Name Haris Ur RAHMAN
ID NO 14574
Paper **Wbcs and platelets disorde**
Submitted to **Saima hadi**

Department B.S (MLT)

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Q1`):Write a note on Hodgkin lymphoma?
Ans):

## *Hodgkin Lymphoma:"

*Hodgkin's disease (HD) is a type of,lymphoma which is a blood cancer that starts in the lymphatic system. The lymphatic system helps the immune system get rid of waste and fight infections. HD is also called Hodgkin disease, Hodgkin lymphoma, and Hodgkin's lymphoma.

*HD originates in white blood cells that help protect you from germs and infections. These white blood cells are called lymphocytes. In people with HD, these cells grow abnormally and spread beyond the lymphatic system. As the disease progresses, it makes it more difficult for your body to fight infections.

*HD can be either classic Hodgkin's disease or nodular lymphocytic predominant Hodgkin's lymphoma (NLPHL). The type of HD is based on the types of cells involved in your condition and their behavior.

*The main cause of HD isn't known. The disease has been linked to DNA mutations, or changes, as well as to the Epstein-Barr virus (EBV), which causes Mononucleosis. HD can occur at any age, but it most commonly affects people between ages 15 and 40 and people over age 55.

### *symptoms of Hodgkin Lymphoma :,

*The most common symptom of HD is swelling of the lymph nodes, which causes a lump to form under the skin. This lump usually isn't painful. It may form in one or more of the following areas:

*on the side of the neck
*in the armpit
*around the groin

Other symptoms of HD include:

* night sweats
*itchy skin
*fever
*fatigue
*unintended weight loss
*persistent cough, trouble breathing, chest pain
*pain in the lymph nodes after consuming alcohol
* enlarged spleen

Call your doctor right away if you have any of these symptoms. They can be signs of other conditions, and it's important to get an accurate diagnosis.

## *Hodgkin Lymphoma disease diagnosed:,

To diagnose HD, your doctor will perform a physical exam and ask you about your medical history. Your doctor will also order certain tests so they can make a proper diagnosis. The following tests may be done:

*imaging tests, such as X-rays or CT scans
*lymph node biopsy,which involves removing a piece of lymph node tissue to test for the presence of abnormal cells
*blood tests, such as a complete blood count (CBC), to measure levels of red blood cells, white blood cells, and platelets
*immunophenotyping to determine the type of lymphoma cells that are present
Lungs function tests to determine how well your lungs are working
*an echocardiogram to determine how well your heart is working
*bone marrow biopsy which involves the removal and examination of marrow inside your bones to see if the cancer has spread

### *Staging:,

Once an HD diagnosis has been made, the cancer is assigned a stage. Staging describes the extent and severity of the disease. It will help your doctor determine your treatment options and outlook.

There are four general stages of HD:

***Stage 1 (early stage)**means that cancer is found in one lymph node region, or the cancer is found in only one area of a single organ.
***Stage 2 (locally advanced disease)**means that cancer is found in two lymph node regions on one side of the diaphragm, which is the muscle beneath your lung, or that cancer was found in one lymph node region as well as in a nearby organ.
***Stage 3 (advanced disease)** means that cancer is found in lymph node regions both above and below your diaphragm or that cancer was found in one lymph node area and in one organ on opposite sides of your diaphragm.
***Stage 4 (widespread disease)** means that cancer was found outside the lymph nodes and has spread widely to other parts of your body, such as your bone marrow, liver, or lung.

## *How is Hodgkin's disease treated:,

*Treatment for HD typically depends on the stage of the disease. The main treatment options are chemotherapy and radiation.

*Radiiattion therapy uses high-energy beams of radiation to destroy cancer cells. chemothrapyinvolves the use of medications that can kill cancer cells. Chemotherapy drugs may be given orally or injected through a vein, depending on the specific medication.

*Radiation therapy alone may be sufficient for treating early stage NLPHL. If you have NLPHL, you may only need radiation since the condition tends to spread more slowly than classic HD. In advanced stages, targeted therapeutic drugs may be added to your chemotherapy regimen.

*Immunotherapy or a stem cell transplant may also be used if you don't respond to chemotherapy or radiation. A stem cell transplant infuses healthy cells called stem cells into your body to replace the cancerous cells in your bone marrow.

*After treatment, your doctor will want to follow up with you on a regular basis. Be sure to keep all your medical appointments and follow your doctor's instructions.

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

Ans:-Hemostasis :-

*Hemostasis or haemostasis is a process to prevent and stop bleeding, meaning to keep blood within a damaged blood vessel (the opposite of hemostasis is hemorrhage). It is the first stage of wound healing. This involves coagulation, blood changing from a liquid to a gel.

