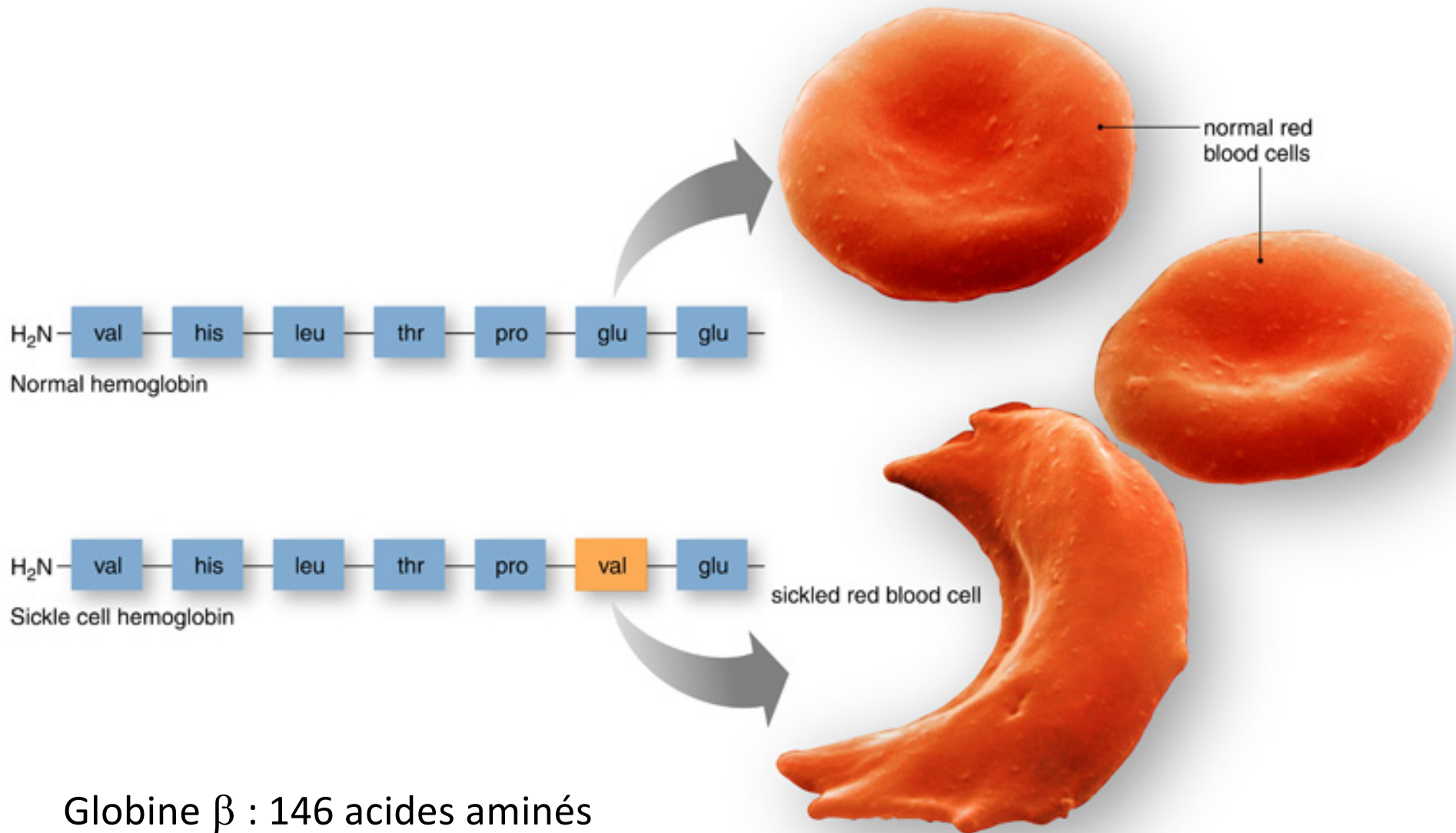
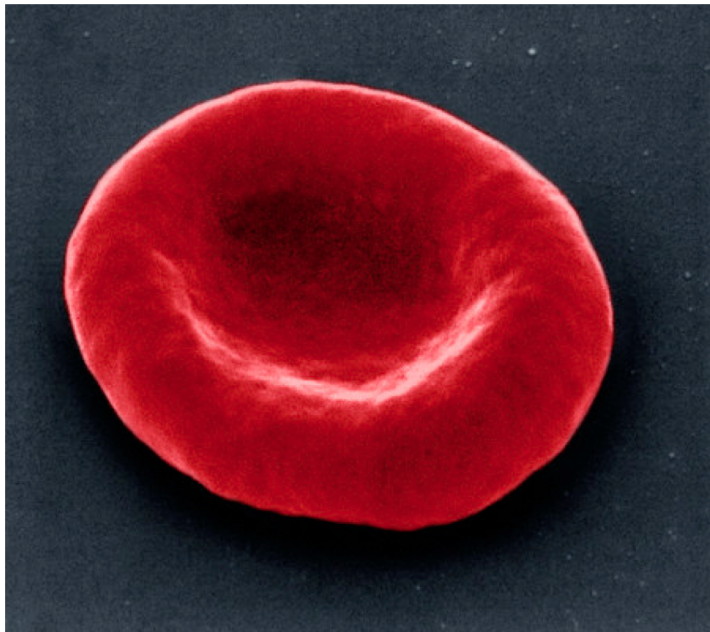
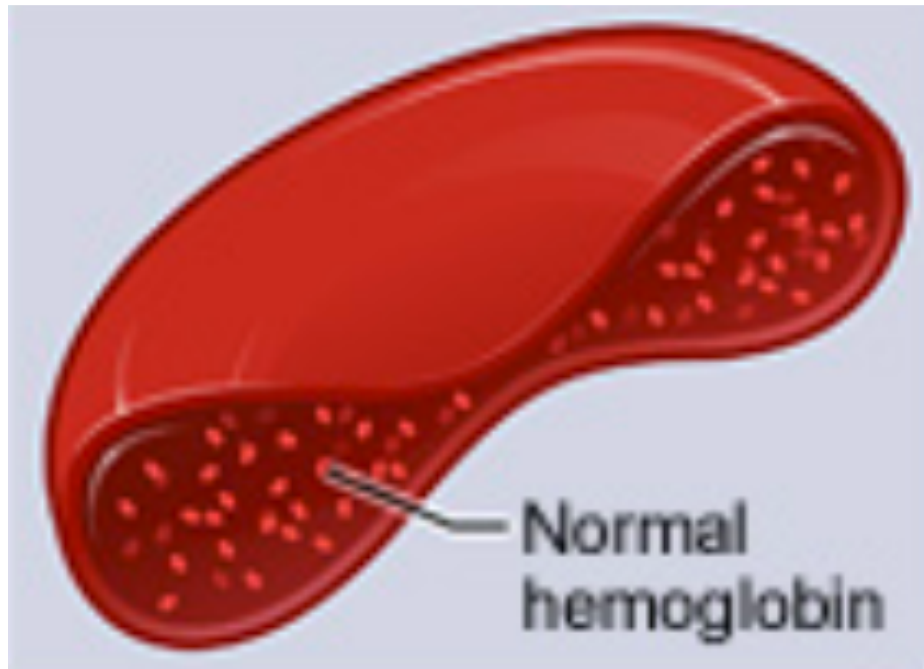


