WHERE'S MARY?

GIRLS AND BLEEDING DISORDERS:
SYMPTOMS, DIAGNOSIS AND TREATMENT
To all the girls who suffer in silence

Approximately 50,000 girls in Sweden have a mild bleeding disorder, but only about a thousand have received a diagnosis. Many of them are living with a bleeding disorder without even realizing it. Von Willebrand disease is the most common cause of congenital bleeding tendency and the number of undiagnosed cases is high. So is the number of undiagnosed cases of girls with platelet function defects. There are many other types of bleeding disorders, as well.

In February 2015, the Swedish Hemophilia Society launched a three-year information project to help girls, women and healthcare professionals become more aware of common symptoms that may be signs of a bleeding disorder. Most common symptoms include:

– Heavy menstruation, nosebleeds, bruises, prolonged bleeding after surgery or childbirth.

The type and severity of the disease determines how pronounced a bleeding disorder is in an individual. Investigating the underlying cause is important, because even a mild bleeding disorder entails an increased risk of bleeding in other contexts, such as accidents, surgery or childbirth.

With this booklet, we hope to reach out to all the girls who suffer in silence. We also hope this material will help support primary care and school health professionals in identifying girls suspected of having bleeding disorders.

Mary, the fictional main character, is based on the stories and experiences of Swedish women who have been diagnosed with von Willebrand disease. Girls, women and health care professionals all over the world are awakening to this information and Sweden is taking awareness to the next level in producing this booklet.

With an investigation, a proper diagnosis and treatment, most girls and women with bleeding disorders will be better prepared for the challenges that may arise and live great lives.

Read more about The Women’s project:
www.facebook.com/kvinnorochblodarsjuka (click translate or ask questions in English)
I’m a 17-year-old girl who hopes to be able to help others by blogging about my “bloody” story. Here, you can read about how I was diagnosed with von Willebrand disease, a type of bleeding disorder that, in my case, caused episodes of long-lasting and heavy periods and also a lot of nosebleeds. At first, I thought bleeding that much was normal, but when I started avoiding things I used to enjoy, I realized something wasn’t right …

Even if YOU can’t relate to my story, maybe you have a girlfriend with problems similar to mine? If you have questions, you can contact mary@fbis.se /Mary

Who can get von Willebrand disease (VWD)?
VWD is the most common congenital bleeding disorder and it affects both women and men. VWD is caused by a defect or deficiency of a blood protein called von Willebrand factor that is necessary for normal coagulation. Women with VWD tend to have more symptoms than men due to their menstrual cycle and also after giving birth to their children. According to the World Federation of Hemophilia up to 1% of the world's population suffers from VWD.

How serious is it?
Von Willebrand disease is characterized as mild, moderate and severe. How serious the disease is depends on the type and degree of severity. The mild form is most common and comprises around 75 percent of all cases. The most severe form of von Willebrand disease is rare; currently, only around 50 cases have been diagnosed in Sweden.

Symptoms
Common symptoms include heavy menstrual bleeding, nosebleeds, bleeding from the oral mucosa and bruises. Menstrual bleeding is sometimes so heavy that it affects a woman’s quality of life. In cases of moderate and severe VWD, spontaneous bleeding in the joints and muscles, as well as bleeding in the stomach and intestines can occur. Even people with a mild bleeding disorder may have heavy bleeding after surgery or childbirth. It is therefore important for people suspected of having a bleeding disorder to be assessed.
I used to bleed an absolute ton. My period would practically gush through sanitary towels, and I would regularly wake up several times each night because my sheets were totally drenched with blood. For a while I was so tired that my friends started to wonder if I was doing okay. Finally, I decided to make an appointment at the youth clinic, where I met with a nurse. /Mary

Heavy period? (checklist)
If one or more of the following points applies to you, it may be a sign of a bleeding disorder. Contact a Healthcare Center and request an investigation!