*Platelets are key players in hemostasis, the process by which the body seals a ruptured blood vessel and prevents further loss of blood. Although rupture of larger vessels usually requires medical intervention, hemostasis is quite effective in dealing with small, simple wounds. There are three steps to the process: vascular spasm, the formation of a platelet plug, and coagulation (blood clotting). Failure of any of these steps will result in hemorrhage—excessive bleeding

STEPS

**3 STEPS IN HEMOSTASIS,**

**1)VASOCONSTRICTION:-(VASCULAR SPASM)**

When a vessel is severed or punctured, or when the wall of a vessel is damaged, vascular spasm occurs. In **vascular spasm**, the smooth muscle in the walls of the vessel contracts dramatically. This smooth muscle has both circular layers; larger vessels also have longitudinal layers. The circular layers tend to constrict the flow of blood, whereas the longitudinal layers, when present, draw the vessel back into the surrounding tissue, often making it more difficult for a surgeon to locate, clamp, and tie off a severed vessel. The vascular spasm response is believed to be triggered by several chemicals called endothelins that are released by vessel-lining cells and by pain receptors in response to vessel injury. This phenomenon typically lasts for up to 30 minutes, although it can last for hours.

***HEMOSTASTIC PLUG/FORMATION OF THE PLATELET PLUG:-**

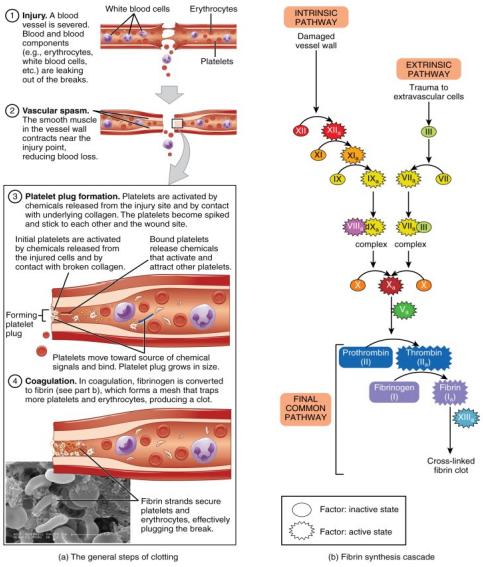
In the second step, platelets, which normally float free in the plasma, encounter the area of vessel rupture with the exposed underlying connective tissue and collagenous fibers. The platelets begin to clump together, become spiked and sticky, and bind to the exposed collagen and endothelial lining. This process is assisted by a glycoprotein in the blood plasma called von Willebrand factor, which helps stabilize the growing **platelet plug**. As platelets collect, they simultaneously release chemicals from their granules into the plasma that further contribute to hemostasis. Among the substances released by the platelets are:

- adenosine diphosphate (ADP), which helps additional platelets to adhere to the injury site, reinforcing and expanding the platelet plug
- serotonin, which maintains vasoconstriction
- prostaglandins and phospholipids, which also maintain vasoconstriction and help to activate further clotting chemicals, as discussed next

A platelet plug can temporarily seal a small opening in a blood vessel. Plug formation, in essence, buys the body time while more sophisticated and durable repairs are being made. In a similar manner, even modern naval warships still carry an assortment of wooden plugs to temporarily repair small breaches in their hulls until permanent repairs can be made.

## *COAGULATION OF BLOOD:-

*Those more sophisticated and more durable repairs are collectively called **coagulation**, the formation of a blood clot. The process is sometimes characterized as a cascade, because one event prompts the next as in a multi-level waterfall. The result is the production of a gelatinous but robust clot made up of a mesh of **fibrin**—an insoluble filamentous protein derived from fibrinogen, the plasma protein introduced earlier—in which platelets and blood cells are trapped. Figure 1 summarizes the three steps of hemostasis.




## *CLOTTING FACTORS:-

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.

*Both of these merge into a third pathway, referred to as the common pathway All three pathways are dependent upon the 12 known clotting factors, including Ca2+ and vitamin K (Table 1). Clotting factors are secreted primarily by the liver and the platelets. The liver requires the fat-soluble vitamin K to produce many of them. Vitamin K (along with biotin

and folate) is somewhat unusual among vitamins in that it is not only consumed in the diet but is also synthesized by bacteria residing in the large intestine.

* The calcium ion, considered factor IV, is derived from the diet and from the breakdown of bone. Some recent evidence indicates that activation of various clotting factors occurs on specific receptor sites on the surfaces of platelets.

