

# Disorders of hemostasis

- **Haemostasis** – stoppage of bleeding, takes place at three levels:
  - primary hemostasis – vessel vasoconstriction, platelet aggregation and platelet degranulation,
  - secondary hemostasis – hemocoagulation,
  - fibrinolysis – dissolution of the thrombus.
- **Disorders of hemostasis are the result of an imbalance between pro- and anti-coagulation factors**, they can be twofold:
  - hemorrhagic diatheses - bleeding,
  - thromboembolic disease – thrombophilic conditions (Thrombosis and Embolism).

## Hemorrhagic diatheses

- **Disorders of primary hemostasis**,
  - Vasculopathy (vasculitis, congenital connective tissue disorders, vitamin C deficiency)
  - Thrombocytopenia (autoimmune – ITP, TTP, with hypersplenism)
  - Thrombocytopathy.
- **Disorders of secondary hemostasis**
  - congenital (lack of one factor) – hemofilie A (VIII), hemofilie B (IX),
    - afibrinogenemia,
  - acquired (mostly several coagulation factors are missing) – Vitamin K, Liver insufficiency.

 For more information see *Bleeding conditions (pediatrics)*.

## Thrombophilic conditions

- **Congenital** – defective hemocoagulation or fibrinolytic factors
  - **Leiden Mutation** (factor V mutation),
  - factor II mutation,
  - deficiency of antithrombin III, proteins C and S, Hyperhomocysteinemia.
- **Acquired** – ( **Virchow's trias** ), these are actually risk factors for phlebothrombosis:
  - stasis (postoperative period, postpartum period, pregnancy), vascular wall damage (injuries, burns, varicosities, Sepsis, post-thrombotic syndromes),
  - blood abnormalities (pregnancy, oral contraceptives, malignancies, Nephrotic syndrome, trauma, burns, infections),
  - the main risk factors are age (over 40) and gender (women – estrogens, contraceptives), as well as smoking and Obesity.

## Hemocoagulation examination

- **INR** (formerly Quick, PT) - **external system**, to control warfarinization, norm 0.8-1.2.
- **APTT** – **internal system**, for checking heparinization, standard 30–45 s.
- **TT** thrombin time) – the last phase of coagulation (thrombin catalyzing the conversion of fibrinogen to fibrin), the norm is 10–20 s.
- **Euglobulin fibrinolysis** – dissolution of fibrin clot in euglobulin plasma, standard 120–240 min.
- **FDP, D-dimers**.
- **Fibrinogen** – norm 2–4 g/l.

## Overview of antithrombotic treatment

- **Anticoagulants** – act against the formation of a thrombus,
  - direct (they inactivate the coagulation factors present) heparin (UFH), LMWH
  - indirect (acts against the synthesis of coagulation factors)- warfarin, ethyl biscum acetate (Pelentan),
- **Fibrinolytics** – they dissolve an already formed thrombus - both venous and arterial
  - streptokinase, urokinase,
  - rt-PA (alteplase)... "Actilyse"
- **Antiaggregant** - acts against the aggregation of platelets (prevention of the formation of a platelet thrombus - mainly as a prevention of arterial thrombosis, not as a prevention of deep vein thrombosis)
  - ASA, dipyridamole, ticlopidine, clopidogrel, abciximab, pentoxifylline.

## Antidote

- **Protamine** - the antidote of heparin.
- **Vitamin K** - antidote to coumarin derivatives (warfarin).
- **Aprotinin, aminocaproic acid** – an antidote to fibrinolytics.

# Links

## related articles

- Bleeding conditions (pediatrics) • Hemorrhagic diatheses (pathology) • Hereditary coagulopathy • Acquired coagulopathy • Thrombocytopathy
- Hemostasis • Hemocoagulation • Examination of blood coagulation • Examination of bleeding

## Source

- BENEŠ, Jiří. *Studijní materiály* [online]. [cit. 29.6.2010]. <<http://jirben.wz.cz>>.