# Base moléculaire de l'anémie falciforme

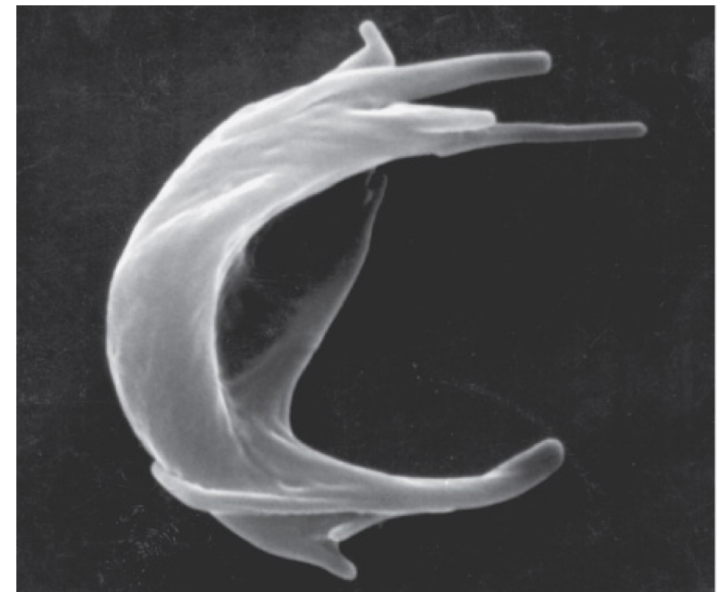
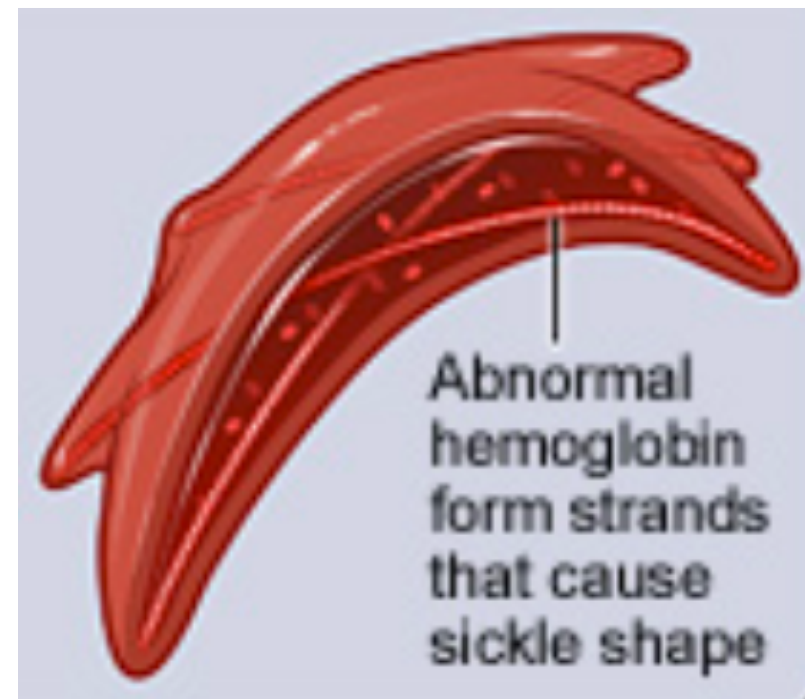


Globine  $\beta$  : 146 acides aminés  
1 seule différence, à la position 6

globule rouge normal :

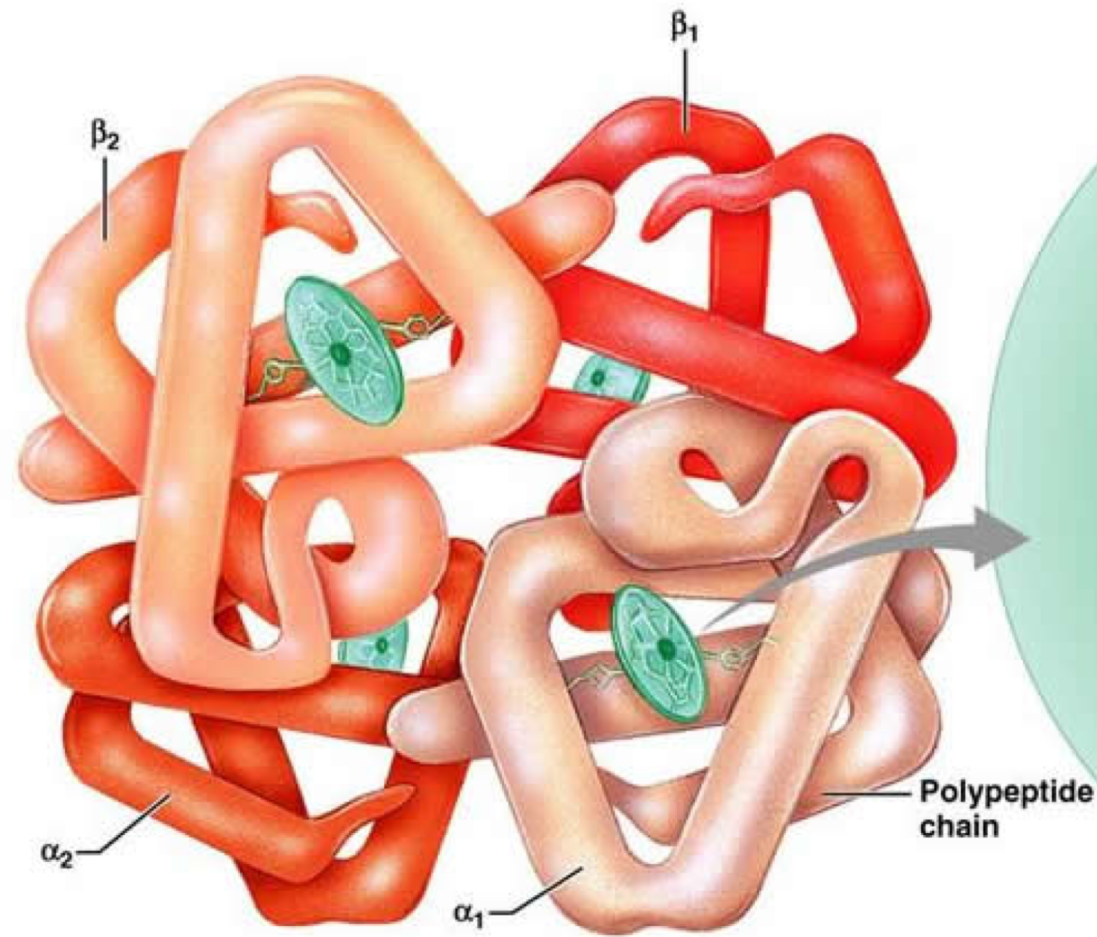
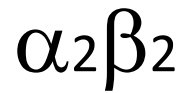


globule rouge en faucille :



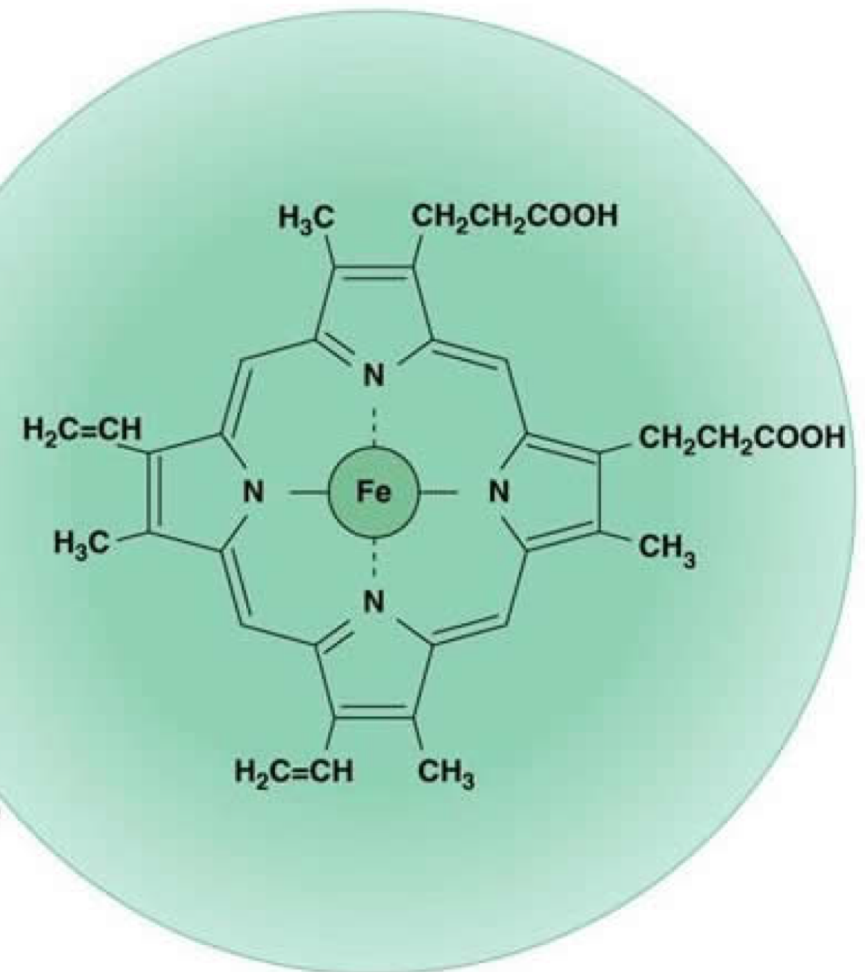
Source: Mescher AL: *Junqueira's Basic Histology: Text and Atlas*, 12th Edition: <http://www.accessmedicine.com>  
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La protéine **hémoglobine** est constituée de 4 polypeptides.



