Von Willebrand Disease

Normally vW Factor (vWF) binds to exposed collagen in areas of endothelial damage then allowing platelets to bind via their GpIb receptor. Without vWF, platelets are unable to bind and function in their normal capacity. Von Willebrand Disease is a unique platelet disorder because there is a concurrent decrease in Factor VIII since vWF plays a role in carrying and protecting this factor.

PLAY PICMONIC

Mixed platelet and coagulation disorder

Mixed Plates and Clogs

Von Willebrand Disease results in a functional platelet disorder and a deficiency of Factor VIII of the coagulation cascade.

Mechanism

Autosomal Dominant Condition Leading to Reduced vWF

Auto Domino with Down-arrow vW

vWD is the most common inherited bleeding disorder and is inherited in an autosomal dominant fashion.

Defect in platelet plug formation

Broken Plate Plug

Without vWF to facilitate the interaction between platelets and exposed collagen on damaged endothelium, the platelet plug does not form.

Labs

Increased Bleeding Time

Up-arrow Blood Clock

In the Bleeding Time test, rarely performed today (but still tested on!), a small incision is made on the underside of the patient's forearm and the time until bleeding ceases is recorded. Because there is a defect in platelet plug formation with vWD, the bleeding time is classically increased.

PTT increased

Up-arrow PTT clotting-hourglass

Von Willebrand Disease has three main subtypes and depending on the subtype the PTT may be Increased or Normal. Since vWF acts to protect Factor VIII from proteolysis, vWF deficiency results in decreased levels of Factor VIII. Factor VIII plays a role in the intrinsic coagulation pathway which is monitored using PTT. Without Factor VIII to act in this pathway, PTT is increased.

Factor VIII decreased

Down-arrow (8) Ball

vWF acts to protect Factor VIII from proteolysis. Thus if vWF is decreased Factor VIII will not be protected and will be decreased.

Normal platelet count

Normal Plates

Although the platelets do not function properly since they cannot form the platelet plug, the platelet count is normal. This lab finding helps distinguish vWD from other diseases such as Idiopathic Thrombocytopenic Purpura (ITP) where the platelet count is decreased but the bleeding time is increased like in vWD.

Treatment

DDAVP (synthetic vasopressin)

Designated Driver Vase-present

This is a synthetic analog of vasopressin (antidiuretic horome) that causes vWF to be released from endothelium in an indirect manner. It is categorized a non-transfusional compound and primarily used in mild to moderate variants of vWF deficiency.