

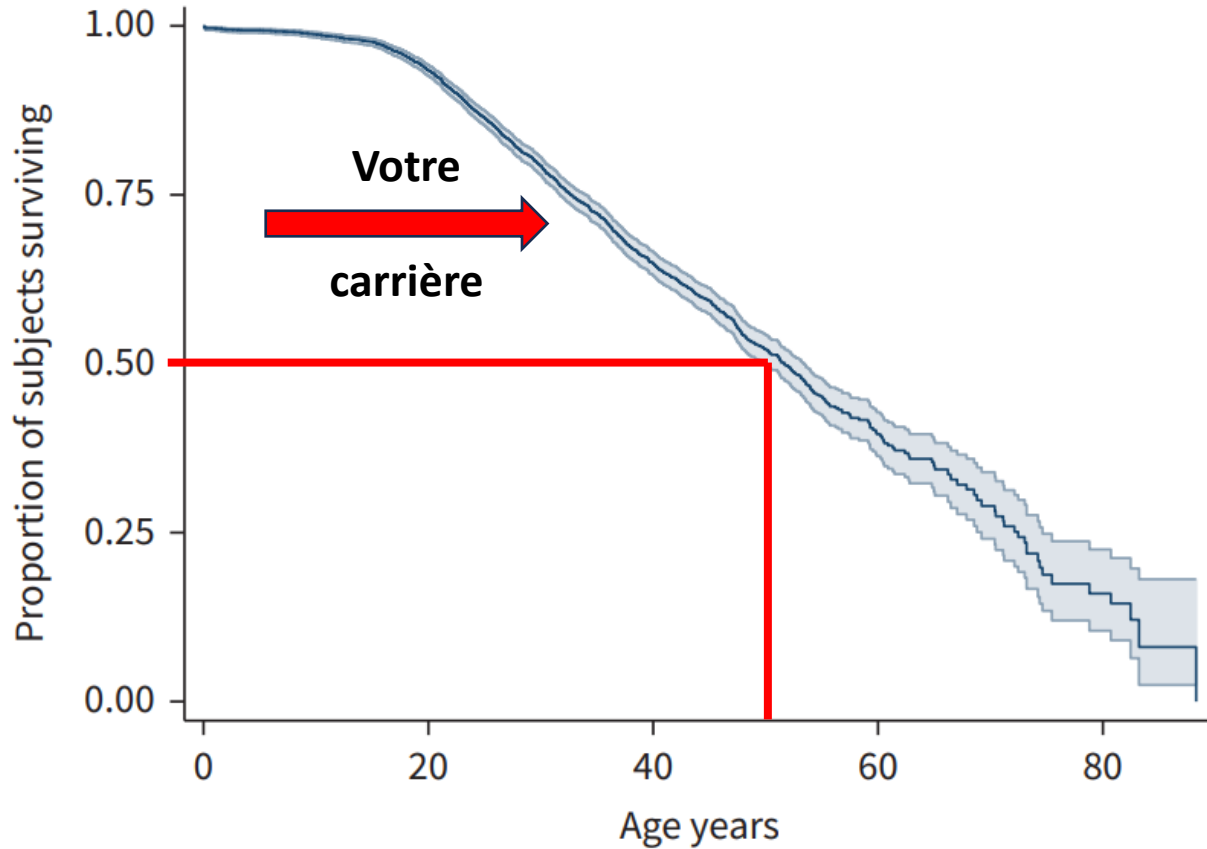
# Manifestations gastrointestinales de la mucoviscidose- de la physiopathologie au symptôme

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Unité de gastroentérologie, hépatologie et nutrition pédiatriques

Dépt Femme Enfant Adolescent

# Espérance de vie



## Causes de décès

- Pulmonaires
- Hépatiques
- Gastrointestinales
- Malnutrition / déficit énergétique
- Infectieuses

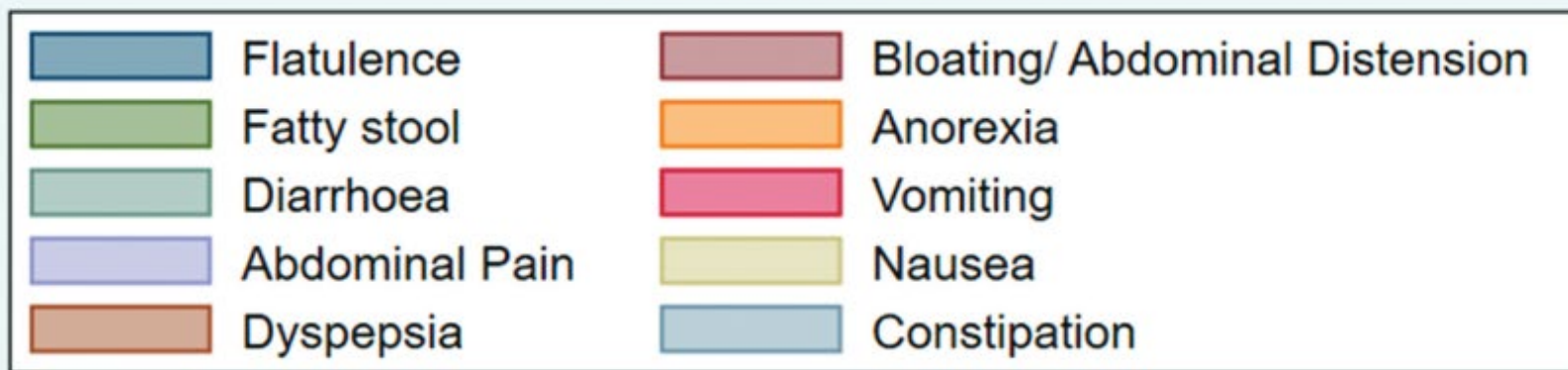
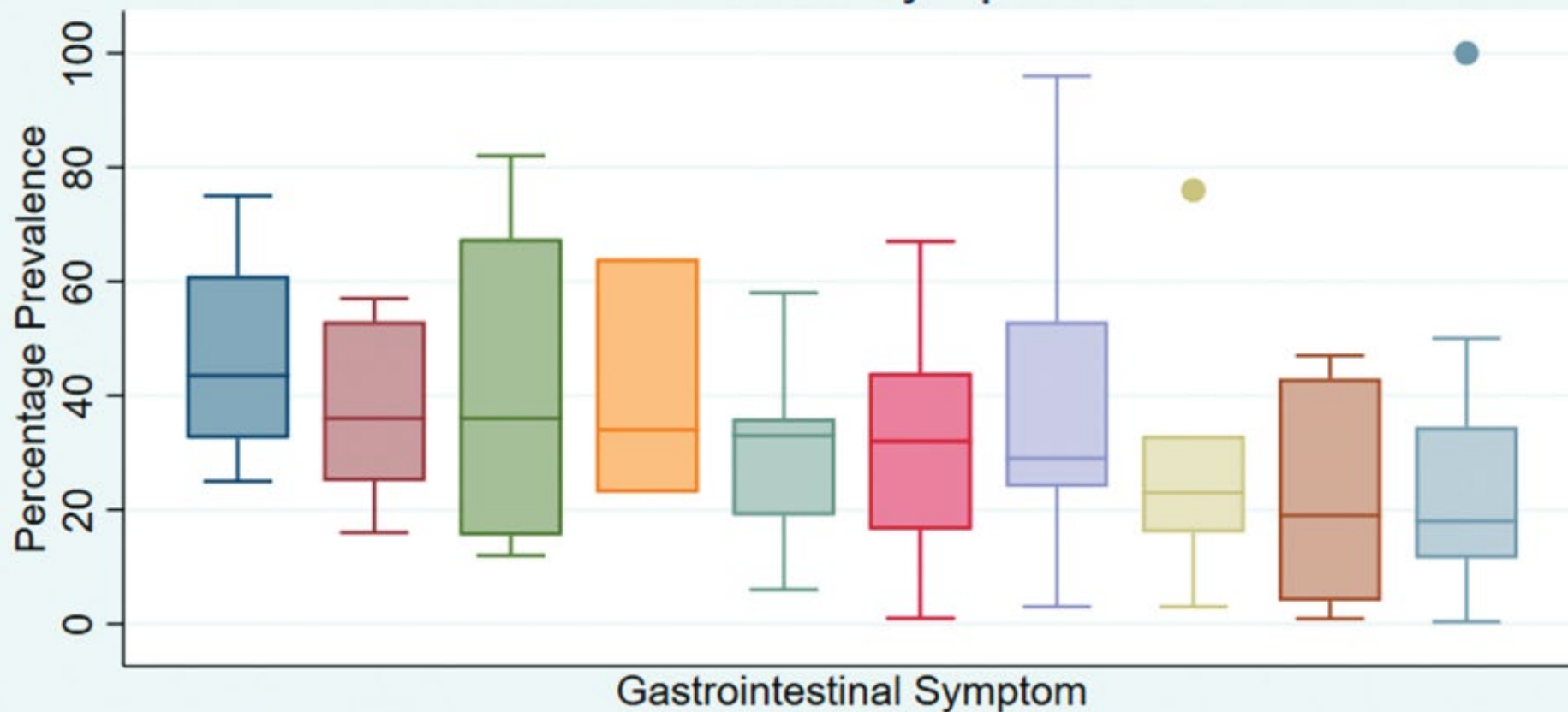
**FIGURE 1** Estimated survival (95% CI) for European cystic fibrosis patients: all-cause mortality.

# Qualité de vie

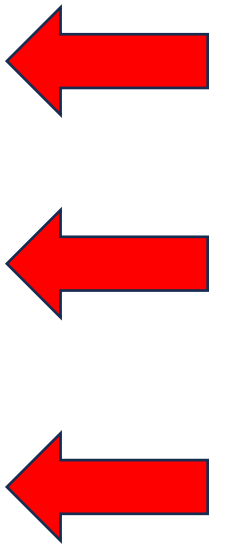
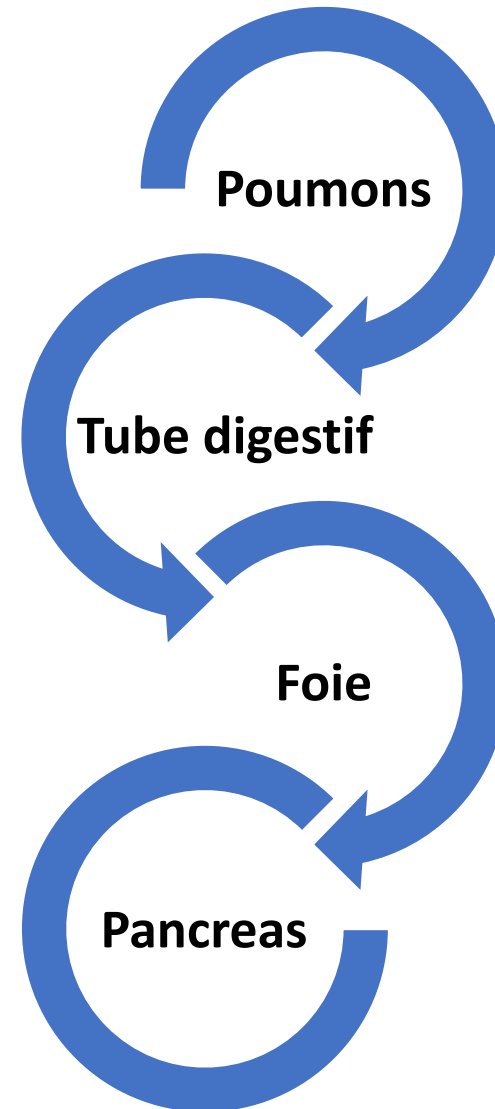
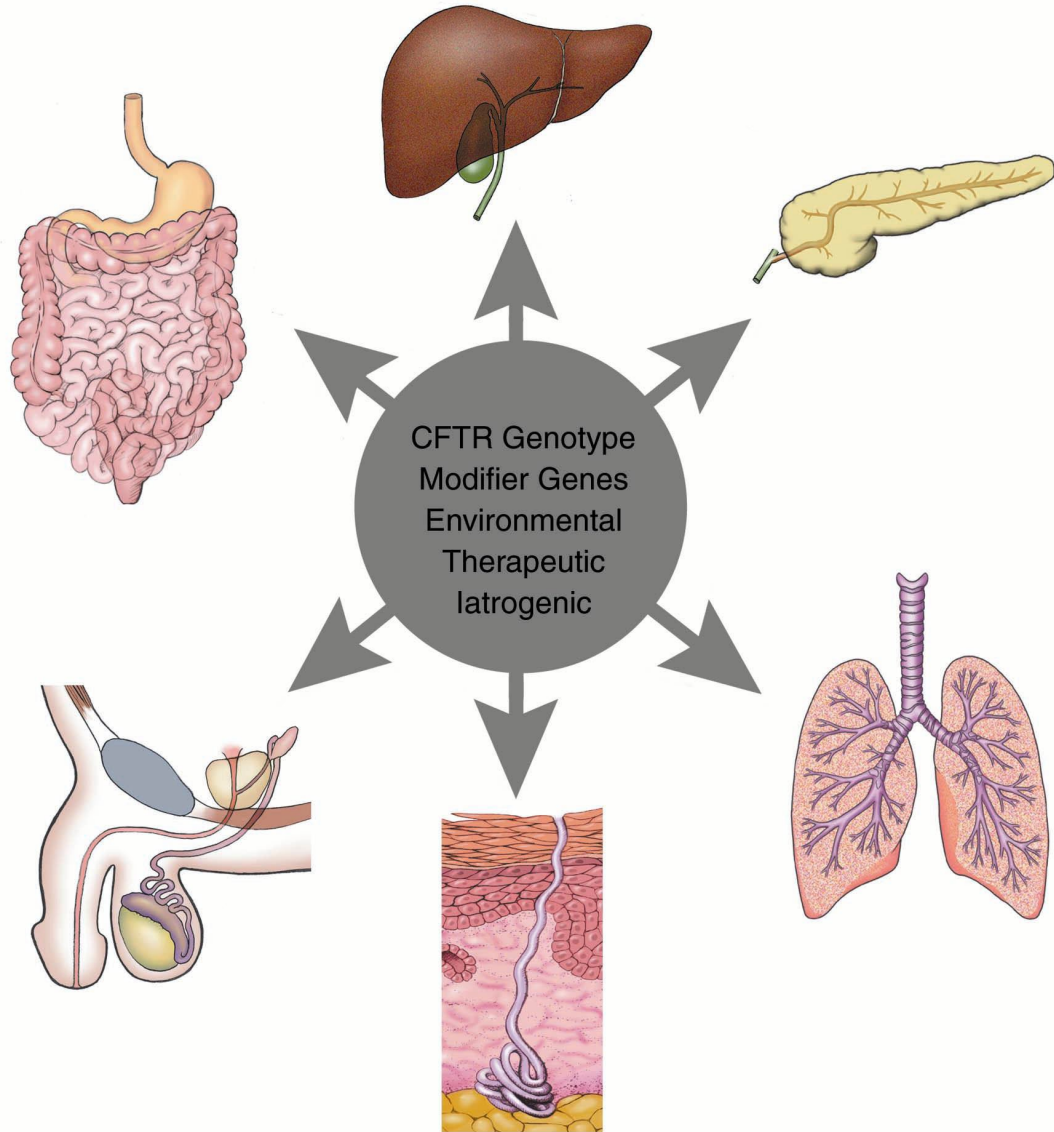


