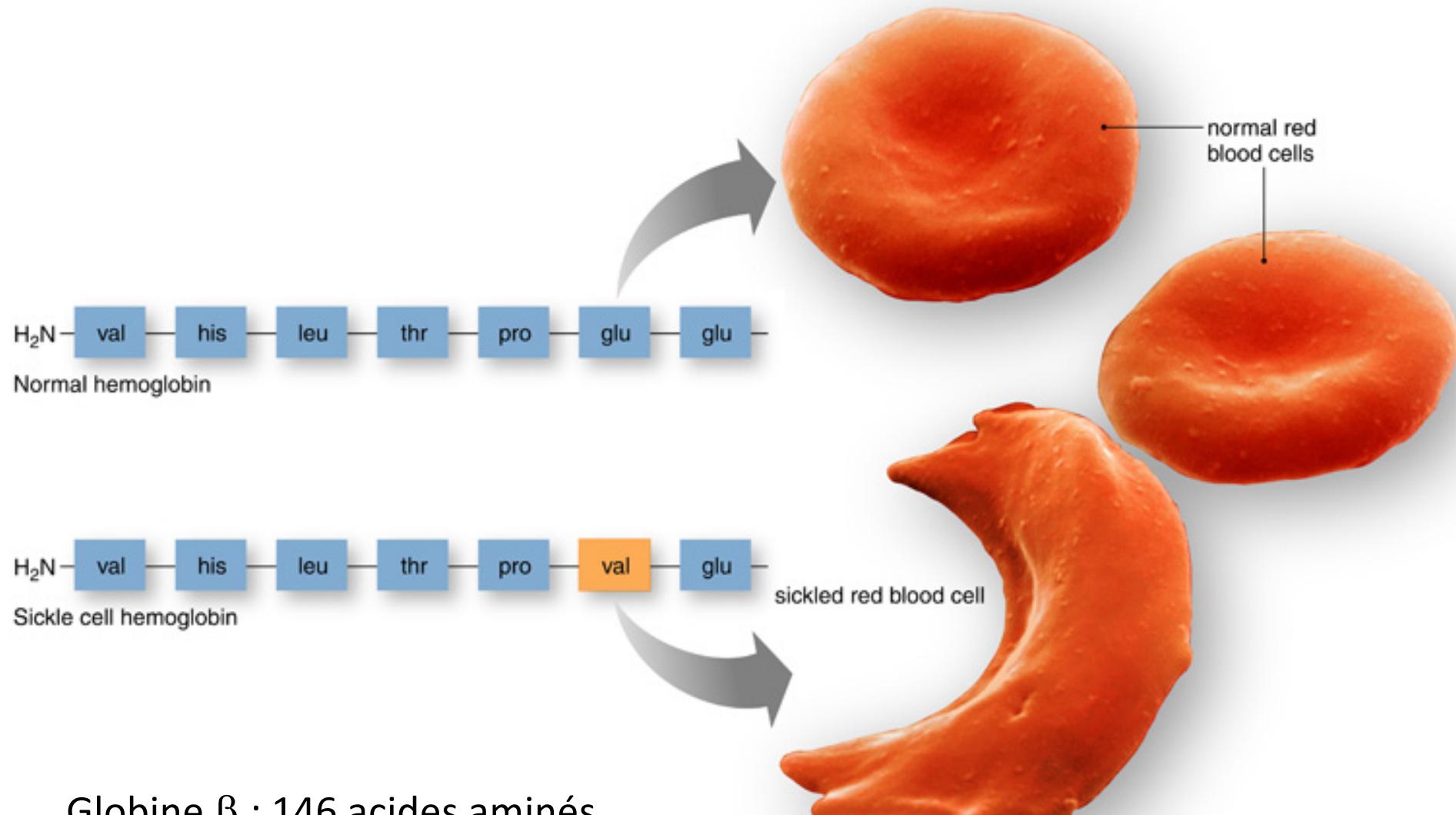
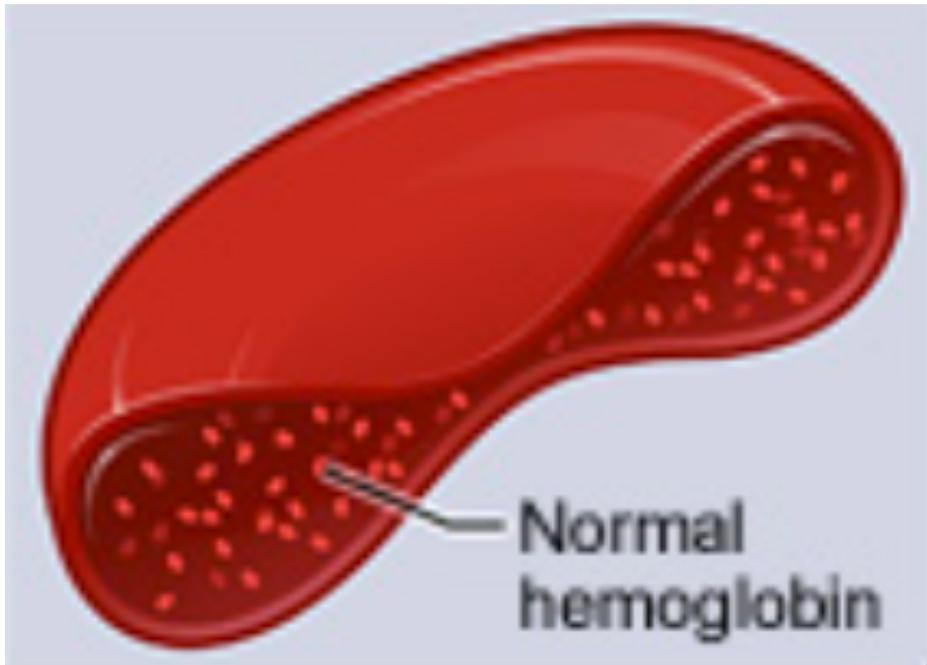


Base moléculaire de l'anémie falciforme

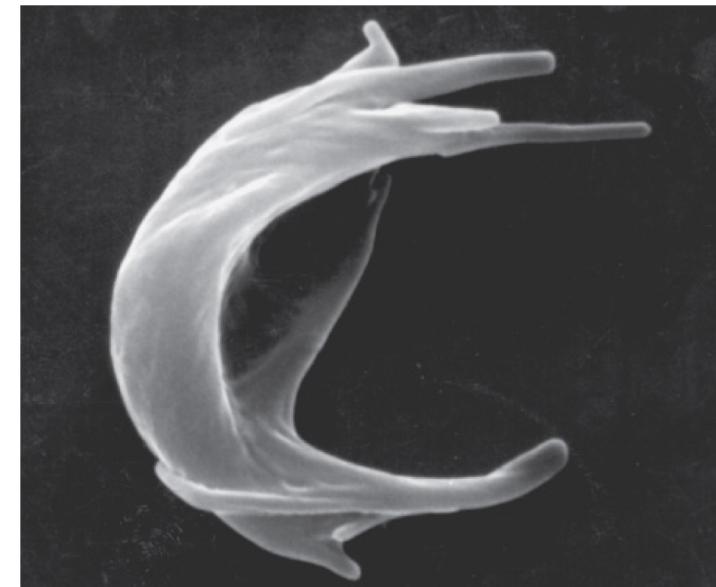
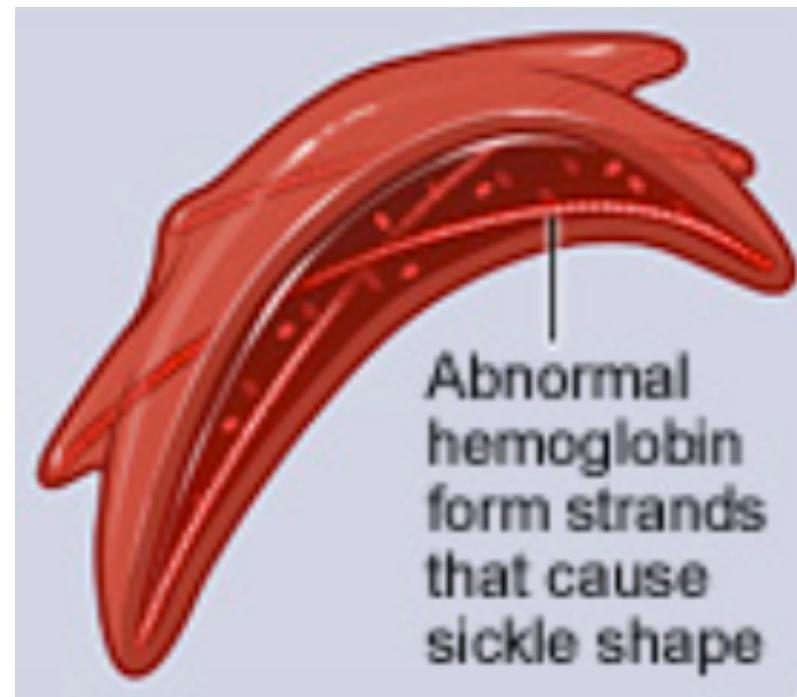