*The 12 clotting factors are numbered I through XIII according to the order of their discovery. Factor VI was once believed to be a distinct clotting factor, but is now thought to be identical to factor V. Rather than renumber the other factors, factor VI was allowed to remain as a placeholder and also a reminder that knowledge changes over time.

*Vitamin K required.

**Clotting Factors (Table 1)**

| Factor number | Name | Type of molecule | Source | Pathway(s) |
|---|---|---|---|---|
| I | Fibrinogen | Plasma protein | Liver | Common; converted into fibrin |
| II | Prothrombin | Plasma protein | Liver* | Common; converted into thrombin |
| III | Tissue thromboplastin or tissue factor | Lipoprotein mixture | Damaged cells and platelets | Extrinsic |
| IV | Calcium ions | Inorganic ions in plasma | Diet, platelets, bone matrix | Entire process |
| V | Proaccelerin | Plasma protein | Liver, platelets | Extrinsic and intrinsic |
| VI | Not used | Not used | Not used | Not used |
| VII | Proconvertin | Plasma protein | Liver * | Extrinsic |
| VIII | Antihemolytic factor A | Plasma protein factor | Platelets and endothelial cells | Intrinsic; deficiency results in hemophilia A |
| IX | Antihemolytic factor B (plasma thromboplastin component) | Plasma protein | Liver* | Intrinsic; deficiency results in hemophilia B |
| X | Stuart–Prower factor (thrombokinase) | Protein | Liver* | Extrinsic and intrinsic |
| XI | Antihemolytic factor C (plasma thromboplastin antecedent) | Plasma protein | Liver | Intrinsic; deficiency results in hemophilia C |
| XII | Hageman factor | Plasma protein | Liver | Intrinsic; initiates clotting in vitro also activates plasmin |
| XIII | Fibrin-stabilizing factor | Plasma protein | Liver, platelets | Stabilizes fibrin; slows fibrinolysis |

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

# *Hemophilia:-

*Hemophilia A ar B similar in both clinical and pathological feature  is a difrence being.
*a medical condition in which the ability of the blood to clot is severely reduced, causing the sufferer to bleed severely from even a slight injury. The condition is typically caused by a hereditary lack of a coagulation factor, most often factor VIII.
*This means the person tends to bleed for a longer time after an injury, and they are more susceptible to internal bleeding.
*have enough of a certain kind of clotting factor. This makes it harder for bleeding to stop. People with hemophilia may bleed a lot after cuts, during surgery, or even after a fall. Some people have abnormal bleeding inside their bodies for no clear reason.

## *Types:-

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

## *Type A and B:-

*Hemophilia A: Caused by a lack of the blood clotting factor VIII; approximately 85% of hemophiliacs have type A disease.of hemophilia cases. About 70 percent of people with hemophilia A have the severe form.

*Hemophilia B: Caused by a deficiency of factor IX.

*Hemophilia B, also known as “Christmas disease,” the person lacks clotting factor IX.
*Hemophilia occurs in around1 in every 20,000males 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 4041 percent of normal clotting factor is considered mild, 1 to 5 percent is moderate, and less than 1 percent is severe.

*Hemophilia is classified by its level of severity. Hemophilia may be mild, moderate, or severe, depending on the level of the blood clotting factors in the blood.

## * Symptoms:-

*Hemophilia symptoms include 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, vomiting, lethargy, behavioral changes, clumsiness, vision problems, paralysis, and seizures.

## *Hemophilia lab diagosis:-

*Diagnosis includes screening tests and clotting factor tests. Screening tests are blood tests that show if the blood is clotting properly. Clotting factor tests, also called factor assays, are required to diagnose a bleeding disorder. This blood test shows the type of hemophilia and the severity

In addition to a complete medical history and physical examination, your doctor may perform numerous blood tests including clotting factor levels, a complete blood count, (CBC) assessment of bleeding times, and/or DNA testing. Your doctor may also request details about your family history

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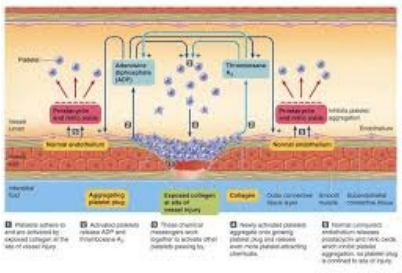
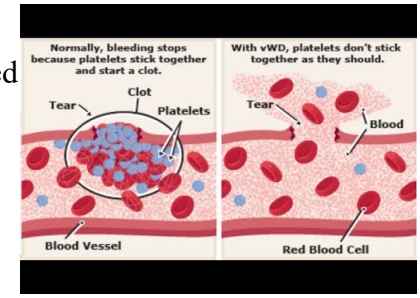
Q4 .Describe Von Wille Brand disease?

