

PLATELET FUNCTION DEFECT

Introduction

Blood is carried throughout the body within a network of blood vessels. When tissues are injured, damage to a blood vessel may result in leakage of blood through holes in the blood vessel wall. Normally, bleeding stops through two processes that work together to form a blood clot: 1) the formation of a platelet plug, and then 2) stabilization of that platelet plug by the formation of a fibrin clot.

What are Platelets

Platelets are small disc shaped cells that circulate in the blood. Platelets play an important role in the clotting of blood and the beginning repair of injured blood vessels. Platelets stick to, and spread on, areas of damaged blood vessel walls (platelet adhesion). These spreading platelets release substances that activate other nearby platelets which clump at the site of injury to form a platelet plug (platelet aggregation). The surface of these activated platelets then provides a site for blood clotting to occur. Clotting proteins that circulate in the blood are activated on the surface of the platelets to form a mesh-like fibrin clot.

Description of the Disorder

There are several different types of platelet function defects, or PFD, as well as varying degrees of severity. The *number* of platelets is usually normal; however, the *function* of the platelets is abnormal.

Most PFDs diagnosed are of the mild hereditary type. These PFDs do not usually “go away”. A patient diagnosed with the hereditary type of PFD will most likely have it for the rest of their life. There are also acquired PFDs. These include PFDs caused by certain medical conditions or from the use of medications, which inhibit platelet function such as aspirin, non-steroidal anti-inflammatory drugs (such as ibuprofen and naproxen), blood thinners, and some antibiotics, antidepressants, anesthetics, and heart drugs. Platelet function returns to normal when these medications are stopped. Bernard-Soulier Syndrome is a rare hereditary PFD that results in an inability of platelets to stick and spread at sites of blood vessel injury. Glanzmann’s thrombasthenia is also a rare hereditary PFD, which results in an inability of platelets to aggregate.

Signs and Symptoms of a Platelet Function Defect

Easy bruising; bruising with little or no known injury; bruising that occurs in more than one part of the body; bruises which are 2 inches or larger; firm or tender bumps in center of bruises (hematomas)

Heavy or prolonged menstrual periods; periods which last longer than 1 week; need to change tampon or pad every 1-2 hours during the first day of menstrual cycle; use more than 2 dozen (24) tampons or pads in 1 month; frequent need to miss work/school because of heavy menstrual flow

Frequent prolonged nosebleeds; nosebleeds that occur more than once a year and are difficult to control or take more than 10 minutes to stop even with pressure

Prolonged or unusual bleeding after injury, surgery, childbirth or dental work; bleeding that has caused your doctor to tell you that you bleed more than expected or required blood transfusions

Severity of symptoms can vary even among family members who have the same genetic platelet function defect. The majority of people will have mild symptoms and many individuals have no symptoms unless they have a serious injury or an operation. In a few individuals, platelet function disorders can cause severe bleeding.

Diagnosing the Disorder

When a bleeding disorder is suspected, diagnosis is made by obtaining a thorough personal history, family history, physical examination, and laboratory testing (blood tests). These blood tests include:

- Complete blood count: to make sure the platelet count (number of platelets in the blood) is adequate to provide normal clotting.
- Bleeding time: measured by inflating a blood pressure cuff on the upper arm and using a device to make a very small cut on the forearm. The time it takes this cut to stop bleeding is measured using a stopwatch. The bleeding time *might* be longer than normal in patients with abnormal platelet function. However, quite often the bleeding time is normal in patients with abnormal platelet function.
- Platelet aggregation study: Platelets are collected from a blood sample and stirred in tubes where they can be monitored for clumping (aggregation). Activating substances are added to stimulate aggregation and the effects are recorded on a chart. Failure of platelets to aggregate under these conditions suggest that there are abnormalities in platelet release or aggregation.

Treating the Disorder

The treatment of PFDs will depend on the particular type of PFD, as well as location and severity of bleeding. Most patients require NO treatment on a regular basis, but DO need treatment at the time of surgical procedures (including dental procedures), or following accidental injury.

IV DESMOPRESSION (DDAVP) - This is a medication that is injected intravenously (into a vein) before surgery or after an accident, to prevent or decrease excessive bleeding. The response to DDAVP varies among individuals. A trial dose may be recommended by your hematologist, in order to determine a patient's response to the medication. Patients who do not respond to DDAVP may require platelet transfusion in the event of bleeding. Side effects of DDAVP may include headache, facial flushing. DDAVP causes fluid retention and careful monitoring of fluid intake after use of DDAVP is very important to avoid the possibility of fluid overload with resultant seizures. (IV DDAVP is usually not effective in treating severe bleeding in patients with Glanzmann's thrombasthenia or Bernard-Soulier syndrome.)

STIMATE - This is the nasal spray form of DDAVP. This medication is often used by patients with PFD at home to control nosebleeds, heavy menstrual periods, and other bleeding symptoms. Some patients with Glanzmann's or Bernard-Soulier may respond to Stimite for minor bleeding. (Note: There is an intranasal spray form of desmopressin by the brand name "DDAVP". This should NOT be used to treat bleeding because this particular brand is too dilute. The brand "STIMATE" must be used.) (See description of DDAVP above.)

PLATELET TRANSFUSION - Some of the more severe types of PFD, including Bernard-Soulier syndrome or Glanzmann's, may not respond to DDAVP and may require treatment with platelets for bleeding or before surgery.

AMICAR - This medication is used to treat mouth bleeding (and sometimes to treat nosebleeds) along with DDAVP or Stimite. Amicar works to stabilize the blood clot in the mouth or nose, to allow the area to heal and to prevent rebleeding. It is used four times per day for several days.

OINTMENT - For frequent nosebleeds, Vaseline applied to the nasal passages may help prevent drying of the nasal tissues. Saline nasal spray may also help to keep the nose moist.

BIRTH CONTROL PILLS - Some women with PFD may have heavy menstrual bleeding. Birth control pills may help control the heavy bleeding. 4

RECOMBINANT FACTOR VIIa - For patients with more severe PFDs (Glanzmann's or Bernard-Soulier) who do not respond to treatment with platelets, recombinant factor VIIa is usually helpful. It is a genetically engineered activated clotting factor which may bypass the need for platelets to help form a stable clot.



Comprehensive Center for Bleeding Disorders

Things to Remember

- Avoid aspirin and ibuprofen products! Learn to read medication labels. Ask your pharmacist.
- Regular preventative dental care can decrease the need for dental surgery.
- Carry medical identification to alert emergency personnel to your specific bleeding disorder.
- Always wear a helmet when biking, skating, or participating in activities with a risk of head injury.

When to Call Your Hematologist

- Before any surgical procedures or major dental procedures or extractions. You may need pre- and post-procedure treatment to prevent or control bleeding.
- In the event of serious accidental injury.
- If experiencing nosebleeds that are difficult to control, or occur very frequently.
- If experiencing other bleeding that seems prolonged (i.e., mouth bleeding, cuts, etc.)
- If experiencing heavy menstrual bleeding, i.e., longer than 7 days, or requiring a change of protection more often than every 2-3 hours.
- With **any** questions or concerns.

If you have any questions regarding this information, please call your CCBD Nurse Coordinator at (414) 257-2424