Von Willebrand disease

Von Willebrand's disease is one of the most common congenital blood clotting disorders. It affects 1 in 1,000 people (women as often as men). Mostly, however, these are mild diseases without more serious clinical manifestations. It was first described by the Finnish physician Erik von Willebrand in 1924, but it was not until the 1950s that deficiency or malfunction of von Willebrand factor (vWF) was shown to be the cause.

Causes

- **autosomal dominant inheritance mutation of the gene encoding the amount, functionality and structure of vWF →** impaired platelet function, inability to bind plasma factor VIII
- the obtained forms were also described

Symptoms

- vWF is not among the plasma factors → it is not a coagulopathy
- impaired blood clotting – frequent nosebleeds (epistaxis), increased formation of bruises, heavier menstrual bleeding, there may be blood in the urine and stools, in severe forms bleeding into the joints

Basic examination

- blood count
- APTT
- Duke bleeding test
- examination of factor VIII
- tests performed may not be pathological in milder forms

Duke:

Classification

Type 1
: light quantitative defect – the most common

Type 2
: qualitative defect

Type 3
: severe qualitative defect – the most severe form

Links

Related Articles

- Hemostasis disorders: Hereditary coagulopathies • Acquired coagulopathies • Bleeding conditions (pediatrics) • Hemorrhagic diatheses (pathology)
- Hemostasis • Hemocoagulation • Blood coagulation test • Bleeding test
- Von Willebrand factor

Resources

- Wikipedia article on von Willebrand disease