Insights into type 2B von Willebrand disease

November 25 2013

In response to blood vessel damage, von Willebrand factor (vWF) binds to the exposed extra cellular matrix, recruits platelets to the site of injury, and activates platelets, which promotes thrombis formation. Patients with von Willebrand disease type 2B (vWD-type 2B) produce a vWF protein that has a high binding affinity for platelets; however, these patients exhibit a bleeding tendency that is thought to be due to loss of vWF multimers.

In this issue of the *Journal of Clinical Investigation*, Marijke Bryckaert and colleagues at the Hôpital Kremlin Bicêtre determined that the bleeding phenotype associated with vWD-type 2B might be due to platelet dysfunction.

Evaluation of platelets treated with vWD-type 2B-associated vWF revealed the mutatnt vWF was able to bind platelets, but was unable to activate them, thereby inhibiting thrombus formation. In an accompanying commentary, Jerry Ware of the University of Arkansas discusses the implications of this study for treatment of vWD-type 2B.

More information: von Willebrand factor mutation promotes thrombocytopathy by inhibiting integrin αIIbβ3, J Clin Invest. DOI: 10.1172/JCI69458

Provided by Journal of Clinical Investigation