- Do you often bleed through your sanitary protection?
- Do you need to double up on sanitary protection?
- Do you have to change your sanitary protection more than every other hour?
- Do you feel limited in your daily activities while on your period?
- Does your period last for more than 7 days? (the flow may vary)
- Does your period have a significant impact on your quality of life?
- Does your period cause iron deficiency and anemia?

Before my visit at the youth clinic, I received a chart that I had to fill in from the first day of menstruation. It’s called a blood loss assessment chart and it can show whether you are bleeding too much. All you have to keep track of is how many tampons and sanitary towels you use every day. If you experience pain during your period, you also write that down. At the youth clinic, I met a doctor who asked a ton of questions and evaluated my blood chart. She quickly determined that I bleed more than what’s considered normal. /Mary

What counts as a heavy period?

Heavy menstrual bleeding includes both regular and irregular bleeding. 80–90 percent of women with von Willebrand disease have heavy periods, compared with just 10 percent of women in the general population.

Menstruation is considered heavy when a woman bleeds more than 80 ml per menstrual period – a measurement that can be difficult to assess. Comparing yourself with other family members can be misleading, because they might also have an undiagnosed bleeding disorder.

The blood loss assessment chart is a tool used to clinically assess the level of menstrual bleeding. Women enter the number of tampons and sanitary towels used during the course of a menstrual period. This provides a measurement to determine whether menstruation can be considered heavy.

Download the chart at fbis.se/fbis-kvinnokommitten/dokument
**Blood loss assessment chart**

Sometimes it’s hard to know how much you bleed during a menstrual period. To determine how heavy your period is, you have been given a blood loss assessment chart to fill in.

1. Start filling in the chart the day before the first day of your menstrual period.
2. Note the number of sanitary towels and/or tampons used per day by writing a dash for each tampon/sanitary towel in the box under the picture that best illustrates how saturated with blood the tampon/sanitary towel is.
3. There are also columns to report the number of blood clots passed and the number of times you bleed through your sanitary protection per day. Write dashes to indicate the number of blood clots.
4. In the last column, write whether you have experienced pain due to menstruation during the day. Indicate the intensity of the pain by drawing an X on the line.

The first row on the blood loss assessment chart is an example of how to fill it in.

### CALCULATION OF POINTS

**Tampon:**
- 1 blood clot = 1 P
- MEDIUM = 5 P
- A LOT = 10 P

**Sanitary towel:**
- 1 blood clot = 1 P
- MEDIUM = 5 P
- A LOT = 10 P

**Blood clots:**
- NO PAIN
- 1 P
- 5 P
- 10 P

**Bleeding through sanitary protection:**
- NO PAIN
- 1 P
- 5 P
- 10 P

**Pain:**
- NO PAIN
- 1 P
- 5 P
- 10 P

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**EXAMPLE**

<table>
<thead>
<tr>
<th>Date</th>
<th>Tampon</th>
<th>Sanitary towel</th>
<th>Blood clots beside sanitary protection</th>
<th>Bleeding through sanitary protection</th>
<th>Pain</th>
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<tr>
<td>Day 1</td>
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**Over 100 points is considered a heavy period (menorrhagia)**
ARE YOU BLOODY?

In addition to my heavy period, I bruise fairly easily. I’ve also had a lot of nosebleeds, especially during wintertime. So there are more signs of bleeding disorders. Even though my period was the biggest hassle for me, it can vary from person to person.

If you answer yes to any of the questions below, I think you should contact a healthcare center, youth clinic or the school nurse. Then they will refer you to a specialist.

Mary

Symptoms suggesting a bleeding disorder:

☐ Are you troubled by heavy menstrual periods?
☐ Are you often pale and tired?
☐ Do you bruise easily?
☐ Do you have frequent nosebleeds?
☐ Do your gums bleed often?
☐ Do you have heavy bleeding after surgery, childbirth or a miscarriage?
☐ Do you have a relative who “bleeds easily”?