Globine β : 146 acides aminés
1 seule différence, à la position 6

globule rouge normal :



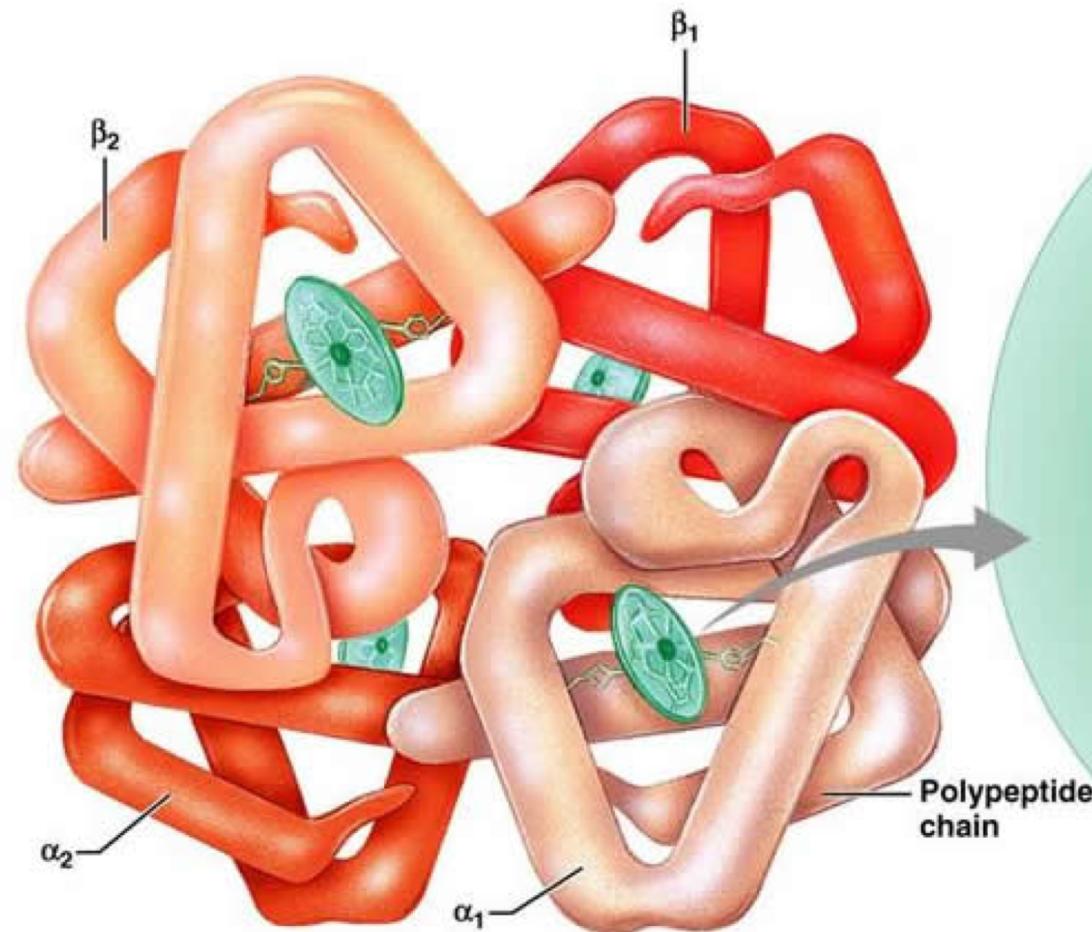
globule rouge en faucille :



Source: Mescher AL: Junqueira's Basic Histology: Text and Atlas, 12th Edition: <http://www.accessmedicine.com>
Copyright © The McGraw-Hill Companies, Inc. All rights reserved.

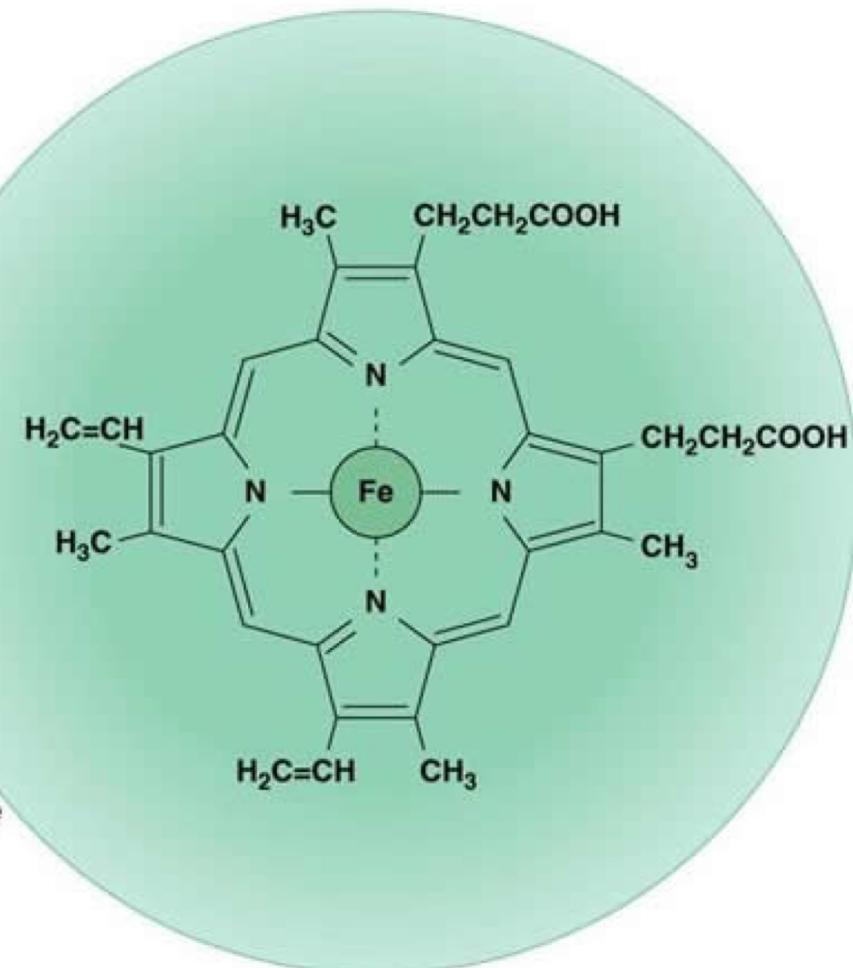
La protéine **hémoglobine** est constituée de 4 polypeptides.

