Blood Counts

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NSPBCP- Celebrates 10 Year Anniversary

This year marks the 10th anniversary of the creation of the Nova Scotia Provincial Blood Coordinating Program (NSPBCP). Our mission remains as relevant today as it did a decade ago and the Program's many successes are attributed to valued partnerships resulting in quality improvements and innovation in transfusion medicine in Nova Scotia.

The NSPBCP is a provincial program of the Quality, Safety and Wait Time Improvement branch of the Nova Scotia Department of Health and Wellness. The program provides leadership in promoting excellence in transfusion medicine and collaborates with health care providers in the DHAs/IWK and Canadian Blood Services (CBS) in order to support the appropriate management and safe administration of blood and blood products to patients in Nova Scotia.

The NSPBCP will host a reception at Halifax Marriott Harbourfront on November 8, 2013 immediately following Blood Matters. We hope you will be able to join us.

Blood Matters 2013

Power of the Past - Force of the Future is the theme for the 4th annual Blood Matters conference being held on Friday, November 8, 2013. In order to accommodate more participants, Blood Matters 2013 will be held at the Halifax Marriott Harbourfront. The agenda is shaping up to offer some interesting presentations from a variety of great speakers such as Dr. Chantale Pambrun, Dr. Dana Devine, Dr. Alan Tinmouth and Dr. Jeannie Callum. Registration remains free and lunch is provided. Please contact Shelley Strickland at shelley.strickland@cdha.nshealth.ca for further details and registration forms.

Marina Hamilton
Program Manager

Rh Antigen Expression - Weak and Partial D's Determining the Difference and It's Significance

The request to aid in the development of a provincial testing and reporting procedure for Rh testing was brought forward to the NSPBCP to eliminate discrepant Rh patient results between hospitals within the province. Hospitals in Nova Scotia are currently working on a standard Rh testing and reporting algorithm and have made some big strides forward. Information was presented to the Quality Specialist group of the province indicating the importance of determining not only weak D individuals but partial D individuals as well. This determination is especially important in those who are pregnant to avoid the risk of Hemolytic Disease of the Newborn (HDN).

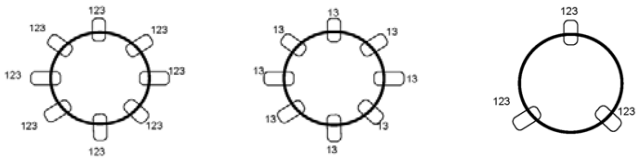
Who are Weak D individuals?

Weak D individuals are those defined as having a reduced amount of D antigen, requiring an indirect antiglobulin test (IAT) for detection. (AABB Technical Manual, Seventeenth edition)

Who are Partial D individuals?

Red cells with partial D have historically been classified as such based on the fact that the red cells type as D positive or negative depending on the reagent used, but individuals make anti-D when exposed to the conventional D antigen. The majority of partial D phenotypes are due to hybrid genes in which portions of RhD are replaced by the corresponding RhCE which results in the loss of D epitopes. These changes are in contrast to weak D, as the changes are predicted to be located on the exterior membrane surface. (AABB Technical Manual, Seventeenth edition)

The difference between Normal D, Partial D and Weak D individuals



Normal D | **Partial D** Lacks an epitope (2) | **Weak D** Reduced number of D Antigens

Dr. Chantale Pambrun, Head, Division of Hematopathology IWK, Children's & Women's Health Centre uses a horse, pony and zebra analogy to describe the difference. If you consider normal D as horses, weak D individuals would be considered ponies. These individuals still exhibit all characteristics of the horse just at a reduced amount. The immune system does not differentiate between horses and ponies. Partial D individuals would be considered zebras; they are built like a horse but are different on the surface. The immune system can differentiate between horses and zebras. If a 'horse' walks into a pasture full of 'zebras', the horse would be detected and the 'zebras' would then form an anti-D.

Why is determining Weak and Partial D patients important?

Determining Weak D and Partial D indviduals is important to avoid alloimmunization of females and to avoid unnessary exposure of RhIG. Weak D patients do not produce an anti-D if exposed to Rh positive cells and therefore determining these patients by testing methods will aid in conservation of Rh neg blood products as well as the unnecessary use of RhIg in these individuals. Partial D individuals on the other hand can produce an anti-D and determining these patients is very important in regards to pregnant females. If these individuals are deemed as Rh postive via routine testing methods they could be transfused with Rh pos red cells and develop an anti-D which could potentially harm the fetus.

