



Schweizerische Hämophilie-Gesellschaft  
Association Suisse des Hémophiles  
Società Svizzera Emofilia



Journée Romande, Centre Loewenberg, Morat, 25 mai 2019

# Corriger l'hémophilie sans facteur plasmatique?

Lorenzo ALBERIO

# Programme

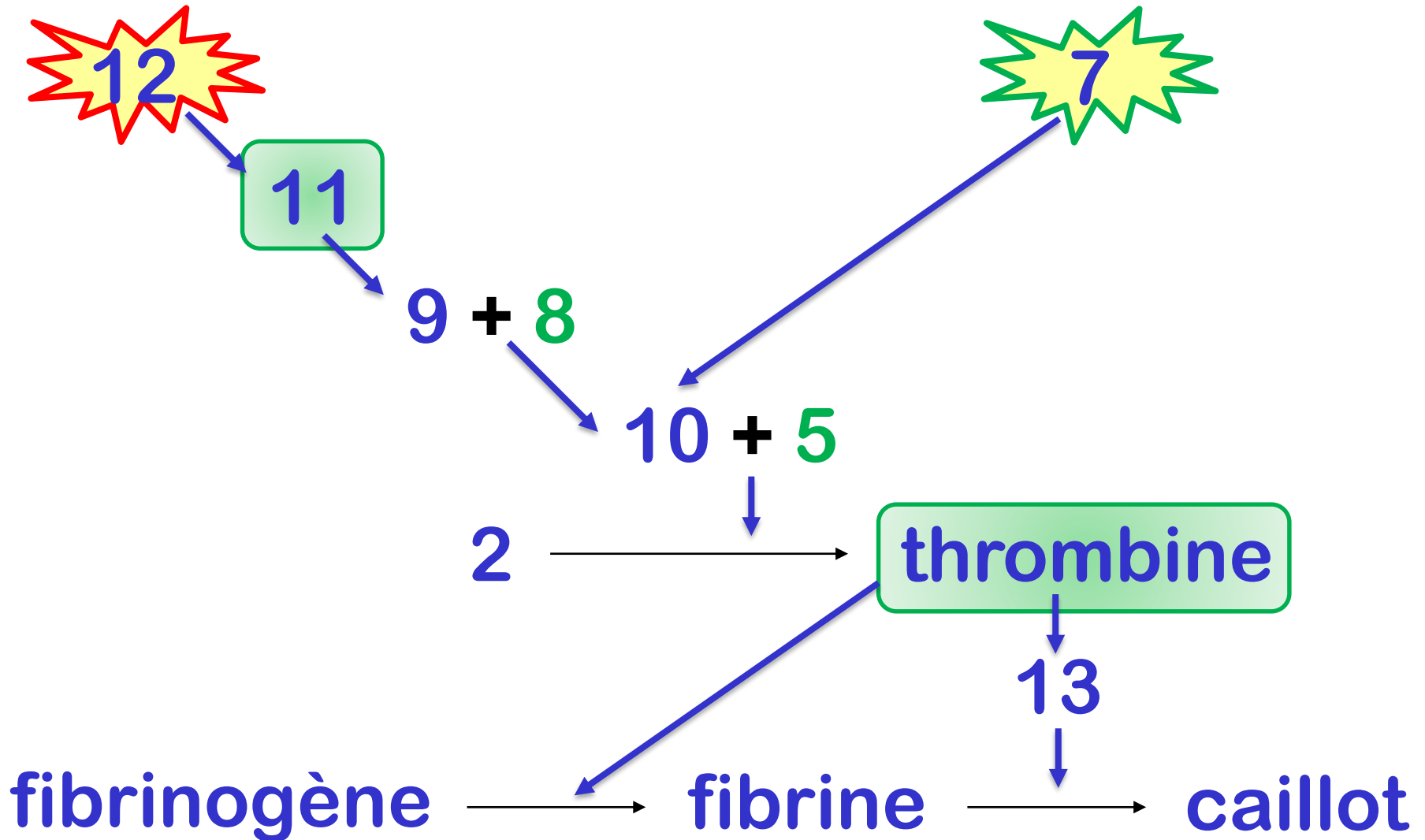
La «cascade» de la coagulation

Les inhibiteurs naturels de la coagulation

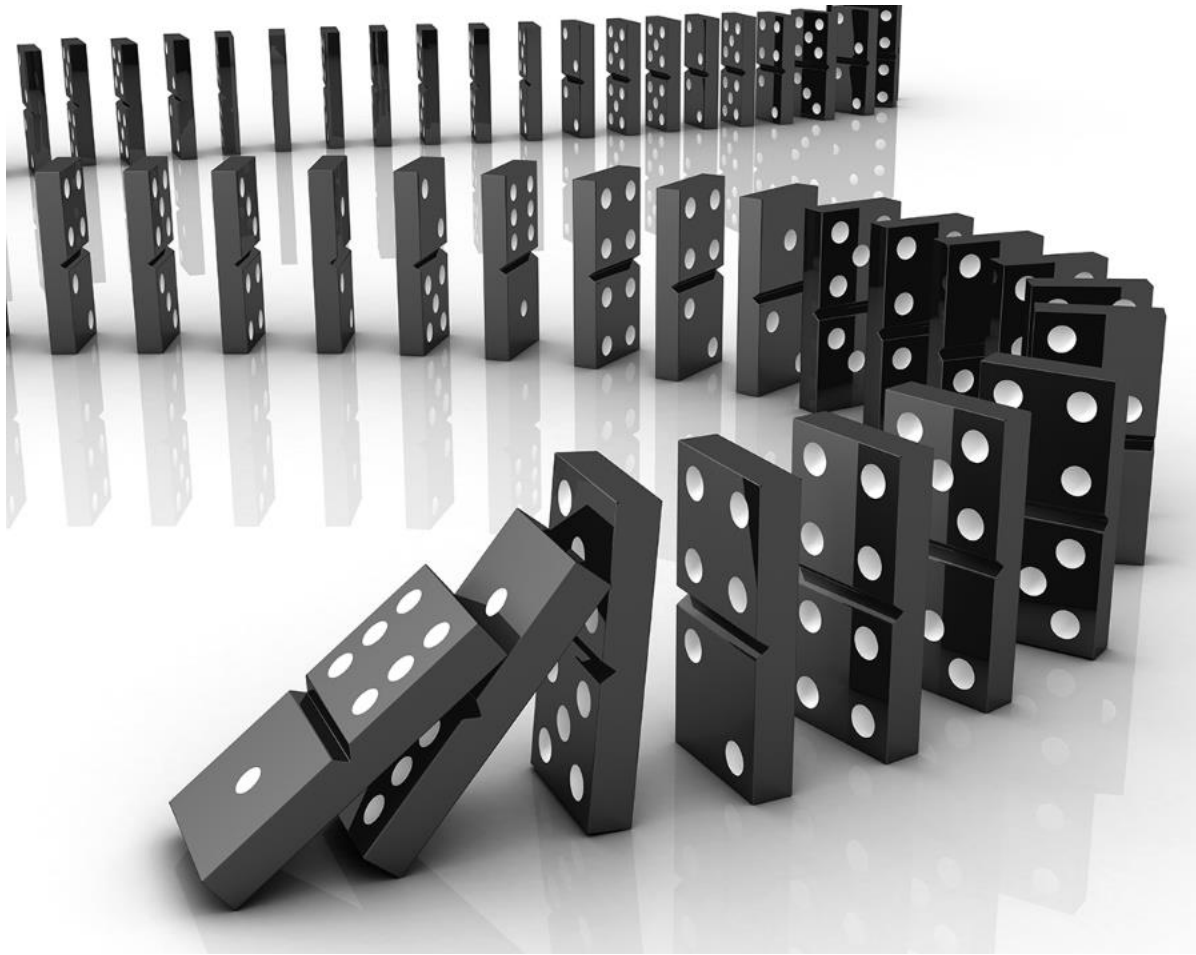
Comment corriger l'hémophilie sans facteurs de coagulation:

