TTP treatment

1. **start as instantly as possible**
2. **Fresh Frozen Plasma (FFP) (30ml/kg body weight)**
3. **plasma exchange (3-4 l) until 3 days after LDH and thrombocyte count normalization, with the purpose of deleting the inhibitor (antibody) and the ultralarge vWF multimers, and adding ADAMTS13 protease.**
4. **Rituximab (possibly Vincristin)**
5. ***corticosteroids (1-2 mg/kg body weight)***

CAUTION

TTP is a contraindication for the administration of platelet concentrates!!

Information Portal and support group

The Internet is an information portal with further information for doctors and sufferers under:

www.ttp-forum.de

On this website you will also find various contact options for sufferers.

In addition, there is a TTP support group within the German Haemophilia Society (DHG). Contact in both cases is Prof. Dr. I. Scharrer.

Contact

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Thrombotic-thrombocytopenic Purpura

MORBUS MOSCHCOWITZ

Information portal for doctors and sufferers

www.TTP-Forum.de

What is TTP

Thrombotic-thrombocytopenic Purpura (TTP/Morbus Moschkowitz) is counted among the rare hematological diseases.

In TTP patients, ADAMTS-13 protease activity is reduced. In patients with acquired TTP, this reduction is caused by antibodies.

Due to an increased synthesis of ultralarge von Willebrand multimers (vWF multimers) triggered by various infections and medicines, the lacking ADAMTS13 protease activity causes an insufficient or even completely lacking cleavability in these multimers.

The great number of ultralarge vWF multimers resulting from this process, on the other hand, can cause a thrombotic microangiopathy, a thrombocytopenia and a haemolytic anaemia (TTP relapse).

Symptoms of TTP manifestation

- headache (severe)
- aphasia (that can even comprise phrase forming difficulties)
- shifting states of consciousness
- temporary loss of responsiveness (lethargy, collapse, coma)
- pallor
- petechiae
- hematomas
- conspicuous weakness, pronounced fatigability
- dyspnoea
- seizures
- paralysis

The early indications of a TTP relapse are different in every patient, and the complete range will not necessarily develop. **In any case, the patient should be seen in a university hospital upon manifestation of one or more of these symptoms in order to be examined for the occurrence of a relapse.** A rapid treatment start can improve the healing rate and reduce damage incidence!

Diagnostics and potential triggers

Diagnostics

1. thrombocytopenia
2. increased LDH
3. helmet cells
4. reduced ADAMTS13 protease activity
5. ultralarge (vWF) multimers detectable

Potential triggers:

1. bacterial or viral infection
2. medication
3. pregnancy
4. bone marrow transplantation
5. chemotherapy
6. tumors