## Les manifestations digestives

## Prevalence of GI Symptoms in CF

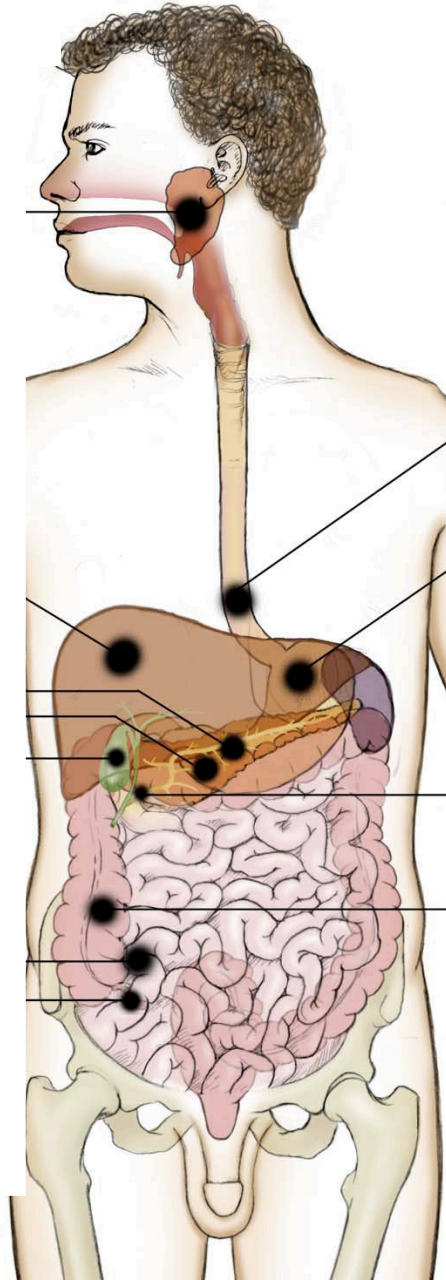


# Maladie multi-système



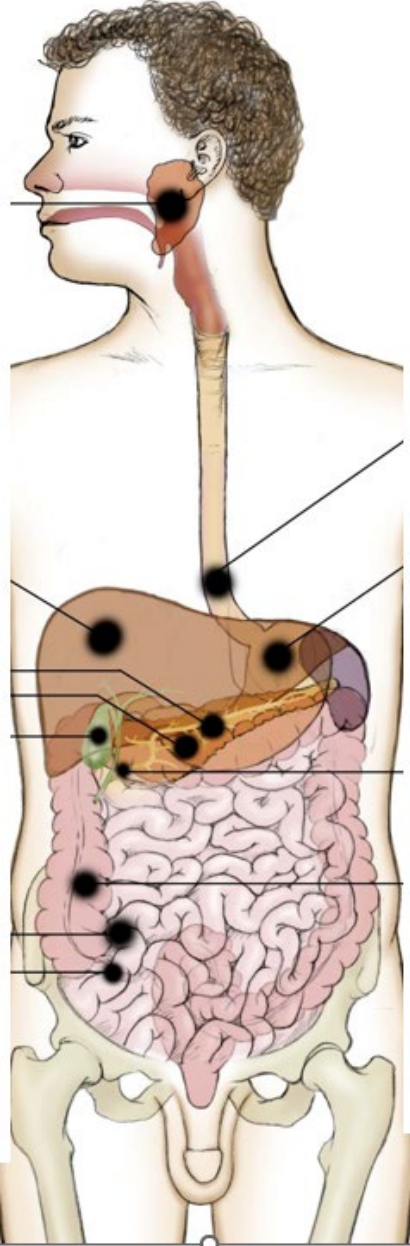
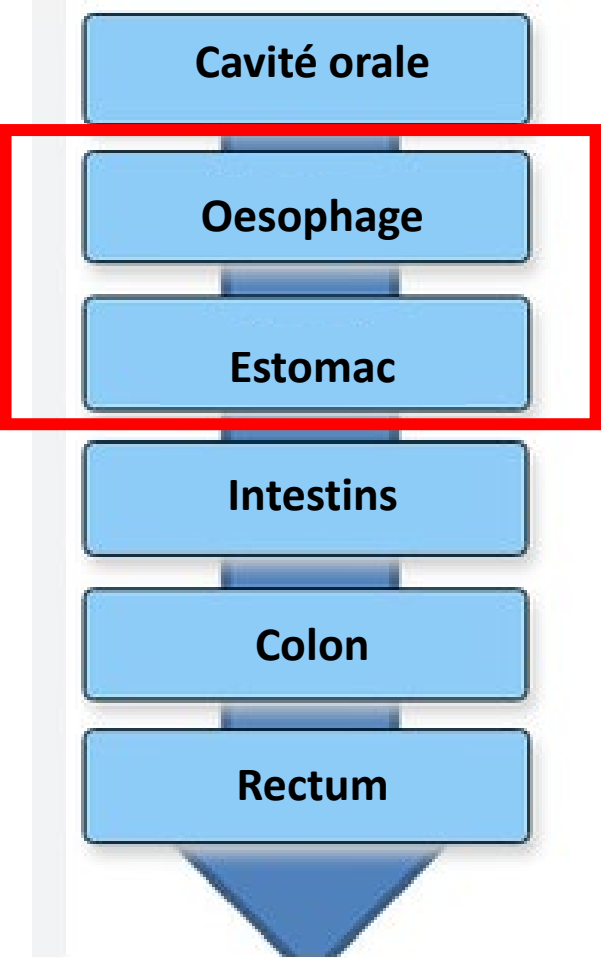
**CFTR dépendantes**  
ou 1°

