About bleeding disorders

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Bleeding disorders are rare health conditions where a person's blood doesn't clot properly.

If someone with a bleeding disorder starts to bleed, it can take longer than other people for their blood to clot and for bleeding to stop. This can happen because there is not enough of an essential clotting factor in the blood, or it doesn't work the way it should. A clotting factor is a protein in the blood that controls bleeding. Many different clotting factors work together in a chain of chemical reactions in the process to stop bleeding.

With current treatments, bleeding disorders can be managed effectively. However, some bleeding episodes can be life-threatening without treatment.

Types of bleeding disorders

- haemophilia
- von Willebrand disorder (VWD)
- rare clotting factor deficiencies
- inherited platelet function disorders.

You cannot 'catch' a bleeding disorder.

Bleeding disorders are nearly always caused by changes to the genes responsible for blood clotting. The altered genes can be passed down from parent to child. People with genetic or inherited bleeding disorders are born with them. They are lifelong conditions.

The exceptions are acquired haemophilia and acquired VWD, which are autoimmune disorders that can occur in adults. These conditions are very rare.

How common are bleeding disorders?

Bleeding disorders are rare but occur in all races and socio-economic groups.

In Australia there are more than 5,900 people diagnosed with haemophilia, von Willebrand disorder and other related bleeding disorders.

Haemophilia

A person with haemophilia has low levels of either clotting factor VIII (8) or factor IX (9).

There are two types of haemophilia, both with the same symptoms:

- **haemophilia A** - factor VIII (8) deficiency
- **haemophilia B** - factor IX (9) deficiency

When factor levels are significantly low, bleeding problems can occur. Haemophilia can be mild, moderate or severe, depending on the percentage of factor VIII or IX in the blood.

Men with haemophilia will pass the gene causing haemophilia on to all their daughters but not their sons. Women who have the gene may pass it on to either daughters or sons. Most females with the gene carry it without bleeding symptoms, but some have low factor levels and have haemophilia.

Haemophilia is usually inherited, but in a third of cases there is no previous family history – it is caused by a spontaneous mutation that has occurred during reproduction. There are genetic tests available to determine whether you have the altered gene causing haemophilia.

Common symptoms

- bruising easily
- more painful swelling and bruising and bleeding for longer after an injury than you would expect
- prolonged bleeding after medical or dental procedures or surgery
- internal bleeding into muscles, joints and organs
- in females – heavy and/or long menstrual periods; heavy or prolonged bleeding after childbirth.

If internal bleeding is not stopped quickly with treatment, it will result in pain and swelling. Repeated bleeding into joints and muscles over time can cause permanent damage, such as arthritis in the joints, and chronic pain.

There is a myth that people with haemophilia bleed to death from a cut. This is NOT true.

Treatment

With current specialised treatments to help blood clot normally, haemophilia can be managed effectively. Treatment may involve factor replacement therapy, which is injected into a vein. This treatment may be given 'on demand' to stop a bleeding episode or before surgery, medical or dental procedures; or as 'prophylaxis', up to 4 times per week to prevent or reduce bleeding. There are also other treatments, depending on what is suitable for the individual, and new treatments are in development and becoming available.

Is there a cure?

Research into gene therapy is currently underway and looks very promising.

Von Willebrand disorder (VWD)

People with VWD (also known as von Willebrand disease) have a problem with a protein in their blood called von Willebrand factor (VWF) that helps control bleeding. They do not have enough of the protein or it does not work the way it should.

VWD is the most common inherited bleeding disorder worldwide. The altered gene causing VWD is passed on from parent to child. VWD affects males and females equally.

Most people with VWD have few or no symptoms and it causes little disruption to their lives, except when they have a serious injury or need surgery. As a result many have not yet been diagnosed. Some people with VWD have bleeding episodes more often, and people with the severe form can often have bleeding into muscles and joints with no obvious cause, similar to severe haemophilia. There can be bleeding problems with all forms of VWD.

Common symptoms

Bleeding in VWD usually involves the mucous membranes, the delicate tissues that line body passages such as the nose, mouth, uterus, vagina, stomach and intestines.

Symptoms vary from person to person, even in the same family, and can include:

- nosebleeds, bleeding from the gums
- bruising easily
- bleeding for a long time with minor cuts
- very heavy or long menstrual periods
- excessive bleeding after injury, surgery or dental work, or after childbirth

Treatment

Several treatments are available, depending on what is appropriate for type of VWD and the individual at the time. This includes synthetic hormones and clotting factor concentrate made with von Willebrand factor and factor VIII.

Rare clotting factor deficiencies

These are other bleeding disorders caused when the body does not produce enough of a specific certain clotting factor, or when the factor does not work properly. They include factor I (1), II (2), V (5), VII (7), X (10), XI (11), or XIII (13) deficiencies and combined factor V (5) and factor VIII (8) deficiency. These disorders are very uncommon.

Platelet function disorders

In platelet function disorders, the platelet plug does not form properly, leading to a tendency to bleed for longer than normal or bruise easily. Since platelets have many roles in blood clotting, platelet function disorders can range from mild to severe. Examples include Glanzmann thrombasthenia and Bernard-Soulier syndrome.

Haemophilia Treatment Centres

Specialist Haemophilia Treatment Centres are available in all states/territories and have a team of health professionals with expertise in treatment and care for people with bleeding disorders.

To find a Centre near you, visit: www.haemophilia.org.au

Where can I find more information?

Talk to your doctor if you think you may be affected by a bleeding disorder. They can then make a referral to a specialist Haemophilia Treatment Centre.

Visit the HFA website for more information about bleeding disorders – www.haemophilia.org.au

Reviewed by representatives of the Australian Haemophilia Nurses Group and the Australian Haemophilia Centre Directors' Organisation.

Haemophilia Foundation Australia (HFA) is the national peak body that represents people with haemophilia, von Willebrand disorder and other bleeding disorders, and their families through:

- representation and advocacy
- education
- promotion of research

HFA works with a network of State and Territory Foundations to ensure everyone with bleeding disorders in Australia has access to the world's best practice treatment and care. Contact HFA for information about bleeding disorders:

W: www.haemophilia.org.au
T: 1800 807 173
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HAEMOPHILIA FOUNDATION AUSTRALIA