$\alpha_2\beta_2$

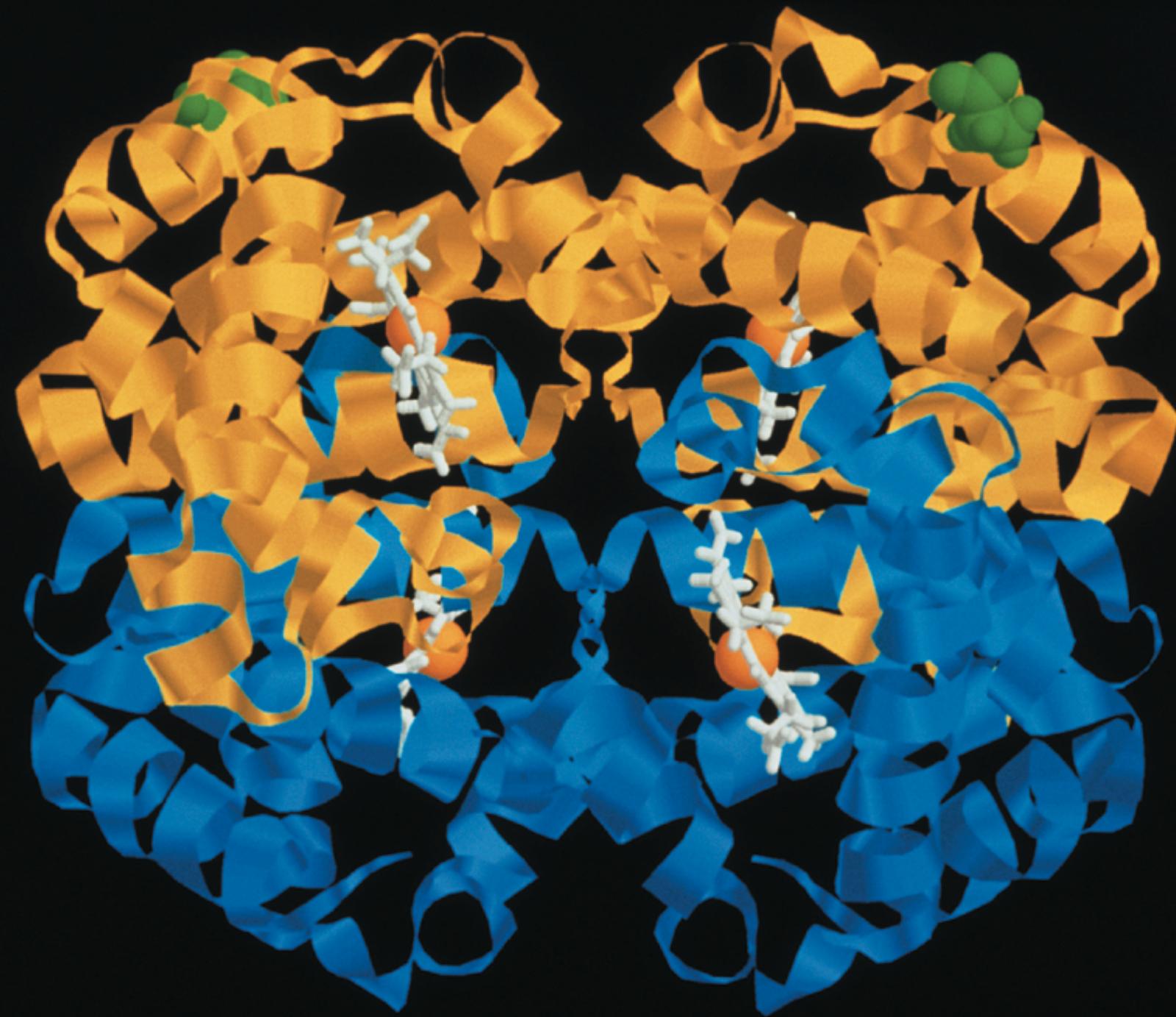


(a) Hemoglobin

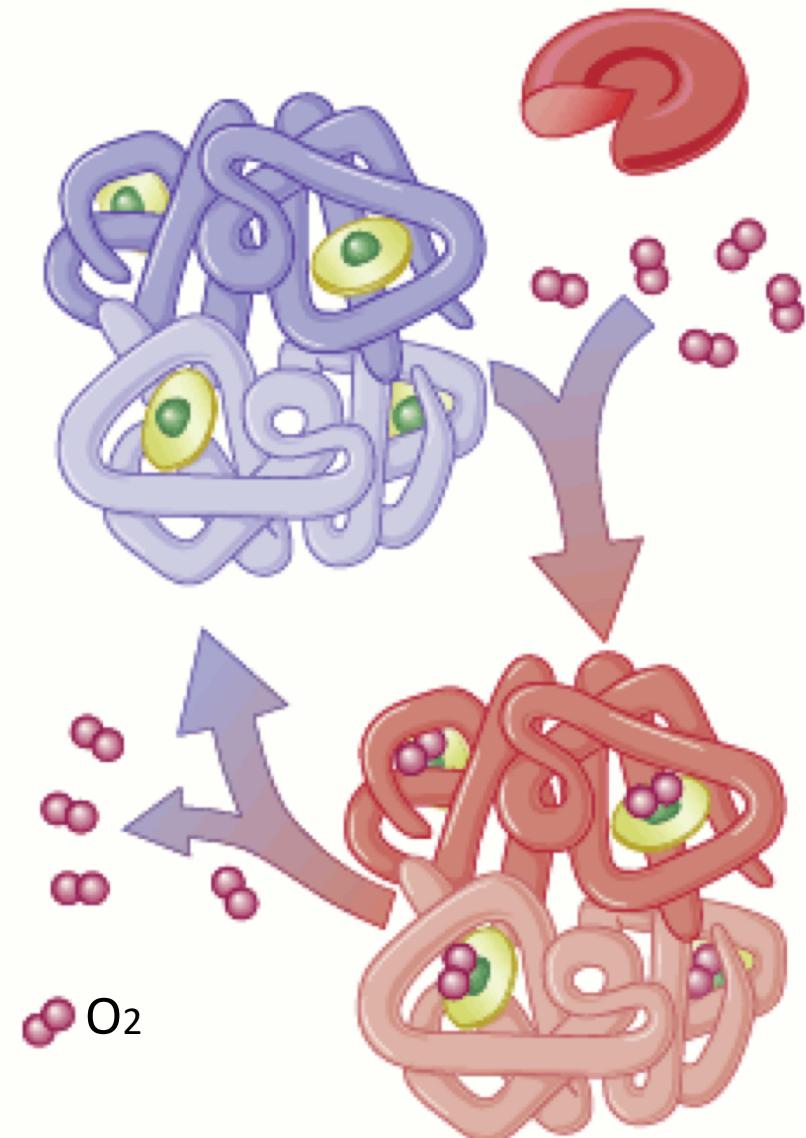
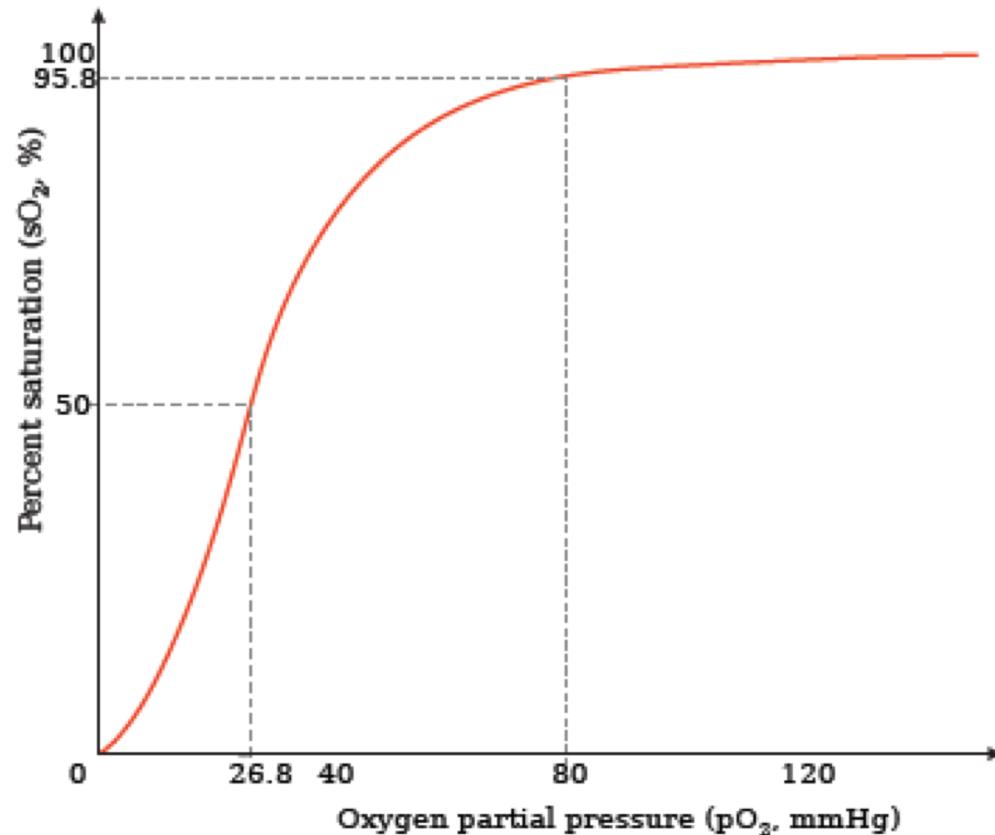
Chaque sous-unité héberge un groupe prosthétique : un **hème**.



