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**Course Title: Wbcs and platelets disorders (MLT 4TH ) Instructor: Saima hadi**

**Marks: 50**

Attempt all questions .Each question carry 10 marks.

**Q1. Write a note on Hodgkin lymphoma?**

**ANSWER:** Hodgkin's lymphoma — formerly known as Hodgkin's disease — is a cancer of the lymphatic system, which is part of your immune system. It may affect people of any age, but is most common in people between 20 and 40 years old and those over 55.In Hodgkin's lymphoma, cells in the lymphatic system grow abnormally and may spread beyond it.Hodgkin's lymphoma is one of two common types of cancers of the lymphatic system. The other type, non-Hodgkin's lymphoma, is far more common.Advances in diagnosis and treatment of Hodgkin's lymphoma have helped give people with this disease the chance for a full recovery. The prognosis continues to improve for people with Hodgkin's lymphoma.

Types

1. [Chronic lymphocytic leukemia](https://www.mayoclinic.org/diseases-conditions/chronic-lymphocytic-leukemia/symptoms-causes/syc-20352428)
2. [Cutaneous B-cell lymphoma](https://www.mayoclinic.org/diseases-conditions/cutaneous-b-cell-lymphoma/cdc-20352953)
3. [Cutaneous T-cell lymphoma](https://www.mayoclinic.org/diseases-conditions/cutaneous-t-cell-lymphoma/symptoms-causes/syc-20351056)
4. [Lymphoma](https://www.mayoclinic.org/diseases-conditions/lymphoma/symptoms-causes/syc-20352638)

**Symptoms**

**Swollen lymph nodesOpen pop-up dialog box**

**Signs and symptoms of Hodgkin's lymphoma may include:**

* Painless swelling of lymph nodes in your neck, armpits or groin
* Persistent fatigue
* Fever
* Night sweats
* Unexplained weight loss
* Severe itching
* Increased sensitivity to the effects of alcohol or pain in your lymph nodes after drinking alcohol

## Causes

Doctors aren't sure what causes Hodgkin's lymphoma. But it begins when an infection-fighting cell called a lymphocyte develops a genetic mutation. The mutation tells the cell to multiply rapidly, causing many diseased cells that continue multiplying.

The mutation causes a large number of oversized, abnormal lymphocytes to accumulate in the lymphatic system, where they crowd out healthy cells and cause the signs and symptoms of Hodgkin's lymphoma.

Various types of Hodgkin's lymphoma exist. Your diagnosis is based on the types of cells involved in your disease and their behavior. The type of lymphoma you are diagnosed with determines your treatment options.

**Risk factors**

Factors that can increase the risk of Hodgkin's lymphoma include:

* **Your age.** Hodgkin's lymphoma is most often diagnosed in people between 15 and 30 years old and those over 55.
* **A family history of lymphoma.** Having a blood relative with Hodgkin's lymphoma or non-Hodgkin's lymphoma increases your risk of developing Hodgkin's lymphoma.
* **Being male.** Males are slightly more likely to develop Hodgkin's lymphoma than are females.

**Past Epstein-Barr infection.** People who have had illnesses caused by the Epstein-Barr virus, such as infectious mononucleosis, are more likely to develop Hodgkin's lymphoma than are people who haven't had Epstein-Barr infections.

**Q2.What is Hemostasis , also explain steps and clotting factors?**

**ANSWER:**

 Hemostasis is the process of how the body stops bleeding from a cut or injury. This involves forming a clot to close the hole in the blood vessel and repairing the blood vessel.

When a blood vessel is injured, platelets stick together to form a plug. Proteins, called clotting factors, interact to form a fibrin mesh to hold the platelets in place. This allows the injury to heal while preventing blood from escaping the blood vessel.

Generally, control of bleeding is achieved very quickly through the formation of a clot. In major trauma or surgery, physicians often need to help patients to achieve adequate hemostasis – in order to minimize blood loss and related injury.

However, some people are born with a bleeding disorder (congenital) that impairs their ability to achieve hemostasis. An example of this is the hereditary disorder, hemophilia.

As well, some people who have never had any bleeding problems can develop a condition that causes them to bleed, known as “acquired hemophilia”.

## Function:

The function of the coagulation pathway is to keep hemostasis, which is the blockage of a bleeding or hemorrhage. Primary hemostasis is an aggregation of platelets forming a plug at the damaged site of exposed endothelial cells. Secondary hemostasis includes the two main coagulation pathways, intrinsic and extrinsic, that meet up at a point to form the common pathway. The common pathway ultimately activates fibrinogen into fibrin. These fibrin subunits have an affinity for each other and combine into fibrin strands that bind the platelets together, stabilizing the platelet plug.

