

A 6-year-old boy is brought to the emergency department with fever, purulent nasal discharge, and epistaxis. He has had daily fever and persistent nasal discharge for the past 10 days without improvement. The parents have had difficulty controlling his intermittent epistaxis, which began 2 days ago. The patient has also had easy bruising for the past 2 months. His medical history is significant for several recurrent pulmonary infections requiring hospitalization for parenteral antibiotic therapy, and previous sweat chloride testing was positive. The boy's father recently lost his job, and his parents report financial difficulties obtaining the patient's prescribed medications. Coagulation study results are as follows:

Prothrombin time	25 sec
Activated partial thromboplastin time	35 sec

Which of the following coagulation factors is most likely to be abnormal in this patient?

- ☐ A. Factor V
- ☐ B. Factor VII
- ☐ C. Factor VIII
- ☐ D. Factor XII
- ☐ E. Fibrinogen

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Prothrombin time	25 sec
Activated partial thromboplastin time	35 sec

Which of the following coagulation factors is most likely to be abnormal in this patient?

- ☐ A. Factor V [5%]
- ☒ B. Factor VII [75%]
- ☐ C. Factor VIII [14%]
- ☐ D. Factor XII [2%]
- ☐ E. Fibrinogen [3%]

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Explanation:

User Id: [REDACTED]

Vitamin K deficiency	
Risk factors	<ul style="list-style-type: none">• Inadequate dietary intake (eg, malnutrition)• Disorders of fat malabsorption<ul style="list-style-type: none">◦ Cystic fibrosis◦ Biliary atresia• Disorders of intestinal inflammation<ul style="list-style-type: none">◦ Celiac disease

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Vitamin K deficiency	
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Clinical features	<ul style="list-style-type: none"> • Easy bruising • Mucosal bleeding • Gastrointestinal bleeding
Laboratory findings	<ul style="list-style-type: none"> • ↑ PT & INR • Normal aPTT (unless severe deficiency)

aPTT = activated partial thromboplastin time; PT = prothrombin time.

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This patient with acute sinusitis, recurrent respiratory infections, and positive sweat test has **cystic fibrosis** (CF). Bleeding diathesis (eg, **easy bruising, mucosal bleeding, epistaxis**) in patients with CF is typically from **vitamin K deficiency**. Vitamin K normally acts as an important cofactor in the hepatic activation of coagulation **factors II, VII, IX, and X**, as well as proteins C and S. Vitamin K deficiency results in low levels of activated vitamin K-dependent coagulation factors and prolonged prothrombin time (PT) due to predominant deficiency of activated factor VII. Activated partial thromboplastin time (aPTT) is typically normal, although aPTT can be prolonged with severe vitamin K deficiency.

Normal sources of vitamin K are both exogenous (eg, leafy vegetables) and endogenous (eg, synthesis from gastrointestinal flora). Due to exocrine pancreatic insufficiency, patients with CF are unable to absorb fats and **fat-soluble vitamins** (A, D, E, and K) and require **pancreatic enzyme replacement** and fat-soluble vitamin supplementation. This patient is at increased risk for vitamin K deficiency due to difficulty obtaining

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(Choice A) Factor V is not dependent on vitamin K for activation. Factor V deficiency is rare and leads to prolonged PT and aPTT.

(Choice C) As part of the intrinsic pathway, factor VIII deficiency results in prolongation of aPTT with normal PT. Hemophilia A is an X-linked recessive disorder of factor VIII deficiency.

(Choice D) Factor XII (Hageman factor) is part of the intrinsic coagulation pathway and is not vitamin K dependent. Patients with inherited factor XII deficiency are typically asymptomatic but have prolonged aPTT.

(Choice E) Fibrinogen normally assists in **coagulation via the final common pathway** through conversion to fibrin clots and binding of platelets. Fibrinogen is not dependent on vitamin K for activation. Disorders of fibrinogen include severe liver disease and disseminated intravascular coagulation.

Educational objective:

Patients with cystic fibrosis are at risk for fat-soluble vitamin (A, D, E, and K) deficiency due to poor absorption from pancreatic insufficiency. Vitamin K is an important cofactor in the activation of coagulation factors II, VII, IX, and X, and deficiency leads to easy bruising, mucosal bleeding, and prolonged prothrombin time (PT).