- Inhibiteur des inhibiteurs
- «Substitut» du facteur VIII

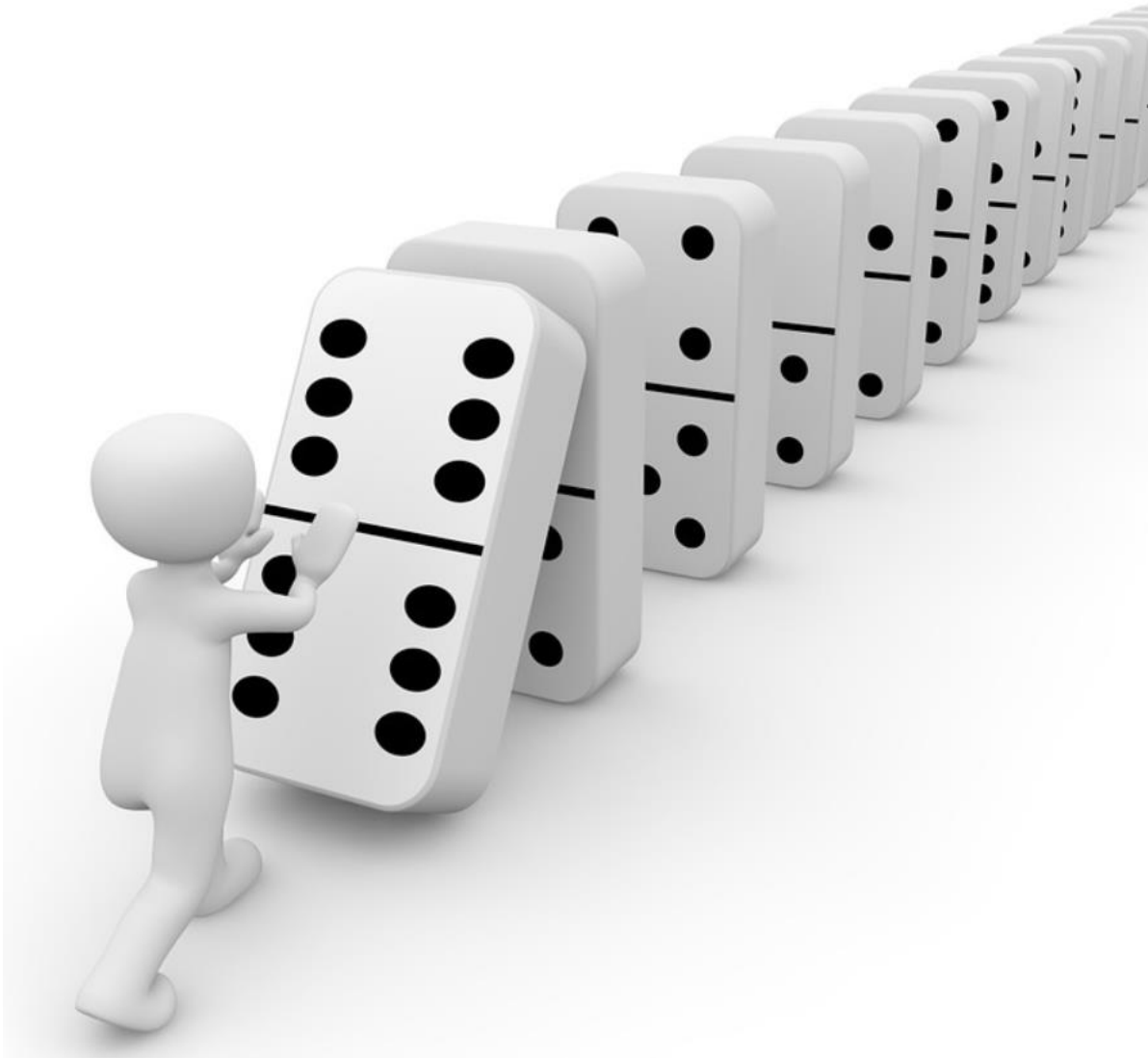
# La «cascade» de la coagulation



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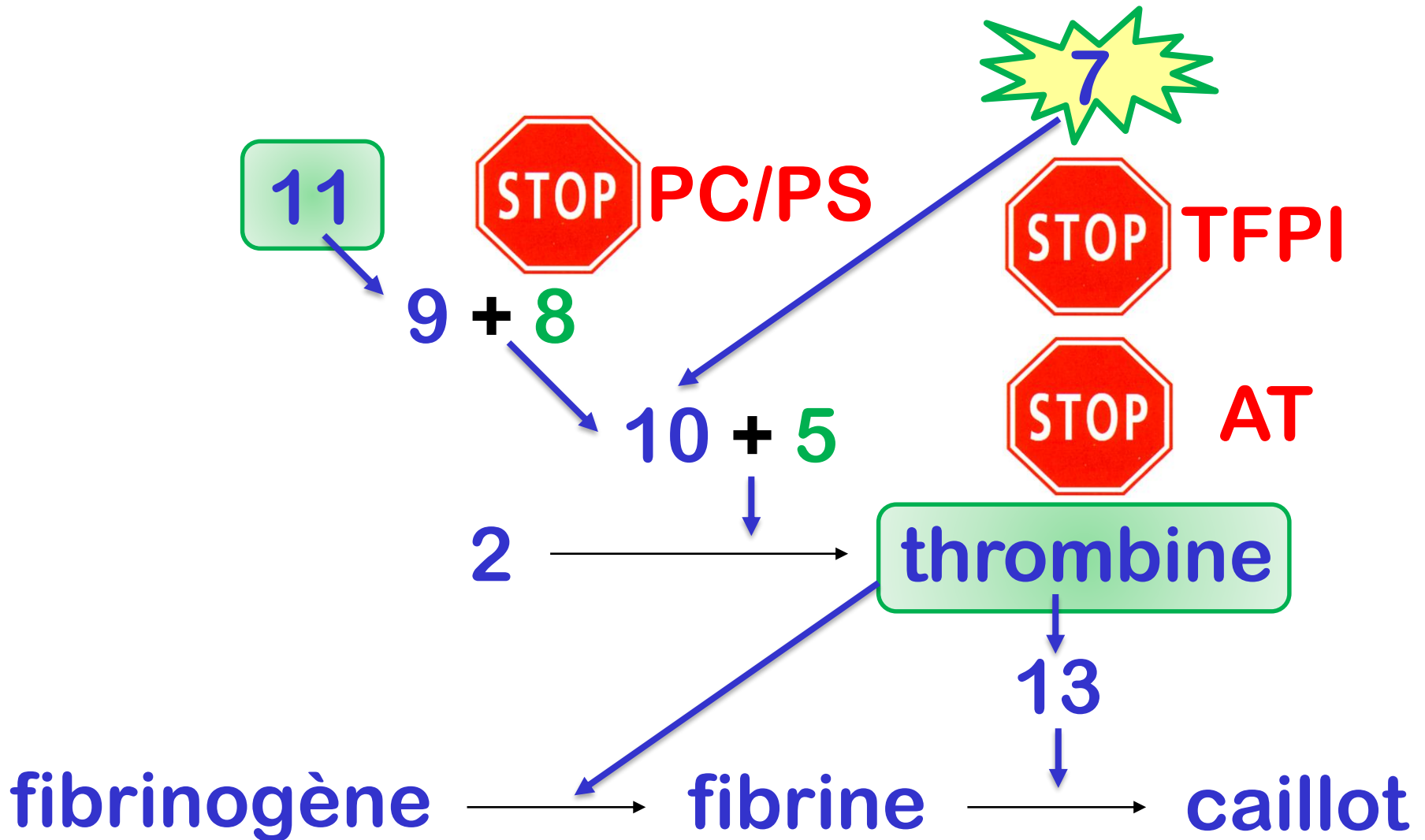
# La «cascade» de la coagulation