Note!
You don’t have to have all the symptoms. With a mild bleeding disorder, heavy menstrual periods may be the only symptom.

VON MARY AT YOUR SERVICE

The doctor at the youth clinic suspected that my heavy periods could be a form of bleeding disorder called something with “von…” They took several blood samples and referred me to a specialist.

Mary

Recommendations for investigation
When von Willebrand disease is suspected, doctors can do a lot to get a good foundation for continued investigation and assessment.

Medical history
☐ What bleeding symptoms does the person have?
☐ Is this person troubled by heavy menstrual bleeding?
☐ Does the person have any known diseases?
☐ Does the person take any medications?
☐ Does the person have any relatives with a bleeding disorder?
☐ Has the person had heavy bleeding associated with surgery, childbirth or dental extraction?

Blood tests
☐ Complete Blood Count
☐ Platelet Function tests
☐ Factor VIII clotting activity
☐ Von Willebrand Factor tests of activity and amount
☐ Fibrinogen
☐ D-dimer

Note!
The level of VWF may vary due to factors such as birth control pills, infection and pregnancy. If suspicion of von Willebrand disease is high, the doctor will refer you to a hematologist, where an investigation can be carried out to make a diagnosis and identify the severity of the disease for proper treatment.
Who should you talk to if you suffer from heavy periods or other bleeding symptoms?

"First you should visit a healthcare center or a gynecologist where the doctor can ask additional questions and take some blood samples. If a bleeding disorder is suspected, the doctor will write a referral for further investigation at a Hemophilia Treatment Center. They have the resources required to investigate and take care of people with different bleeding disorders."

What happens at a Hemophilia Treatment Center?

"A doctor will carry out a thorough assessment of your medical history and ask questions, for example about bleeding symptoms, dental extractions and bruises. By taking several blood samples, the type of bleeding disorder and degree of severity can be established. Von Willebrand disease is hard to diagnose because levels of the von Willebrand factor can be affected by birth control pills, breastfeeding, pregnancy and infections."

How do you inherit von Willebrand disease?

"Von Willebrand disease can be passed to a child from either the mother or the father. If one parent has the mutation, there is a 50% risk that the child has it. If both parents have a mild form of the disease, there is a risk that the child has a severe form of VWD. The severe form of VWD is recessive, which means it takes a double trait for the disease to occur."

Mild and moderate forms:

- Unaffected mother/dad
- Unaffected daughter/son
- Carrier mother/Dad
- Affected/carryer daughter/son
- Carrier daughter

Severe form:

- Unaffected mother/dad
- Affected mother/Dad
- Affected daughter/son
- Carrier daughter

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What is a bleeding disorder and how do you get it?

"Bleeding disorders are the collective name for a number of chronic disorders that limit the body’s ability to coagulate blood. Most bleeding disorders are congenital and can have different degrees of severity, for example hemophilia A and B, von Willebrand disease, platelet function defects and so on. In some cases, they can occur through genetic mutations without previously being present in the family."

What bleeding disorders can women get?

"The most common forms that affect women are von Willebrand disease and platelet function defects. Both have similar symptoms, where bleeding from mucous membranes and bruises are common. Women may carry the trait for hemophilia, which can sometimes result in bleeding easily.

Idiopathic thrombocytopenic purpura (ITP) is a rare bleeding disorder in which the immune system attacks platelets, causing them to fall to such low levels that bleeding occurs. ITP affects women more often than men."