(b) Iron-containing heme group



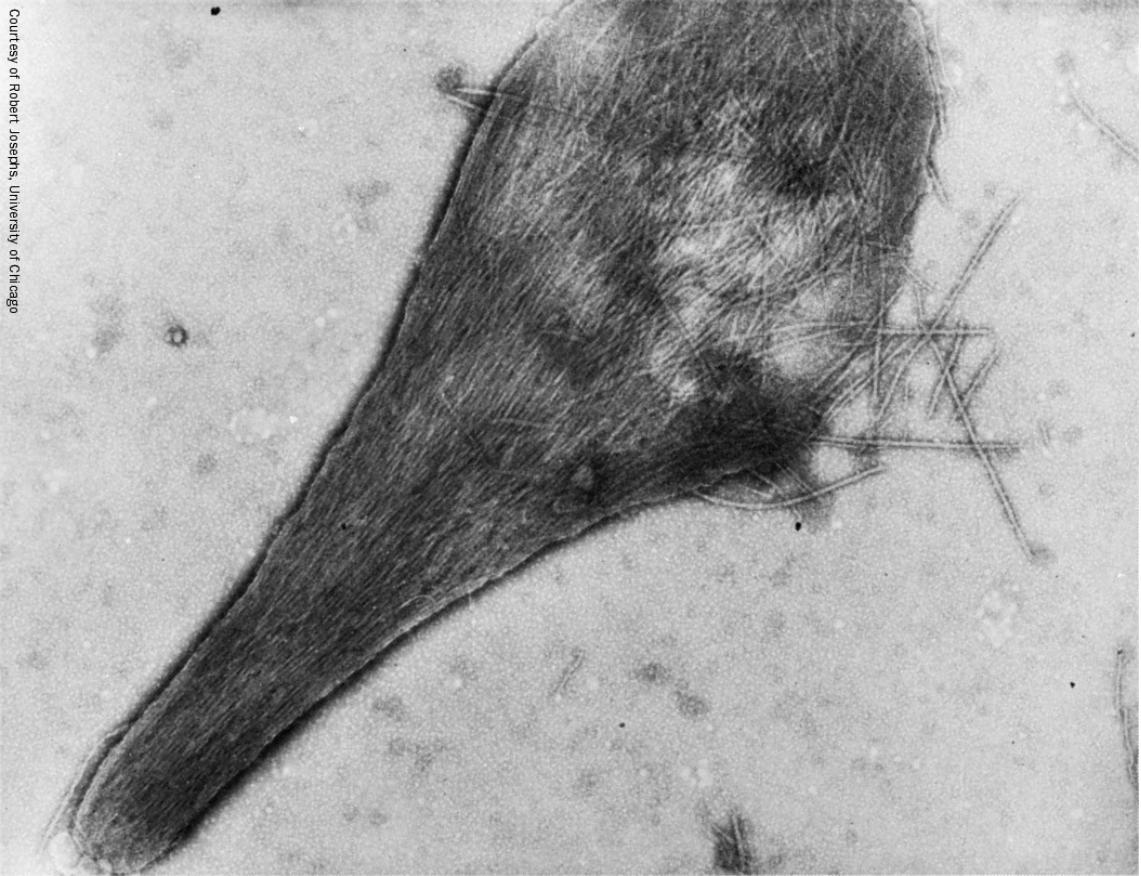
L'hémoglobine est un transporteur d'O₂.



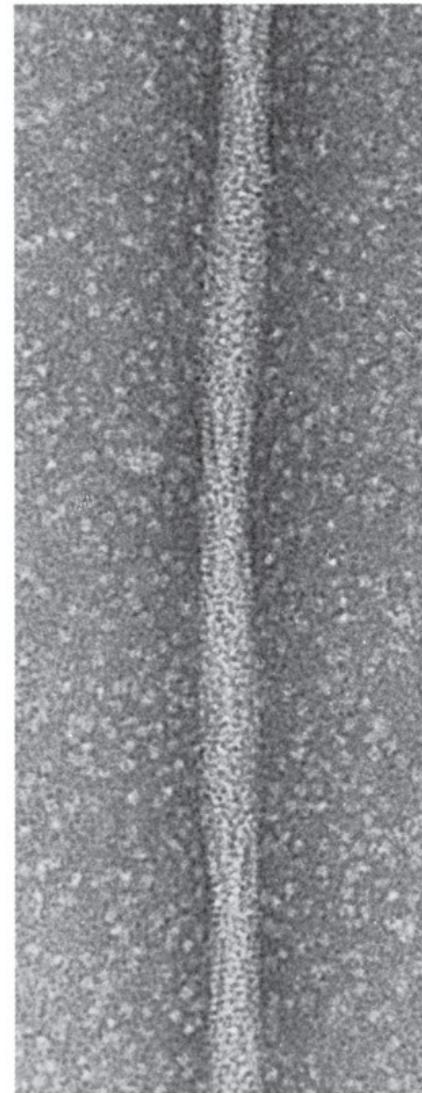
Pourcentage d'hémoglobine ayant fixé du dioxygène, en fonction de la pression partielle en dioxygène. Courbe de Barcroft de type sigmoïde → effet coopératif