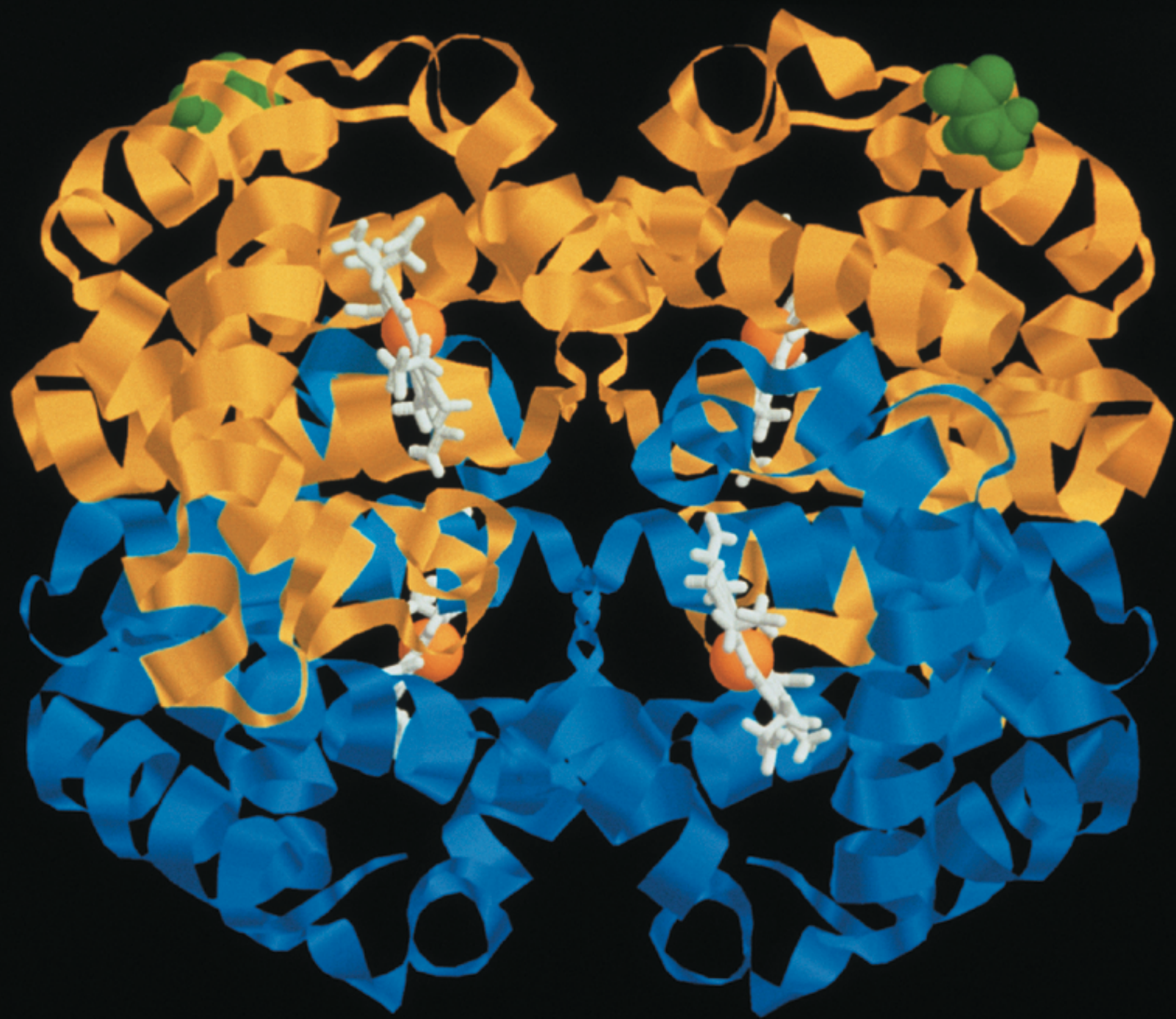
(a) Hemoglobin

Chaque sous-unité héberge un groupe prosthétique :  
un **hème**.