Why use two reagents to determine Partial D individuals?

The use of two anti-D reagents differing by the epitope they bind will allow the technologist to determine if the patient is possibly a partial D individual. Discrepant results between the two reagents is indication that a mutation has occured and the patient lacks an epitope.

What is Nova Scotia doing to identify these patients?

The province of Nova Scotia has developed a Rh Testing working group and has determined that two testing algorithms are necessary. One algorithm for pre-natal specimens and cord samples (for determining mothers RhIG eligibility) and the other for the rest of the patient population.

Pre-natal and cord samples will be tested with two different anti-D reagents. These reagents will differ by which epitope they bind on the D antigen. Discrepant results between the two reagents will require further investigation as this indicates the possibility of a partial D patient. Samples on these patients will be refered to Canadian Blood Services for genotyping to determine their Rh status.

Where do we sit right now?

A draft algorithm has been developed and once approved by all members of the working group an SOP and Clinical Guideline will be developed to support implementation. We are anticipating this project be completed by the fall of 2013.

Jennifer LeFrense
Laboratory Standards Coordinator

Transfusion Tidbits - Test your Knowledge

Take this quiz to find out how well you know blood transfusion. Choose one correct answer from the choices listed. Answers are at the end of the quiz.

1. What types of patients are **most** at risk for developing Transfusion Associated Circulatory Overload (TACO)?
   - A. Elderly patients
   - B. Patients with renal disease
   - C. Infants
   - D. All of the above

2. What are some measures that can be taken to prevent TACO?
   - A. Infuse over 4 hours if possible.
   - B. Pre-emptive diuretics
   - C. Avoid transfusing more than one unit at a time
   - D. All of the above

3. What is the correct action if it is 4 hours since a unit of red cells was started and there is still some in the bag?
   - A. Transfuse the remainder in order not to waste it
   - B. Stop the transfusion and notify ordering healthcare provider that patient did not receive entire unit
   - C. Insert a new IV and transfuse remainder of unit into it

4. What type of reaction might your patient be having if he/she developed chills during the transfusion?
   - A. Minor Allergic
   - B. Febrile Non-Hemolytic
   - C. Acute Hemolytic
   - D. Answers B and C

5. What temperatures must RBCs be maintained at during transport?
   - A. 1-10°C
   - B. 5-15°C
   - C. 2-6°C

6. Why is it important to identify patients that exhibit a partial D?
   - A. They are at risk of developing anti-D if exposed to Rh + red cells
   - B. To ensure partial D individuals still receive RhIG
   - C. Both A and B

7. What size filter must be used in the blood tubing for the administration of red cells?
   - A. 170-260 microns
   - B. 20- 40 microns
   - C. 120- 160 microns

8. IVIG should be administered using a dosing (adjusted) body weight. Which indication is an exception to this?
   - A. Myasthenia Gravis
   - B. Guillain-Barré Syndrome
   - C. Idiopathic Thrombocytopenic Purpura

9. Which of the following requires a type and screen prior to being issued from blood transfusion services?
   - A. Red cells
   - B. Albumin
   - C. Prothrombin complex concentrates

10. What is Berinert® used for?
    - A. Prevention of bleeding
    - B. Treatment of hemolytic uremic syndrome
    - C. Hereditary angioedema

Peggy Wilson
Transfusion Practice Coordinator

Upcoming Events

AATB 37th Annual Meeting National Harbor, MD
October 1-5, 2013

AABB Annual Meeting & CTTXPO 2013 Denver, CO
October 12-15, 2013

"Blood Matters" 4th Annual Transfusion Medicine Conference Halifax Marriott Harbourfront Hotel Halifax, NS November 8, 2013

NSPBCP 10th Anniversary Reception Halifax Marriott Harbourfront Hotel
November 8, 2013