4 O₂ sont liés.

Fibres d'hémoglobine S



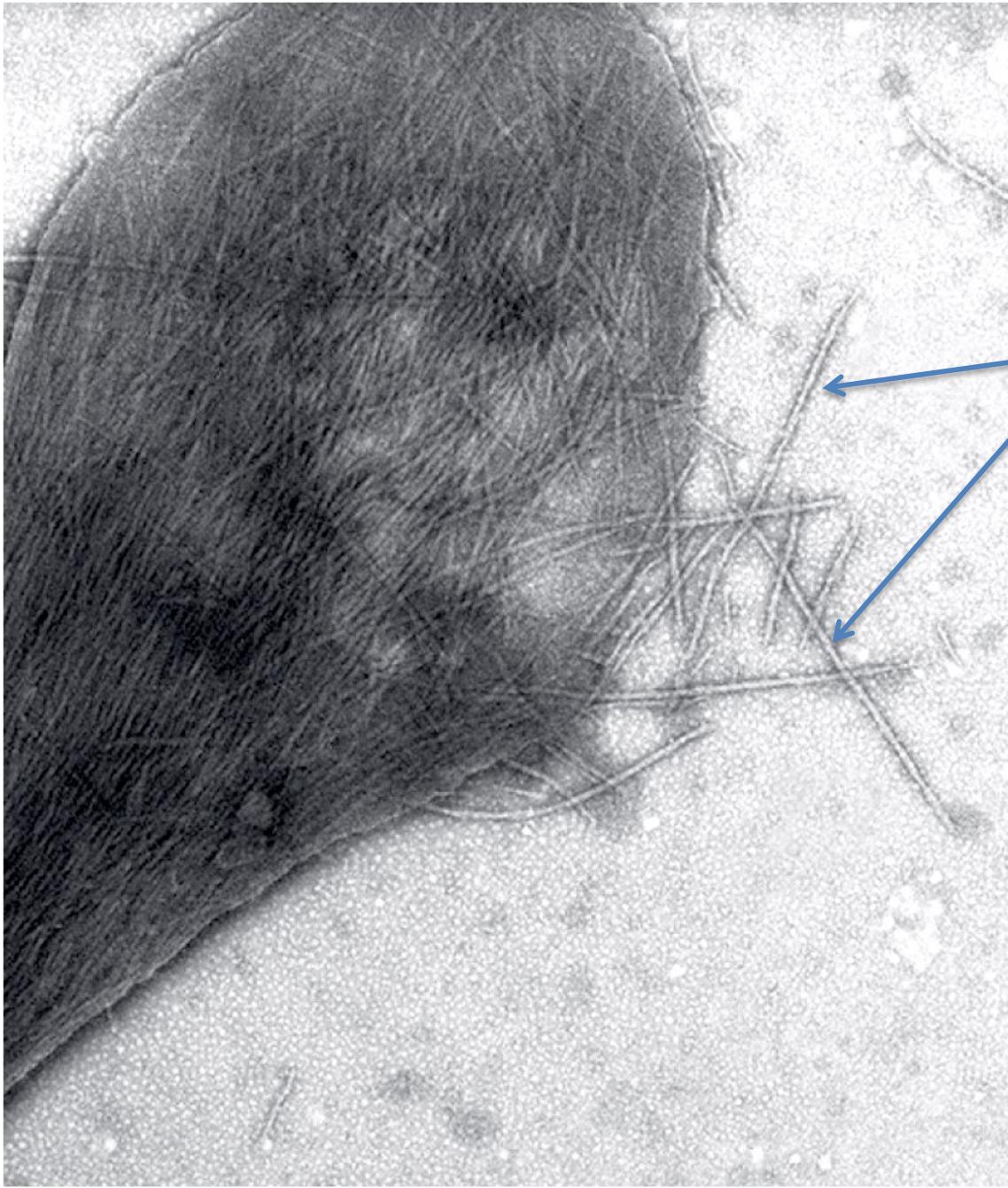
Courtesy of Robert Josephs, University of Chicago



(a)

