

HEMATOLOGY

Primary Care Paramedicine

Module: 15

Section: 02



- Introduction
- General assessment and management
- Specific patient problems
- Blood products and typing
- Transfusion reactions

- Hematology
 - Study of blood and blood forming organs
- Disorders common
 - Rarely the primary problem
 - Genetic or acquired
 - Presentations tend not to point to a specific disease process

- History
 - Hematological disorders are rarely the chief complaint
- Physical assessment
 - Nervous system
 - Skin signs
 - Lymphatic signs
 - Gastrointestinal signs
 - Musculoskeletal signs
 - Cardiorespiratory signs
 - Genitourinary signs

- Maintain ABCs
 - Provide high-flow oxygen or assist ventilations as indicated
 - Consider volume replacement
- Monitor cardiac rhythm and vital signs
 - Treat rhythm disturbances
- Provide reassurance, comfort care, and transport.

Hematological Emergencies

SPECIFIC PATIENT PROBLEMS

- Diseases of the erythrocytes
- Diseases of the leukocytes
- Diseases of the platelets
- Blood clotting disorders
- Hemolytic disease

- Polycythemia
- Anemia
- Impairment of red cell function

- Polycythemia
 - Excess of red blood cells
 - Uncommon but several diseases can cause it
- Anemia
 - Lack of red blood cells or inadequate hemoglobin
- Impairment of red cell function
 - Problems with hemoglobin structure and function
 - Problems with red blood cell membrane

- Overproduction of erythrocytes
 - Primary: due to inherent problems in the process of RBC production
 - Secondary: a response to other factors or underlying conditions that promote RBC production (dehydration, hypoxia etc.)
 - Uncommon but several diseases can cause it
- Results in bleeding abnormalities:
 - Epistaxis, spontaneous bruising, GI bleeding.

Signs and Symptoms

- Can be nonexistent or minimal
- Weakness/fatigue
- Headache
- Itching
- Bruising
- Joint pain
- ABD pain

Management

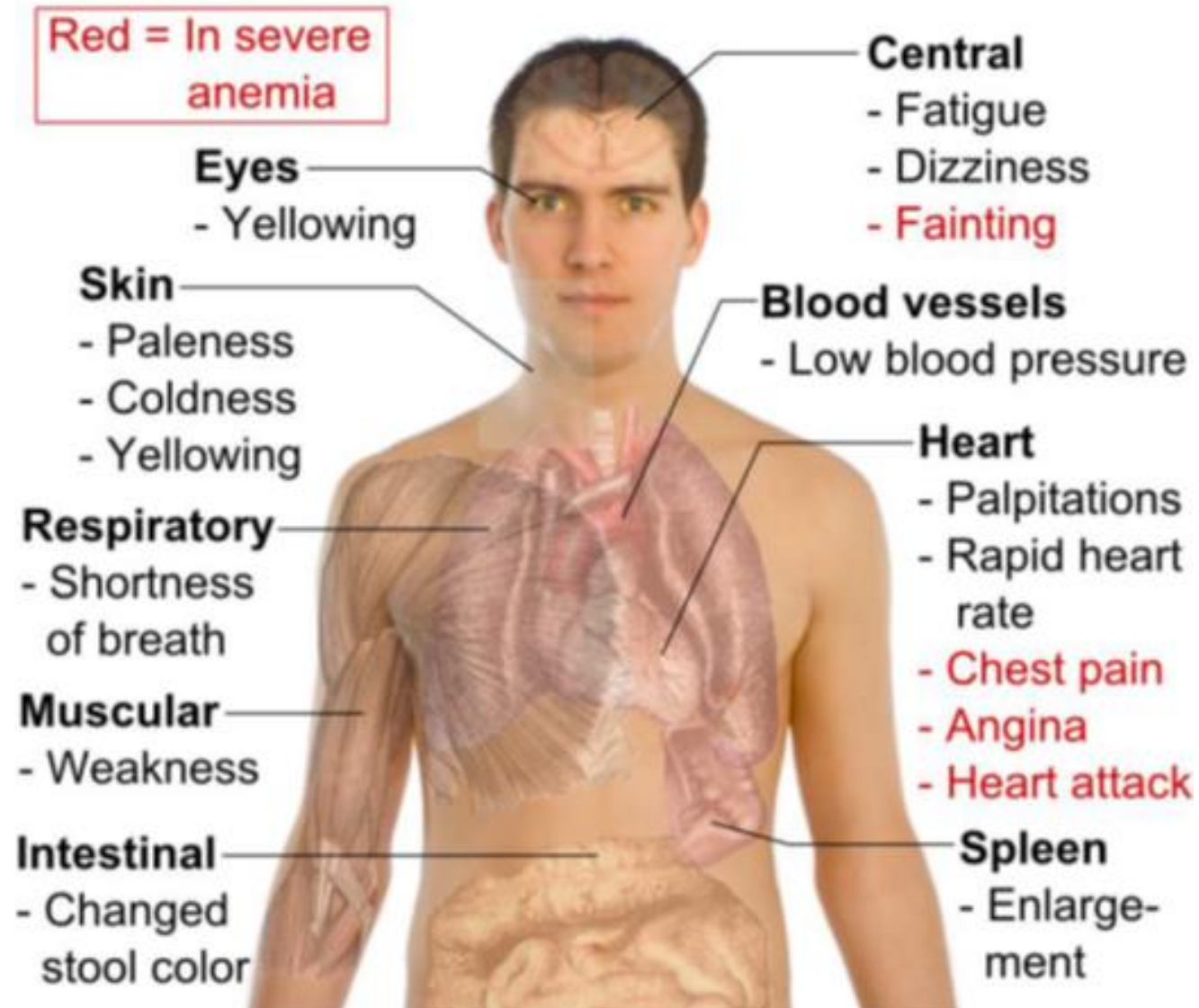
- Follow general treatment guidelines

- A decreased number of RBC's or low amount of hemoglobin
- Leads to a poor oxygen carrying capacity of blood
- Various conditions have anemia as a symptom