References:

activated vitamin K-dependent coagulation factors and prolonged prothrombin time (PT) due to predominant deficiency of activated factor VII. Activated partial thromboplastin time (aPTT) is typically normal, although aPTT can be prolonged with severe vitamin K deficiency.

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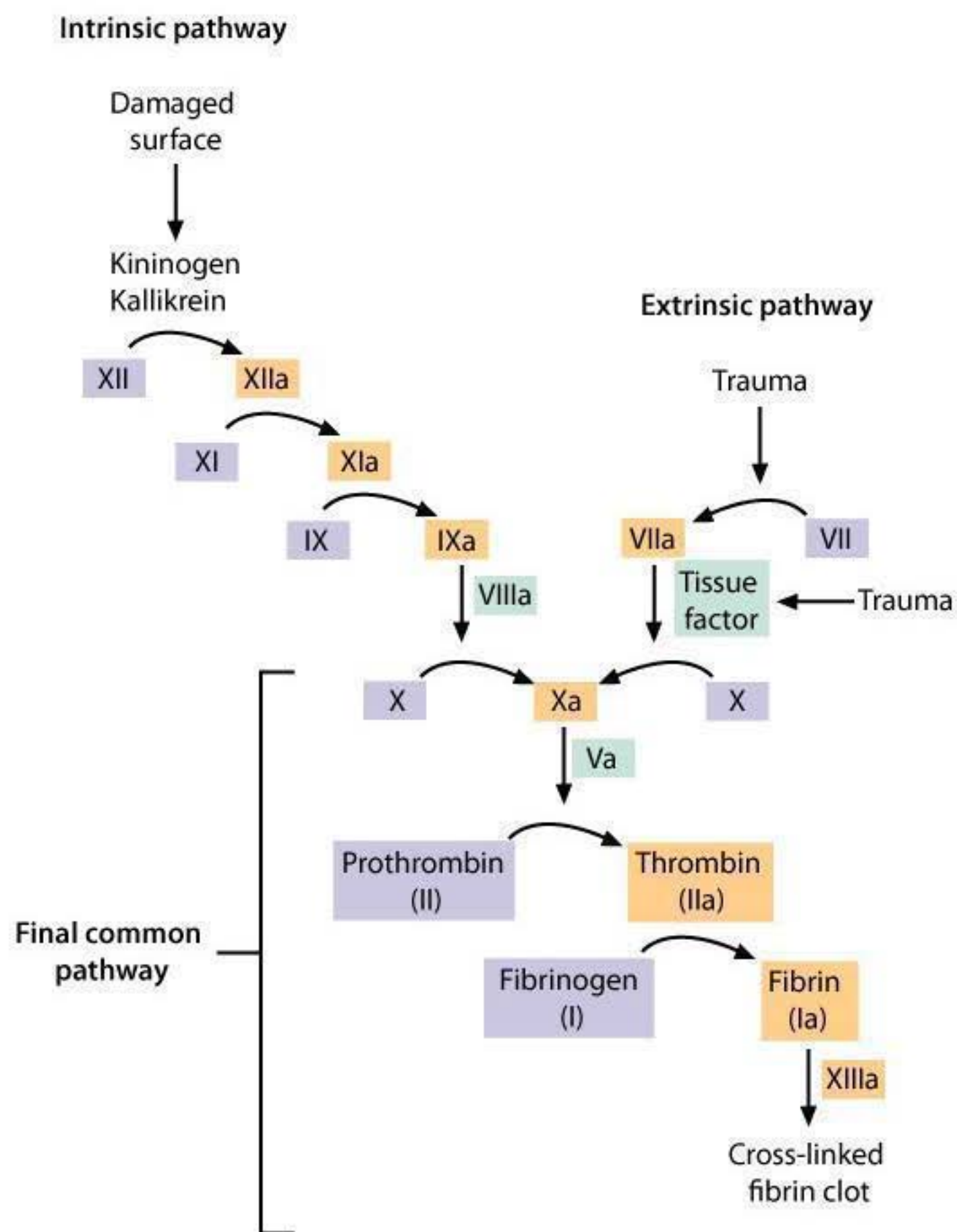
References:

1. [Routine use of daily oral vitamin K to treat infants with cystic fibrosis.](#)
2. [Update in pediatrics: focus on fat-soluble vitamins.](#)
3. [ESPEN-ESPGHAN-ECFS guidelines on nutrition care for infants, children, and adults with cystic fibrosis.](#)

Media Exhibit

ation cascade pathway

Coagulation cascade pathway



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