**WBC and Platelets Disorder final term**

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**Dep:Bs MLT 4TH**

**ANS NO 1:- Note on Hodgkin Lymphoma**

* It is The major subdivision of lymphomas
* Hodgkin's disease is a lymphoma in which RS cells are found in the disease tissue.
* Immunoglobulin gene rearrangement
* the RS cell is of B-lymphoid lineage
* and that is derived from B cell with a 'crippled' Immunoglobulin gene
* caused by the acquisition of mutations
* they prevent synthesis of full-length immunoglobulin.
* Epstein-Barr virus genome has been detected in cases of Hodgkin tissue
* but its role in the pathogenesis is unclear.
* RS cells are found in the disease tissue.
* RS cells, and abnormal mononuclear cells, are neoplastic
* infiltrating inflammatory cells are reactive.

**Staging of Hodgkin's lymphoma**

**Stage I:**

* node involvement in one lymph node area.

**Stage II**:

* involving two or more lymph nodal areas confined to one side of the diaphragm.

**Stage III:**

* involving lymph nodes above and below the diaphragm.

**Stage IV:**

* indicates involvement outside the lymph node areas
* and refers to diffuse disease in the

 bone marrow

 liver

 and other extranodal sites.

**ANS NO2:-**

 **HEMOSTASIS**

* Hemo means BLOOD and stasis means STABLE
* Prevention of blood loss its is a processin which body stop bleeding

Steps­­:-

There are three steps involve in this procedure….

i**)Vasoconstriction**

* They increase the result of Ca ion
* And stimulate cross bridge cycle

ii)**Hemostastic plug**

* also called platelets plug
* in this procedure platelets perform 2 functions:-
1. they Release two chemicals
* thromboxane
* ADP adenosine diphosphate
1. And they fill the injured areas
* Both are help in platelets aggregations
* glycoprotein localize some factors to the sites

**iii)Coaglution of blood**

* it is responsible for preventing and termination of bleeding
* it is process in which a blood clot is formed.
* It is consist of 13 clotting factors
* They help in different coagulation pathways
* But factor **vi** is still missing

**FACTORS:-**

* Fibrinogen,Prothrombin ,Thromboplastin,Calium ,Labile factor

Presence not proved,Stable factor,Antihemophilic ,Christmas, Staurt power factor,

Plasma thromboplastein ,Hagemen and Fibrin stablizing factors

**ANS NO 3;-**

**Hemophilia**

* It is a medical condition
* in which the blood clot is severely reduced,
* causing to bleed severely from a slight injury.
* it is typically caused by a hereditary lack of a coagulation factor,
* most often is factor VIII.

**The three main types of hemophilia include :**

**Hemophilia A:**

* It is Caused by the lack of the blood clotting factor VIII
* approximately 85% of hemophiliacs have type A disease.

**Hemophilia B:**

* Caused by a deficiency of factor IX.

**Hemophilia C**:

* this term refer to a lack of clotting factor XI

**Symptoms of hemophilia include:**

* Bleeding into a joint or muscle

 which causes pain and swelling.

* Abnormal Bleeding after an injury or surgery.
* Frequent nosebleeds.
* Blood in the urine.
* Bleeding after dental work.

**Laboratory Findings**

Similar to hemophilia A

•normal Bleeding time

•prolonge Clotting time

• normal Platelets count.

•normal Prothrombin time

• APTT is Increased.

• Factor IX is decreased.

**ANSNO 5:-**

**Hemolytic uremic syndrome *(HUS)***

* It is a abnormal premature destruction of red blood cells.
* damaged red blood cells start to clog the filtering system in the kidneys,
* which cause the life-threatening kidney failure
* They associated with hemolytic uremic syndrome.
* Most cases of this condition develop in children after two to 14 days of bleedy diarrhea
* Infection coused by a certain strain of Escherichia coli
* Adults also may develop this condition after an E. coli infection,
* but the cause may be certain medications,

TYPES:-

**Typical**

* Infection coused by Shiga toxin
* Toxin producing bacteria *E coli*
* Mostly often with bloody diarrhea

**Atypical**

* It is a rare condition
* Also known is non–Stx-HUS
* The familial form is associated with genetic abnormalities.

**Stx-associated HUS**

* After ingestion, Stx– *E coli* closely adheres to the epithelial cells of the gut mucosa
* Shiga Toxin is transfer by PMNs in the blood,
* because Stx r completely binds to PMNs
* receptor expressed on glomerular endothelial cells
* Infiltrates of inflammatory cells and production of cytokines such
* and tumor necrosis factor contribute to the cytotoxic damage
* Endothelial damage release
	+ prothrombotic-,
	+ vasoactive-,
	+ and platelet-aggregating substances
* Erythrocyte damage primarily occurs in the renal microvasculature.
* toxin attach to endothelial cells
* they exerts a direct toxic effect.
* Toxin has two subunits.

 **B subunits**

* toxin bind to plasma membrane Gb3 receptors

 **A subunits**

* toxin inhibits protein synthesis at the ribosomal level,
* and they leads to cell death.

 **ANS NO :-4**

* **Von willebrand disease**
* first described in 1926 by a Finnish Dr. Erik von Willebrand.
* They estimated to affect 1 in 100 individuals.
* it is the most common genetic bleeding disorder.
* disease pattern is autosomal dominant
* in this disease Males and females are affected equally
* **Functions :**
* it is bind to particular factor VIII and
* Factor VIII is bound to vWF
* That Factor is inactive in circulation;
* they degrades rapidly when not bound to vWF.
* Factor VIII released from vWF
* by the action of thrombin
* They play important role in platelet adhesion to wound sites.
* vWF binds to collagen,

**Pathophysiology**

 There are two forms of VWD:

1. **Hereditary vWD**

 Quantitative :-

* Type I
* Type II
* Type III
* Platelets type

2. Acquired vWD

* Antibodies against vwf

**Clinical Features**

* mild bleeding
* bleeding from mucous membranes
* Excessive bleeding from wounds
* Increase Menses bleeding
	+ Normal Hemoglobin
	+ Normal Hematocrit
	+ Normal Platelet count
	+ decrease type 2b
	+ Normal Prothrombin time
	+ increase thromboplastin time increase
	+ normal Fibrinogen
	+ prolong Bleeding time
	+ Prolong Clotting time
	+ Thrombin time Normal

 **ThE EnD................**