

# **von Willebrand Disease**

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# Epidemiology

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- VWD is the most common inherited bleeding disorder.
- Low VWF levels affect up to 1% of the population.
- 0.1 to 1% of these individuals are symptomatic.(1:10,000-1:100,000)

# Epidemiology

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- In 1926, Erik von willebrand described the first patient with the disease.
- Males and females are equally affected.

# VWD

## ❖ VWF produced in:

➤ Endothelium

➤ Megakaryocytes

- In circulation VWF travels as a complex with F8
- Half life is about 12 hours
- Short arm of chromosome 12

# VWD

## ➤ Quantitative

◆ Partial deficiency: type 1

◆ Total deficiency: type 3

## ➤ Qualitative

◆ Type 2



# VWD classification

- ❖ **Type 1:** AD, 75-85% of all cases
- ❖ **Type 2:** AD
  - **2A:** AD, some AR, 10-15% of all cases
  - **2B:** AD, 5% of all cases
  - **2M:** AD, some AR
  - **2N:** AR
- ❖ **Type 3:** AR
- ❖ **Pseudo-type VWD:** AD
- ❖ **Acquired VWD**

# Clinical Features

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- The disease is often mild and no bleeding symptoms.
- Patients with VWD can become symptomatic at any age.

# Clinical Features

- ❖ **Type 1:** variable, from asymptomatic to serious bleeding (the majority are mild), mucocutaneous
- ❖ **Type 2A:** usually moderate to severe, mucocutaneous
- ❖ **Type 2B:** usually moderate to severe, mucocutaneous
- ❖ **Type 2M:** usually moderate to severe, mucocutaneous
- ❖ **Type 2N:** moderate to severe, joint, soft tissue, GI, or surgical
- ❖ **Type 3:** Severe, mucocutaneous and joint, soft tissue, GI, or surgical; often presents during infancy.



# Clinical Features

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- ❖ Severe bleeding is more common with types 2 and 3.
- ❖ Easy bruising
- ❖ mucocutaneous bleeding
- ❖ Prolonged bleeding from mucosal surfaces
- ❖ Heavy menstrual bleeding (menorrhagia)
- ❖ Postpartum bleeding
- ❖ GI bleeding
- ❖ Epistaxis
- ❖ Hemarthrosis: 2N, type 3
- ❖ Muscle hematoma: 2N, type 3

# Laboratory testing

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- ◆ **CBC:** normal, anemia(IDA), thrombocytopenia (2B)
- ◆ **PT:** normal
- ◆ **PTT:** normal or prolonged
- ◆ **BT:** normal or prolonged
- ◆ **BG,RH**
- ◆ **VWF:Ag**
- ◆ **VWF:Act**
- ◆ **FVIII activity**

# Laboratory testing

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◆ **VWF multimer analysis**

◆ **RIPA**

◆ **VWF:FVIII B**

◆ **VWF:PP**

◆ **Response to DDAVP**

◆ **Genetic testing**

# Differential diagnosis

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1. Mild hemophilia A
2. BSS
3. Platelet type (pseudo) VWD
4. Acquired VWD



*Thank You..*





# TREATMENT

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- **VWF concentrate**
  - **Plasma derived VWF concentrate**
  - **Recombinant VWF**
- **FFP**
- **Cryoprecipitate**
- **FVIII**
- **DDAVP**
- **Platelet Tx**
- **Antifibrinolytic agents**