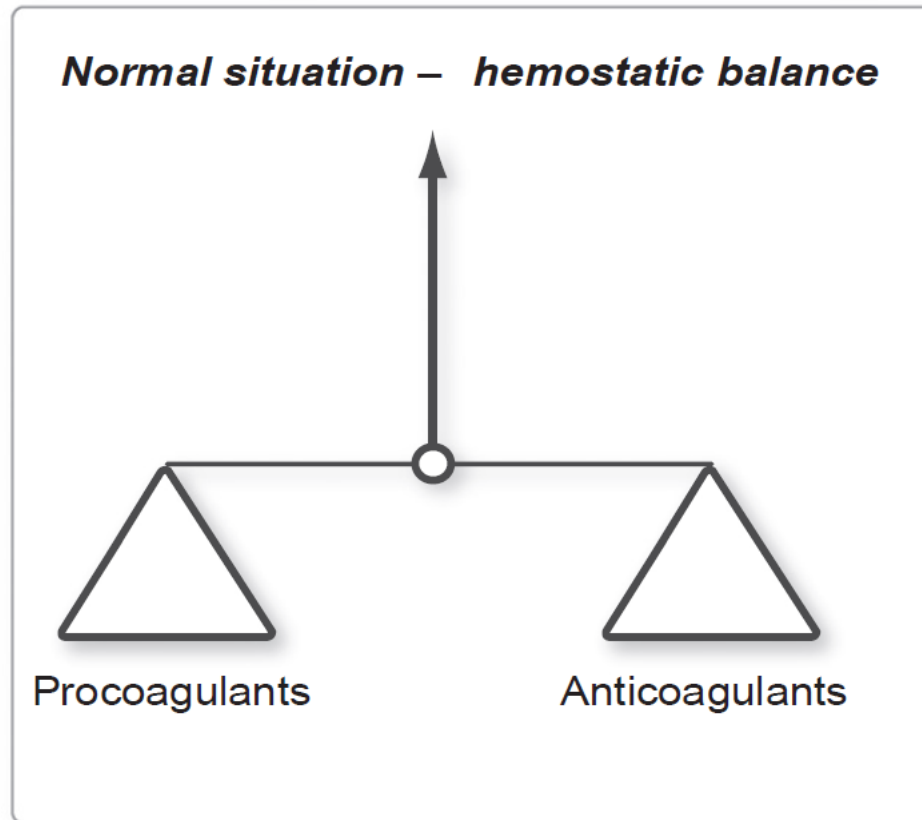
# Inhibiteurs naturels de la coagulation



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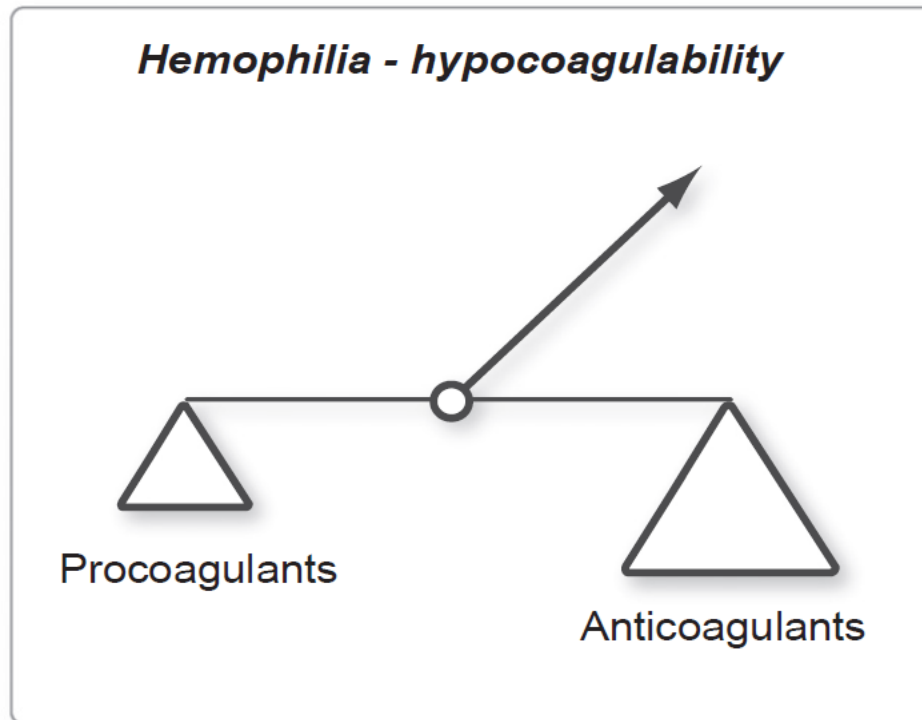