## Mechanism

In the coagulation cascade, chemicals called clotting factors (or coagulation factors) prompt reactions that activate still more coagulation factors. The process is complex, but is initiated along two basic pathways:

* The extrinsic pathway, which normally is triggered by trauma.
* The intrinsic pathway, which begins in the bloodstream and is triggered by internal damage to the wall of the vessel.

## EXTRINSIC PATHWAY

The quicker responding and more direct **extrinsic pathway** (also known as the **tissue factor** pathway) begins when damage occurs to the surrounding tissues, such as in a traumatic injury. Upon contact with blood plasma, the damaged extravascular cells, which are extrinsic to the bloodstream, release factor III (thromboplastin). Sequentially, Ca2+ then factor VII (proconvertin), which is activated by factor III, are added, forming an enzyme complex. This enzyme complex leads to activation of factor X (Stuart–Prower factor), which activates the common pathway discussed below. The events in the extrinsic pathway are completed in a matter of seconds.

## INTRINSIC PATHWAY

The **intrinsic pathway** (also known as the contact activation pathway) is longer and more complex. In this case, the factors involved are intrinsic to (present within) the bloodstream. The pathway can be prompted by damage to the tissues, resulting from internal factors such as arterial disease; however, it is most often initiated when factor XII (Hageman factor) comes into contact with foreign materials, such as when a blood sample is put into a glass test tube. Within the body, factor XII is typically activated when it encounters negatively charged molecules, such as inorganic polymers and phosphate produced earlier in the series of intrinsic pathway reactions. Factor XII sets off a series of reactions that in turn activates factor XI (antihemolytic factor C or plasma thromboplastin antecedent) then factor IX (antihemolytic factor B or plasma thromboplasmin). In the meantime, chemicals released by the platelets increase the rate of these activation reactions. Finally, factor VIII (antihemolytic factor A) from the platelets and endothelial cells combines with factor IX (antihemolytic factor B or plasma thromboplasmin) to form an enzyme complex that activates factor X (Stuart–Prower factor or thrombokinase), leading to the common pathway. The events in the intrinsic pathway are completed in a few minutes.

## COMMON PATHWAY

Both the intrinsic and extrinsic pathways lead to the **common pathway**, in which fibrin is produced to seal off the vessel. Once factor X has been activated by either the intrinsic or extrinsic pathway, the enzyme prothrombinase converts factor II, the inactive enzyme prothrombin, into the active enzyme **thrombin**. (Note that if the enzyme thrombin were not normally in an inactive form, clots would form spontaneously, a condition not consistent with life.) Then, thrombin converts factor I, the insoluble fibrinogen, into the soluble fibrin protein strands. Factor XIII then stabilizes the fibrin clot.

**Q3.Explain Hemophilia its types, symptoms, and lab diagnosis ?**

**ANSWER:**

**Types**

There are two major types of hemophilia, type A and type B.

In hemophilia A, there is a lack of clotting factor VIII. This accounts for about [80 percent](https://www.nhlbi.nih.gov/health/health-topics/topics/hemophilia) of hemophilia cases. About 70 percent of people with hemophilia A have the severe form.

In hemophilia B, also known as “Christmas disease,” the person lacks clotting factor IX. Hemophilia occurs in around [1 in every 20,000](https://ghr.nlm.nih.gov/condition/hemophilia#statistics) males born worldwide.

Both A and B can be mild, moderate, or severe, depending on the amount of clotting factor that is in the blood. From [5 to 40 percent](https://www.nhlbi.nih.gov/health/health-topics/topics/hemophilia/diagnosis) of normal clotting factor is considered mild, 1 to 5 percent is moderate, and less than 1 percent is severe.

## Symptoms

Hemophilia [symptoms include](https://www.nhlbi.nih.gov/health/health-topics/topics/hemophilia/signs) excessive bleeding and easy bruising. The severity of symptoms depends on how low the level of clotting factors is in the blood.

Bleeding can occur externally or internally.

Any wound, cut, bite, or dental injury can lead to excessive external bleeding.

Spontaneous nosebleeds are common.

There may be prolonged or continued bleeding after bleeding previously ceased.

Signs of excessive internal bleeding include blood in the urine or stools, and large, deep bruises.

Bleeding can also happen within joints, like knees and elbows, causing them to become

swollen, hot to the touch, and painful to move.

A person with hemophilia may experience internal bleeding in the brain following a bump on the head.

Symptoms of brain bleeding can include [headaches](https://www.medicalnewstoday.com/articles/73936.php), vomiting, lethargy, behavioral changes, clumsiness, vision problems, paralysis, and seizures.

**Diagnosis**

Medical history and blood tests are key to diagnosing hemophilia.