A few weeks after visiting the Hemophilia Treatment Center, I went back to see the doctor again. She told me I have a mild form of von Willebrand disease. It felt good to know why I bleed so much. My entire family will also get tested, because it’s hereditary. Since I was already there, I took the chance to ask my doctor a few questions about bleeding disorders. /Mary
Now that I know what’s causing my bleeding and I’m getting treatment, I don’t feel sick, even though I’ve received a diagnosis. I feel much better than before, and I take medication during my period or when I have a nosebleed. I’ve also spoken with a gynecologist about possibly starting with birth control pills. /Mary

Choice of treatment depends on the severity of the disease
In mild cases of VWD, treatment when bleeding is often enough, which means you take medicine (like pills or nasal spray) when bleeding episodes occur. Women of childbearing age often take birth control pills, which reduce the amount of menstrual bleeding. Another option is a hormonal intrauterine device (IUD). For more severe bleeding disorders, preventive treatment is required, which means injections of factor concentrate. Women with mild bleeding disorders may also need preventive treatment before surgery and childbirth.

Desmopressin
The effects of desmopressin (Minirin®, Octostim®), a medication that releases VWF from endothelial cells, are often tested in conjunction with an investigation. Desmopressin can be administered intravenously, subcutaneously or as a nasal spray. One side effect is fluid retention and a disturbed salt balance; doctors therefore do not recommend Desmopressin for small children or elderly people with congestive heart failure.

Tranexamic acid
Tranexamic acid (Cyklokapron®, Cyklo-F®) effectively treats mucosal membrane bleeding. Plasmin is an enzyme found in the body that degrades plasma proteins. Tranexamic acid inhibits production of this enzyme. The medication can be administered intravenously, orally and topically, for example in the event of bleeding gums.

VWF concentrate
VWF concentrate is prepared from donor plasma and administered through intravenous injection. This means the person is given the deficient or defective protein. Some concentrates, such as Haemate® and Wilate®, also contain coagulation factor VIII.
"Where is Mary?" A question teachers often asked my friends when I didn’t show up for class. I thought it was embarrassing to say I had my period. And I didn’t think anyone would take it seriously. My heavy periods started interfering with my schoolwork and always being tired made me feel really sad. I feel much happier now and the diagnosis has helped me better understand my disease.

Today I told the whole class and several teachers about von Willebrand disease and how it has affected my life. It feels good that they now know more about the disease, in case something serious would ever happened. I also feel safe now that my health records show that I have an increased risk of bleeding if I ever need surgery. /Mary

Contact the Swedish Hemophilia Society
If you or someone you know has been diagnosed, contact the Swedish Hemophilia Society, a non-profit organization that provides a support network for individuals with bleeding disorders, carriers and their relatives.

Women’s Committee of the Swedish Hemophilia Society
The Women’s Committee proactively works to call attention to and spread information about bleeding disorders that affect girls and women specifically. The committee has representatives for various bleeding disorders that can affect women, for example von Willebrand disease, ITP, hemophilia and carriers.

You can ask questions or contact the Women’s Committee here:
this.se/kvinnokommitten
facebook.com/kvinnorochblodarsjuka
mary@fbis.se

THE COMFORT OF A DIAGNOSIS
Girls also have bleeding disorders

What counts as a heavy period? Are there tools for measuring the amount of menstrual bleeding? What other symptoms are signs of a bleeding disorder in girls and women? Where do you go for help? And what happens during an investigation?

In this booklet, we’ve assembled facts about girls and women with bleeding disorders for people who want to know more or who work in primary care and school health. Mary, the fictional main character, is based on stories from women of the Swedish Hemophilia Society.

THANK YOU to Arvsfonden, the Women’s Committee, Evelina Ahlström, Cheryl Nineff D’Ambrosio, Måns Edlund and partners at the Hemophilia Treatment Center, Karolinska University Hospital in Solna. Project management: Maneka Ghosh Idea and copywriting: Maneka Ghosh and Anna Tollwé Layout: Paizano.se Printing: Norrköpings Tryckeri AB Translation of the booklet is sponsored by Baxalta Incorporated.


This booklet is published by the Swedish Hemophilia Society, 2016, Box 1386, 172 27 Sundbyberg, Phone: 00468-546 405 10, fhus.se