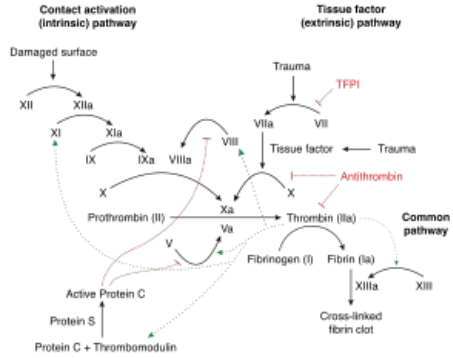
Coagulation disorders

Physiology of hemocoagulation

Hemostasis = a condition in which blood flow is ensured on the one hand, while on the other hand in case of bleeding it stops (x hemocoagulation = function of plasma coagulation factors)

- 1) clot formation - clot
- 2) activation - pledge (limited to the required section)
- 3) breakdown of the clot (so as not to interfere with circulation)

- parts:
    - primary hemostasis
    - plasma coagulation system
    - clotting inhibitors
    - fibrinolytic system
    - fibrinolysis inhibitors



Coagulation full

Hemostasis disorders - coagulopathy

It is a bleeding condition caused by a decreased concentration or activity of plasma coagulation factors.

Congenital coagulopathy

Hemophilias

- congenital defect f.VIII (hemophilia A)
- congenital defect f.IX (hemophilia B)
- recessively inherited X-linked disease, transmissible women
- prenatal diagnostics already 10. − 12. week - a collection of chorionic villi, DNA examination in high-risk families, later blood collection from the fetal umbilical cord
- clinical picture:
- f.VIII ≤ 1%, f.IX ≤ 2% (spontaneous bleeding into joints, muscles)
    - f.VIII 5−10%, f.IX 2−6% (bleeding after injuries or operations)
    - f.VIII ≥ 10%, f.IX ≥ 6% (light form)

- consequences: typical hemophilic arthropathy to joint amylosis, disability, extensive muscle hematomas, psoatic syndrome
- lab. findings and treatment: aPTT, determination of f.VIII and f.IX; substitution and supportive treatment, prevention of consequences; the therapeutic regime in the hospital - preparation for surgery (treatment of bleeding into the GIT, CNS)
    - hemophilia C: defect f.X
    - parahemophilia: defect f.V

Von Willebrand's disease

- autosomal inherited
- vWF deficiency or malfunction (for platelet adhesion)
- vWF is the carrier for f.VIII
- vWF deficiency can lead to f.VIII deficiency
- bleeding into the skin, mucous membranes, epistaxis, hypermenorrhea
- prolonged bleeding after childbirth, injury, surgical intervention
- at the same time with f.VIII deficiency - bleeding as in hemophilia
- treatment: substitution, in mild cases vasopressin

Acquired coagulopathies

Disorder of resorption and utilization of vit. K

- insufficient synthesis of factors
    - liver cell involvement
    - vit.K transport disorder in obstructive jaundice
    - intestinal resorption disorder

- overdose of dicoumarin treatment (kamavitiv, frozen plasma, prothrombin complex factor concentrate)

Disseminated intravascular coagulation (DIC)

- a necessary evoking moment

- DIC development
  - a) the coagulation system is activated (if it is not meant to be) → microthrombus formation → storage in organs
  - b) consumption of coagulation material → formation of hypocoagulation state
  - c) defibrination
  - d) fatal bleeding
- cause: endotoxin leaching, tissue thromboplastin release, proteolytic enzyme leaching, antigen-antibody reaction
- accompanies: obstetric complications, injuries, infections, sepsis, anaphylactic shock
- course: activation phase (microthrombolization), compensated and decompensated phase, excessive activation phase of secondary fibrinolysis
- treatment: substitution of all components (MP, thrombo, fibrinogen, EM), eliminate the inducing moment

Links

Related articles

- Blood
- Vitamin K
- Hemostasis disorders
- Hemocoagulation

Literature

PECKA, Miroslav. *Laboratorní hematologie v přehledu. [Díl 2.], Fyziologie a patofyziologie krevní buňky.* 1. vydání. Český Těšín : Finidr, 2006. ISBN 80-86682-00-5.