# Hémostase normale = équilibre

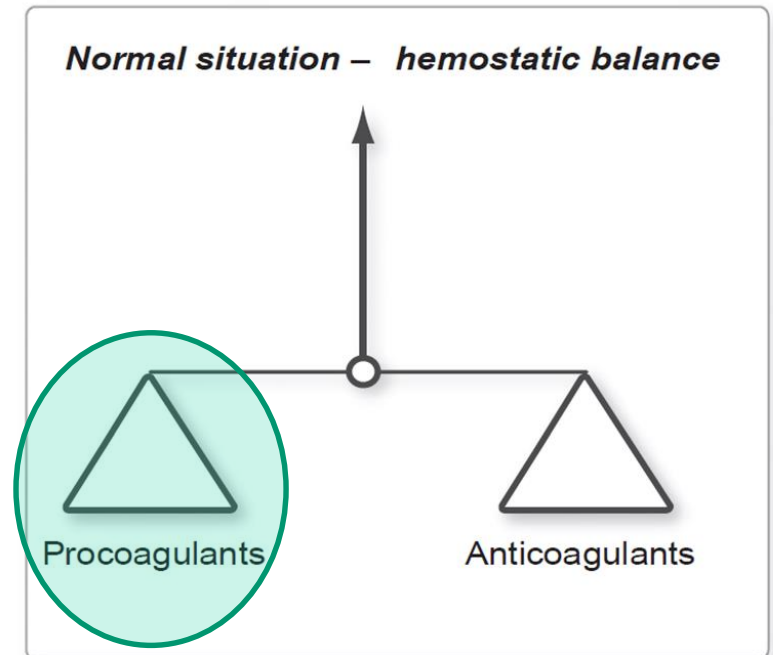
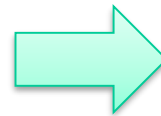
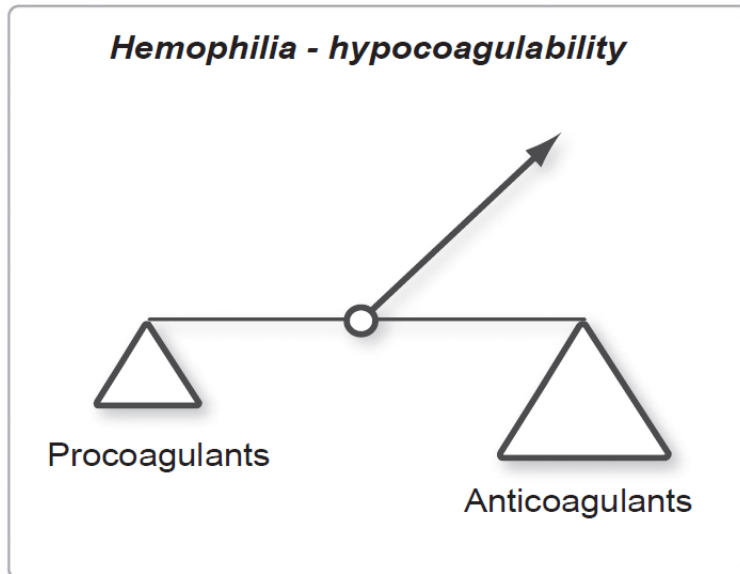




# Hémophilie = des-équilibre → hypocoagulation



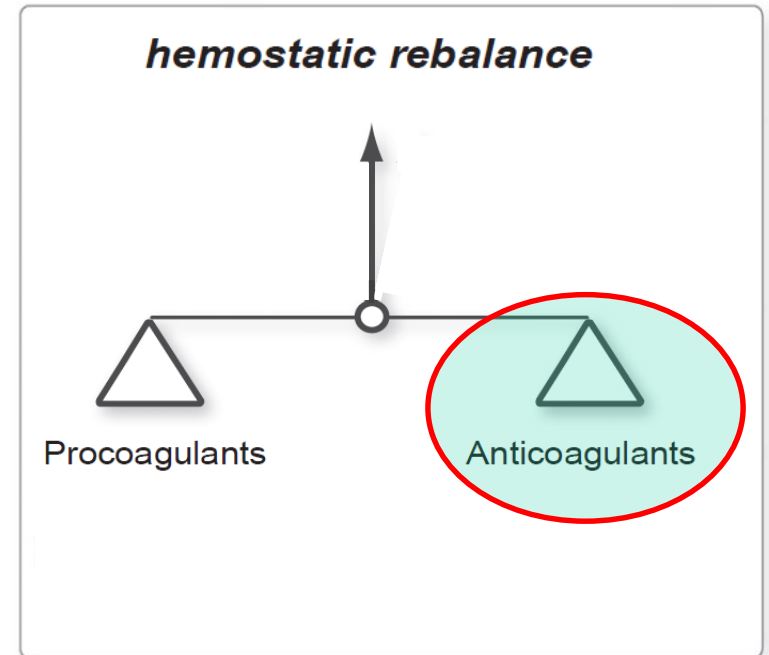
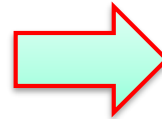
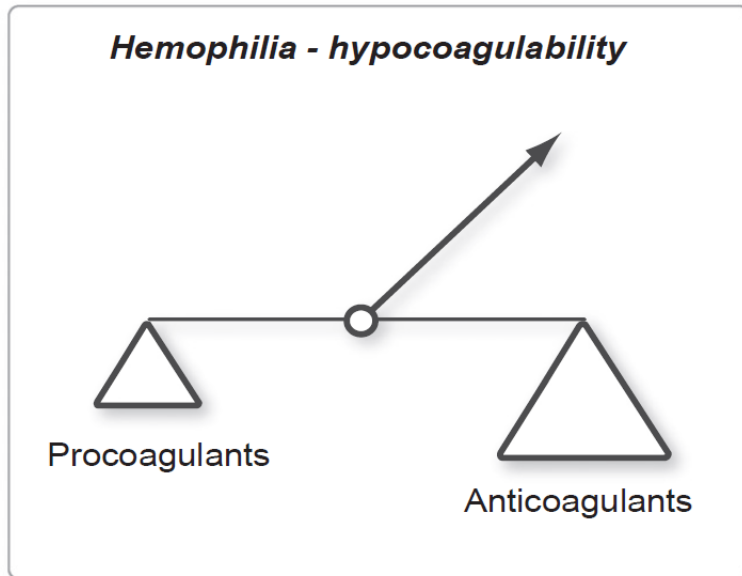
# Thérapie avec facteurs de coagulation