- Idiopathic
 - No clear cause (may be poor nutrition or stress)
- Nutritional
 - Iron deficiency
 - Affects women more than men
 - Vitamin B12 deficiency
 - Needed for RBC production (known as pernicious anemia and can also cause neurological symptoms of paralysis and cognitive problems);
- Hemorrhagic
 - Due to blood loss; typically a slow leak (ulcers or kidney disease)
- Aplastic
 - Bone marrow disorder; poor to no production of formed elements
 - Typically autoimmune; (myelodysplastic anemia is production of abnormal cells and may be a indication of cancer development)
- Secondary
 - A complication of a disorder, such as: ulcers (blood loss), kidney disease (leaking RBC's or low production of EPO), hepatitis (not breaking down RBC's therefore less products available for RBC production), acute infection (especially pneumonia), cancer (leukemia, myeloma, or lymphoma)

Table 35-2 TYPES OF ANEMIA

Cause	Type	Pathophysiology
Inadequate production of red blood cells	Aplastic	Failure to produce red blood cells
	Iron deficiency	Iron is primary component of hemoglobin
	Pernicious	Vitamin B ₁₂ is necessary for correct red blood cell division during its development
	Sickle cell	Genetic alteration causes production of a hemoglobin that changes shape of red blood cell to a C, or sickle, in low oxygen states
Increased red blood cell destruction	Hemolytic	Body destroys red blood cells at greater rate than production; red blood cell parts interfere with blood flow
Blood cell loss or dilution	Chronic disease	Hemorrhage leads to cell loss while excessive fluid leads to a dilution of red blood cell concentration



Signs and Symptoms

- May not be present until the body is stressed
- Differentiate chronic anemia from acute episode

Management

- Treat signs and symptoms
 - Maximize oxygenation and limit blood loss
 - Establish IV therapy if indicated

- Body's principle defence system
 - Leukopenia/neutropenia
 - Leukocytosis

- Leukopenia/neutropenia
 - Too few white blood cells or neutrophils.
 - Bacterial infections are a real risk.
 - Follow general treatment guidelines and provide supportive care.
- Leukocytosis
 - An increase in the number of circulating white blood cells, often due to infection.
 - Leukemoid reaction

- Affect platelets or clotting
- Hereditary or acquired
- Thrombocytosis
 - Increased platelets
- Thrombocytopenia
 - Reduced platelets

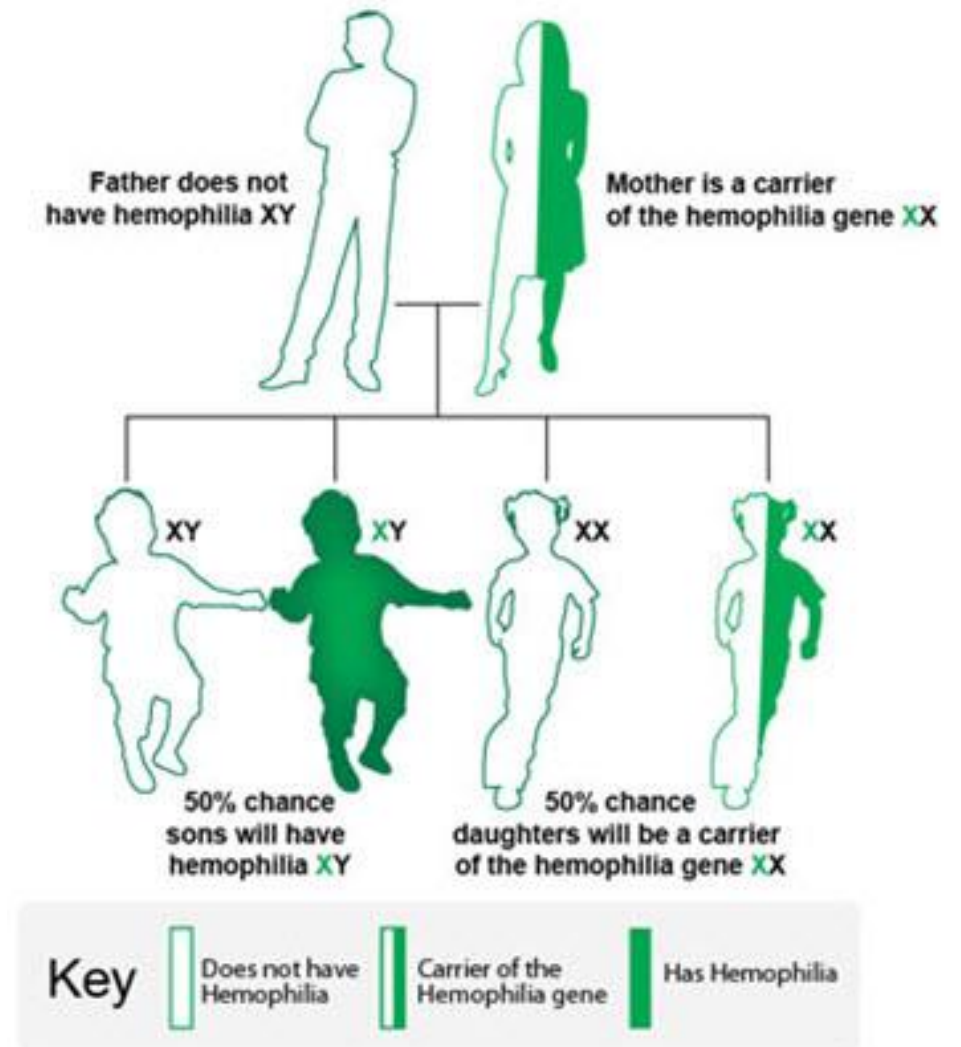
- Increased platelets
 - Usually due to increased production
 - Also seen in polycythemia
- Often a complication of:
 - Malignant diseases
 - Hemolytic diseases
 - Hemorrhagic and inflammatory diseases
- Asymptomatic
 - Management usually supportive