Courtesy of Stuart Edelstein, University of Geneva





Anémie
falciforme

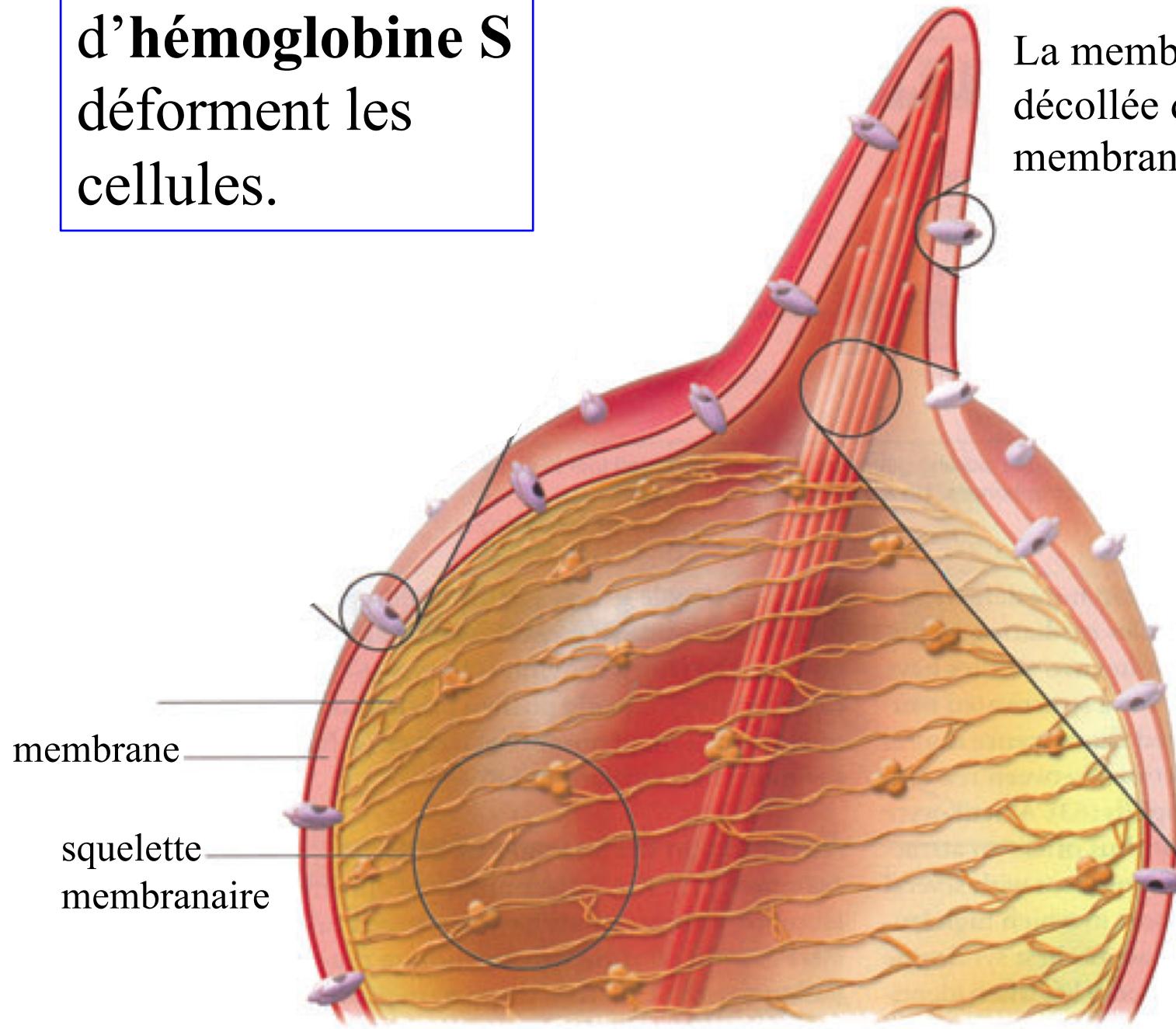
Fibres
d' hémoglobine S

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

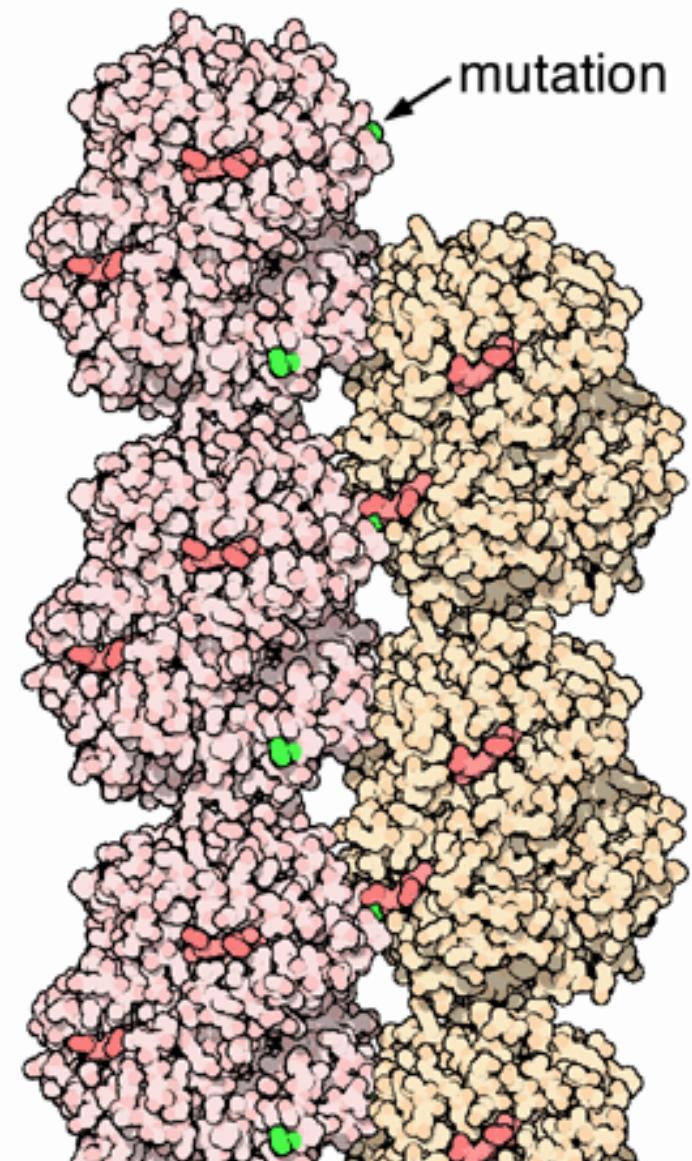
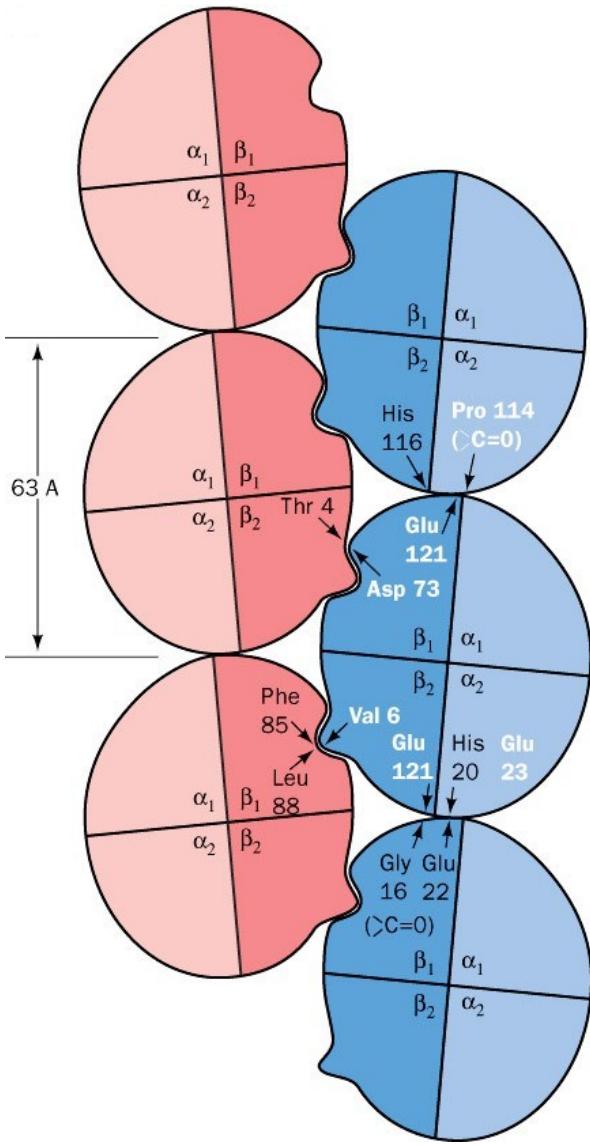
Microscopie électronique à transmission

Les fibres
d'hémoglobine S
déforment les
cellules.

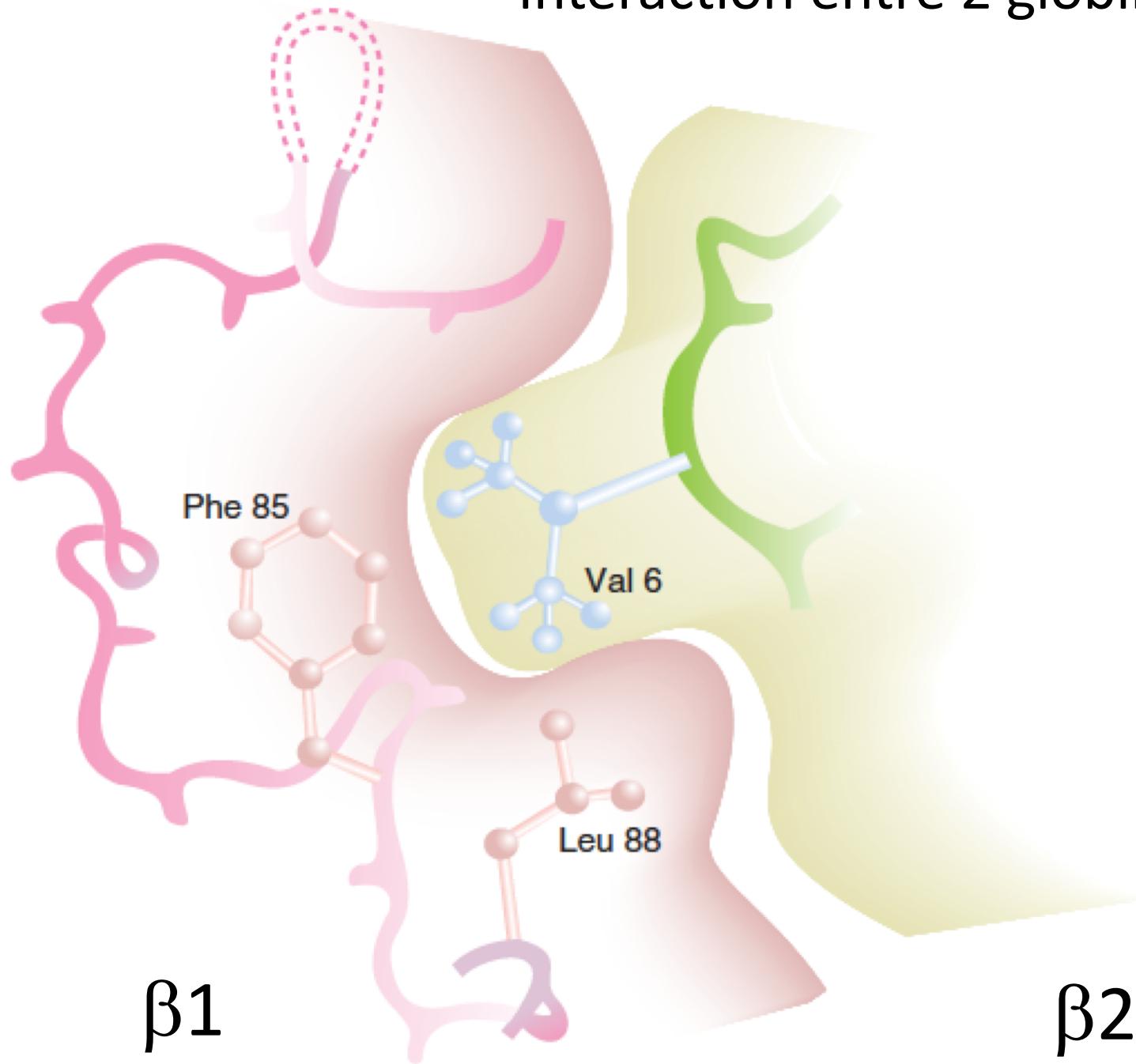
La membrane est
décollée du squelette
membranaire.