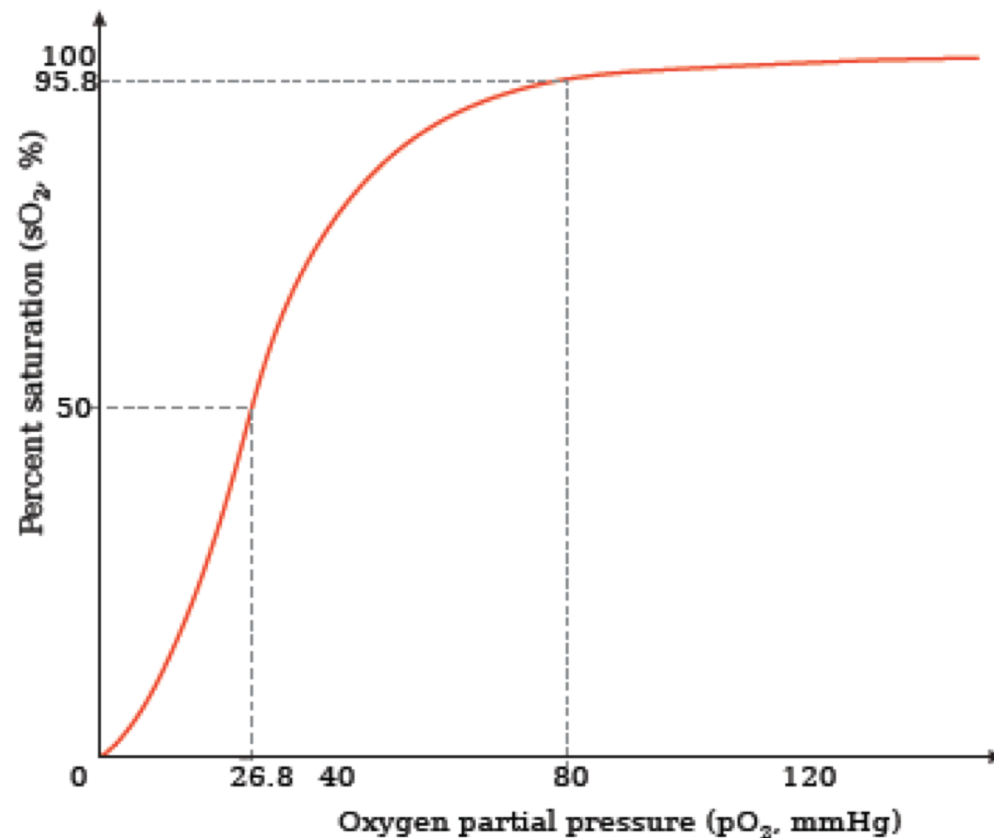
(b) Iron-containing heme group



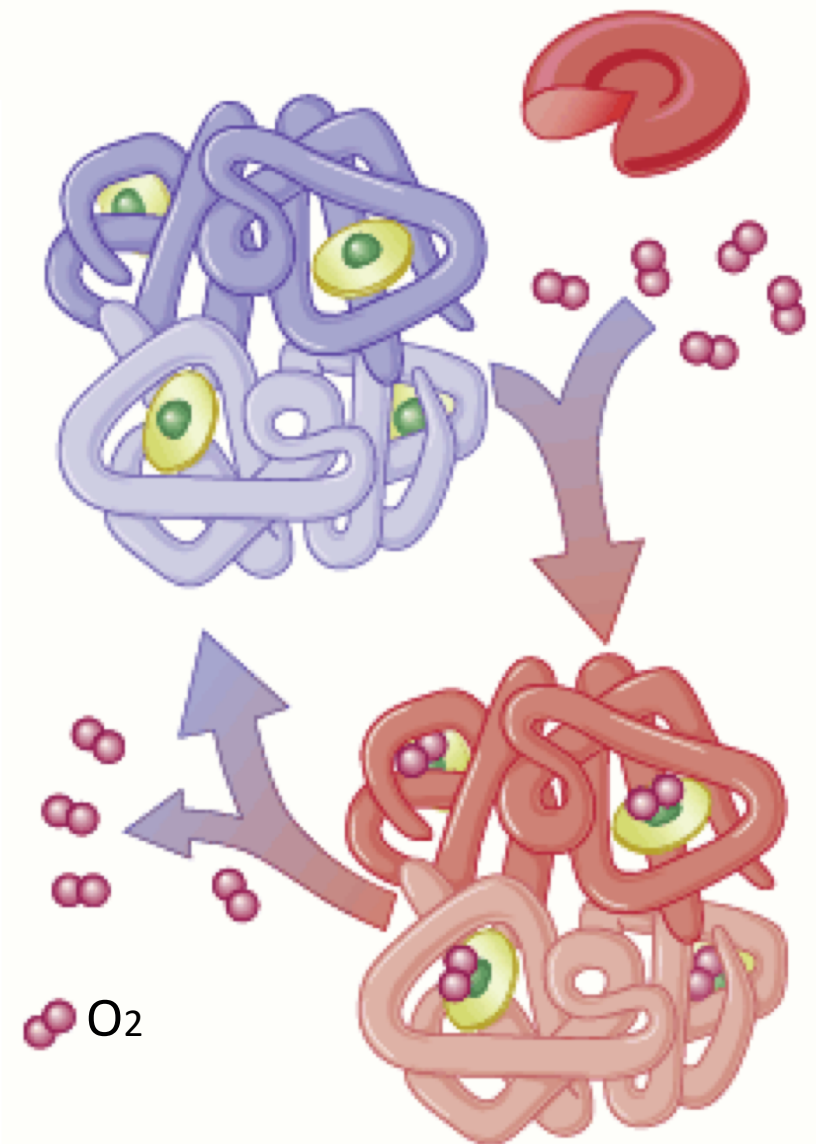




L'hémoglobine est un transporteur d'O<sub>2</sub>.

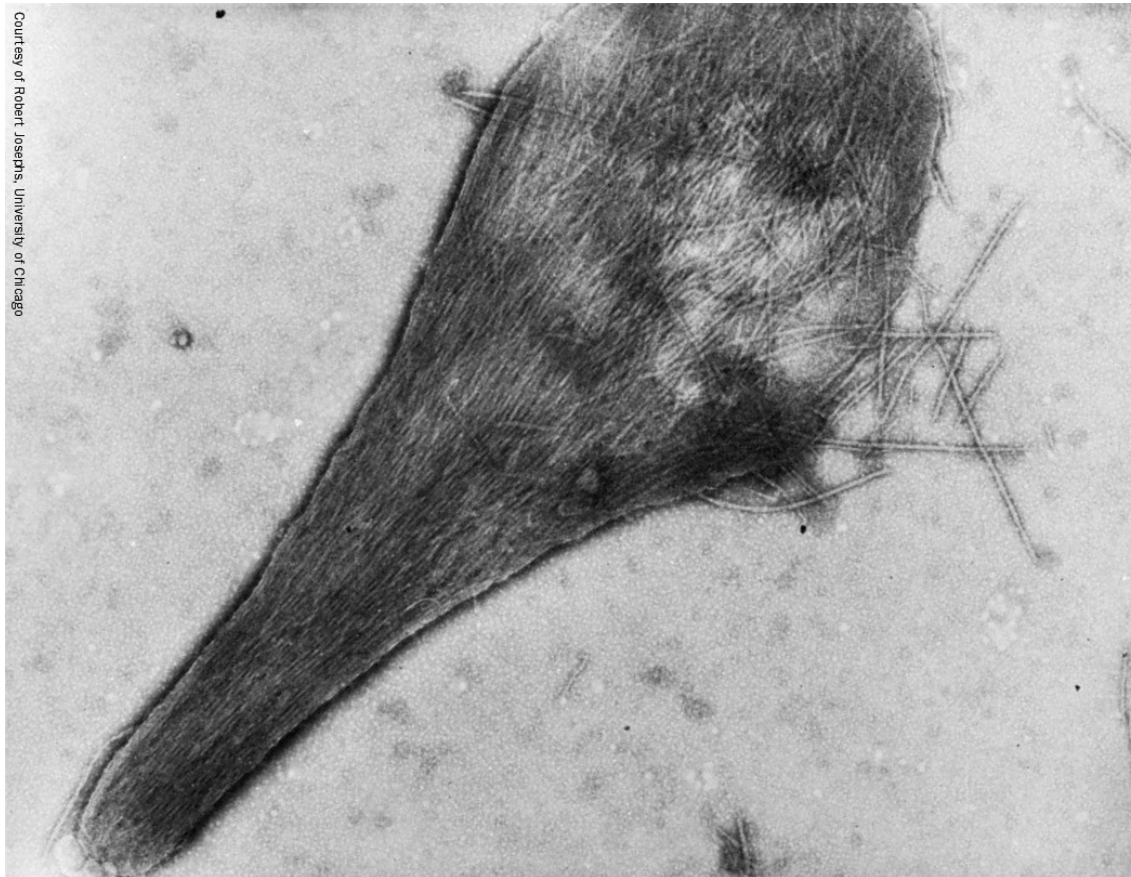


*Pourcentage d'hémoglobine ayant fixé du dioxygène, en fonction de la **pression partielle** en dioxygène. Courbe de **Barcroft** de type sigmoïde → effet coopératif*