If a person has bleeding problems, or if hemophilia is suspected, a physician will ask about the person’s family and personal medical history, as this can help to identify the cause.

A physical examination will be carried out.

Blood tests can provide information about how long it takes for blood to clot, the levels of clotting factors, and which clotting factors, if any, are missing.

Blood test results can identify the type of hemophilia and its severity.

For pregnant women who are carriers of hemophilia, doctors are able to test the fetus for the condition after [10 weeks](https://www.cincinnatichildrens.org/health/h/hemophilia-test) of pregnancy.

**Q4 .Describe Von Wille Brand disease?**

**ANSWER:**

Von Willebrand disease (VWD) is a blood disorder in which the blood does not clot properly. Blood contains many proteins that help the body stop bleeding. One of these proteins is called von Willebrand factor (VWF). People with VWD either have a low level of VWF in their blood or the VWF protein doesn’t work the way it should.

Normally, when a person is injured and starts to bleed, the VWF in the blood attaches to small blood cells called platelets. This helps the platelets stick together, like glue, to form a clot at the site of injury and stop the bleeding. When a person has VWD, because the VWF doesn’t work the way it should, the clot might take longer to form or not form the way it should, and bleeding might take longer to stop. This can lead to heavy, hard-to-stop bleeding. Although rare, the bleeding can be severe enough to damage joints or internal organs, or even be life-threatening.

### Who is Affected

VWD is the most common bleeding disorder, found in up to 1% of the U.S. population. This means that 3.2 million (or about 1 in every 100) people in the United States have the disease. Although VWD occurs among men and women equally, women are more likely to notice the symptoms because of heavy or abnormal bleeding during their menstrual periods and after childbirth.

### Types of VWD

##### **Type 1**

This is the most common and mildest form of VWD, in which a person has lower than normal levels of VWF. A person with Type 1 VWD also might have low levels of factor VIII, another type of blood-clotting protein. This should not be confused with [hemophilia](https://www.cdc.gov/ncbddd/hemophilia/index.html), in which there are low levels or a complete lack of factor VIII but normal levels of VWF. About 85% of people treated for VWD have Type 1.

##### **Type 2**

With this type of VWD, although the body makes normal amounts of the VWF, the factor does not work the way it should. Type 2 is further broken down into four subtypes―2A, 2B, 2M, and 2N―depending on the specific problem with the person’s VWF. Because the treatment is different for each type, it is important that a person know which subtype he or she has.

##### **Type 3**

This is the most severe form of VWD, in which a person has very little or no VWF and low levels of factor VIII. This is the rarest type of VWD. Only 3% of people with VWD have Type 3.

### Causes

Most people who have VWD are born with it. It almost always is inherited, or passed down, from a parent to a child. VWD can be passed down from either the mother or the father, or both, to the child.

While rare, it is possible for a person to get VWD without a family history of the disease. This happens when a “spontaneous mutation” occurs. That means there has been a change in the person’s gene. Whether the child received the affected gene from a parent or as a result of a mutation, once the child has it, the child can later pass it along to his or her children. Rarely, a person who is not born with VWD can acquire it or have it first occur later in life. This can happen when a person’s own immune system destroys his or her VWF, often as a result of use of a medication or as a result of another disease. If VWD is acquired, meaning it was not inherited from a parent, it cannot be passed along to any children.

### Signs and Symptoms

The major signs of VWD are:

##### **Frequent or Hard-to-Stop Nosebleeds**

People with VWD might have nosebleeds that:

* Start without injury (spontaneous)
* Occur often, usually five times or more in a year
* Last more than 10 minutes
* Need packing or cautery to stop the bleeding

##### **Easy Bruising**

People with VWD might experience easy bruising that:

* Occurs with very little or no trauma or injury
* Occurs often (one to four times per month)
* Is larger than the size of a quarter
* Is not flat and has a raised lump

##### **Heavy Menstrual Bleeding**

Women with VWD might have [heavy menstrual periods](https://www.cdc.gov/ncbddd/blooddisorders/women/menorrhagia.html) during which:

* Clots larger than the size of a quarter are passed
* More than one pad is soaked through every 2 hours
* A diagnosis of anemia (not having enough red blood cells) is made as a result of bleeding from heavy periods

##### **Longer than Normal Bleeding After Injury, Surgery, Childbirth, or Dental Work**

People with VWD might have longer than normal bleeding after injury, surgery, or childbirth, for example:

* After a cut to the skin, the bleeding lasts more than 5 minutes
* Heavy or longer bleeding occurs after surgery. Bleeding sometimes stops, but starts up again hours or days later.
* Heavy bleeding occurs during or after childbirth

People with VWD might have longer than normal bleeding during or after dental work, for example:

* Heavy bleeding occurs during or after dental surgery
* The surgery site oozes blood longer than 3 hours after the surgery
* The surgery site needs packing or cautery to stop the bleeding

The amount of bleeding depends on the type and severity of VWD. Other common bleeding events include:

* Blood in the stool (feces) from bleeding into the stomach or intestines
* Blood in the urine from bleeding into the kidneys or bladder
* Bleeding into joints or internal organs in severe cases (Type 3)

### Diagnosis

To find out if a person has VWD, the doctor will ask questions about personal and family histories of bleeding. The doctor also will check for unusual bruising or other signs of recent bleeding and order some blood tests that will measure how the blood clots. The tests will provide information about the amount of clotting proteins present in the blood and if the clotting proteins are working properly. Because certain medications can cause bleeding, even among people without a bleeding disorder, the doctor will ask about recent or routine medications taken that could cause bleeding or make bleeding symptoms worse.

### Treatments

The type of treatment prescribed for VWD depends on the type and severity of the disease. For minor bleeds, treatment might not be needed.

The most commonly used types of treatment are:

##### **Desmopressin Acetate Injection**

This medicine DDAVP is injected into a vein to treat people with milder forms of VWD (mainly type 1). It works by making the body release more VWF into the blood. It helps increase the level of factor VIII in the blood as well.

##### **Desmopressin Acetate Nasal Spray**

This high-strength nasal spray Stimate is used to treat people with milder forms of VWD. It works by making the body release more VWF into the blood.

##### **Antifibrinolytic Drugs**

These drugs (for example, Amicar, Lysteda) are either injected or taken orally to help slow or prevent the breakdown of blood clots.

##### **Birth Control Pills**

Birth control pills can increase the levels of VWF and factor VIII in the blood and reduce menstrual blood loss. A doctor can prescribe these pills for women who have heavy menstrual bleeding.

**Q5.Explain Hemolytic uremic syndrome and its types?**

**ANSWER:**

Hemolytic uremic syndrome (HUS) is a condition that affects the blood and blood vessels. It results in the destruction of blood platelets (cells involved in clotting), a low red blood cell count (anemia) and kidney failure due to damage to the very small blood vessels of the kidneys. Other organs, such as the brain or heart, may also be affected by damage to very small blood vessels.

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In HUS the tiny filter units in the kidneys known as glomeruli become clogged with platelets and damaged red blood cells. This leads to problems with the kidney's ability to filter and eliminate waste products.

 HUS is considered a syndrome because it is a combination of findings that may have different causes. In most cases, HUS occurs after a severe bowel infection with certain toxic strains of the bacteria called E. coli. It may also occur in response to certain medicines, but this is rare. Even more rarely, HUS occurs for unknown reasons. This fact sheet primarily focuses on the type of HUS that occurs in infants and children as a result of an E. coli infection.

HUS from E. coli infections results when bacterial toxins cross from the intestines into the bloodstream and damage the very small blood vessels. The toxic E. coli may come from eating spoiled, undercooked or poorly processed food products, or from exposure to contaminated water. HUS occurs most often in certain regions, such as the Pacific Northwest and Argentina.

HUS is generally treated with medical care in the hospital. Close attention to fluid volume is very important. This potentially includes intravenous (IV) fluids and nutritional supplementation by IV or tube feeding. A transfusion of blood may also be needed. In about 50 percent of cases, short-term kidney replacement treatment in the form of dialysis is necessary. Most patients who need dialysis will recover kidney function and ultimately be able to discontinue dialysis treatment. At times a special form of treatment called plasmapheresis may also be necessary.

**Symptoms**

The signs and symptoms of HUS may vary, depending on the cause. Most cases of HUS are caused by infection with certain strains of E. coli bacteria, which first affect the digestive tract. The initial signs and symptoms of this form of HUS may include:

* Diarrhea, which is often bloody
* Abdominal pain, cramping or bloating
* Vomiting
* Fever

All forms of HUS — no matter the cause — damage the blood vessels. This damage causes red blood cells to break down (anemia), blood clots to form in the blood vessels and kidney damage. Signs and symptoms of these changes include:

* Pale coloring, including loss of pink color in cheeks and inside the lower eyelids
* Extreme fatigue
* Shortness of breath
* Easy bruising or unexplained bruises
* Unusual bleeding, such as bleeding from the nose and mouth
* Decreased urination or blood in the urine
* Swelling (edema) of the legs, feet or ankles, and less often in the face, hands, feet or entire body
* Confusion, seizures or stroke
* High blood pressure

See your doctor immediately if you or your child experiences bloody diarrhea or several days of diarrhea followed by:

* Decreased urine output
* Swelling
* Unexplained bruises
* Unusual bleeding
* Extreme fatigue