Assemblage de la désoxyhémoglobine S en brins.



Interaction entre 2 globines β^S



Assemblage en fibres de l'hémoglobine S.

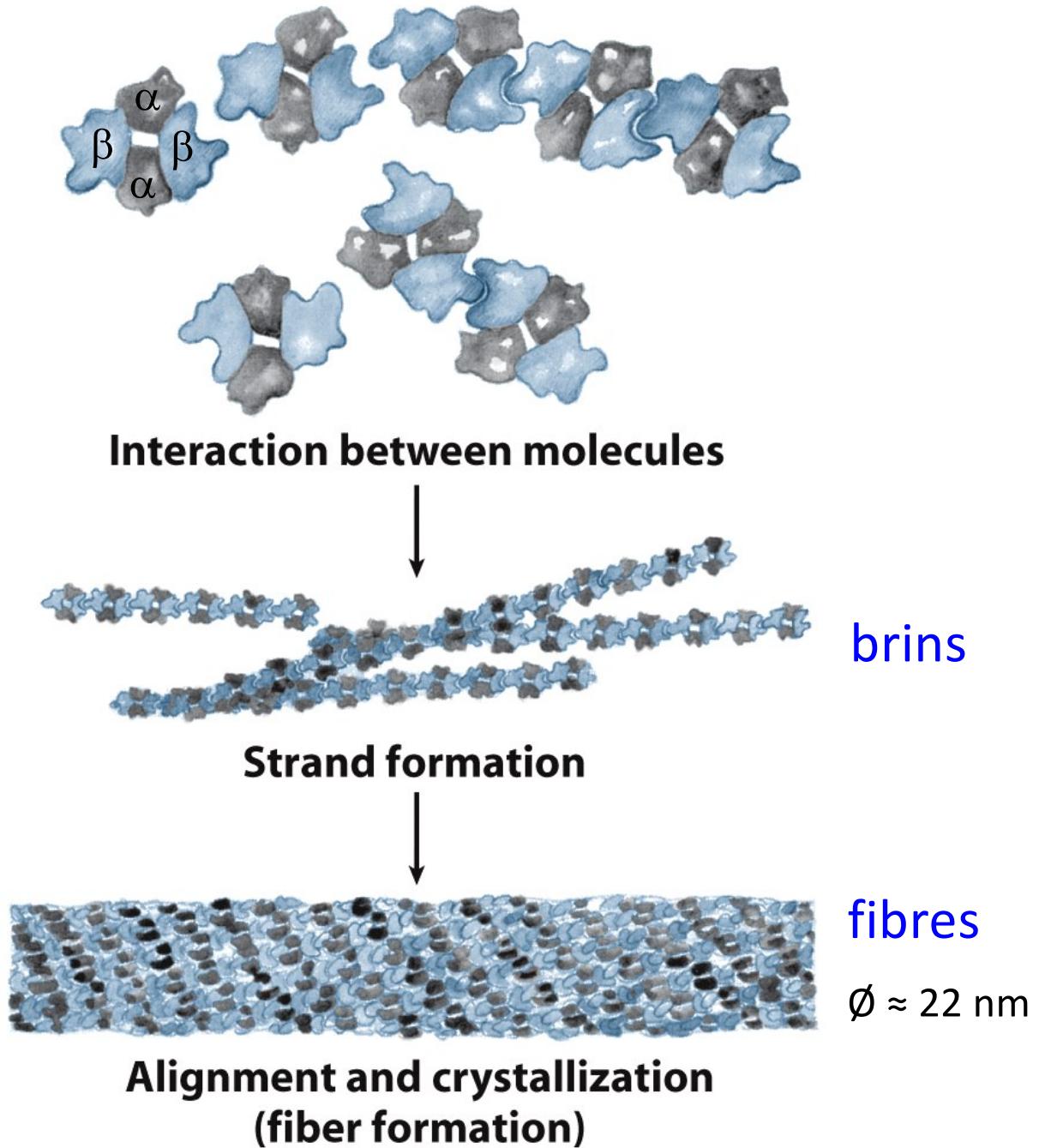
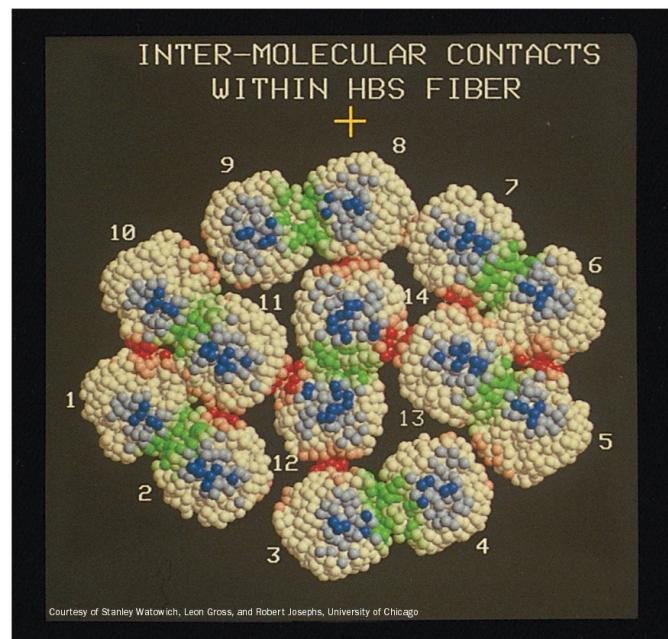
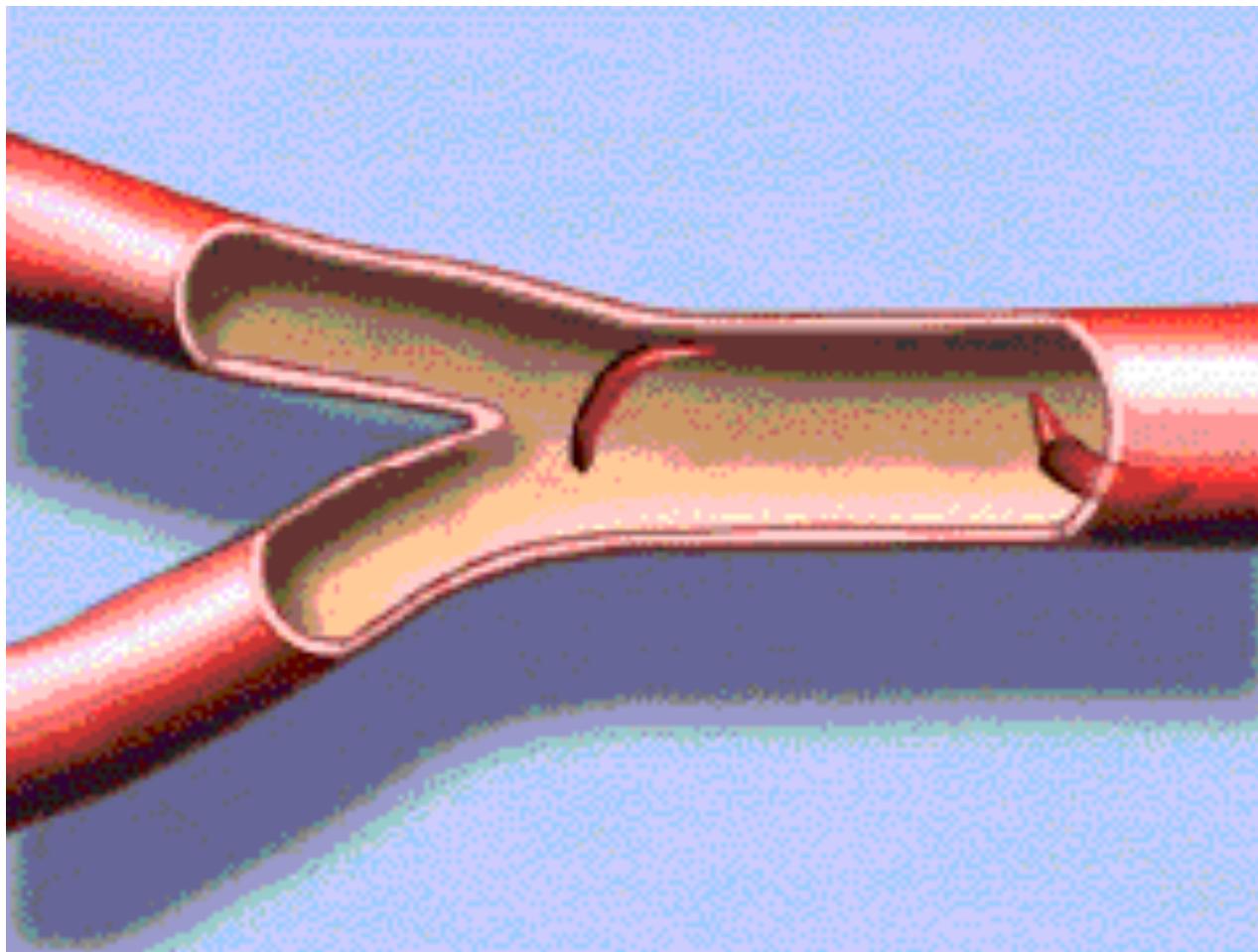
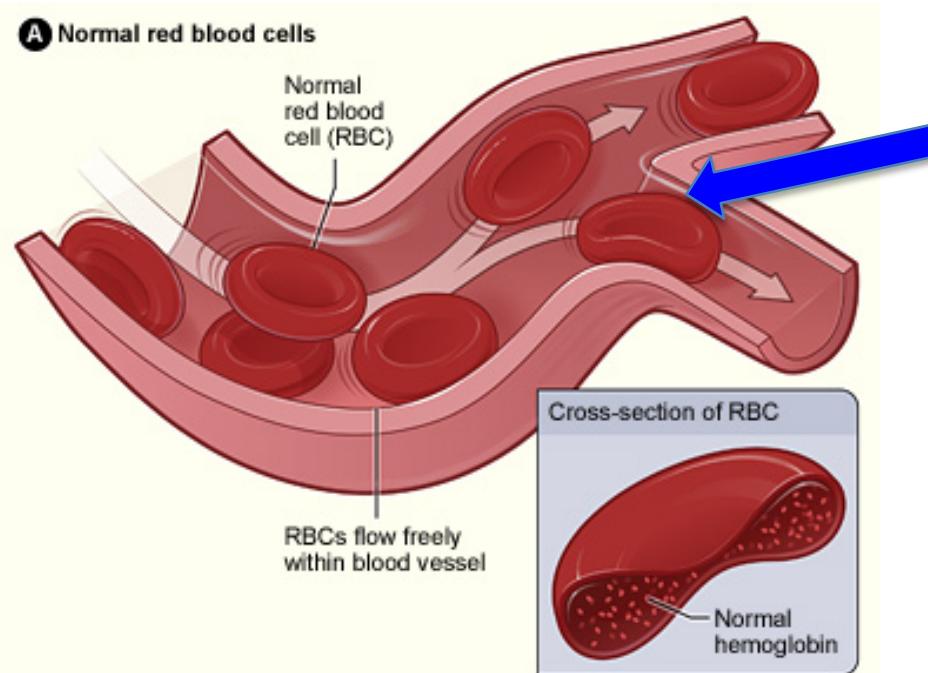