- Abnormal decrease in number of platelets
 - Decreased production
 - Sequestration of spleen
 - Destruction of platelets
- Causal factors
 - Drug induced
 - Idiopathic thrombocytopenia purpura
- Asymptomatic
 - Management usually supportive

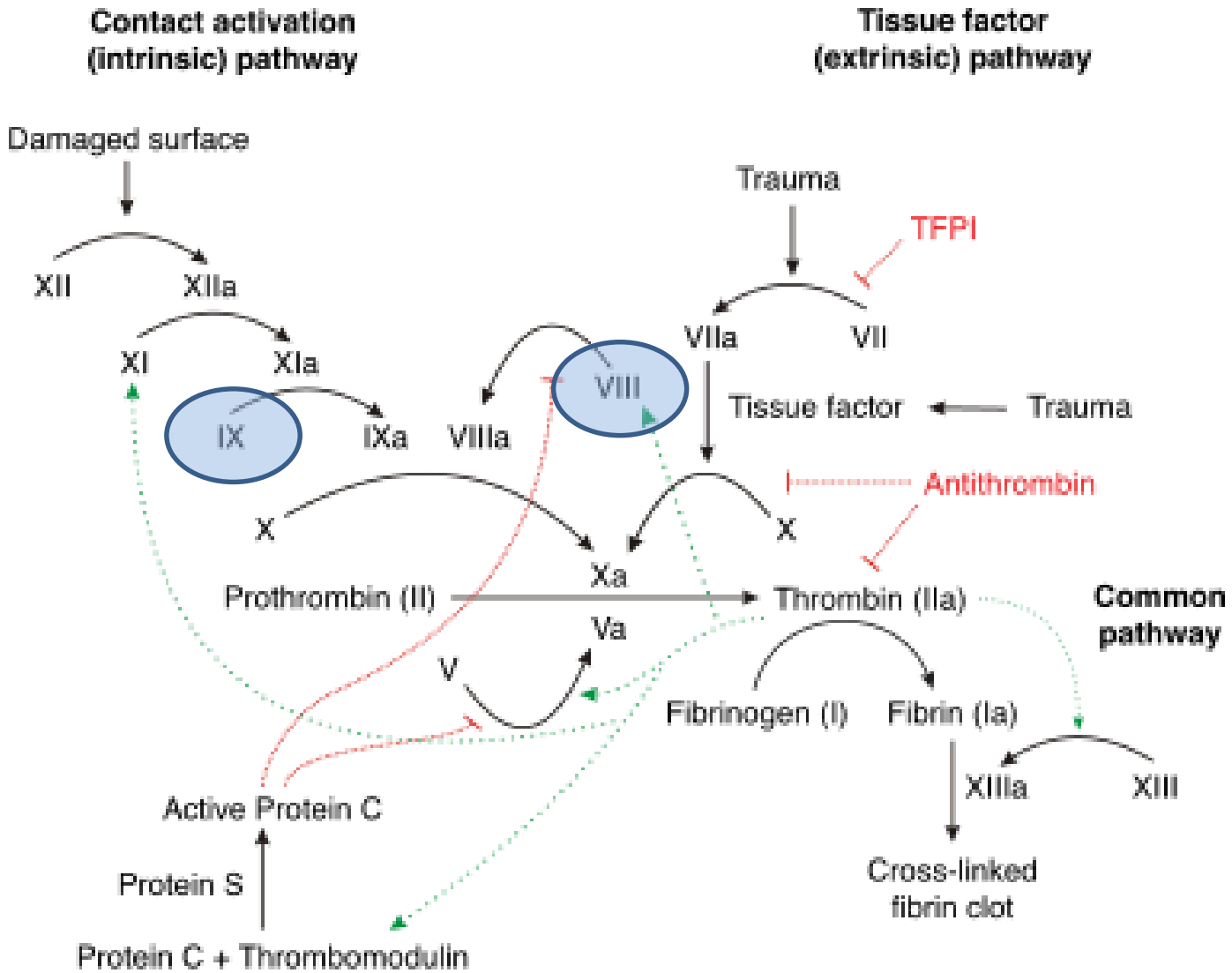
- Hemophilia
- Disseminated Intravascular Coagulopathy

- Genetic disorders characterized by lack of, or insufficiency of, certain clotting proteins
 - Leads to difficulty clotting blood and therefore excessive bleeding from even minor wounds (internally or externally)
 - Can be mild, moderate or severe depending on the normal percentage of clotting factors present in the blood
 - Severe hemophilia will present with less than 1% of clotting factors in the blood, and is the most common type of hemophilia

- Deficiency is a sex-linked, inherited disorder.
 - Defective gene is carried on the X chromosome.
 - But it is much more common in males



- Three main types:
 - Type A Hemophilia
 - Most common
 - Deficiency of clotting factor VIII
 - Type B Hemophilia
 - AKA Christmas disease
 - Deficiency of clotting factor IX
 - ~15% of all cases



Injury Occurs

- 1 Injury to blood vessel results in bleeding.
- 2 Vessel constricts and clotting factors are activated.