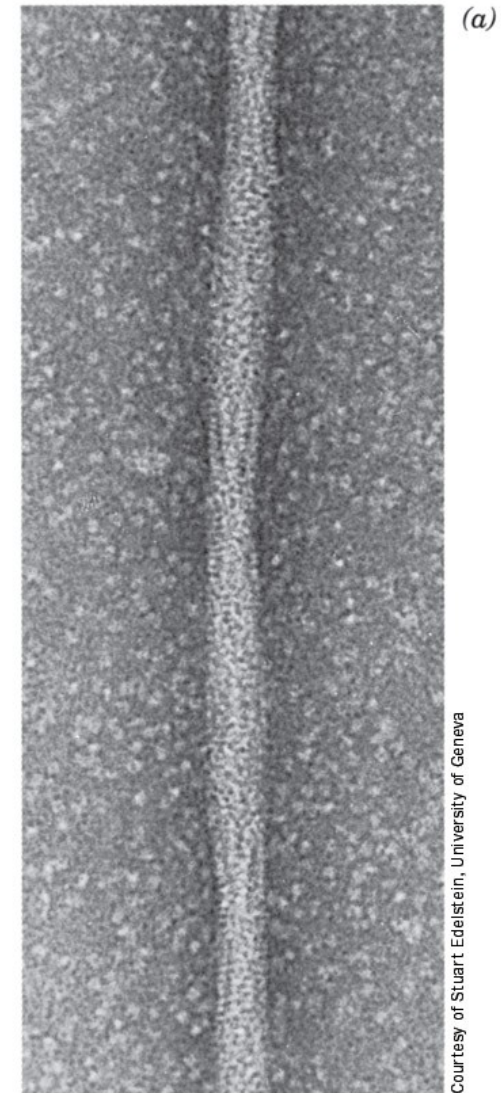


4 O<sub>2</sub> sont liés.

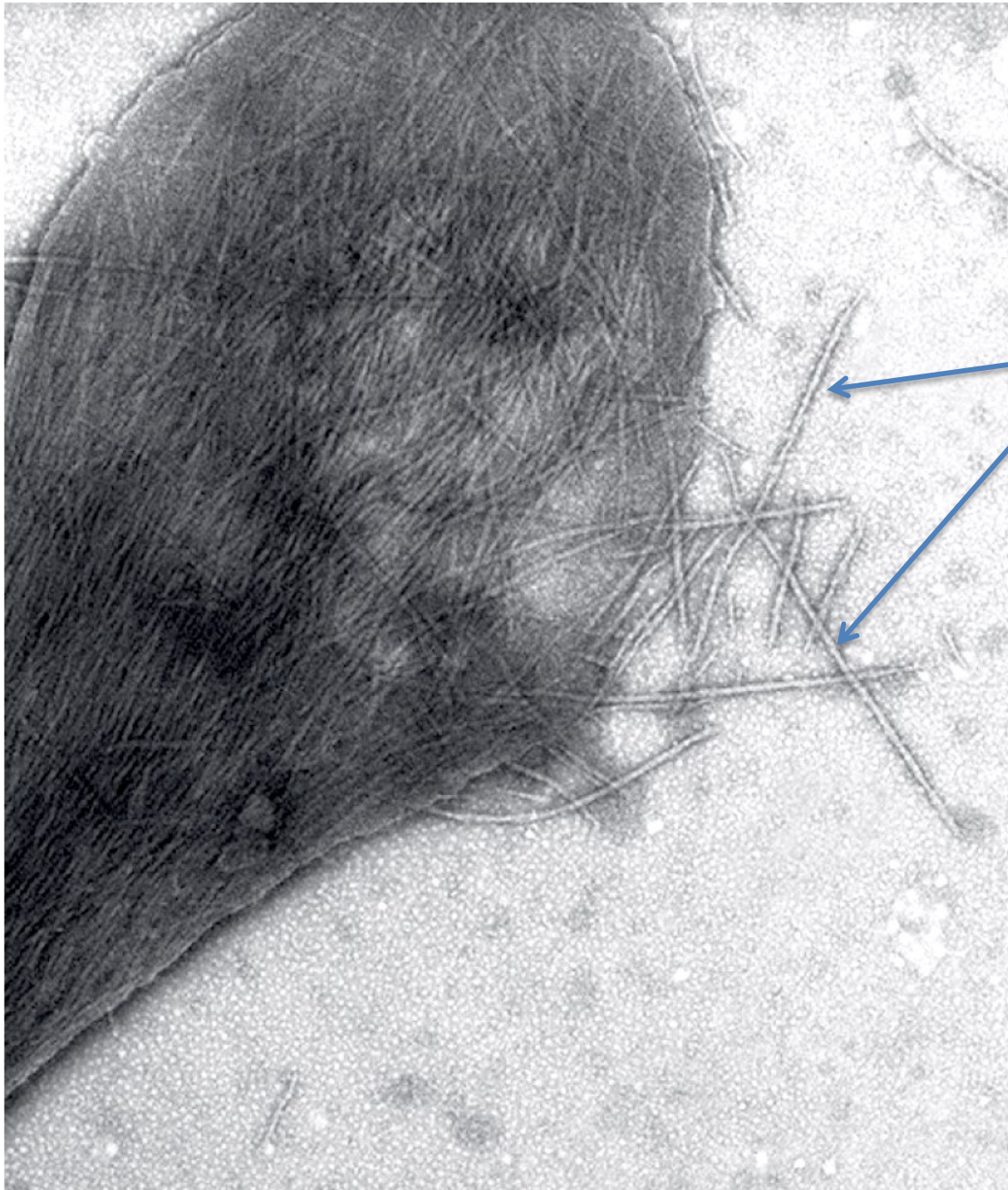
# Fibres d'hémoglobine S



Membrane cellulaire rompue :  
fibres d'hémoglobine S en dehors  
de la cellule.