**CFTR indépendantes**  
ou 2°

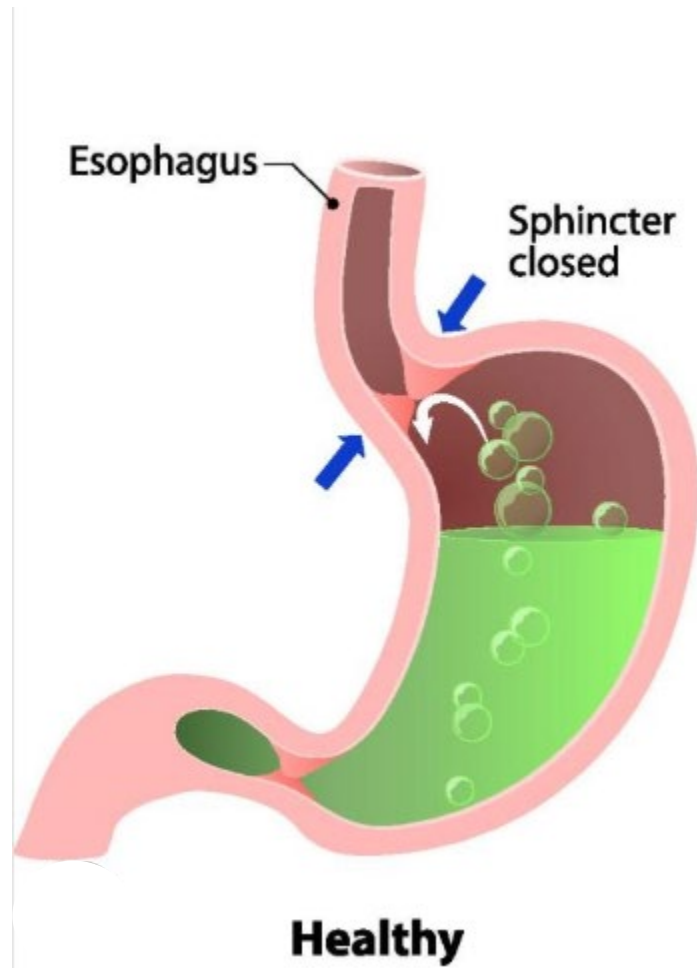


# Tube digestif

**Proximal**

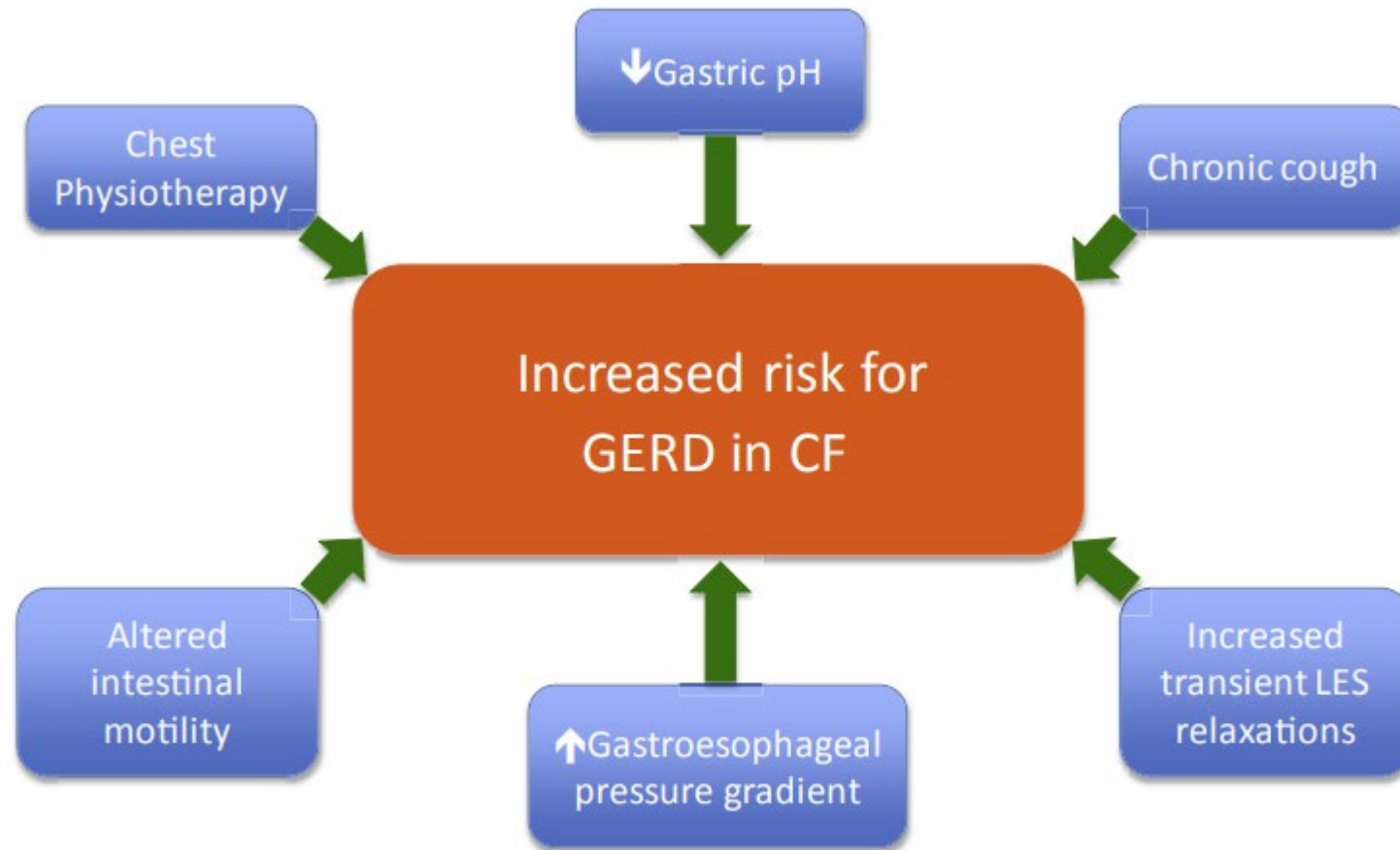


# Physiopathologie → symptôme

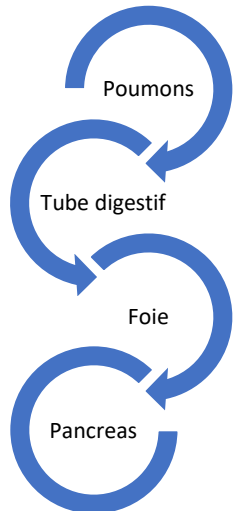


**25% enfants**  
**66% adultes**

# Reflux gastro-oesophagien - physiopathologie

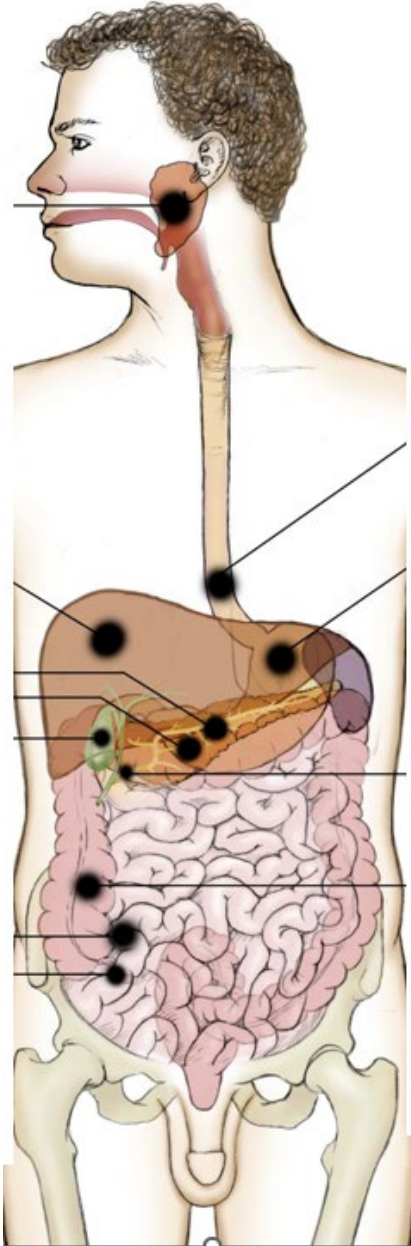
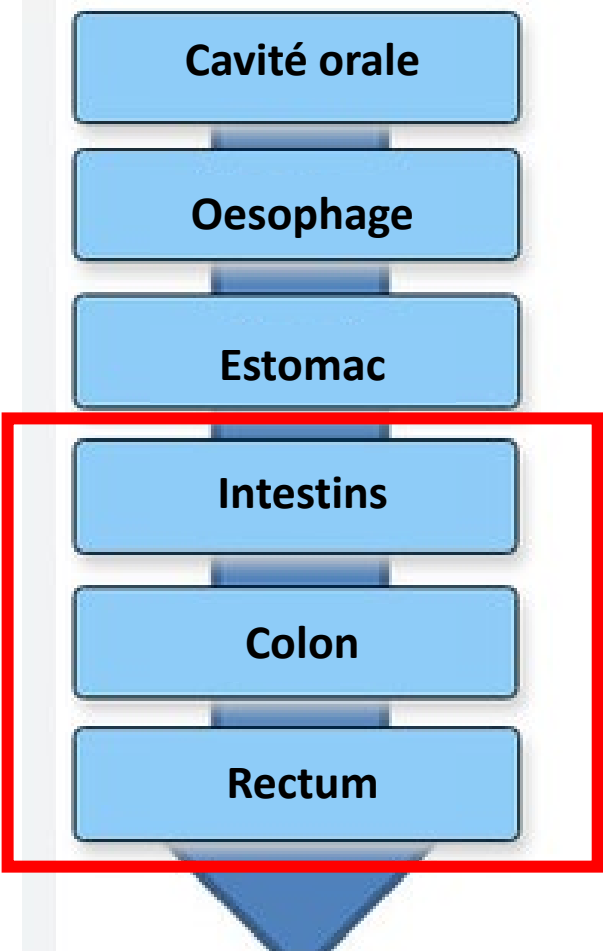


**Fig. 3.** Primary and secondary factors that increase the risk for a patient with CF developing GERD.



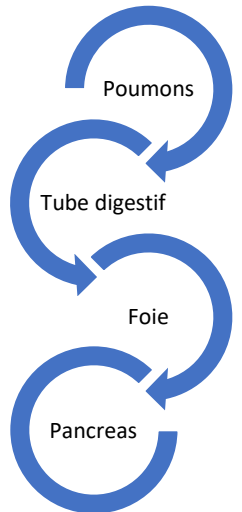
# Tube digestif

**Distal**



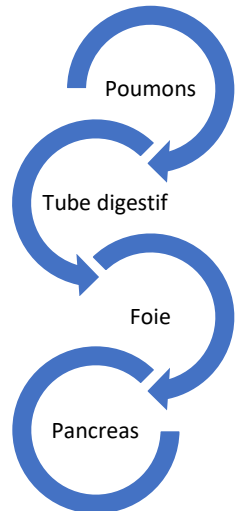
# Tube digestif distal - pathophysiologie

- **Chyme épais** (pH, contenu en H<sub>2</sub>O) → motricité
- **Réflex gastro-iléal** ralenti → motricité
  - Repas riches en graisse
- **Pullulation bactérienne** (grêle, colique) → distension
  - Antécédents antibiothérapie
- **Inflammation** → motricité
  - pH
  - Acides biliaires
  - Espèces bactériennes (C.difficile)
- **(Colopathie fibrosante)**

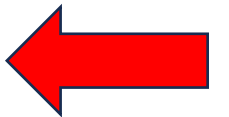
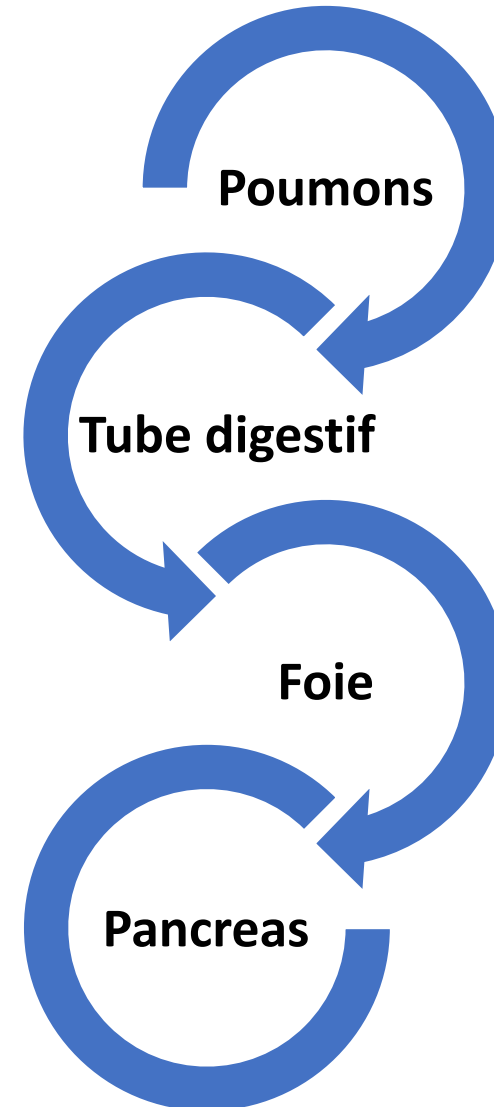
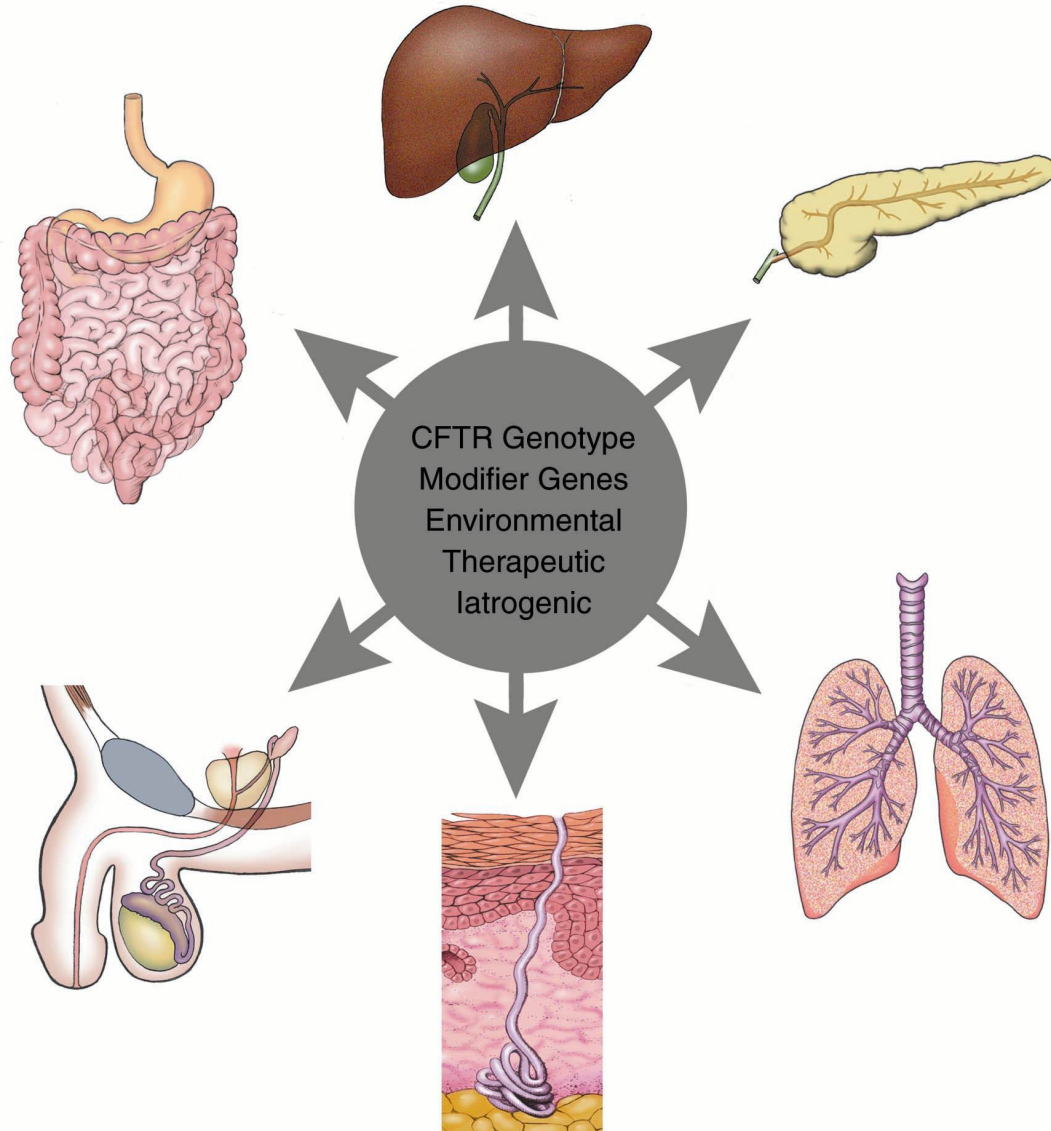