Normal

- 3 Along with other substances, clotting factor VIII causes a strong platelet plug to form.
- 4 A stable fibrin clot forms over the platelet plug as a final seal on the injury, and the bleeding stops.



Hemophilia A

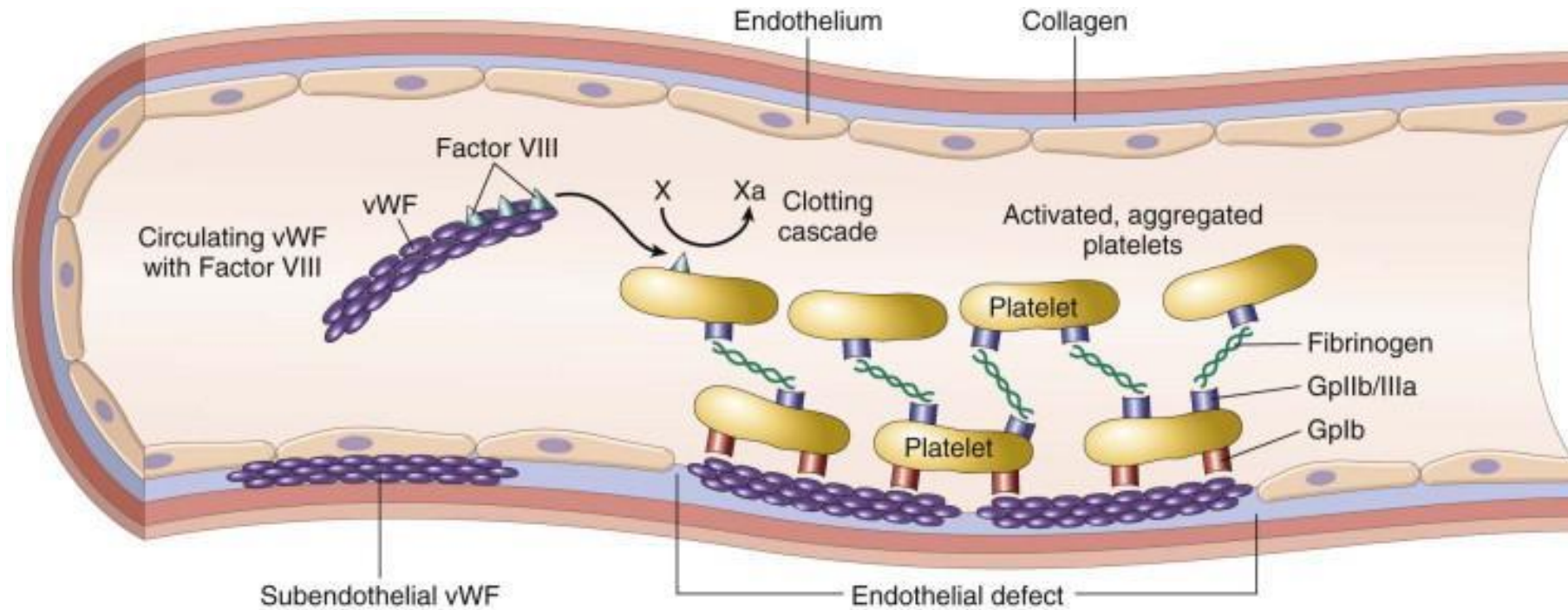
- 3 Lack of clotting factor VIII causes a weak platelet plug to form.
- 4 Incomplete and/or delayed fibrin clot allows bleeding to continue.



