How is this going to affect
MY PERSONAL & PROFESSIONAL LIFE?

Except the fact that you will have frequent blood tests to check the stability of your situation, this disease may not impact your everyday life. However, this can be different for each patient.

CONTACT *When should I* MY DOCTOR?

Do not hesitate to contact your doctor again if you have any bleeding (from nose, gums), an unusual fatigue, persisting headaches, or if you have marks on your skin (bruising, purpura, petechiae). Please also contact your doctor if you want to get pregnant.

Do you have SOME QUESTIONS?

More information on www.itn-profile.eu or contact Prof. Dr. Karen Vanhoorelbeke (karen.vanhoorelbeke@kuleuven.be)

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YOU HAVE BEEN URGENTLY HOSPITALIZED. THE DOCTOR IDENTIFIED YOUR DISEASE AS TTP.

TTP: *What is*

THROMBOTIC THROMBOCYTOPENIC PURPURA

WHY A STAY *in Intensive Care?*

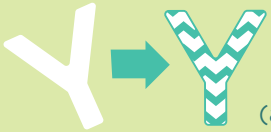
TTP is a disease characterized by a sudden clumping of platelets in the body, leading to a decrease of platelets in the circulation, anemia and sometimes organ failure. It is a severe disease that needs a specialized management typically in an intensive care unit to monitor the involved organs.
Sometimes, flares of the disease occur during treatment. This warrants further detailed monitoring. It is therefore crucial to stay in the hospital until the disease stabilizes.

In most patients, the formation of autoantibodies (see below) is the mechanism of the disease. This is called immune-mediated TTP (iTTP). In a minor part of the patients, the disease is caused by a mutation in the ADAMTS13 gene (congenital TTP).

What's happening IN MY BODY?

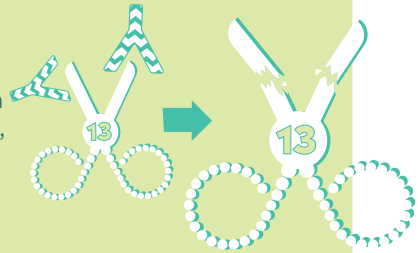
Some vocabulary you may need

TTP is a rare disorder of the blood clotting system.

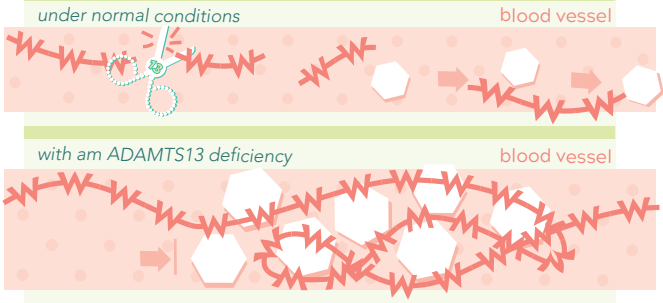


One of your antibodies has an unusual (autoimmune) behaviour.

Instead of fighting against diseases, it attacks your own ADAMTS 13 protein, and makes it inactive.



Because of the reduced ADAMTS13 function, the size of von Willebrand factor is too large.



So, due to the presence of this very large vWF, platelets spontaneously form clumps all over your body, what is problematic for two reasons:

- these clumps obstruct blood circulation and thus oxygenation of many organs.
- platelets can no longer perform their initial role of stopping bleeding.

some VOCA-BULARY

Adamts13 a large protein involved in regulation of platelet clumping, that acts as a scissor and cleaves the very large vWF molecules

von Willebrand factor (vWF) a large blood factor, acting as a glue to clump platelets together

Antibody a protein produced by B-cells in the blood that fights diseases by attacking and killing harmful bacteria and viruses.

Platelets very small cells in the blood that are crucial in stopping bleeding upon injury

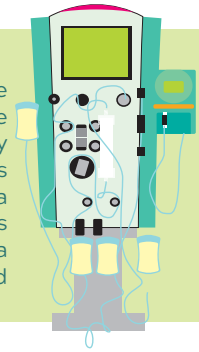
B-Cells or B-lymphocytes are a type of white blood cells. They defend us against viruses and bacteria that enter the blood and lymph by secreting antibodies.

WHO IS MORE SUSCEPTIBLE TO DEVELOP TTP?

The incidence of TTP is about 2-3 cases per million people per year. TTP occurs more often in women. People of African descent are more likely to develop TTP for unclear reasons. TTP can be associated with other types of autoimmune disease, such as systemic lupus erythematosus. TTP is also more frequent in pregnant women. Consequently, patients with a history of TTP will require a special follow-up in case of pregnancy.

TREATMENT

In all cases of iTTP, plasma exchange is the basic treatment of choice. Plasma exchange involves the use of automated machinery which permits the removal of the patient's plasma and replacement with donor plasma during a 3 to 4 hour treatment. This makes treatment quite long and heavy. Plasma exchange both removes antibodies and replenishes normal plasma proteins.



What is B-CELL DEPLETING THERAPY?

B lymphocytes are the cells that produce harmful antibodies against ADAMTS13. So, by depleting B lymphocytes, you eliminate these antibodies, which results in the restoration of a normal ADAMTS13 activity after 2 to 3 weeks. B cell depletion is performed in many but not all cases.

WHY PREVENTIVE TREATMENT

even if I'm not feeling sick?

It is really important to have regular blood tests to check ADAMTS13 activity levels, because the decrease in its activity is often unnoticed, and does not have immediate effects. With the B-cell therapy, we can act before symptoms occur.