# Tube digestif distal - symptômes

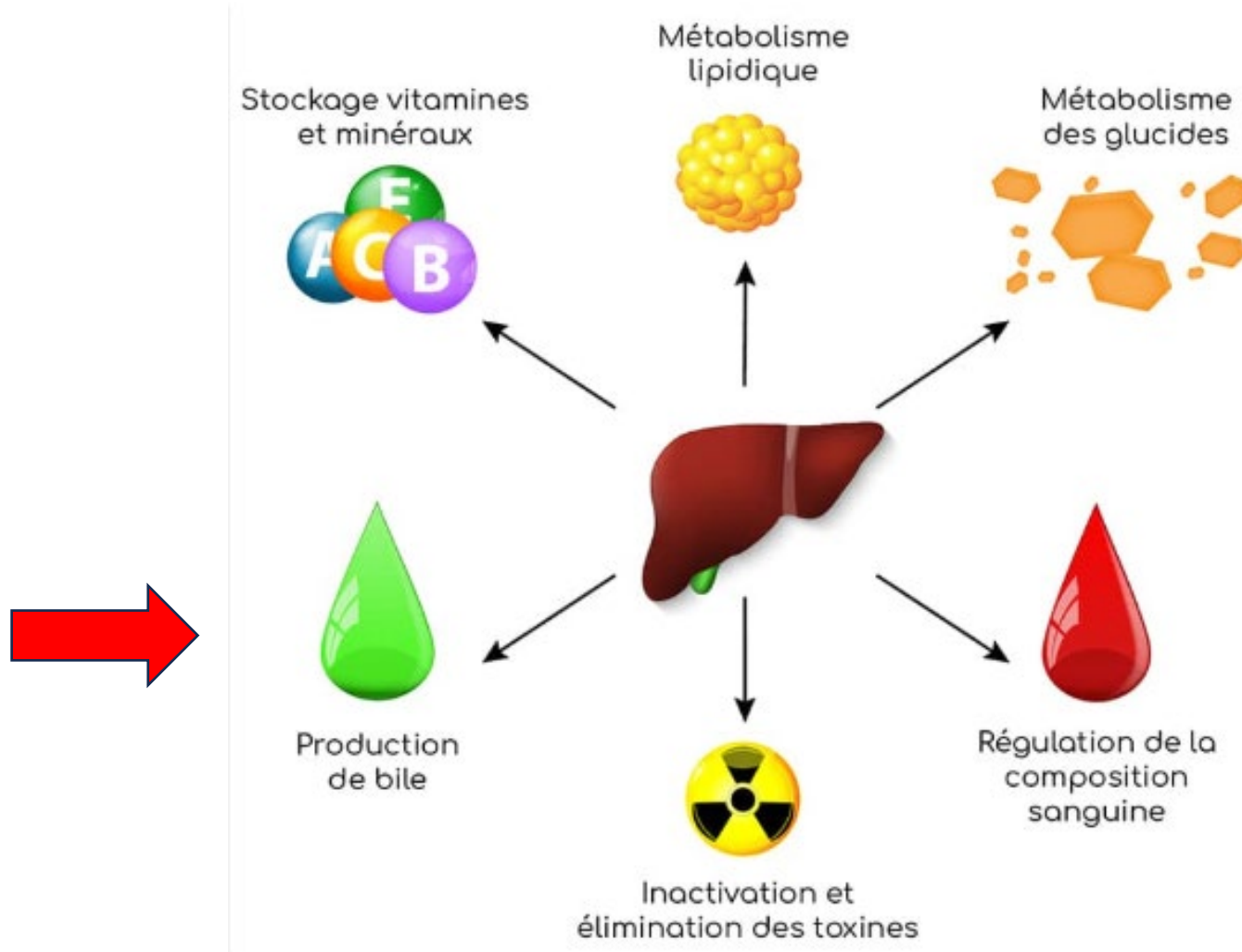
- Iléus méconial (nouveau né)
- Obstruction intestinale distale (enfant, adulte)
- Constipation
- Distension abdominale
- Flatulence
- Augmentation du risque des Ca: grêle, colique



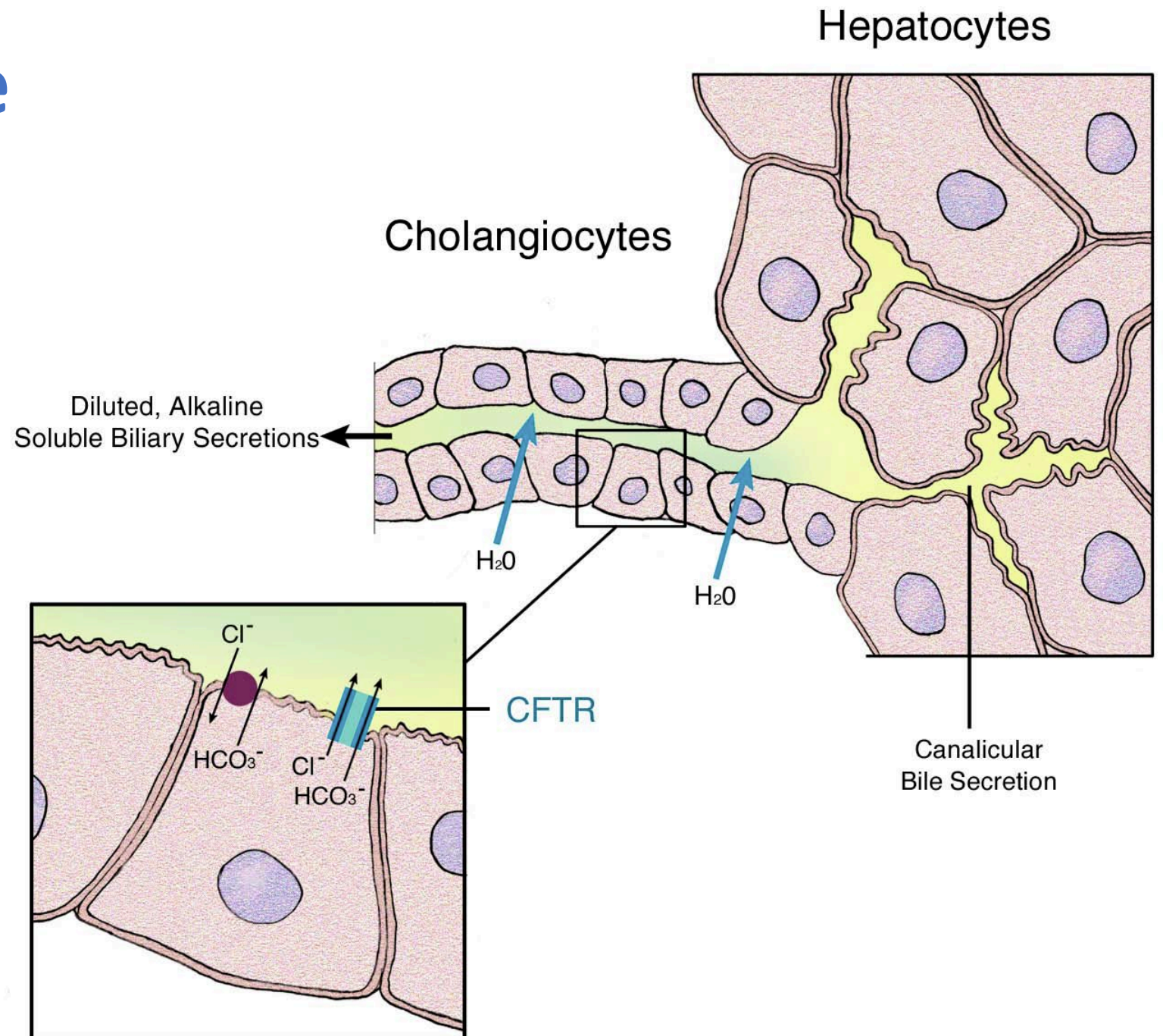
# Maladie multi-système



# Foie- fonctions

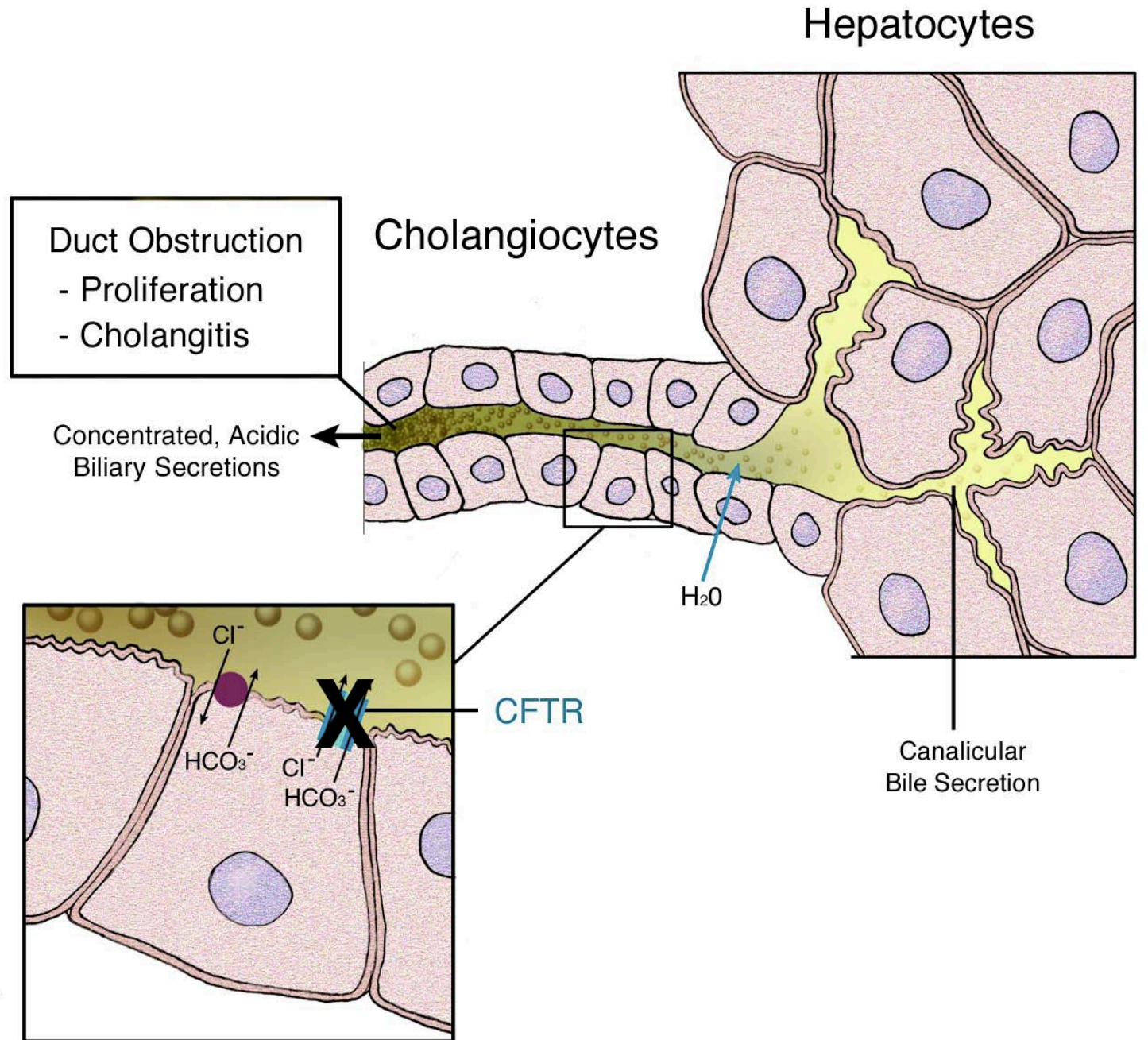


# Atteinte hépatique Physiologie



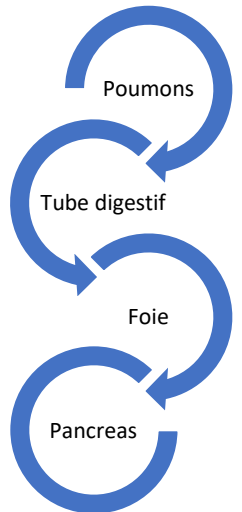
# Atteinte hépatique Pathophysiologie

## Cholestase



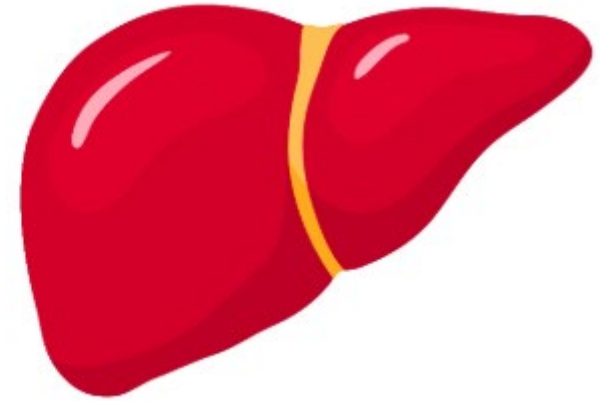
# Atteinte hépatique - symptômes

- Hépatomégalie
- Hypertension portale
- Impact sur la fonction respiratoire → syndrome restrictif