Management

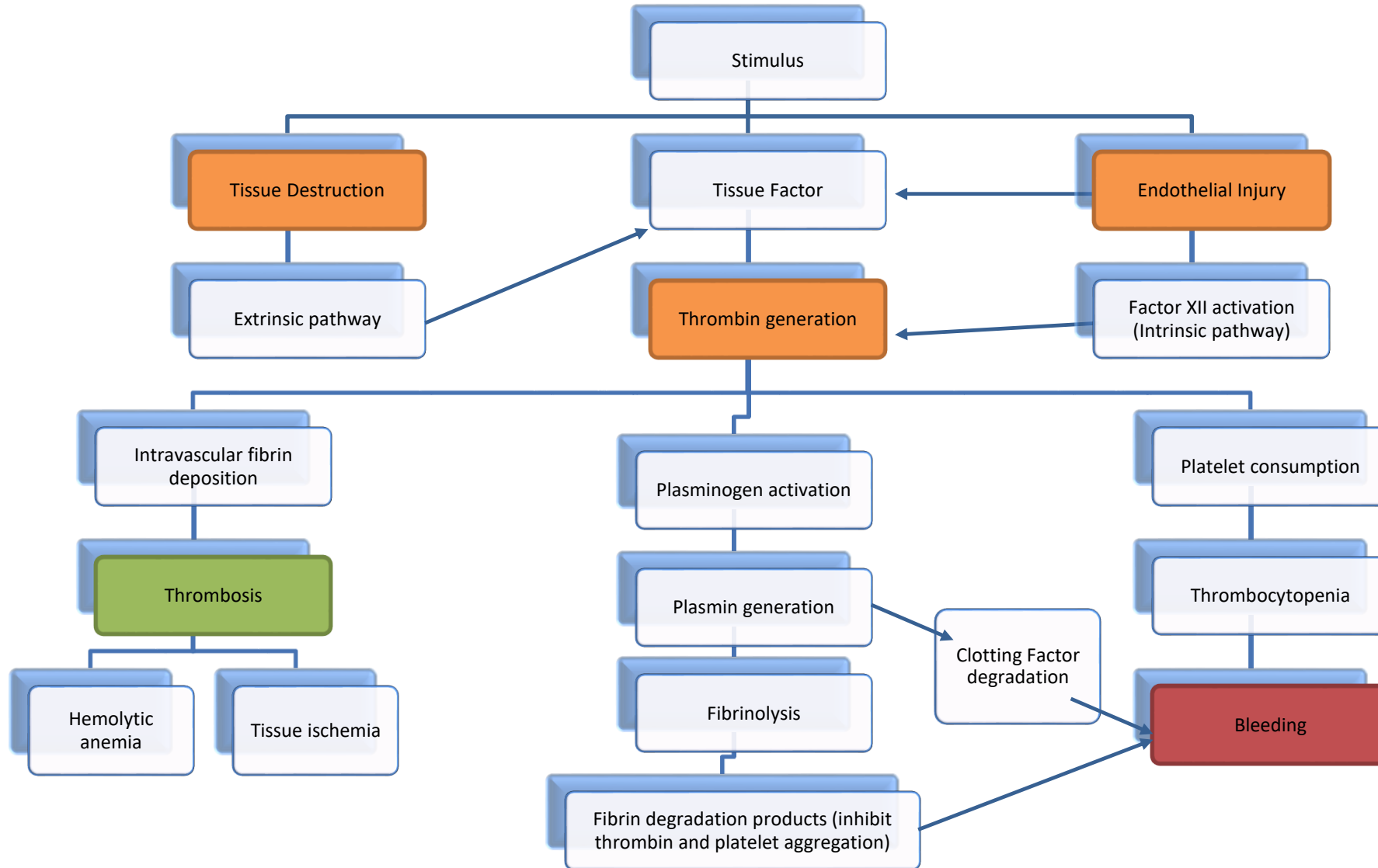
- Treat the patient similarly to others.
- Administer supplemental oxygen.
- Establish IV access.
- Be alert for recurrent or prolonged bleeding.
- Prevent additional trauma.

- Still inherited (but not linked to X chromosome)
 - Not a problem with production of clotting factors
- Endothelial lining of blood vessels does not produce Von Willebrand factor
 - VWF is typically secreted by damaged endothelial cells to improve platelet adhesion
- Generally results in excessive bleeding
- Generally is not serious
 - Provide supportive care



- System activation of coagulation cascade.
 - Widespread thrombosis
 - Occasionally end-organ ischemia
- Causal factors
 - Sepsis, hypotension, OB complications
 - Severe tissue or brain injury
 - Cancer, and major hemolytic reactions.