## *Von Willebrand disease:-



*Von Willebrand disease is a bleeding disorder. It's caused by a deficiency of von Willebrand factor (VWF). This is a type of protein that helps your blood to clot. Von Willebrand is different from hemophilia, another type of bleeding disorder.

*Bleeding happens when one of your blood vessels breaks. Platelets are a type of cell that circulates in your blood and clumps together to plug broken blood vessels and stop bleeding. VWF is a protein that helps platelets clump together, or clot. If your levels of functional VWF are low, your platelets won't be able to clot properly. This leads to prolonged bleeding.




## * Types of von Willebrand disease:-

*Three main types of von Willebrand disease exist:

### *Type 1

Type 1 is the most common type of von Willebrand disease. It causes lower-than-normal levels of VWF to occur in your body. You still have small amounts of VWF in your body to help clot blood. You'll likely experience mild bleeding problems but be able to live a normal life.

### *Type 2

If you have type 2 von Willebrand disease, you have normal levels of VWF but it won't work properly due to structural and functional defects. Type 2 is divided into subtypes, including types:

- 2A
- 2B
- 2M
- 2N

## *Type 3

Type 3 is the most dangerous type of von Willebrand disease. If you have this type, your body won't produce any VWF. As a result, your platelets won't be able to clot. This will put you at risk of severe bleeding that's difficult to stop.

## *Symptoms of von Willebrand disease:-

If you have von Willebrand disease, your symptoms will vary depending on which type of the disease you have. The most common symptoms that occur in all three types include:

*easy bruising
*excessive nose bleeds
*bleeding from your gums
*abnormally heavy bleeding during menstruation

## *causes von Willebrand disease:-

*A genetic mutation causes von Willebrand disease.

*The type of von Willebrand disease that you have depends on whether one or both of your parents have passed a mutated gene on to you.

* For example,

*you can only develop type 3 Von Willebrand if you've inherited a mutated gene from both of your parents. If you've only inherited one copy of the mutated gene, you'll develop type 1 or 2 von Willebrand disease.

## *von Willebrand disease diagnosed:-

*Your doctor will ask you questions about your personal and family history of abnormal bruising and bleeding. Type 3 tends to be the easiest to diagnose. If you have it, you'll likely have a history of severe bleeding starting early in life.

*Along with taking a detailed medical history, your doctor may also use laboratory tests to check for abnormalities in your VWF levels and function. They may also check for abnormalities in clotting factor VIII, which can cause hemophilia. Your doctor can also use blood tests to learn how well your platelets function.

*Your doctor will need to collect a sample of your blood to conduct these tests. Then, they'll send it to a laboratory for testing. Because of the specialized nature of these tests, it may take up to two or three weeks to receive your results.

Q5Explain Hemolytic uremic syndrome and its types?

Ans:- *Hemolytic uremic syndrome:-

*Hemolytic uremic syndrome (HUS) is a condition that can occur when the small blood vessels in your kidneys become damaged and inflamed. This damage can cause clots to form in the vessels.reduction in the clotting cells called platelets (thrombocytopenia). Organs most commonly affected include the kidneys (kidney failure) and the brain (confusion, seizures).

HUS was previously grouped with another diagnosis called thrombocytopenic thrombotic purpura (TTP) and referred to as “HUS/TTP” because they had similar symptoms. However, it is now known that they are separate diseases and should not be confused. The term “HUS” is not reserved for the process described below when it is caused by an infection of the GI tract with E coli. Other causes of this presentation are referred to as “atypical” HUS, and make up less than 10% of cases.

Hemolytic uremic syndrome is a common cause of acute kidney injury in children.

The diarrhea caused by these bacteria is severe and often bloody.

*Symptoms:-

*Symptoms of E. coli gastroenteritis are:

- •Diarrhea (usually bloody)
- •Fever
- •Stomach pain
- •Vomiting

*Types of (HUS):-

*Typical and Atypical-HUS

*Typical:>

*infection by shiga toxin-producing bacteria E-coli serotype O157:H7 is cause *mostly with diarrhea often bloody diaeehea (D+HUS).

*ATYPICAL:>

Atypical HUS (non-stx-HUS)is rare .

*as the name implies,infection by stx-producing bacteria is not the cause,and disease may occur year-round without diarrhea(D-HUS)

*The familial from is associated with genetic abnormalities.