ALTERATIONS OF HEMOSTASIS

(CHAPTER 22)

QUESTION 1

Describe disorders of primary hemostasis.

Primary hemostasis refers to the initial phase of **platelet aggregation** that occurs during **early** clot formation.

Primary hemostatic disorders cause a disruption in the formation of the platelet plug, either due to **decreased** platelets or **failure** to aggregate.

Some examples of primary hemostatic disorders:

- **aplastic anemia** – decreased production of platelets & blood cells
- **immune thrombocytopenic purpura** (ITP) – immune system attacks platelets
- **von Willebrand Disease** – lack of "glue" to adhere clot to site

QUESTION 2

Describe disorders of secondary hemostasis.

Secondary hemostasis refers to the second phase of clot formation where the platelet plug is **strengthened** with fibrin and turned into a full clot.

Secondary hemostatic disorders, usually associated with a deficit of one or more **clotting factors**.

Some examples of secondary hemostatic disorders:

- **Hemophilia** (both A and B)
- Underproduction due to **vitamin K** deficiency
- Underproduction of clotting factors due to **liver failure**

QUESTION 3

Describe von Willebrand disease.

von Willebrand Factor (vWF) is a glycoprotein in the blood which plays an important role in **platelet adhesion** during the early stages of clot formation.

The underproduction of vWF is known as **von Willebrand Disease** (vWD,) which is also hereditary, but unlike hemophilia, affects men and women equally.

Symptoms of vWD are similar to those of hemophilia, although the most common forms of vWD tend to have only mild symptoms such as bruising and epistaxis (nosebleeds.)

Internal bleeding, such as bleeding into the joints, is uncommon in most forms of von Willebrand disease but can occur in the more severe types.

QUESTION 4

Describe the consequences of thrombocytopenia.

Thrombocytopenia, or a decreased number of platelets, can cause a variety of symptoms: but most of them are **bleeding**.

These include **petechiae** and **purpura** (small, dot-like bruises,) as well as **expstaxis** (nosebleeds,) **hematochezia**, **hematuria**, increased **menstrual flow**, and **slowed clotting**.

QUESTION 5

List some causes of thrombocytopenia.

Thrombocytopenia can have a few different causes.

First, **production** could be decreased, as in aplastic anemia, leukemia, nutritional deficiencies, etc.

Second, platelets could be prematurely **destroyed**, as occurs in immune thrombocytopenic purpura (ITP.)

Third, platelets could be **used up** by improper clotting, as in diffuse intravascular coagulation (DIC) and thrombotic thrombocytopenic purpura (TTP.)

QUESTION 6

Compare hemophilia A and hemophilia B.

Hemophilia is a group of **hereditary bleeding disorders** due to deficiency of clotting factors, which result in impaired coagulation.

Mild symptoms include **bruising**, **epistaxis** (nosebleeds,) increased **bleeding time**, etc.

More pronounced symptoms include severe **internal bleeding** and **hemarthrosis** (bleeding into the joint space.)

Hemophilia **A** is linked to impaired production of clotting factor VIII (anti-hemophilic factor/AHF,) whereas hemophilia **B** is due to impaired production of factor IX (Christmas factor.)

Hemophilia A is about **6 times** as common as hemophilia B. Both occur **almost exclusively** in male patients.

QUESTION 7

Why is sickle cell anemia called an anemia? Why is it a hyper-coagulatory disease?

Sickle cell anemia is a recessive genetic defect in the DNA coding for hemoglobin, resulting in RBCs that collapse into a "moon" or "sickle" shape when they become deoxygenated. It occurs almost exclusively in **black patients**.

These malformed cells can become tangled with each other and stuck in the arteries, leading to sudden episodes of pain referred to as **sickle cell crisis**.

Once they malfunction, these RBCs are targeted for hemolysis and broken down, resulting in a **hemolytic** anemia.

The reason for **hypercoagulation** in sickle cell disease is not fully understood, but one contributing factor is blood **stasis** due to the obstruction from tangles of sickled RBCs.

QUESTION 8

What is disseminated intravascular coagulation (DIC?)
Why can bleeding occur in this state?

Disseminated intravascular coagulation (DIC) is an often-acute state of impaired coagulation caused by platelets and clotting factors being **used up** to create small, unnecessary blood clots all over the body.

It's often caused by **sepsis** (systemic infection,) and results in tiny clots forming all over the body, which deplete the body's stores of materials for coagulation.

QUESTION 9

Describe thrombocythemia. How is it related to von Willebrand factor?

Thrombocythemia (or, more commonly **thrombocytosis**) is the **opposite** of thrombocytopenia—having **too many** platelets in the blood.

Paradoxically, this can actually cause a **decrease** in coagulation, as there is no longer enough vWF to adhere the platelets to each other.

You can think of this as being too many "bricks" and not enough "mortar." Without enough vWF to stick the larger number of platelets together, there is difficulty forming a complete **platelet plug**.