Anémie  
falciforme

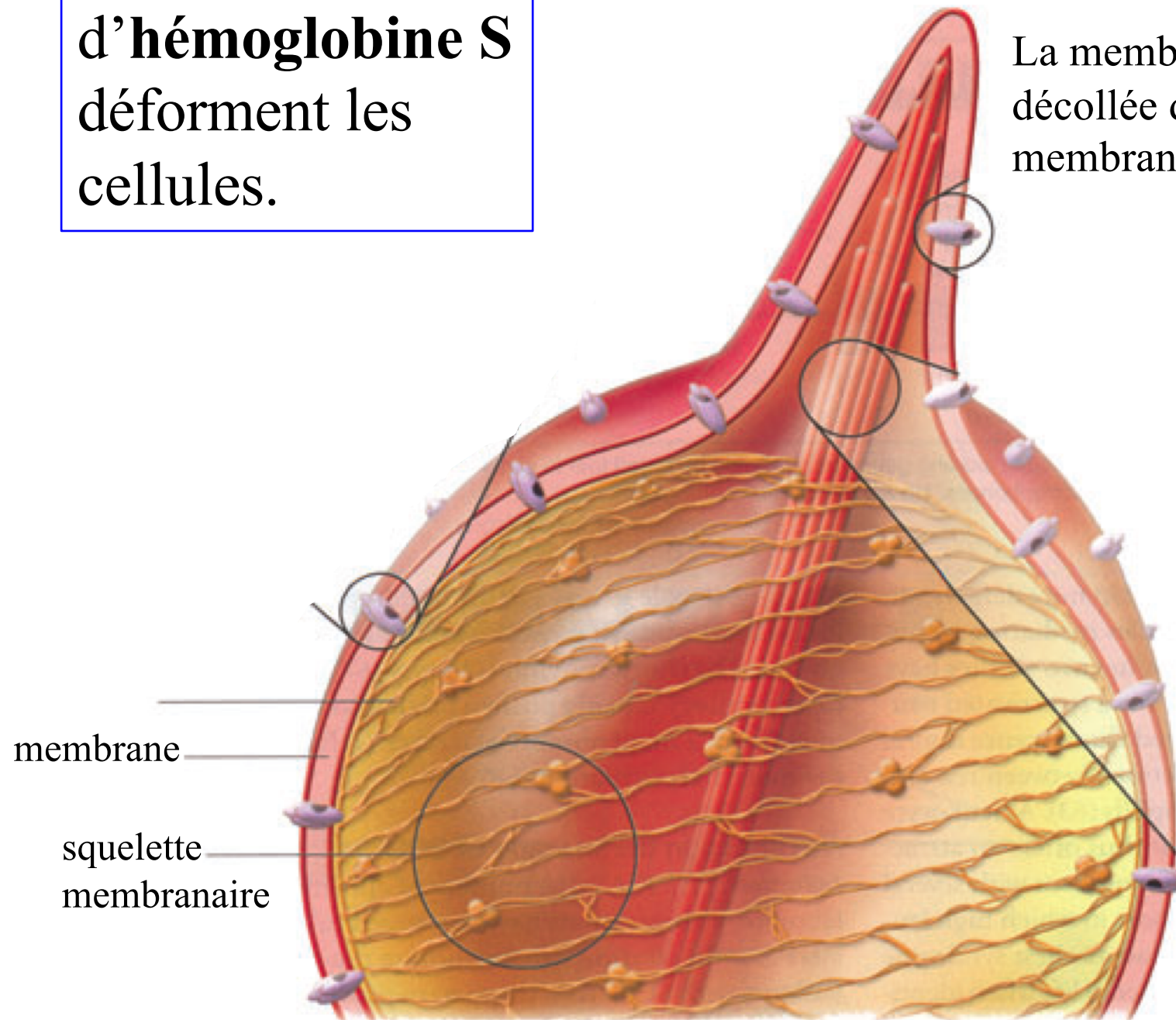
Fibres  
d'hémoglobine S

**Figure 7-24**  
*Biochemistry, Sixth Edition*  
© 2007 W. H. Freeman and Company

Microscopie électronique à transmission

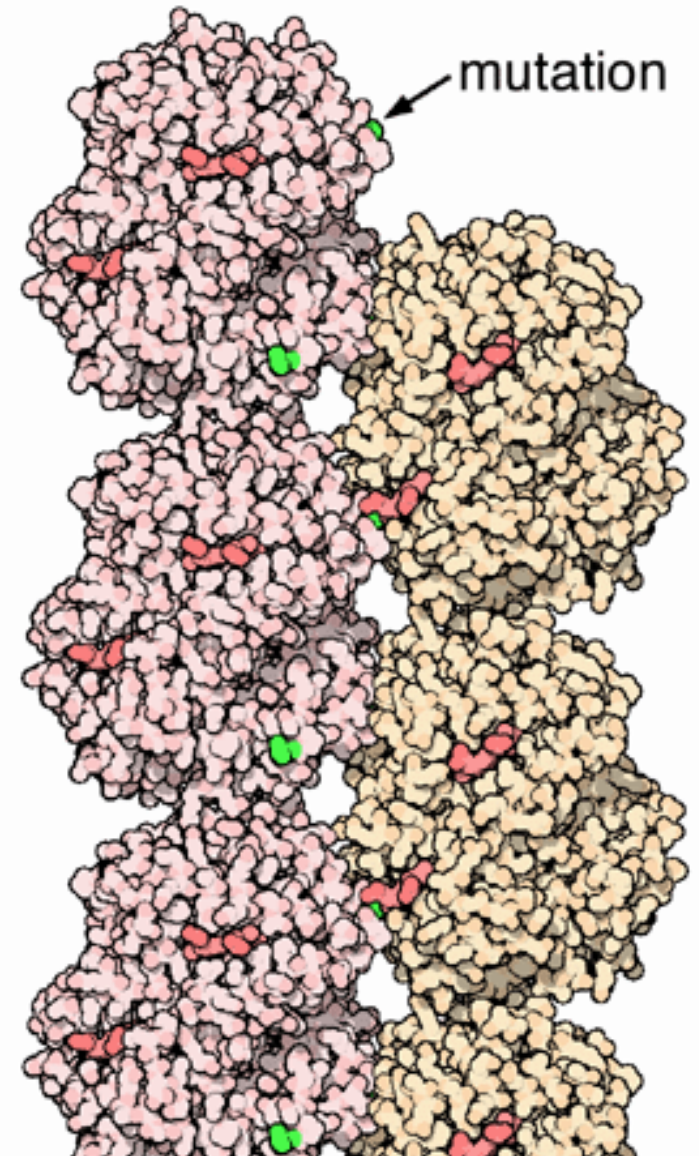
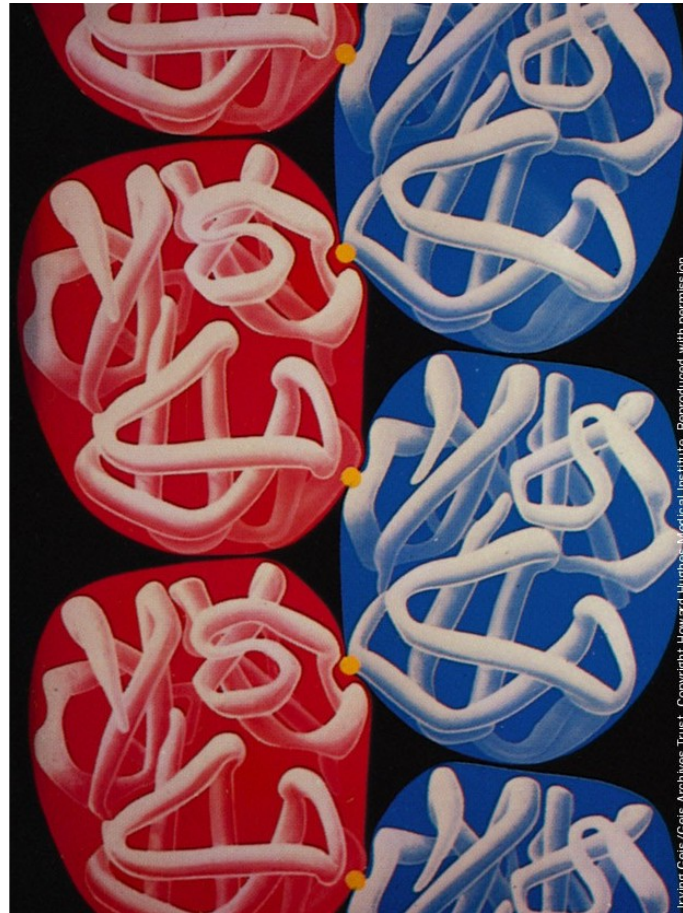
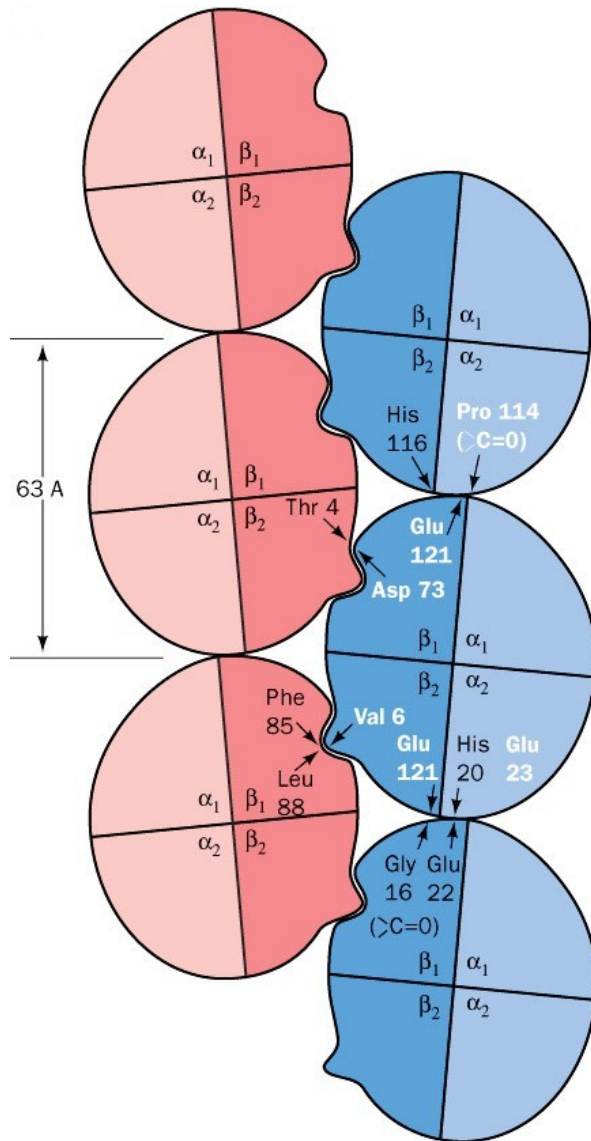
Les fibres  
d'**hémoglobine S**  
déforment les  
cellules.

La membrane est  
décollée du squelette  
membranaire.