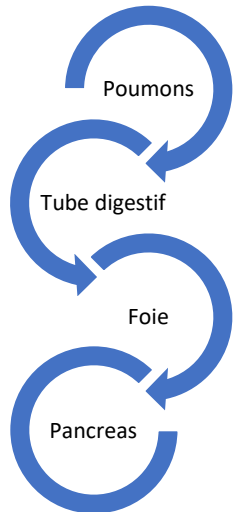
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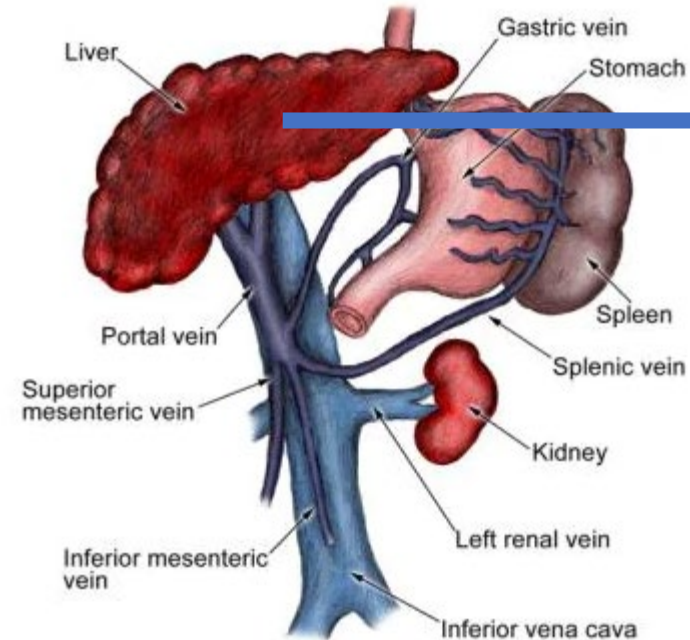


# Atteinte hépatique - symptômes

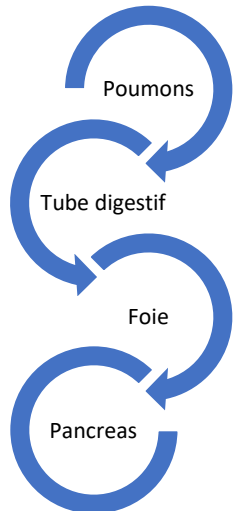
- Hépatomégalie

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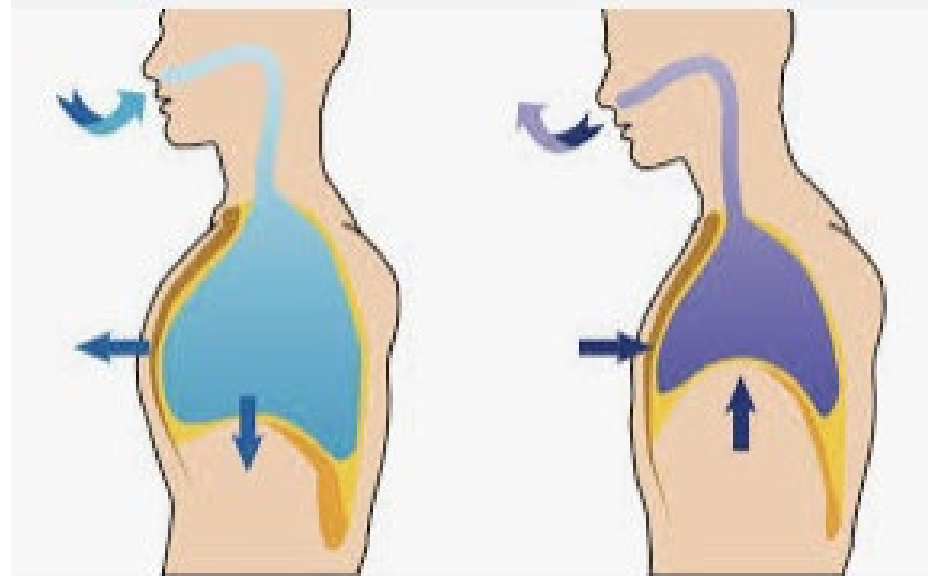


Complications  
pulmonaires de  
la cirrhose



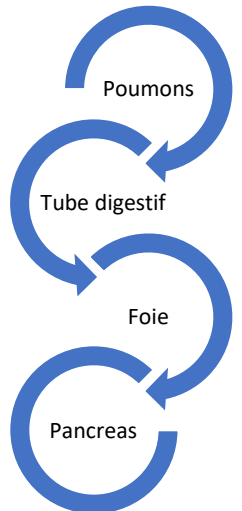
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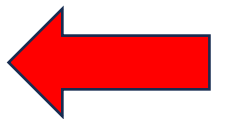
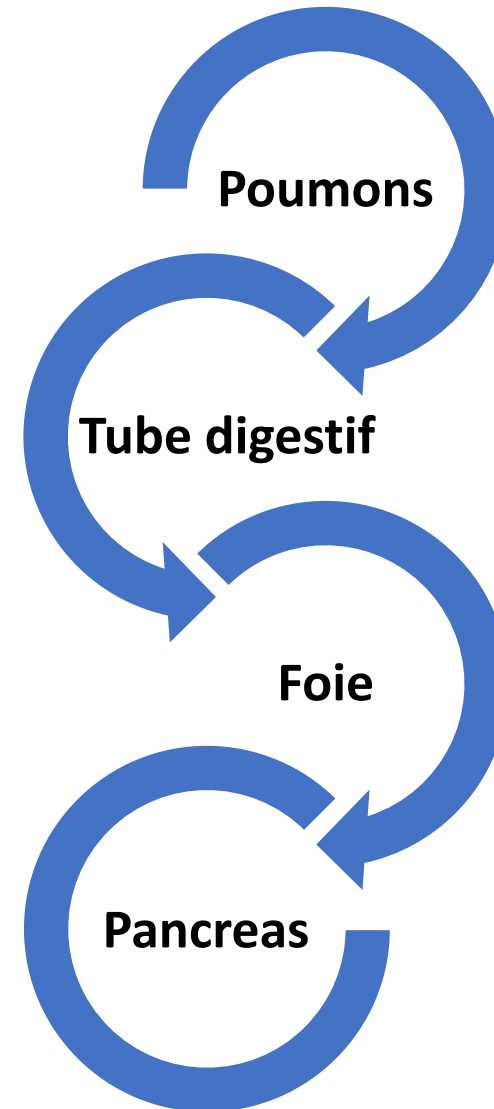
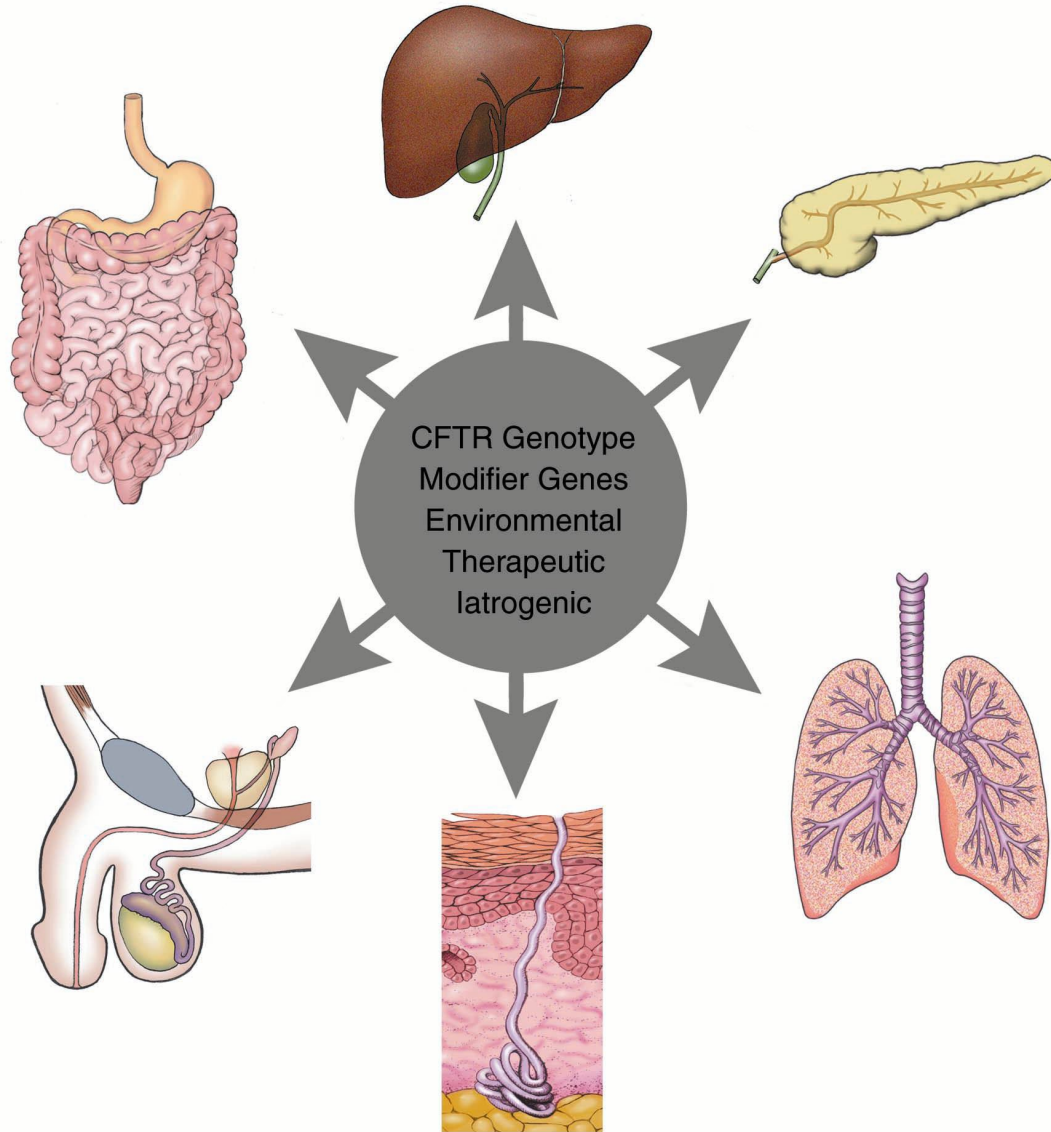