## Facteurs de coagulation:

- Plasmatiques
- Recombinants
- Demi-vie prolongée

# Thérapie avec inhibiteurs des inhibiteurs naturels



**Inhibiteurs des inhibiteurs naturels:**

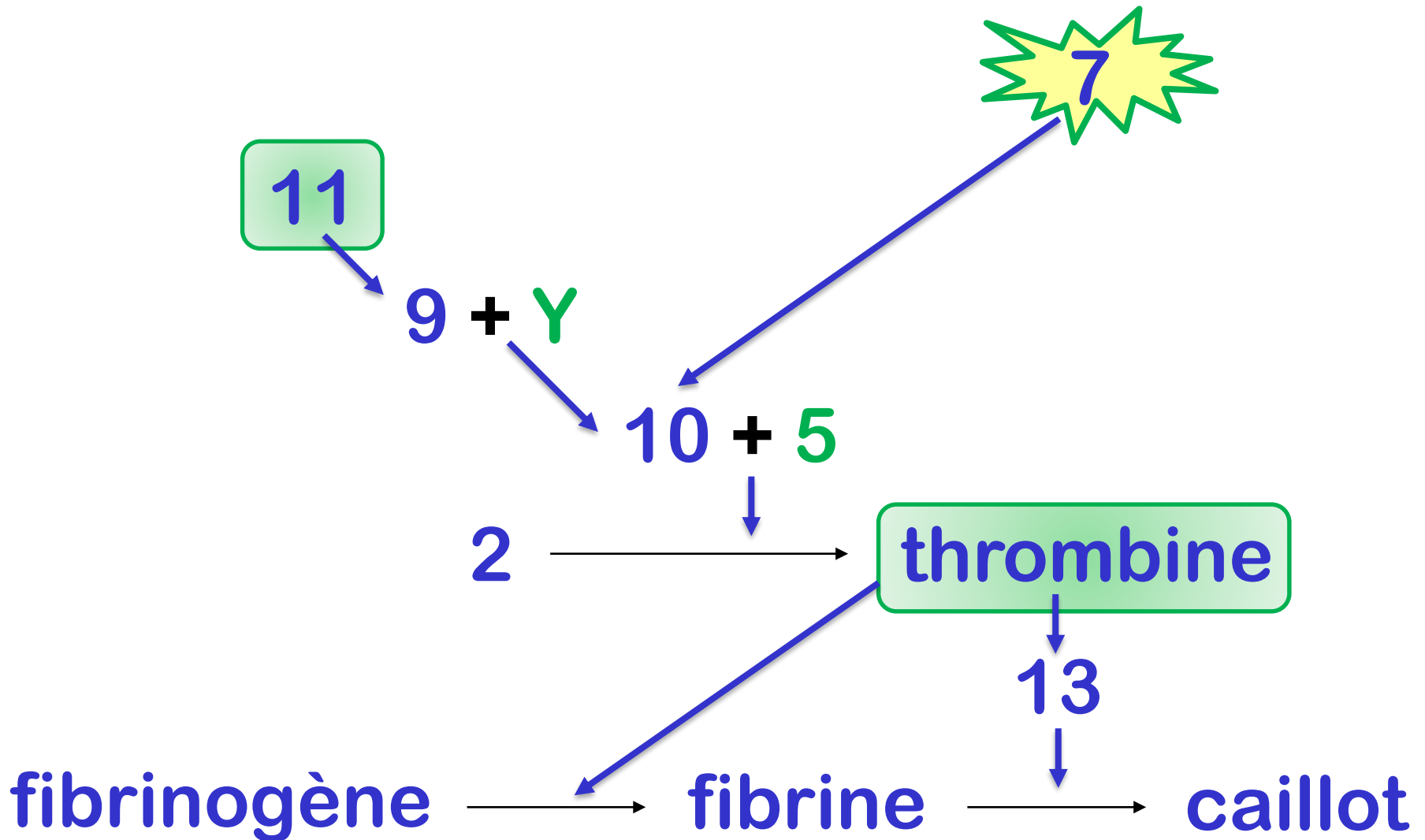
- TFPI (anticorps)
- AT (fitusiran)
- PC/S (anticorps, serpines)