CSTM 2014 Quebec City, QC May 1-4, 2014

Answers

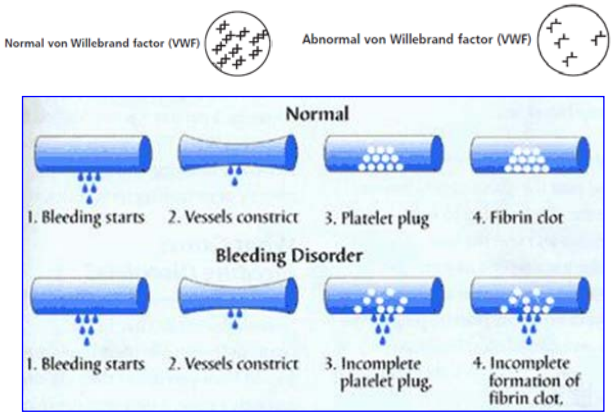
1. D 6. C
2. D 7. A
3. B 8. B
4. D 9. C
5. A 10. C

von Willebrand Disease

Von Willebrand Disease (VWD) was first described in 1925 by the Finnish physician, Erik von Willebrand. 1 in 100 of the general population will have low levels of von Willebrand Factor (VWF) while 1 in 1000 will be diagnosed with the disease. In Nova Scotia, VWD is the most common hereditary bleeding disorder seen in the adult and pediatric Hereditary Bleeding Disorder Clinics (BDC).

What is von Willebrand Disease?

VWD is a bleeding disorder where the VWF is deficient (quantitative) or defective (qualitative). VWF, a glycoprotein, plays a major role in blood coagulation, acting like a glue to hold platelets in place when damage to the blood vessel has occurred. The surface of these platelets then provides an area where blood clotting can occur. The patient with VWD does not have the VWF to hold the platelets in the damaged location thereby not allowing the platelets to adhere to the lining of the blood vessel.



Source: http://www.health-reply.com/willebrand-disease-inheritance/

VWF carries factor VIII (FVIII), another protein required to make a stable clot. When the VWF is low, so may be FVIII.

There are 3 types of VWD:

- Type I – the most common form where people have low levels of VWF and possibly low levels of FVIII
- Type II – the quality of VWF is poor. Type II has 4 subtypes which may have different treatment strategies.
- Type III – this is very rare and the most serious type. These patients do not have any VWF and the FVIII levels are very low.

VWD is usually inherited. Some patients acquire VWD later in life due to other medical conditions but this is a rare occurrence. VWD affects both males and females equally.

What are the Signs and Symptoms of VWD?

The signs and symptoms can vary greatly from person to person. In Type I and Type II, bleeding episodes may be mild to moderate - easy bruising, heavy menstrual periods, frequent or prolonged bleeding from the nose and gums, and prolonged bleeding after injury, surgery, childbirth, or dental work.

In Type III, patients will have the same symptoms however they may also have episodes of severe bleeding for no apparent reason. Patients with Type III may also experience bleeding into soft tissue or joints, causing swelling and pain.

How is VWD diagnosed?

Diagnosing this disease can be very challenging as a patient's blood test results may fluctuate from day to day, with some results being normal. People with blood group O have naturally lower levels of VWF. The diagnosis is based on the following:

- patients bleeding history
- family history of bleeding
- physical examination
- CBC, PT/PTT, INR, fibrinogen
- Factor VIII:C
- VWF: antigen
- VWF activity (Ristocetin cofactor)

Other tests may include...

- VWF multimers
- Platelet function tests

What is the treatment for VWD?

There is no known cure for von Willebrand Disease. Treatment is based on the type of VWD. Minor bleeding episodes may not require treatment. Patients with the severe type may require emergency treatment to stop the bleeding before it becomes life-threatening. Treatment options are:

- Desmopressin acetate (DDAVP) (not effective in Type III)
- FVIII/VWF concentrate (Humate P® or Wilate®)
- Tranexamic acid (Cyklokapron®)
- Fibrin glue

Women may require additional treatment for heavy menstrual bleeding.

Most people with VWD have only occasional mild bleeding problems, others bleed more frequently. With proper medical care, these bleeding episodes can be successfully managed and controlled. Patients are advised to wear a MedicAlert® bracelet or necklace and carry a FactorFirst wallet card.

Source: http://www.hemophilia.ca/files/ER%20CARD%20E_%20Jan%2009.pdf

The FactorFirst wallet card should be recognized and followed as an important tool to treat patients. The card has been provided to patients by the local BDC and is signed by the BDC physician. It contains instructions for the plan of care in case of an emergency. Additional information about VWD is available at http://www.hemophilia.ca.

Reference:
Canadian Hemophilia Society (2011) *All About von Willebrand Disease...for people with von Willebrand Disease and their families* Third Edition. Montreal, Quebec

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