- **Impact sur la fonction respiratoire → syndrome restrictif**

Atteinte hépatique → répercussions respiratoires multiples



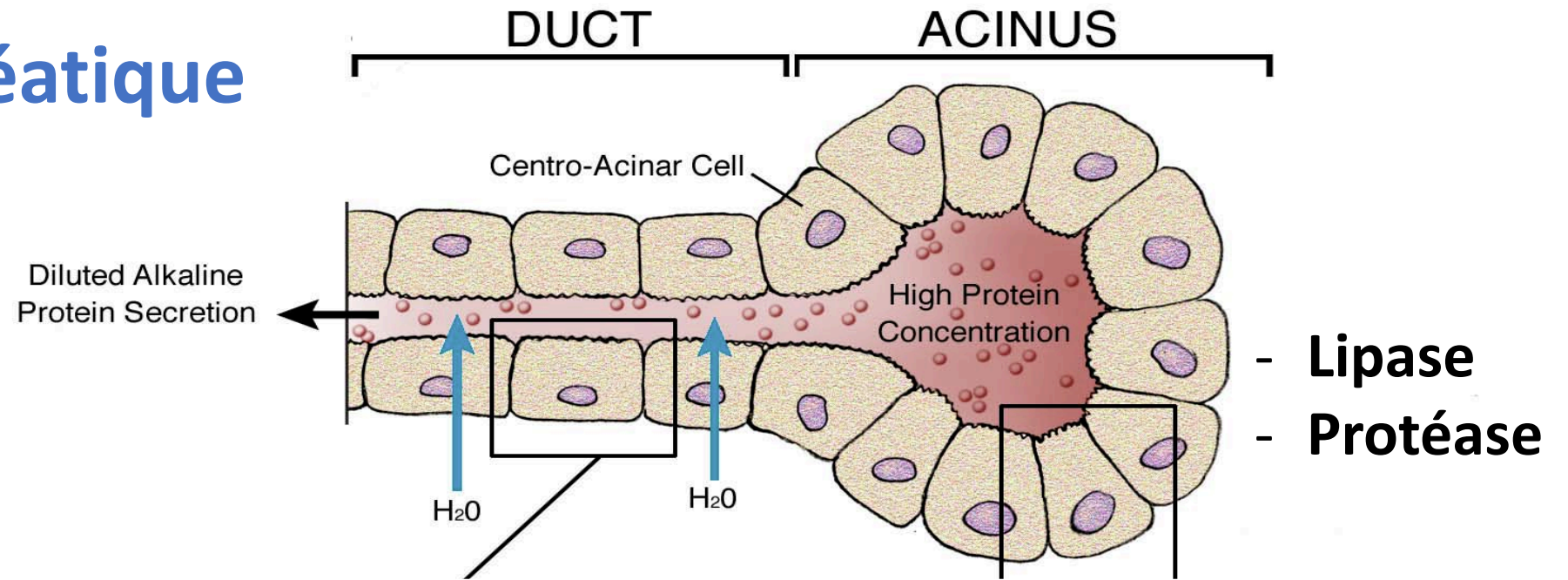
# Maladie multi-système



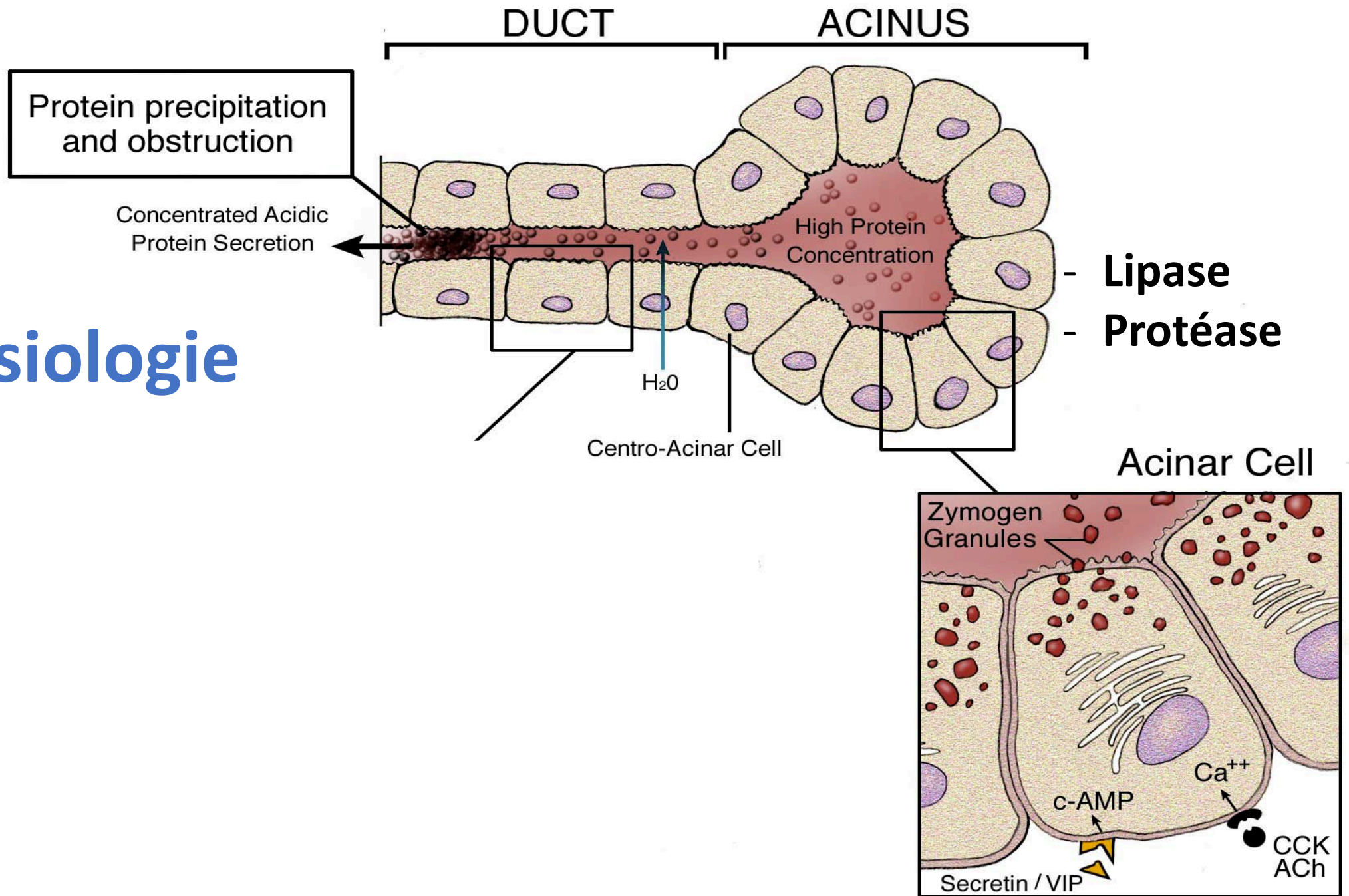
# Atteinte pancréatique

## Physiologie

- Endocrine
- Exocrine



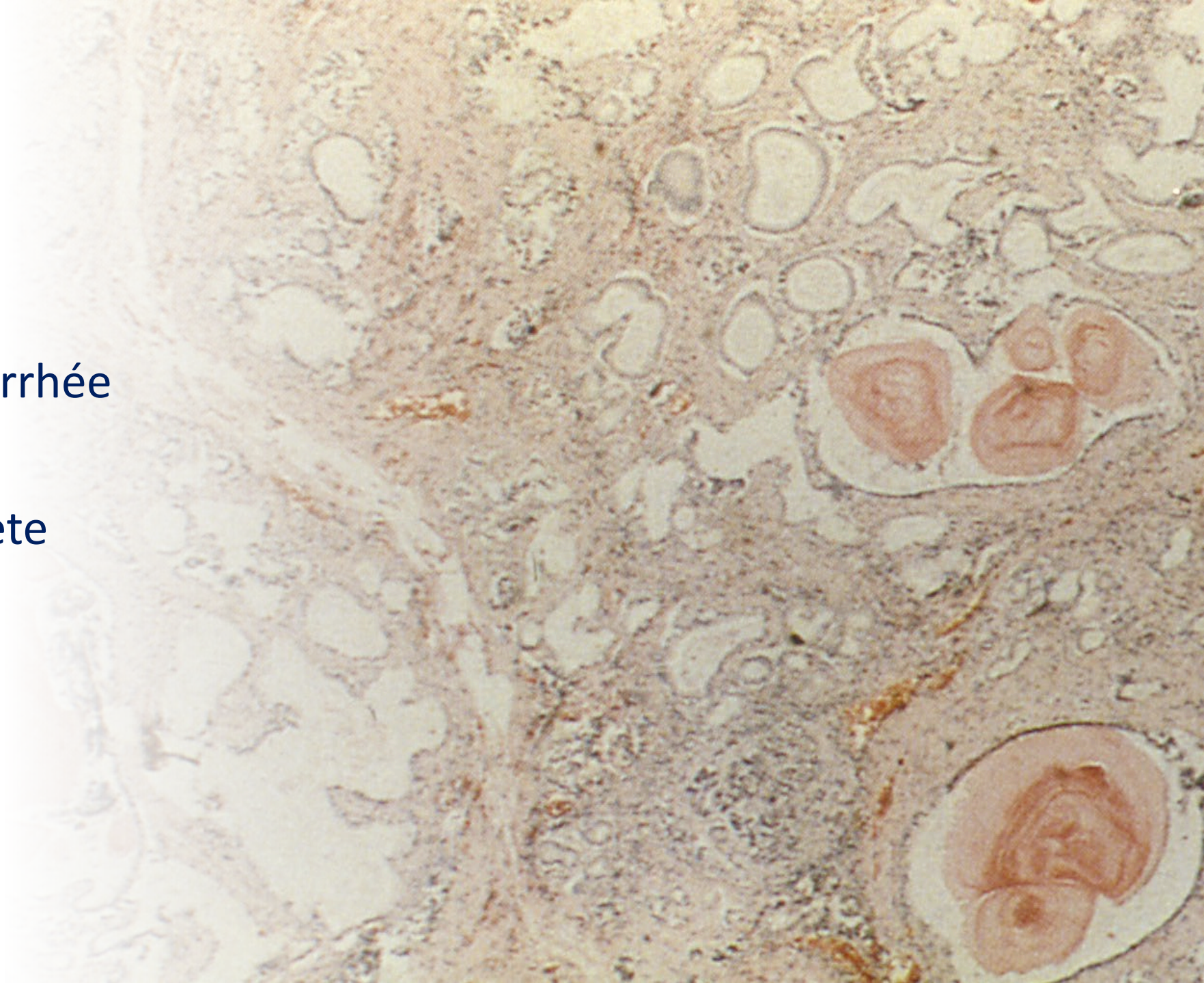
