patients with chronic myeloleucosis:
- Pronounced intoxication on the background of hyperleucocytosis of over 50-100 x 10^9/l and hyperthrombocytosis of above 350 x 10^9/l, which bring about stasis in cerebral vessels
- Developed resistance to polychemotherapy (with progressing stage and blastic crisis), when the procedure is applied in order to remove a big tumoral mass, thus allowing to intensify the proliferative activity of the bone marrow and increase the efficiency of further courses of PChT

Indications for performing LCA in patients with ChLL:
- Hyperleucocytosis of over 150-200 x 10^9/l, matching with anemia, thrombocytopenia, splenomegaly and inefficacy of the different schedules of cytostatic treatment
- LCA is performed in blood separators under the LCA program, with a course of up to 2-3 procedures and an interval of 7-10 days. Administration of heparin and protein blood substitutes is to be realized according to indications

CRITERIA OF EFFICIENCY

- Reduction of the level of leucocytes
- Reduction of the signs of intoxication (weakness, sweatiness and alike)
- Reduction of the size of the spleen
- Improvement of the indicators of hemoglobin and erythrocytes with a preceding increase of the number of reticulocytes
- Increase of the number of thrombocytes

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