

WOMEN WITH BLEEDING DISORDERS

Introduction

Nature has blessed us with a number of things. Clotting of blood after bleeding is one of these blessings. But there are a few people who are deprived of this blessing i.e. they bleed continuously and their blood does not clot itself. These people also have the problem of spontaneous bleeding. Bleeding can be external and internal. External bleeding can cause blood deficiency, weakness and exertion. Internal bleeding episodes can occur anywhere in the body and cause the area to be immobile. Women with bleeding disorders are often ignored because bleeding disorders are thought to be a male disease. It is very important to understand that bleeding disorders are present in both men and women.

Advancement in medical technology has led to a number of new techniques of diagnosis and treatment of bleeding disorders. von Willebrand Disease is one of the kinds of bleeding disorders and according to a survey, 1-3% people are diagnosed with it.

According to statistics of Pakistan's Haemophilia Centers, von Willebrand Disease is the second most common bleeding disorder after Haemophilia. Men and women are both diagnosed with vWD and therefore the perception that bleeding disorders are only diagnosed in men is wrong.

The causes of ignorance of women with bleeding disorders are misconception of bleeding disorders to be a male disease, little number of patients with bleeding disorders, and unawareness among doctors and society. Deficiency of clotting factors in blood is so rare that even doctors have not seen a patient with such a disease. The diagnosis of deficiency of clotting factors in blood has been discovered in the past 40 years.

Blood and Bleeding Disorders

To understand bleeding disorders better, it is important to know more about blood and how it functions in the body. Blood is a complex fluid with many parts. It is carried through the body in tubes or vessels called arteries, veins and capillaries. When one of these tubes is injured, a small hole forms, allowing blood to flow out. This is called bleeding. Bleeding is stopped when a **clot**, or jelly-like plug, forms to fill the hole. Bleeding can happen anywhere in the body. Bleeding outside the body from a cut or scrape is easy to see. It is also easy to see the result of bleeding under the skin because a bruise forms. Sometimes a person can bleed from nose or mouth. Bleeding also can occur inside the body where it can not be seen at all, such as joints or organs.

Certain proteins found in the body are needed to stop bleeding and are known as **clotting factors**. Deficiency of these clotting factors cause bleeding disorders.

Clotting factor missing or deficient in people with haemophilia is either factor VIII or factor IX. Whereas deficiency of von Willebrand Factor causes von Willebrand Disease. Clotting factors are used to clot blood in time of bleeding. Clotting factors flowing in the blood past the affected area interact with the platelet plug to form a solid clot and seal the hole.

Symptoms

Some common symptoms of bleeding disorder are:

- Recurrent nose bleeds
- Easy bruising
- Heavy or prolonged menstrual flow
- Bleeding from the digestive tract or urinary tract
- Bleeding into joints, muscles and even brain
- Bleeding can be spontaneous without any apparent cause

Diagnosis

A doctor who suspects that a woman or girl has a bleeding disorder will often make a referral to a haematologist. After carefully reviewing the person's medical and family histories and conducting an examination, the haematologist orders laboratory tests to determine if the person has a bleeding disorder. The tests include a bleeding time test (BTT), Platelet Count and Coagulation Profile. These tests help to diagnose the disease to some extent but in case of abnormal results, several other tests are done which includes Platelet Function Test. Deficiency of vWD factors can also affect functions of factor VIII.

The level of vWF continuously changes in the blood which makes it difficult to make the correct diagnosis of the disease. Several other tests are done to make the diagnosis.

There are a number of situations that can make it difficult to diagnose a bleeding disorder:

- Birth control pills
- Hormone replacement therapy
- Pregnancy or recent delivery of a baby
- Blood group O is associated with lower factor VIII and vWF than group A, B or AB

Bleeding disorders in women and men

von Willebrand Disease

von Willebrand Factor is a protein in the blood essential to the formation of the platelet plug. When there is not enough vWF in the blood or the factor does not work correctly, clots form slowly and poorly. This is especially true when bleeding occurs from nose, mouth and digestive tract. Occasionally bleeding can occur without apparent cause. Person with vWD type 3 is considered to have a severe vWD. Such individuals generally have inherited a gene for vWD from both parents. Some of the most commonly used medications for the treatment of vWD include:

- FFP

- Factor VIII concentrate
- Cryoprecipitate
- vWF Concentrate, Haemalte P, Alphanate
- DDAVP
- Use of oral contraceptive pills

Factor I deficiency

Many individuals with Factor I deficiency are diagnosed at birth because of excessive bleeding from umbilical cord. Children with Factor I deficiency might bleed after minor bumps and scrapes or when baby teeth come out, and they might have frequent nose bleeds. Cryoprecipitate and FFP are used to treat the deficiency of Factor I.