# Pathophysiologie



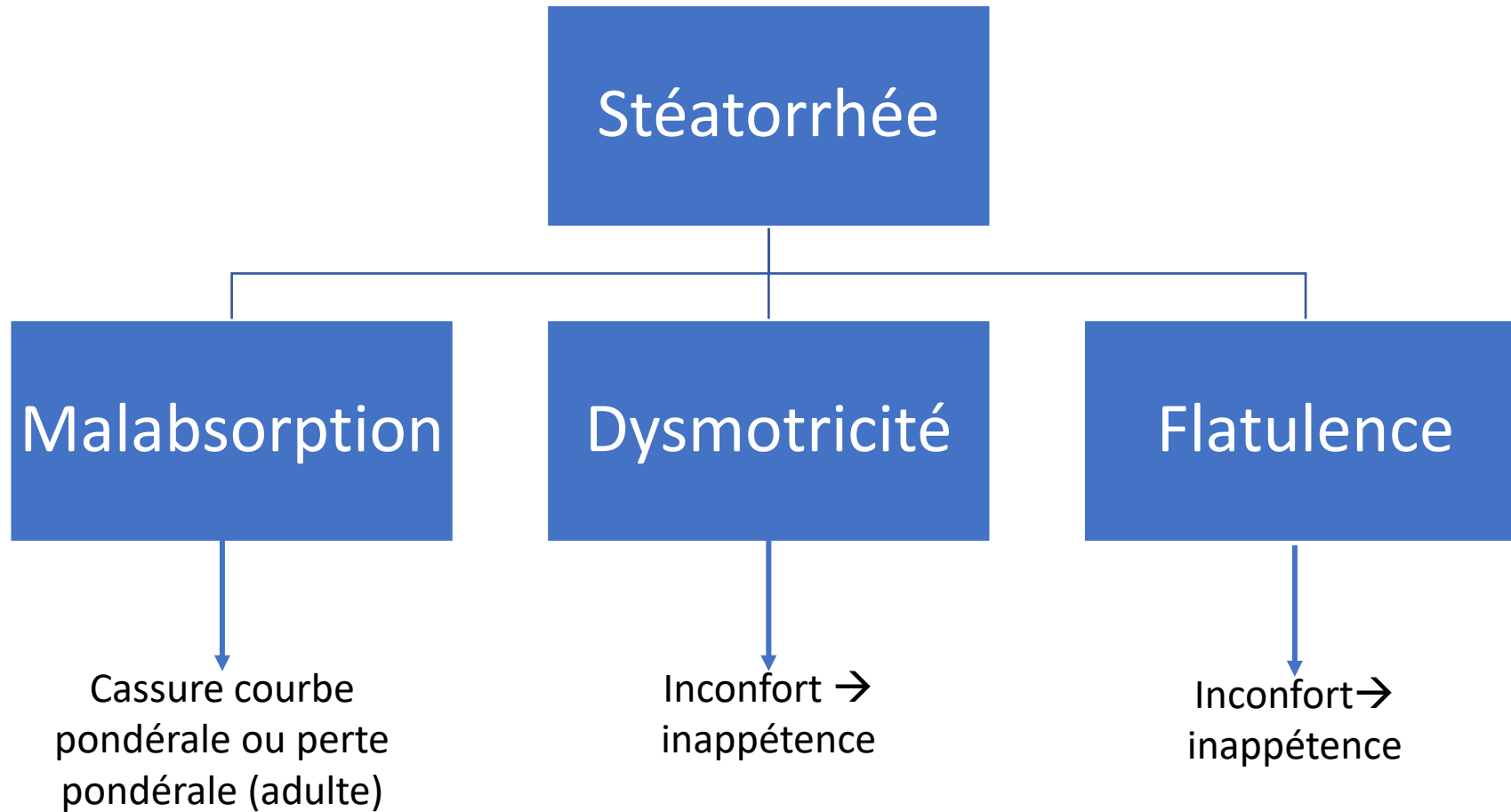
# Insuffisance pancréatique terminale

- Exocrine → stéatorrhée
- Endocrine → diabète

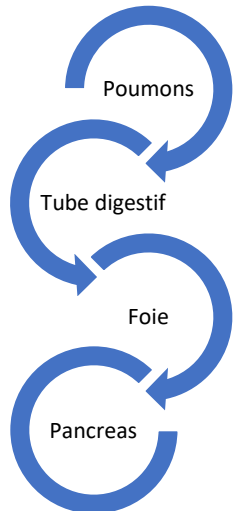
Delta F508



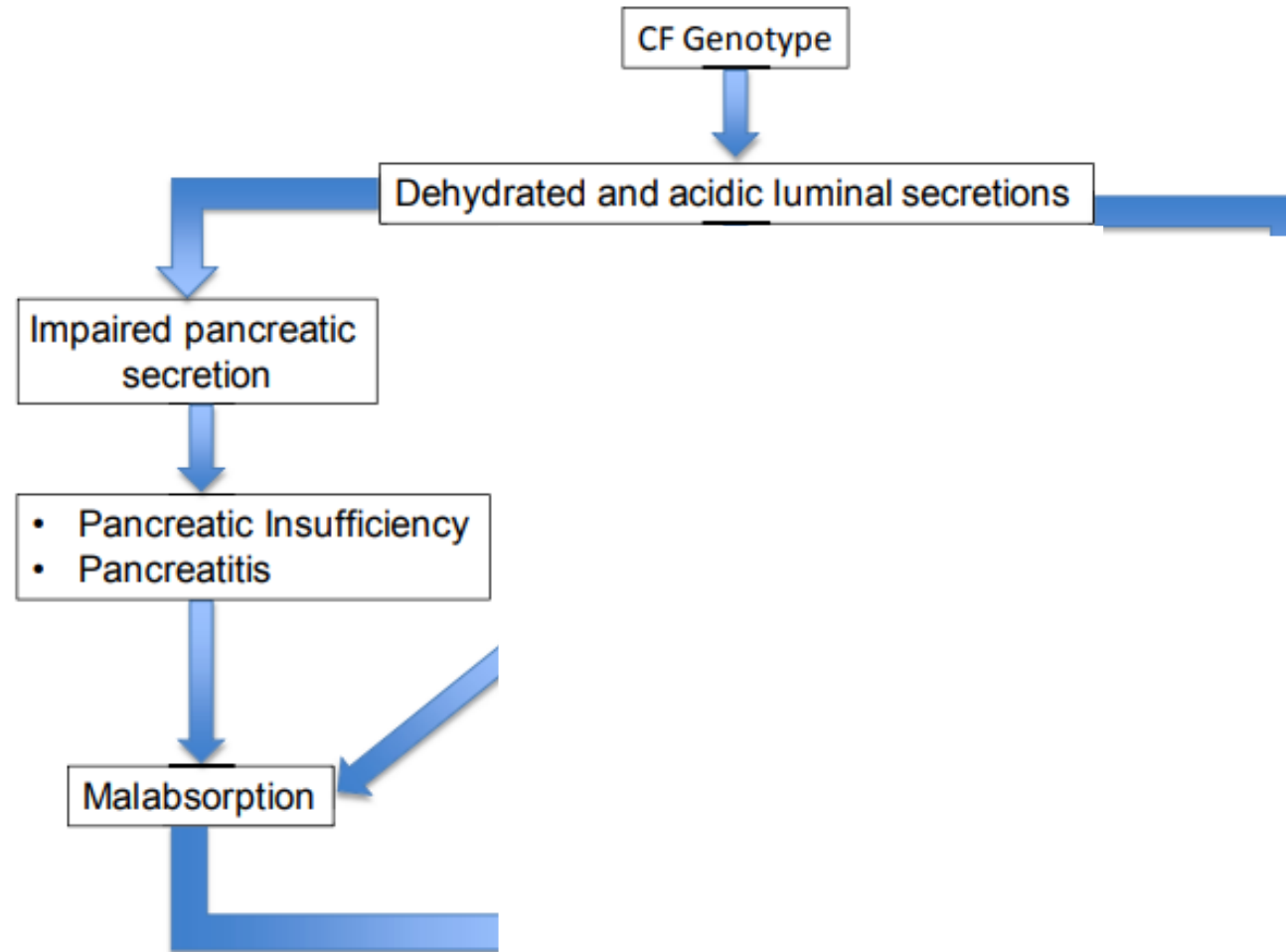
# Atteinte pancréatique exocrine - symptômes



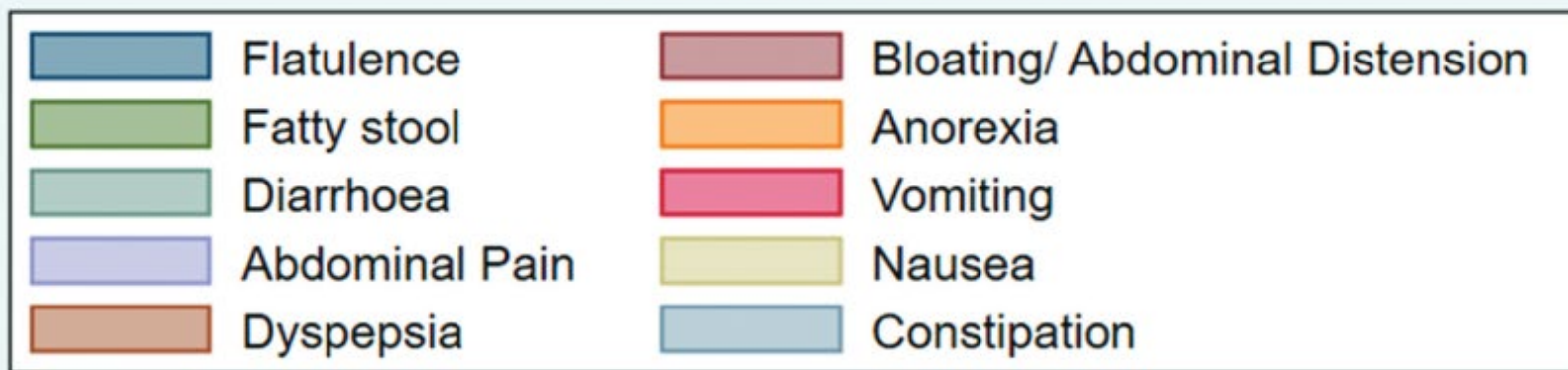
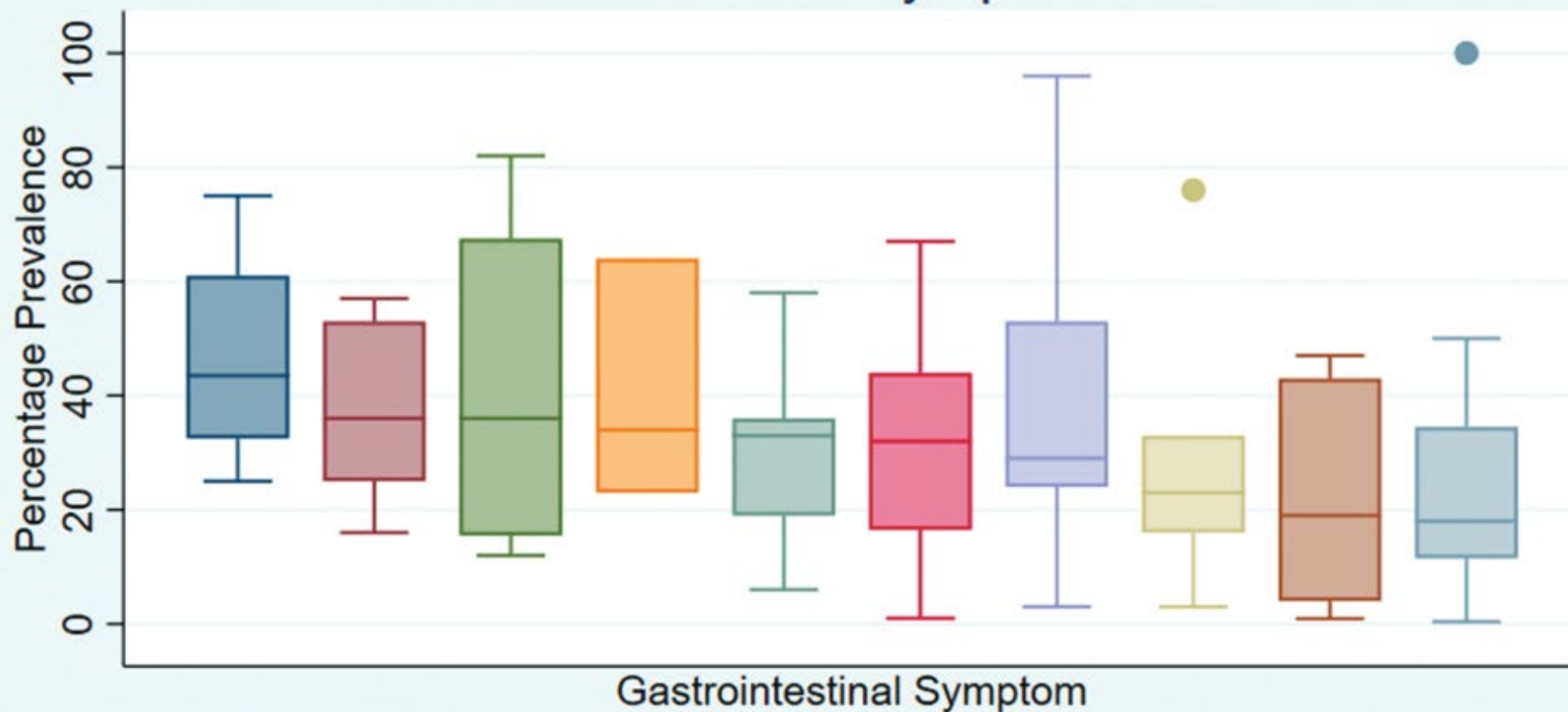
**> 85% patients**



