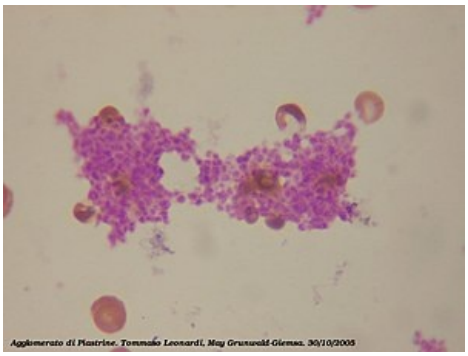
Trombocytopathy

Plateletopathy is a condition in which the function of platelets fails due to an internal disorder. It may be the cause of the primary disorder of hemostasis



Clumping platelets

Congenital plateletopathies

Two glycoprotein complexes are important for platelets: gpIb-IX and gpIIb-IIIa, with which they bind to subendothelial structures (collagenu I and II) and fibrinogenu, respectively. These glycoprotein complexes have a receptor function and are thus involved in the process of platelet adhesion and aggregation. Their congenital defect leads to congenital thrombocytopathies.

For example, congenital plateletopathies include:

- **Bernard-Soulier syndrome** (deficiency of the platelet glycoprotein complex gpIb-IX); * ***Glanzmann thrombocytopathy** (deficiency of the platelet glycoprotein complex gpIIb-IIIa). Both syndromes are autosomal recessive disorders and cause a clinically significant disorder of primary hemostasis. * ***Von Willebrand disease** affects platelet function secondarily. Thus, it is not a primary deficit of platelets but a decrease in Von Willebrand factor, which is necessary for their adhesion. Independent of Von Willebrand factor, clotting factor VIII is also unstable and its level decreases. Thus, both primary hemostasis and secondary disorders occur. Von Willebrand disease has various forms, most of which are diseases with an autosomal dominant mode of inheritance. Clinically, it manifests as epistaxis (nosebleeds), bleeding gums, bleeding in the GIT and genitourinary tract, more severe bleeding after surgery or trauma. Laboratory findings show decreased vWF level in plasma and decreased factor VIII level. * The storage and degradation function of platelets may also be congenital - this corresponds to, for example: ** **Heřmanský-Pudlák syndrome**; ** syndrome **Chédiak-Higashi**.

Acquired plateletopathies

Acquired plateletopathies are often associated with drug use. For example, inhibiting acetylsalicylic acid (**"aspirin"**) irreversibly inhibits cyclooxygenase, which is responsible for the synthesis of thromboxane (an important factor of aggregation and degranulation), thus reducing the ability to heal minor bleeding mechanism of primary hemostasis. It then takes up to seven days after a single aspirin dose for a significant proportion of affected platelets (lacking thromboxane) to be replaced by new ones with functional cyclooxygenase. Another example is **penicillin**, which at high doses causes disruption of primary hemostasis by coating platelets and disrupting substances released from their granules. Acquired plateletopathies may also occur in association with diseases of certain organs, such as renal dysfunction. == Odkazy == === Související články === * Von Willebrandova choroba * Chédiakův-Higashiho syndrome * Trombocyty * Hemostáza === Použitá literatura === * Template:Navbox - onemocnění krve