Factor II deficiency

People with Factor II deficiency may experience easy bruising, nose bleeds, bleeding after child birth and bleeding after surgery. FFP and Prothrombin complex concentrates are the two treatment options factor II deficiency.

Factor V deficiency

People with Factor V deficiency usually have mild haemophilia and bleeding after a surgery can be the first sign that a problem exists. Those with severe Factor V deficiency may have spontaneous bleeding from the gums, in the stomach or intestines, or even in the brain. Platelet function can also be affected. FFP is used for its treatment.

Factor VII deficiency

Often, people with Factor VII deficiency are diagnosed as newborns because of bleeding into the brain as a result of birth trauma. Women with Factor VII deficiency can experience heavy menstrual bleeding, spontaneous nosebleeds, gum bleeding, bleeding deep within skin, as well as bleeding into the stomach, intestine, and urinary tract. Factor VII and FFP are used for its treatment.

Factor X deficiency

Persons born with severe Factor X deficiency may have bleeding from the umbilical cord at birth. Women with Factor X deficiency might experience heavy menstrual bleeding. Easy bruising, spontaneous nosebleeds and bleeding into the stomach is common in the patients of Factor X deficiency. Prothrombin Complex Concentrate and FFP are used for its treatment.

Factor XI deficiency

Factor XI deficiency is usually diagnosed after injury-related bleeding, and symptoms tend to be mild. Nearly 50% of people with Factor XI deficiency report no bleeding problems.

Factor XIII deficiency

A person with a factor XIII defect fails to form firm, solid blood clots. They can also have problems with wound healing. Bleeding after surgery or injury can be serious and without treatment, bleeding can last for days or weeks. Clot Lysis Test is used for its diagnosis and it is treated by Factor XIII, FFP or Cryoprecipitate.

Women: Problems and Solution

Problems

Menstrual Bleeding

Prolonged and heavy menstrual bleeding is one of the most common symptoms for women with bleeding disorders. Women with these symptoms are in danger of developing iron deficiency anemia and can be counseled to take an iron supplement. Many women find relief with the use of oral contraceptives, other contraceptive hormones or estrogen. When hormone therapy or DDAVP does not result in adequate relief of menstrual bleeding problems, more invasive approaches might have to be considered, including D&C, hysterectomy (surgical removal of the uterus) and uterine ablation. These are extreme measures that should not be considered unless a woman and her physician feel there are no alternatives. In most cases, D&C is ill advised, since it promotes new bleeding from the uterine lining. Mirena Coil is an instrument put inside uterus which produces hormones to minimize time of bleeding. Before using Mirena Coil, consultation with your haematologist is very important.

Painful menstruation/mid-cycle pain

Women and girls with bleeding disorders often complain of pain during their periods and at mid-cycle. Pain can result from the volume of bleeding and incomplete clots forming in the uterus. Some women experience pain in the middle of their cycle during ovulation. This is called mittelschmerz, pain associated with ovulation.

Conception/fertility

If a woman uses hormonal therapy to control bleeding, this treatment will interfere with her ability to conceive. Among women in the bleeding disorder community, there are reports of a higher incidence of miscarriage than in the general population.

Pregnancy and childbirth

Most of the women with bleeding disorders don't have problems in pregnancy and childbirth. But the medications they are on might create problems in conceiving a child. Miscarriage is more common in women with bleeding disorders. Women with bleeding disorders are strongly advised to have regular medical care from the first days of the pregnancy through the postpartum period.

During pregnancy, higher hormone levels may stimulate increased levels of plasma clotting factors. This provides a protective effect for some women with bleeding disorders at the time of delivery. But after the birth of the child, factor level drops again and results in heavy blood loss after delivery. Therefore it is important to go through the delivery in a hospital where the doctors know about the disease. It is very important to consult your obstetrician before delivery so your obstetrician can discuss the problem with your haematologist and make sure that proper treatment is given.

Solutions

Getting past the obstacles

Women and girls with bleeding disorders might experience a sense of isolation because they are unaware that other women struggle with similar conditions or because their complaints, especially if they report heavy periods, are not taken seriously. They might feel alone within the organized bleeding disorders community where, traditionally, the majority of the members have been men and boys with haemophilia. Many women find it difficult to take care of themselves. The most important way to be healthy is to obtain appropriate medical care. But sometimes women with bleeding disorders ignore their condition and avoid making treatment decisions. Bleeding disorders can give rise to a number of questions in your mind and can cause anxiety and fear in you. Women with bleeding must understand the fact that they have a bleeding disorder and by keeping this in mind, they should plan for their future. Communication with other women with bleeding disorders can be helpful and this can be done by making a group of women who have bleeding disorders.