# Résumé



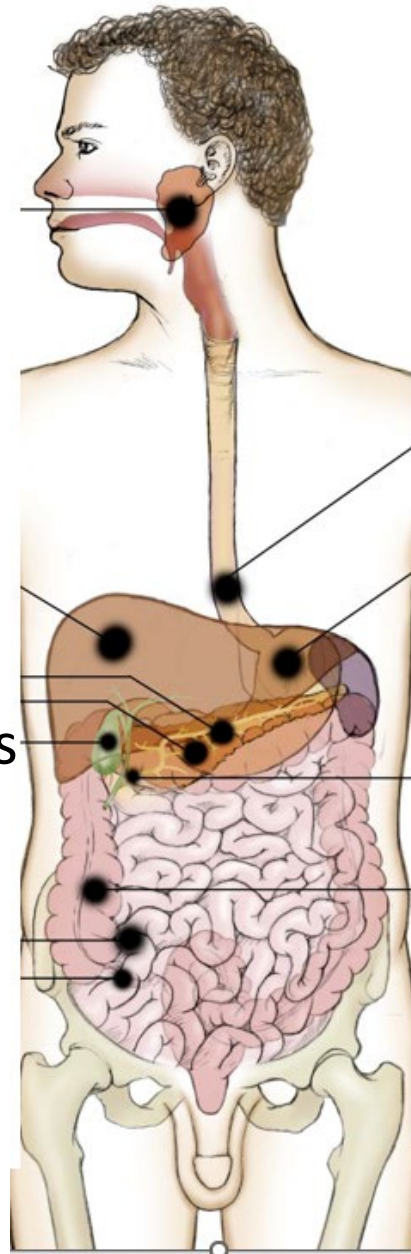
## Prevalence of GI Symptoms in CF



# Traitements

Acide ursodeoxcholique

Enzymes pancréatiques gastrorésistants



Anti-acide – inhibiteurs pompe à proton

Laxatifs (macrogol)

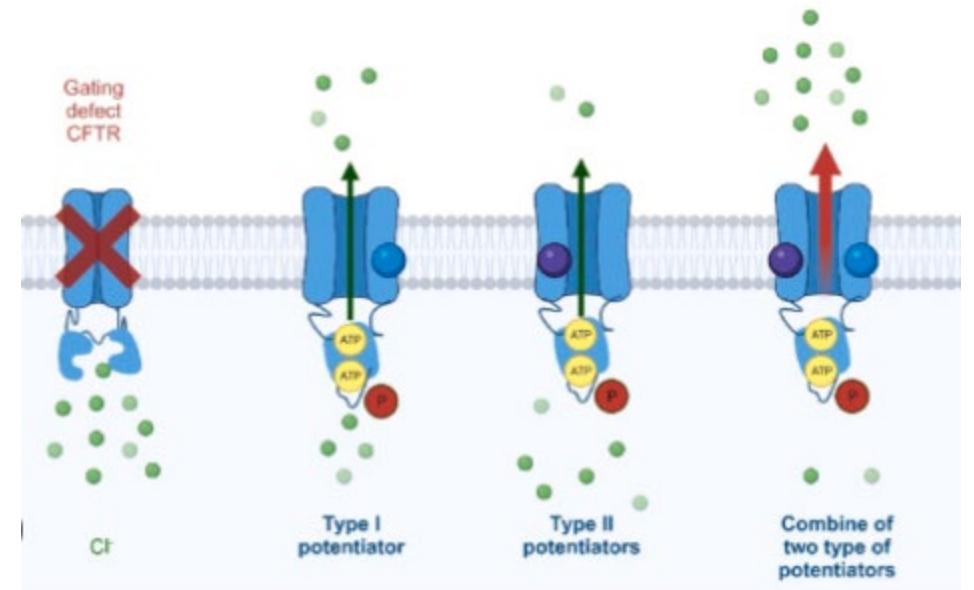
Probiotiques?

**Modulateurs CFTR???**

# Modulateurs du CFTR

- Effets sur épithélium respiratoire **confirmés**

- Effets sur épithélia du système digestif?



Le futur des patients = votre futur!

# Qualité de vie

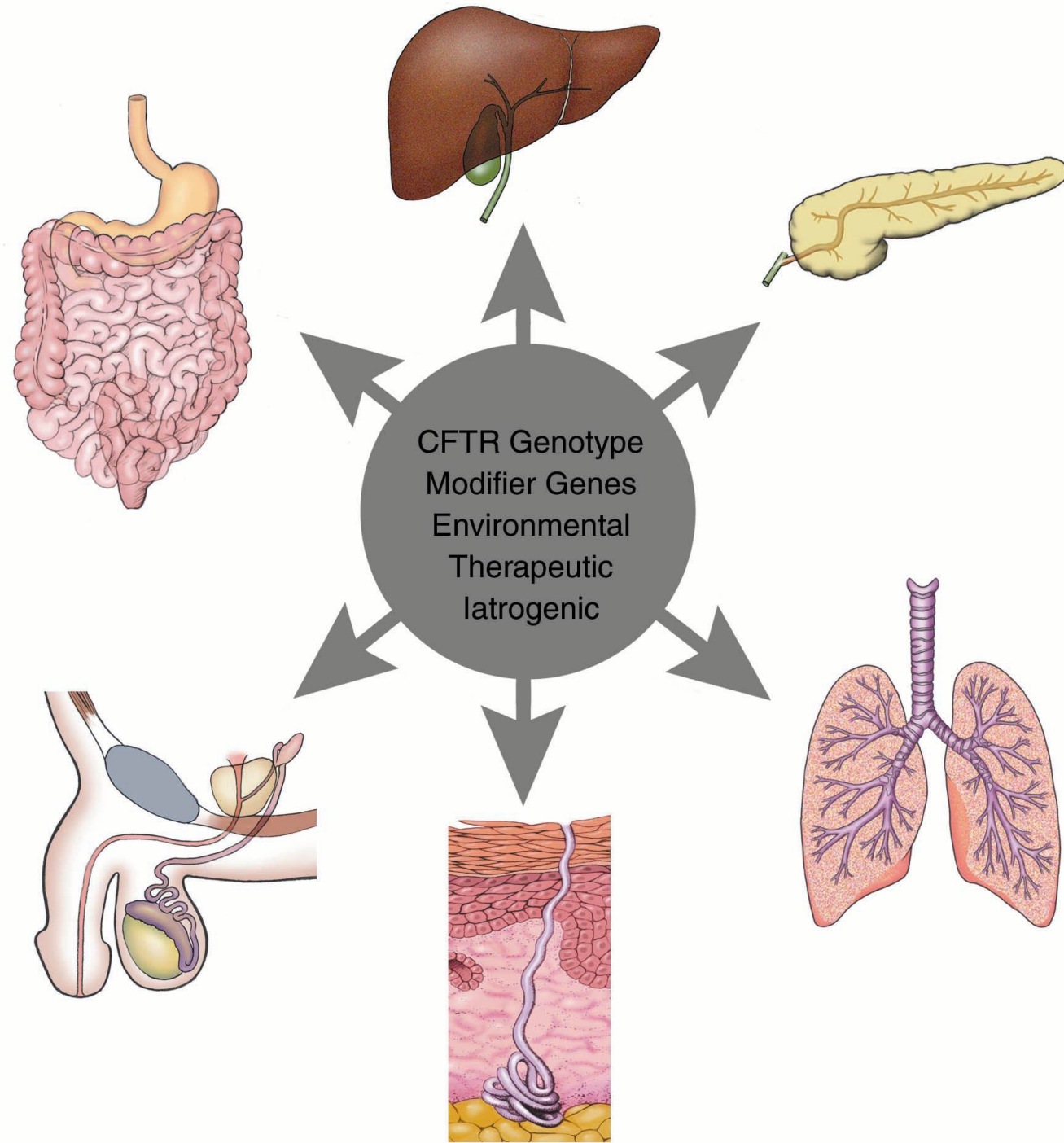


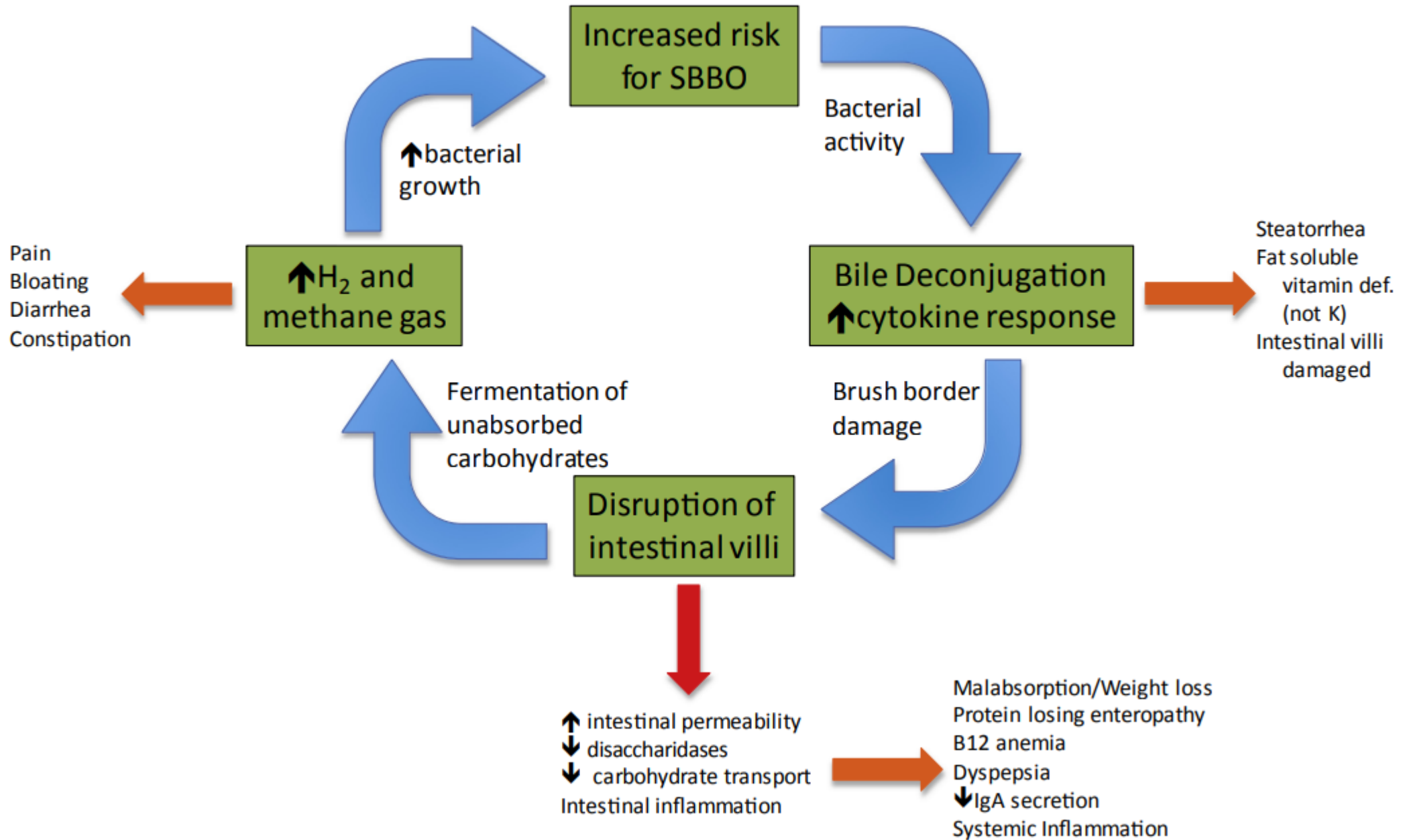
## Les manifestations digestives

**Merci de votre attention**

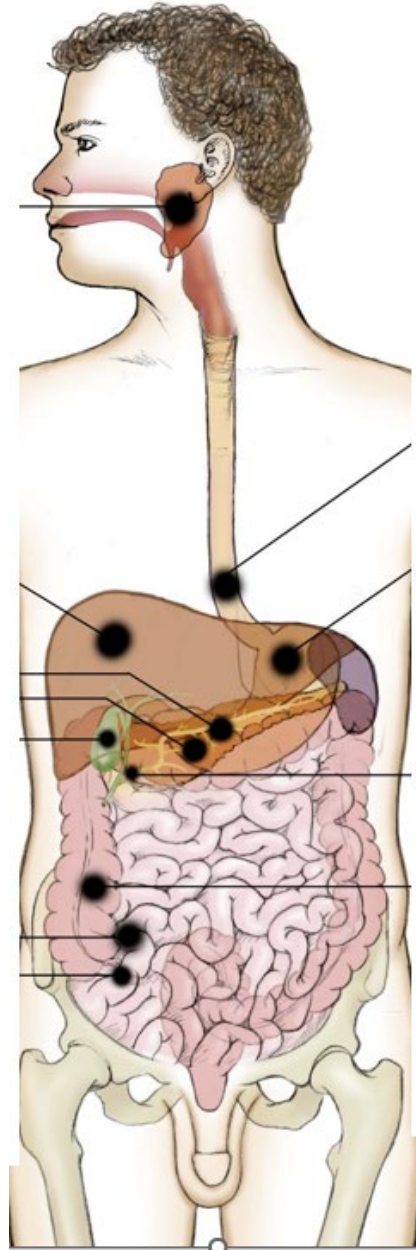








# Résumé



Déficit énergétique