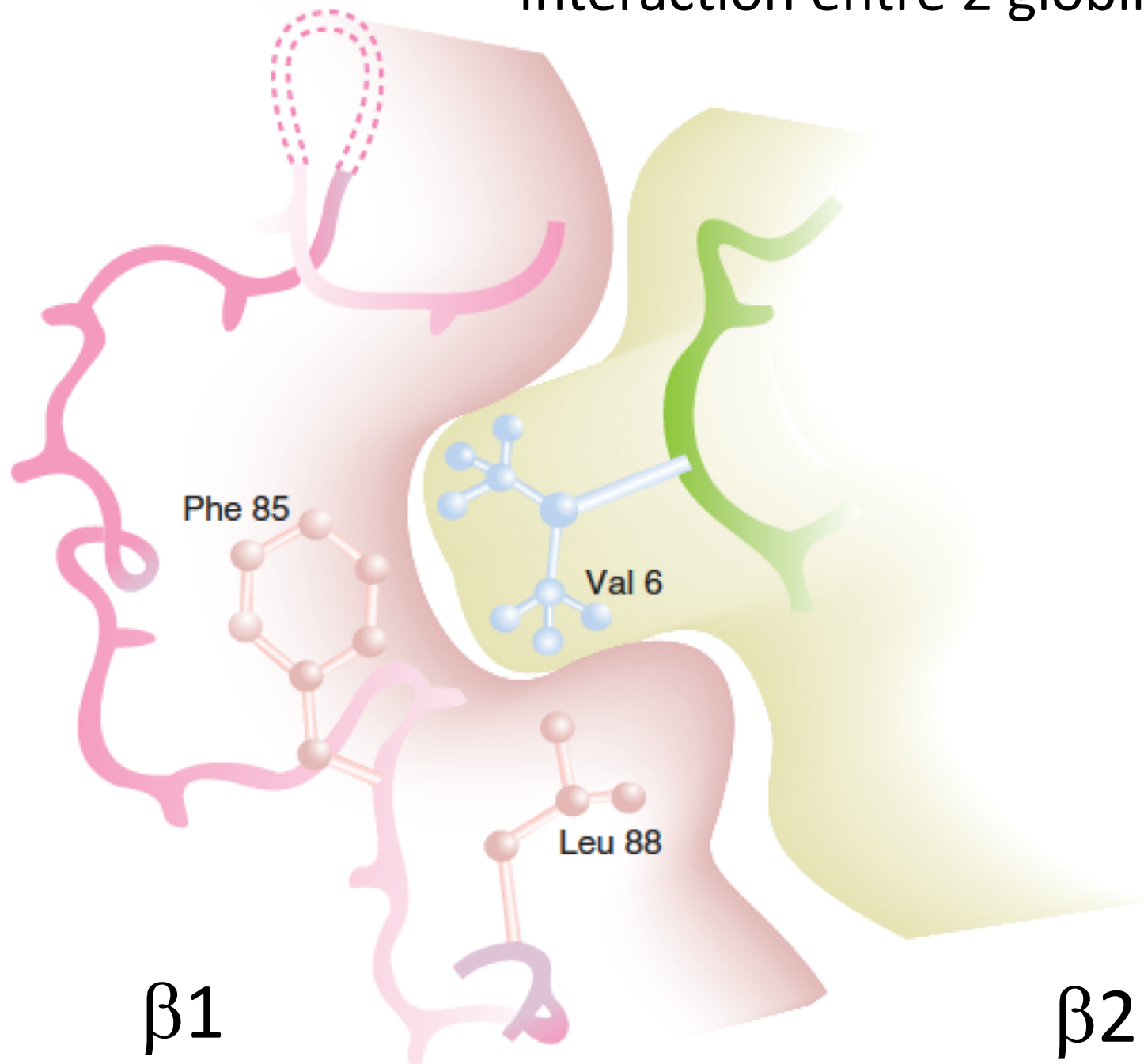




# Assemblage de la **désoxy**hémoglobine **S** en brins.

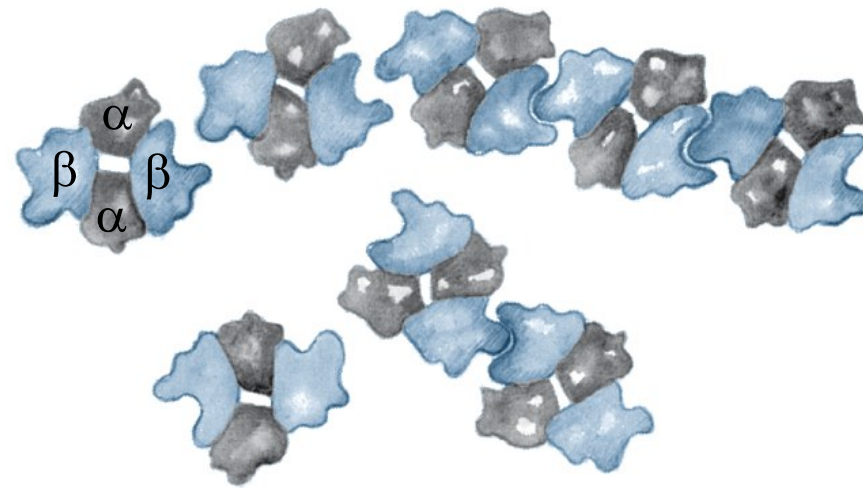


# Interaction entre 2 globines $\beta^S$

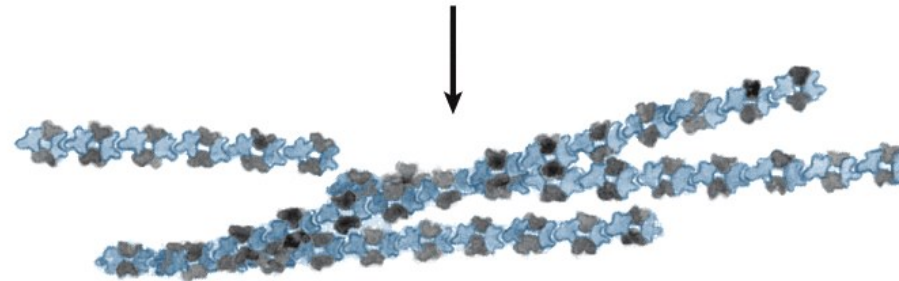




# Assemblage en fibres de l'hémoglobine S.

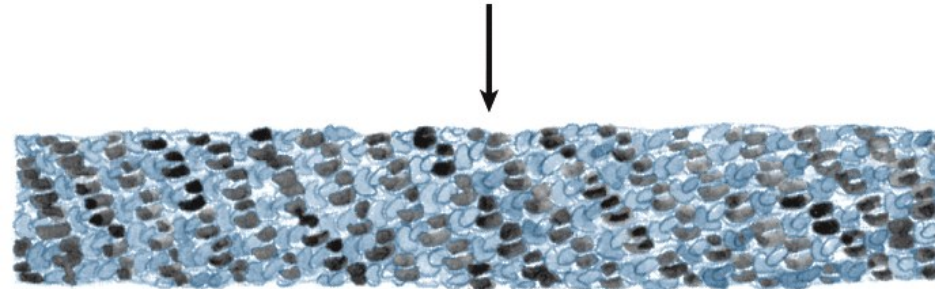


Interaction between molecules



brins

Strand formation



fibres

$\varnothing \approx 22 \text{ nm}$

Alignment and crystallization  
(fiber formation)

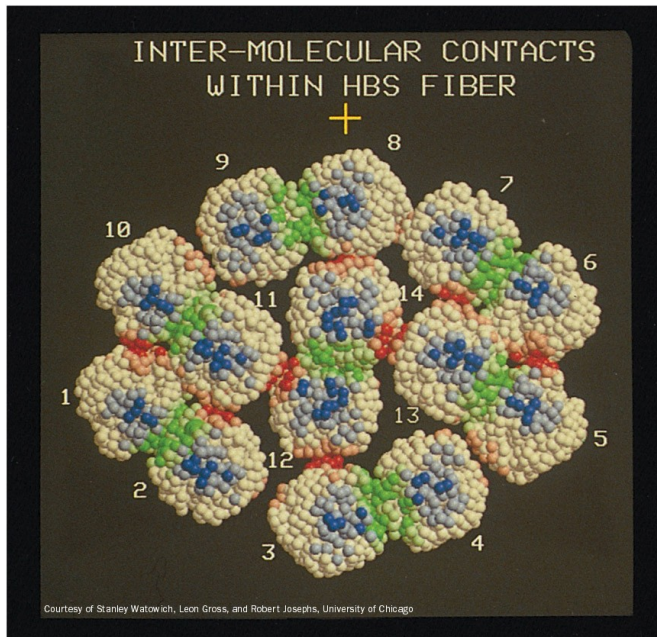
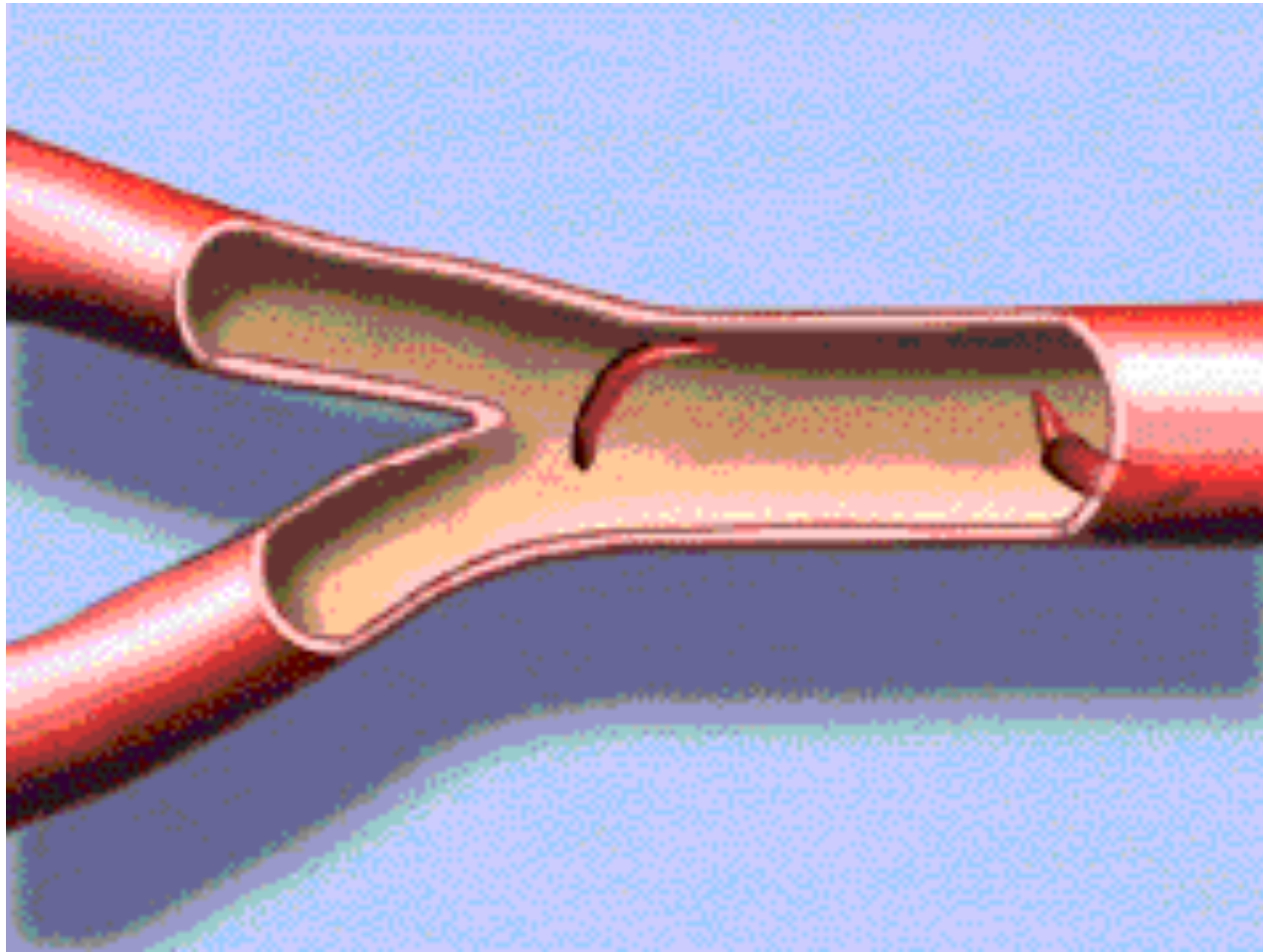