Disseminated Intravascular Coagulation

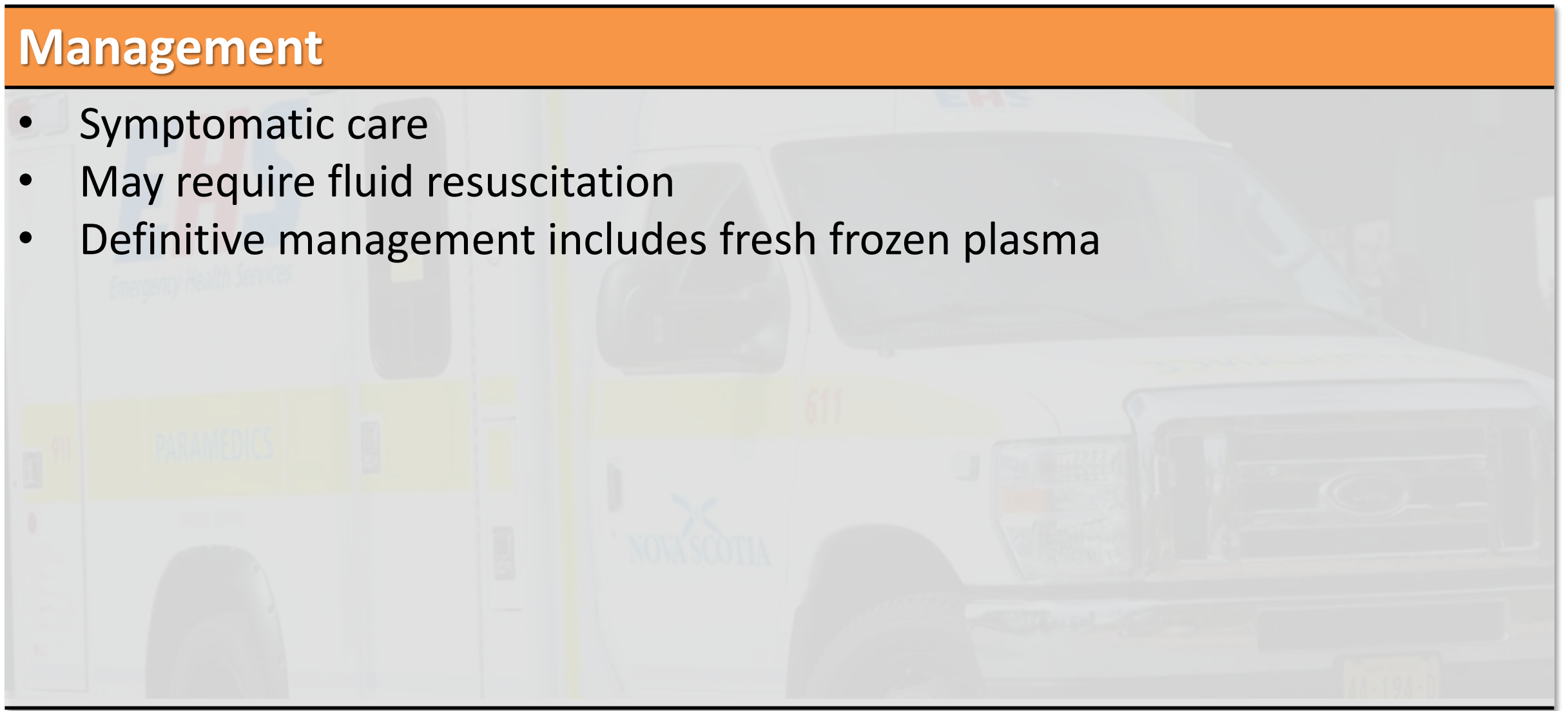


Disseminated Intravascular Coagulation (DIC)

- **Bleeding**
 - Spontaneous bruising
 - Petechiae
 - Prolonged bleeding from venepuncture sites, arterial lines, etc.
 - Bleeding into gastrointestinal tract or lungs
 - Secondary bleeding after surgery
 - Coma (intracerebral bleeding)
- **Clotting**
 - Acute renal failure (ischemia of renal cortex)
 - Venous thromboembolism
 - Skin necrosis or gangrene
 - Liver failure (due to infection and hypotension)
 - Coma (cerebral infarction)
- **Shock**
 - Due to underlying disease together with disseminated intravascular coagulation
- **Central nervous system**
 - Transient neurological symptoms and signs
 - Coma
 - Delirium
- **Lungs**
 - Transient hypoxaemia
 - Pulmonary haemorrhage
 - Adult respiratory distress syndrome

Management

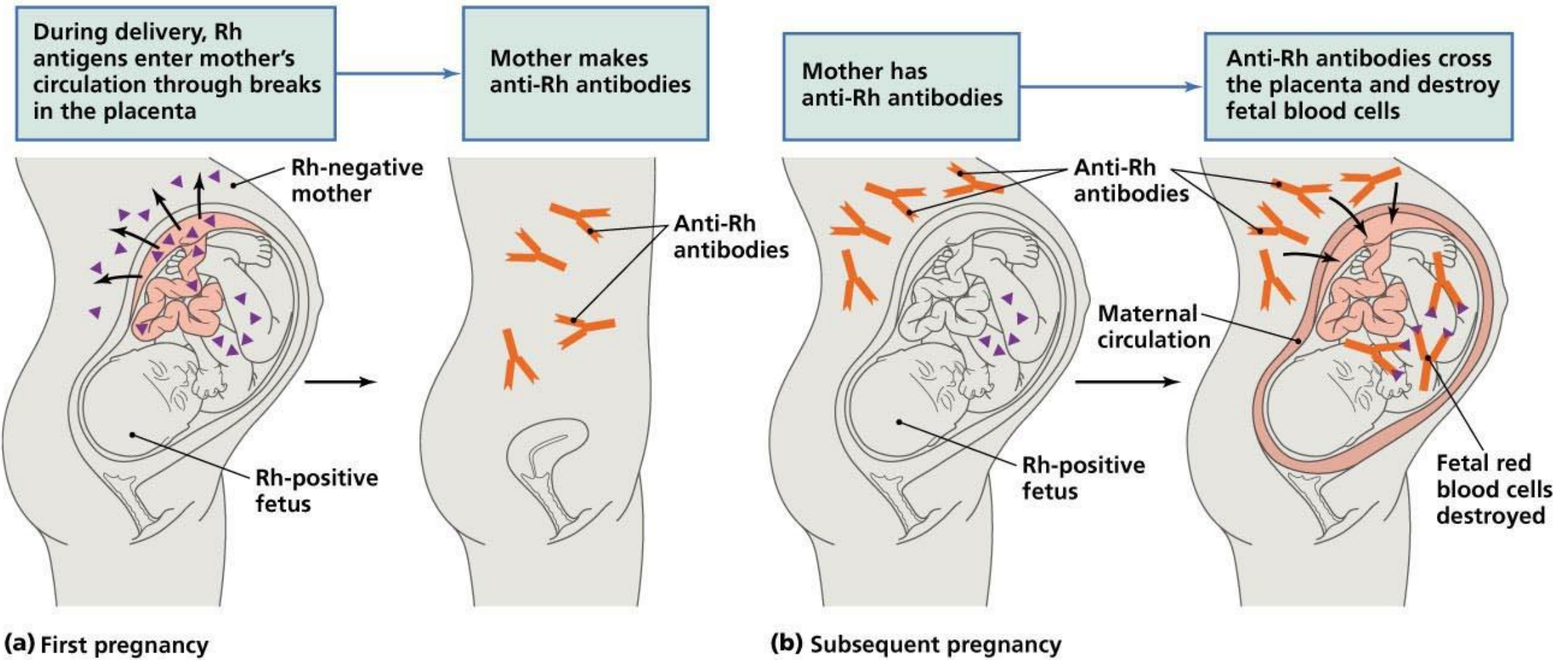
- Symptomatic care
- May require fluid resuscitation
- Definitive management includes fresh frozen plasma



- Rh⁻ Mother and Rh⁺ Father
- Baby inherits father's with Rh⁺
- Mother develops anti-Rh agglutinins
- Next pregnancy with Rh⁺ baby, mother's anti-Rh agglutinins will be activated and cause erythroblastosis fetalis
 - Hemolysis of baby's RBC's occurs