**Études cliniques en cours**

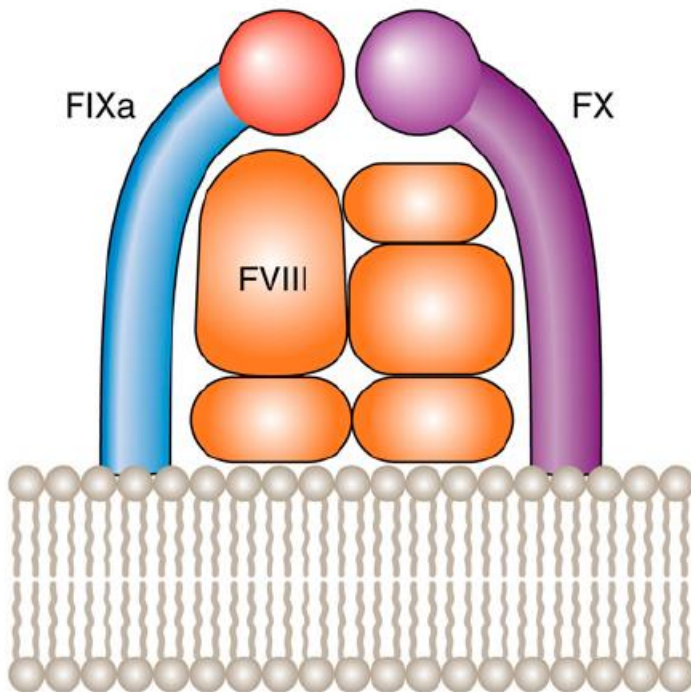
# Traiter l'hémophilie avec un substitut du F.VIII