Figure 5-20b

*Lehninger Principles of Biochemistry, Fifth Edition*

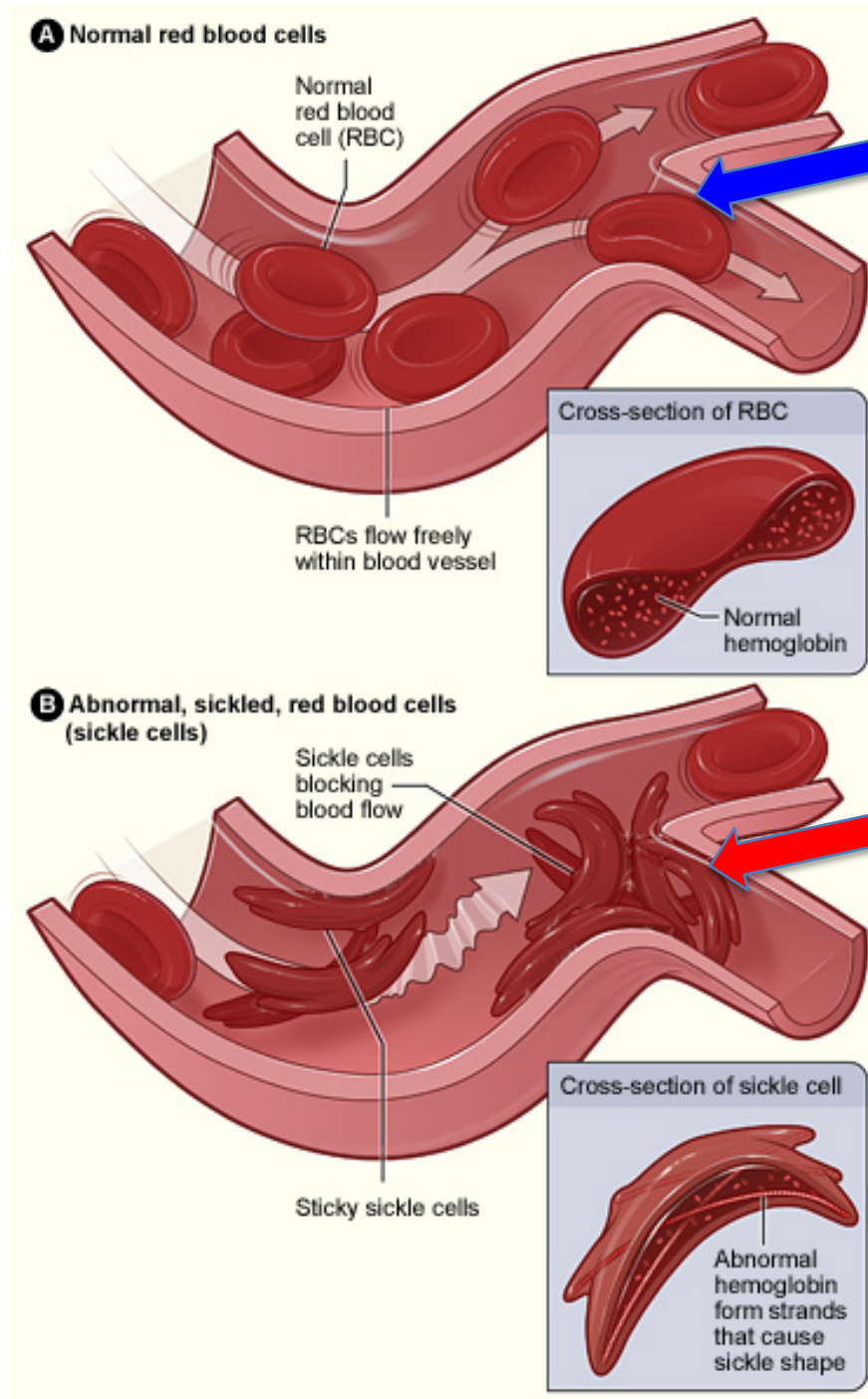
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# Blockage de petits capillaires





Échelle phénotypique :  
macroscopique



Ce globule rouge  
se déforme pour  
passer.

**Obstruction**  
d'un petit  
vaisseau  
→ douleur

# Les échelles du phénotype de l'anémie falciforme :

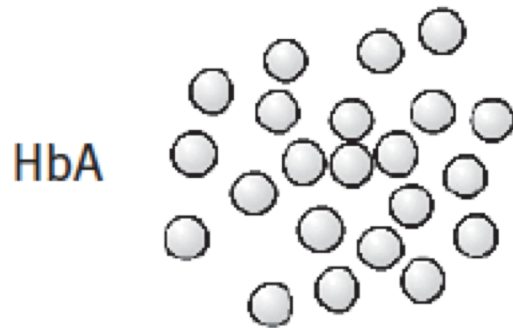
## Échelle cellulaire :



Microscopie  
optique

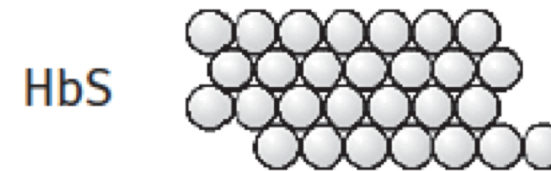


## Échelle moléculaire :



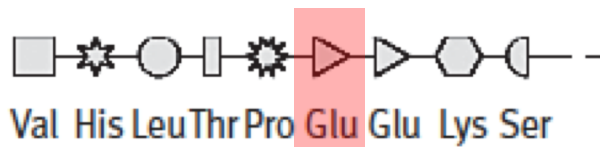
Protéines indépendantes

Microscopie  
électronique

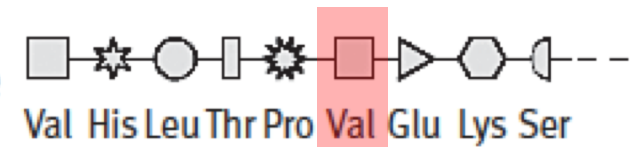


Protéines assemblées en fibres

Chaîne  $\beta$  de  
l'hémoglobine  
HbA



Chaîne  $\beta$  de  
l'hémoglobine  
HbS



Analyse  
biochimique