Figure 5-20b
Lehninger Principles of Biochemistry, Fifth Edition
© 2008 W. H. Freeman and Company

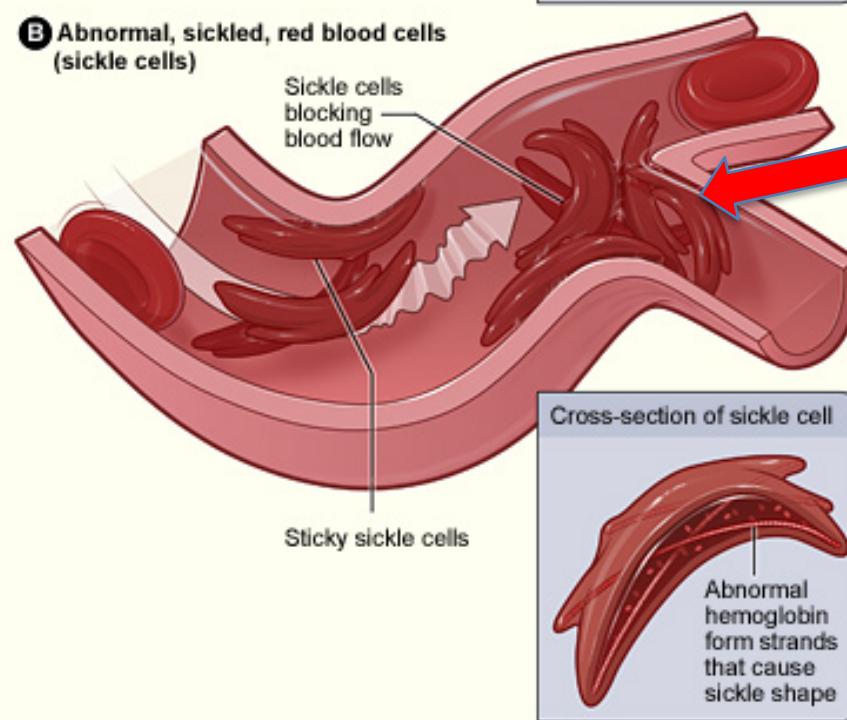
Blockage de petits capillaires



Échelle phénotypique : macroscopique



Ce globule rouge
se déforme pour
passer.



Obstruction
d'un petit
vaisseau
→ douleur

Les échelles du phénotype de l'anémie falciforme :

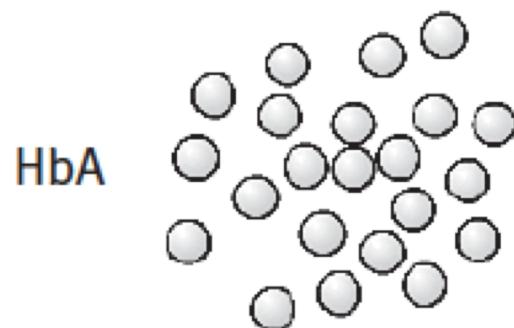
Échelle cellulaire :



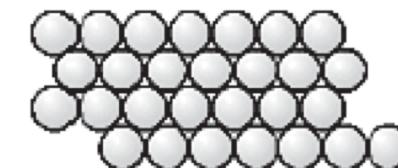
Microscopie
optique



Échelle moléculaire :



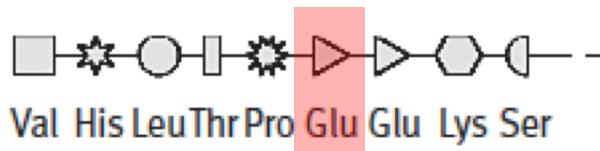
Microscopie
électronique



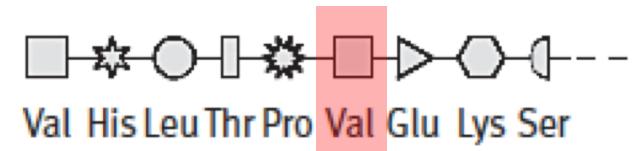
Protéines indépendantes

Protéines assemblées en fibres

Chaîne β de
l'hémoglobine
HbA



Chaîne β de
l'hémoglobine
HbS



Analyse
biochimique