Hemolytic Disease



(a) First pregnancy

(b) Subsequent pregnancy

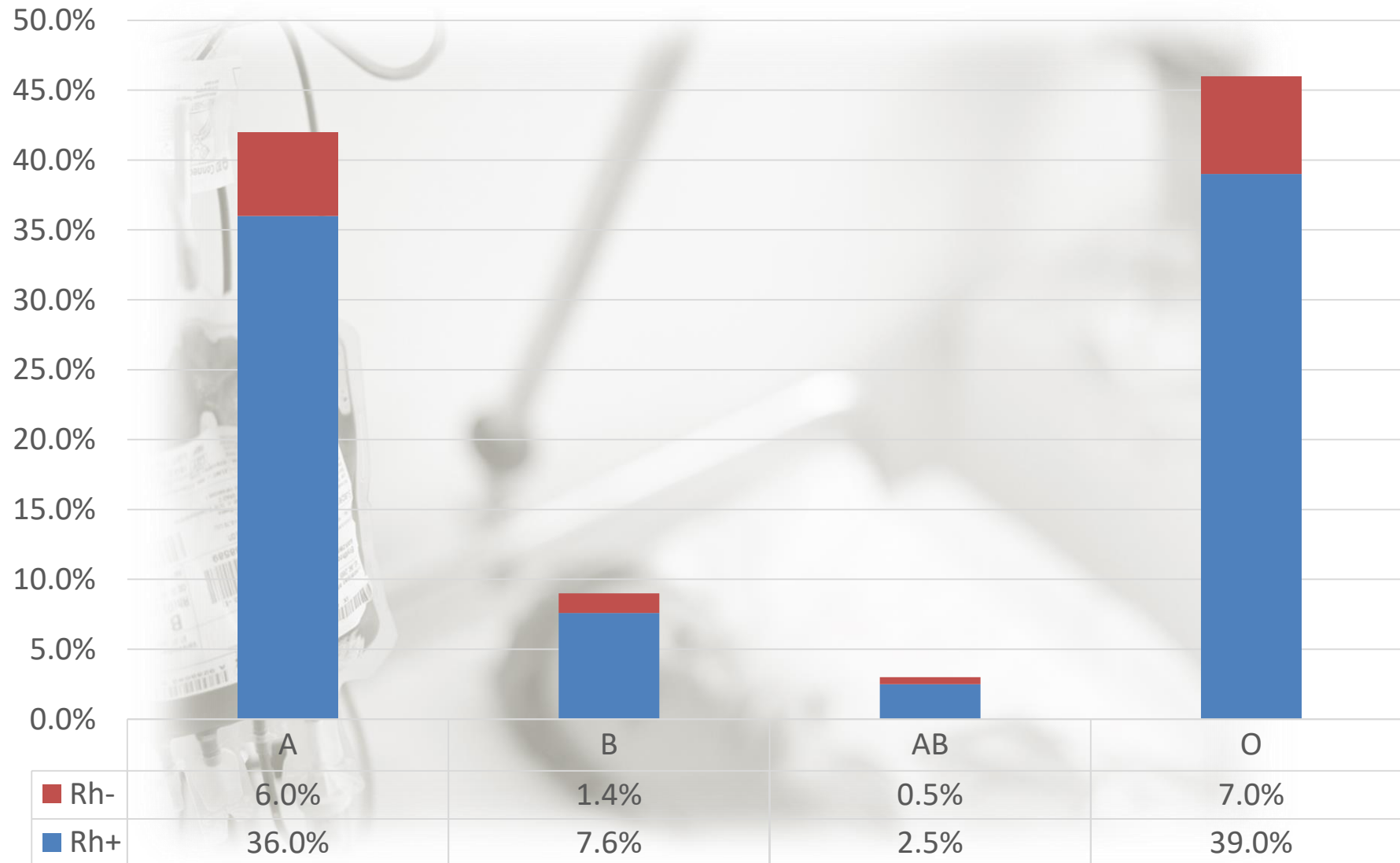
- This decrease O₂ carrying capability and increase bilirubin levels
- This may lead to brain damage
- Rh⁻ Mothers with Rh⁺ babies are given medication to keep her system from forming the anti-Rh antibodies

- Baby will soon replace Rh⁺ with Rh⁻ if hemolytic disease develops
- Rh⁻ depletes through normal life span and as the Rh⁻ transfusion is depleted Rh⁺ is reintroduced through the bone marrow

Hematological Emergencies

BLOOD PRODUCTS

- Transfusion
 - Transplantation of blood or a component of blood
- Antigen
 - Surface proteins that distinguish self from not self
 - Must be compatible for a transfusion to be successful
- Cross-matching
 - Checking donor and recipient samples for compatibility