# Traiter l'hémophilie avec un substitut du F.VIII

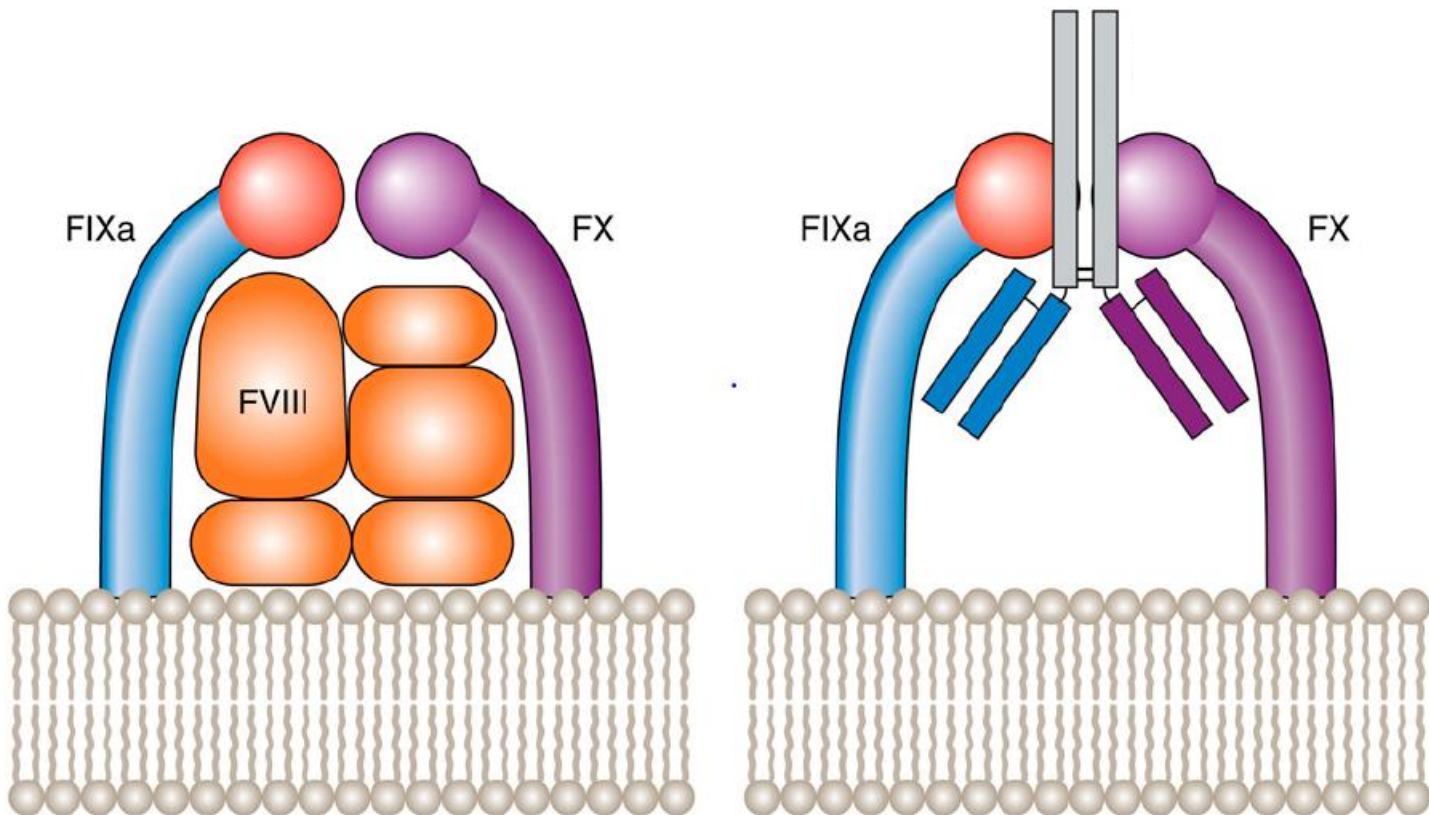


# Facteur VIII



FVIII brings together FIXa and FX, allowing FXa and, ultimately, thrombin generation to proceed. The ACE910 bispecific antibody (against FIXa and FX) achieves the same action in the absence of FVIII. Professional illustration by Patrick Lane, ScEYence Studios.

# Anticorps bispécifique : Emicizumab



FVIII brings together FIXa and FX, allowing FXa and, ultimately, thrombin generation to proceed. The ACE910 bispecific antibody (against FIXa and FX) achieves the same action in the absence of FVIII. Professional illustration by Patrick Lane, ScEYence Studios.

# Nomenclature

**Emicizumab**

anticorps  
monoclonal



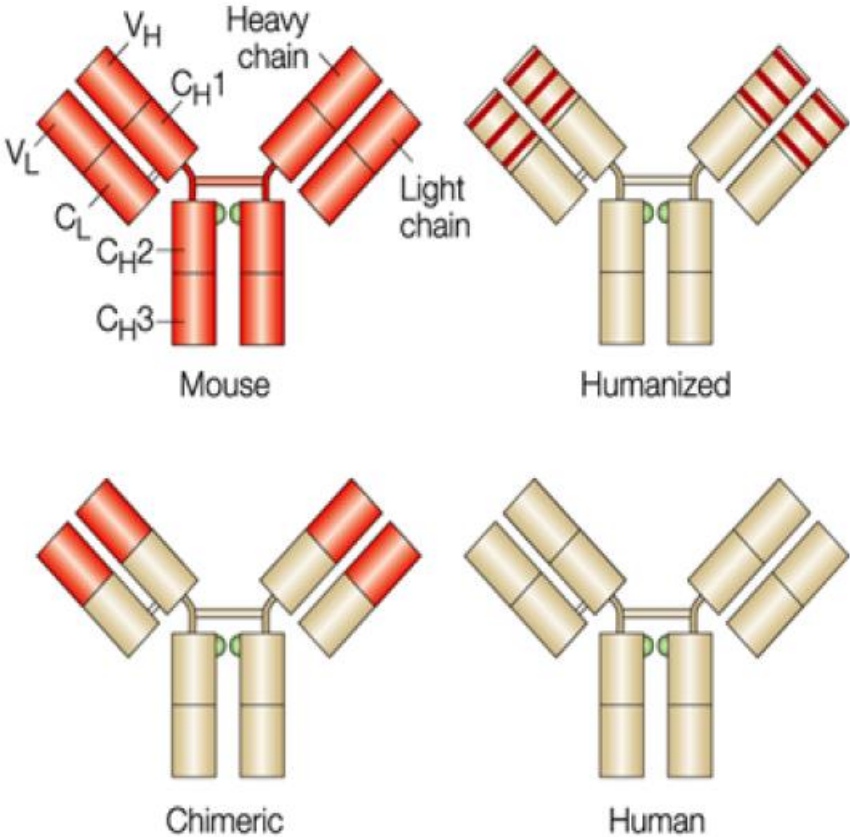
# Nomenclature

## Emicizumab

Source	
	meaning
-a-	<u>rat</u>
-e-	<u>hamster</u>
-i-	<u>primate</u>
-o-	<u>mouse</u>
-u-	<u>human</u>
-xi-	<u>chimeric</u> (human/foreign)
-zu-	<u>humanized</u>
-vet-	<u>veterinary</u>
-xizu-*	chimeric/humanized hybrid
-axo-	rat/mouse hybrid (see <u>trifunctional antibody</u> )

# Nomenclature

# Emicizumab



# Nomenclature

## Emicizumab

Target	
new	meaning
-ami-	serum amyloid protein
—	<u>angiogenesis</u> (inhibitor)
-ba-	<u>bacterium</u>
-ci-	<u>circulatory system</u>
-fung-	<u>fungus</u>
-gros-	<u>growth factor</u>
-ki-	<u>interleukin</u>
—	inflammatory <u>lesions</u>
-li-	<u>immune system</u>

# Nomenclature

Emicizumab

笑み = smile

# Synthèse

La «cascade» de la coagulation = «domino»

Les inhibiteurs naturels de la coagulation = trois

Comment corriger l'hémophilie sans facteurs de coagulation:

- Inhibiteur des inhibiteurs = études cliniques
- «Substitut» du facteur VIII = Emicizumab