Consultation with haematologist

Women with bleeding disorders have very specific needs and concerns that are often not understood by even the best-trained medical professionals. Therefore it is necessary to consult such a doctor who's not only familiar with the disease but also know about the problems faced by these patients.

Lifestyle modifications

Being in good physical condition is important to reduce the potential for bleeding problems. Regular exercise and physical activity help keep joints and muscles strong and can actually raise the plasma level of clotting factor in women with mild vWD and those

who are symptomatic factor VIII carriers. These people should not also involve in activities in which there is a chance of getting injured.

Medical card

People with bleeding disorders must carry some sort of document (e.g. your physician's or HTC's business card) describing your condition, the prescribed treatment(s) to be used during a bleeding episode or an emergency, and the name and telephone number of your doctor or HTC.

Offering information at school and at work

Teachers, nurses and other school personnel who have no experience with bleeding disorders can feel anxious about having a student with a bleeding disorder in the school. It is helpful to meet with the school staff to inform and educate them about your daughter's bleeding disorder and establish open communication. Provide them with the complete set of booklets on haemophilia for their better understanding.

Psychological and emotional problems of women with bleeding disorders

Starting Phase

Women and girls with bleeding disorders might feel a sense of isolation: perhaps because they are unaware that other women struggle with similar conditions or if they are not taken seriously when they talk to their doctors about heavy periods for example. In addition, the process of arriving at a diagnosis can be long and frustrating, and may cause anxiety.

Many women have found it hard to be taken seriously and listened to sensitively. You may have to fight to get diagnosed and treated. Some women get angry and frustrated because it takes so long to diagnose the condition and, in the meantime, they just have to live with it. But it's important to accept that these emotions are normal and allow yourself to be human. At the same time it's good to learn when and how to ask for help and support when you need it.

Women speaking for themselves

Many women and girls want to take control of decisions about their treatment and take responsibility for their own welfare. This empowerment enables women to make joint decisions with their medical team about treatment options and helps them feel more in control of their life.

Talking to those who care about you

Many women don't think their symptoms are at all unusual. They just think that's the way things are. For this – and a lot of other reasons – many women can be hesitant about

sharing information about their condition with friends and family. They may feel uncomfortable or embarrassed, or not know what words to use or how to start the conversation. But there are good reasons to talk with your extended family because bleeding disorders are usually hereditary. The information you share could lead to the diagnosis of a relative, and could also be a great source of strength and support to you. At the beginning of a relationship women are often concerned about the impact of disclosing their condition to their partner. If you are unsure about what to say, and how to say it, you might want to talk to other women with bleeding disorders in the Haemophilia Society's support network.

You are not alone

Many women find it helps to talk to others similarly affected. Talking to other women gives you a chance to share experiences and learn more about living and coping successfully with vWD. If you find it difficult to face your medical problem, a first step might be to talk to other women or girls with bleeding problem. Ask your haemophilia centre to put you in touch with other women with vW, or contact the Haemophilia Society to find out about our support network.

Issues for parents

Psychological pressures

People who have a child with a genetic problem often blame themselves or feel guilty. But you can only be guilty of something you have control over, and you have no control over your genetic make-up.

Psychological fostering

The condition does not affect intellectual properties. Children with vW can take part in all classroom activities and their motivation level and achievement rate will be the same as any other child's. There are only few sports or activities in which the child with vW taking part should be careful.

Students with vW should be treated as any other student. Being seen as different can be harmful to a child's emotional and social well-being. It is only in cases of the severe form of vW that bleeding episodes may mean your child has to miss some school and may need extra help in catching up with schoolwork.

Mental reinstatement

Treatments can be very frightening for a young child, and it is important for parents to be there to reassure them and let them know that everything will be alright. If your daughter has a bleeding condition, it is useful to prepare her for her first menstrual period in a supportive and positive way. Tell her that girls go through this stage of life when they want to discover more about her. In this process, she might have face harsh reactions

from people around her. Therefore supportive attitude by the parents will help her pass this stage easily.

Important thing

To maintain a healthy lifestyle, learn as much as you can about your condition and understand the impact it has on your life. Work to find medical professionals with whom you feel comfortable and who listen to your concerns and needs. Working with your medical team and making decisions together will give you a greater sense of control and choice in managing your life and treating your condition. A healthy attitude also helps. When you are ready to face your disease, your Haemophilia centre can be very helpful.