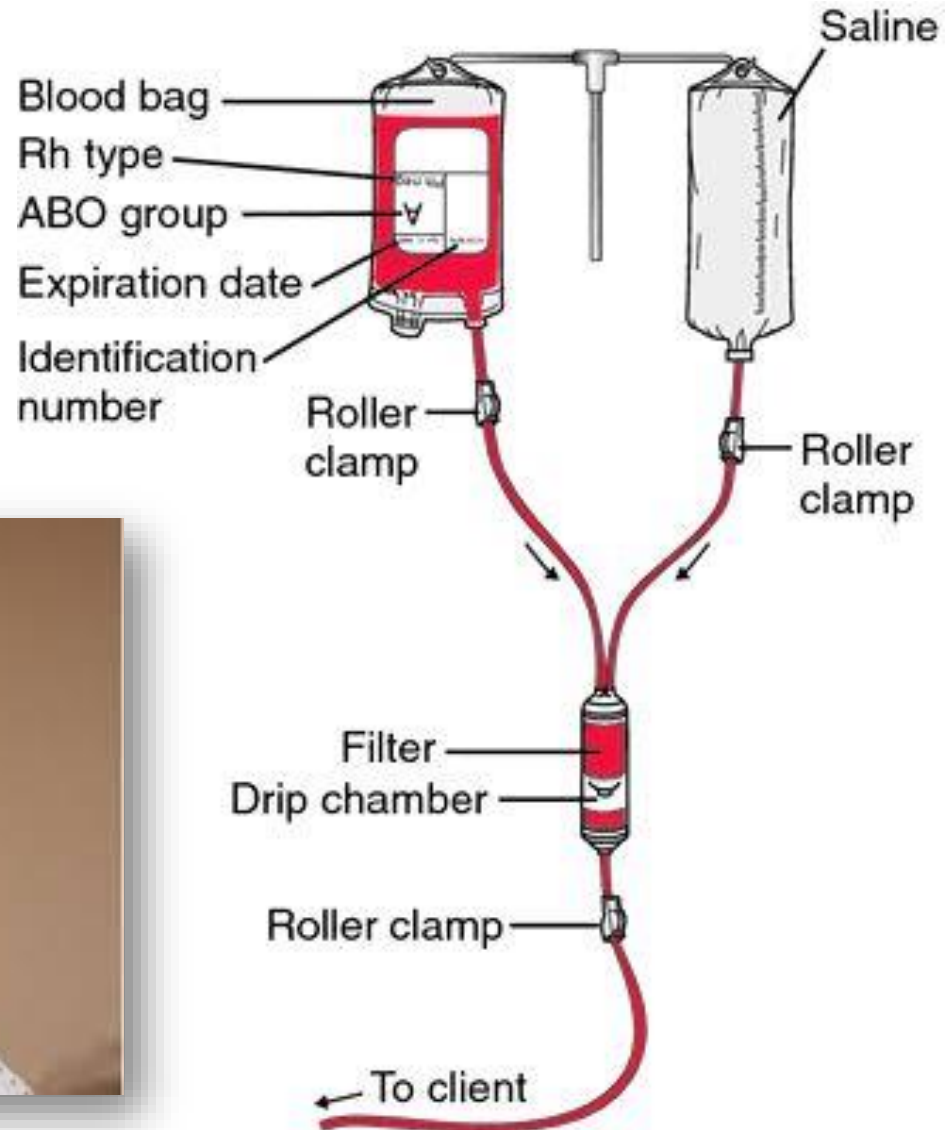


Compatibility of
**BLOOD
 TYPES**

		Donor							
		O-	O+	B-	B+	A-	A+	AB-	AB+
Recipient	AB+	✓	✓	✓	✓	✓	✓	✓	✓
	AB-	✓		✓		✓		✓	
	A+	✓	✓			✓	✓		
	A-	✓				✓			
	B+	✓	✓	✓	✓				
	B-	✓		✓					
	O+	✓	✓						
	O-	✓							



Type	Contents	Use
Whole Blood	All cells, platelets, clotting factors, and plasma	Replace blood loss from hemorrhage
Packed Red Blood Cells (PRBCs)	Red blood cells and some plasma	Replaces RBCs in anemic patients
Platelets	Thrombocytes and some plasma	Replace platelets in patients with thrombocytopenia
Fresh Frozen Plasma (FFP)	Plasma, a combination of fluids, clotting factors, and proteins	Replace volume in a burn patient or in a hypovolemia secondary to low oncotic pressure
Clotting Factors	Specific clotting factors needed for coagulation	Replaces factors missing due to inadequate production (hemophilia)



8 Rights of Transfusion Administration

8 RIGHTS:

- Product
- Patient
- Dose
- Time
- Reason
- Site
- Documentation
- Response



- Hemolytic reaction
- Febrile non-hemolytic reaction
- Allergic reaction
- TRALI (Transfusion Related Acute Lung Injury)
- TACO (Transfusion Associated Circulatory Overload)

Identification and Management

Complication	Signs and Symptoms	Treatment	Notes
Febrile Transfusion Reaction	<ul style="list-style-type: none"> • 1 degree rise in temp • May have chills, malaise 	<ul style="list-style-type: none"> • Supportive • Acetaminophen 	Most Common
Hemolytic Transfusion Reaction	<ul style="list-style-type: none"> • Fever, chills, pain at the site of reaction, N/V, shock, dark urine 	<ul style="list-style-type: none"> • STOP the transfusion • IV Fluid bolus 	Worst reaction. ABO Incompatibility
Allergic Reaction	<ul style="list-style-type: none"> • Urticaria, pruritis, hives • Anaphylaxis is rare 	<ul style="list-style-type: none"> • Symptomatic <ul style="list-style-type: none"> • Antihistamines • Treat anaphylaxis as per protocol • Do NOT stop the transfusion 	They are not actually allergic the blood but secondary to antibodies in the blood
TRALI	<ul style="list-style-type: none"> • Dyspnea, hypoxemia, ARDS 	<ul style="list-style-type: none"> • STOP the transfusion • Airway control, supportive care 	Most common cause of death associated with transfusions
TACO	<ul style="list-style-type: none"> • Dyspnea, edema 	<ul style="list-style-type: none"> • Give blood slowly 	Often occurs in the elderly and chronically anemic

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- Specific patient problems
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- Transfusion reactions