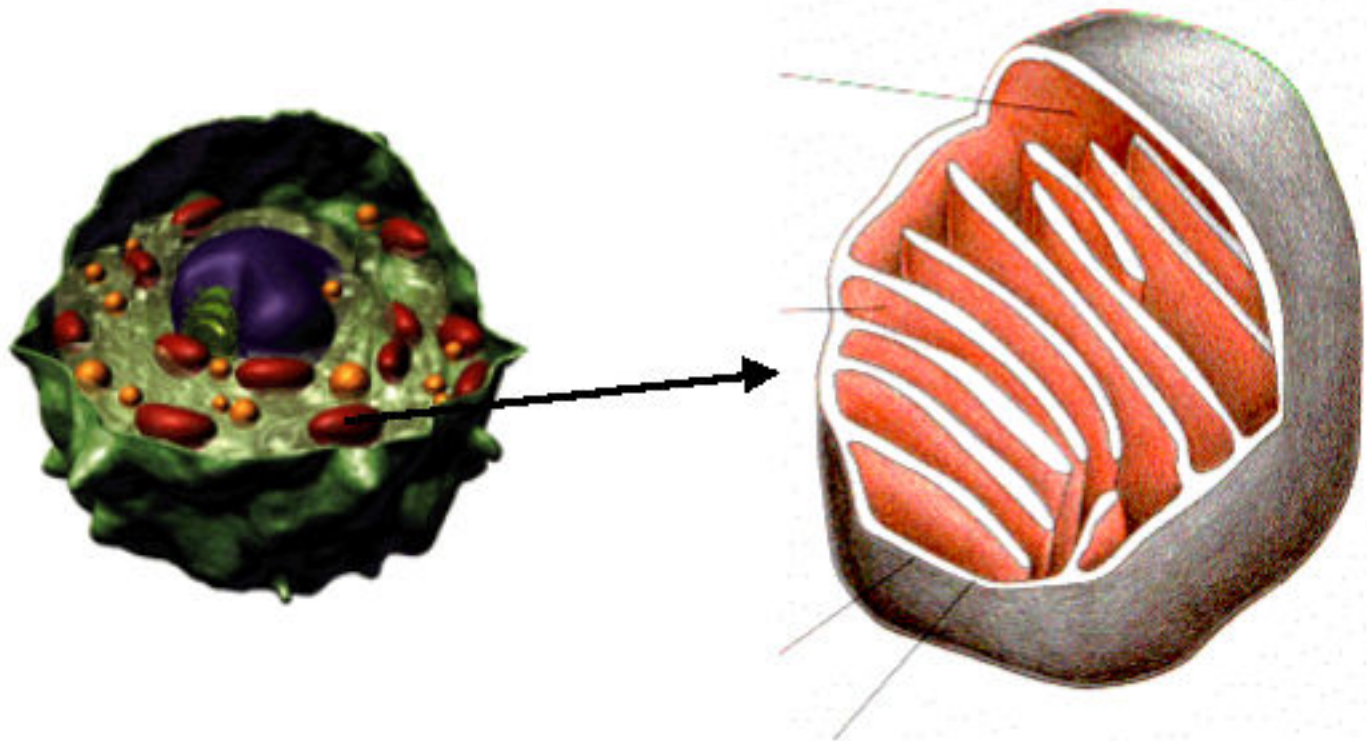
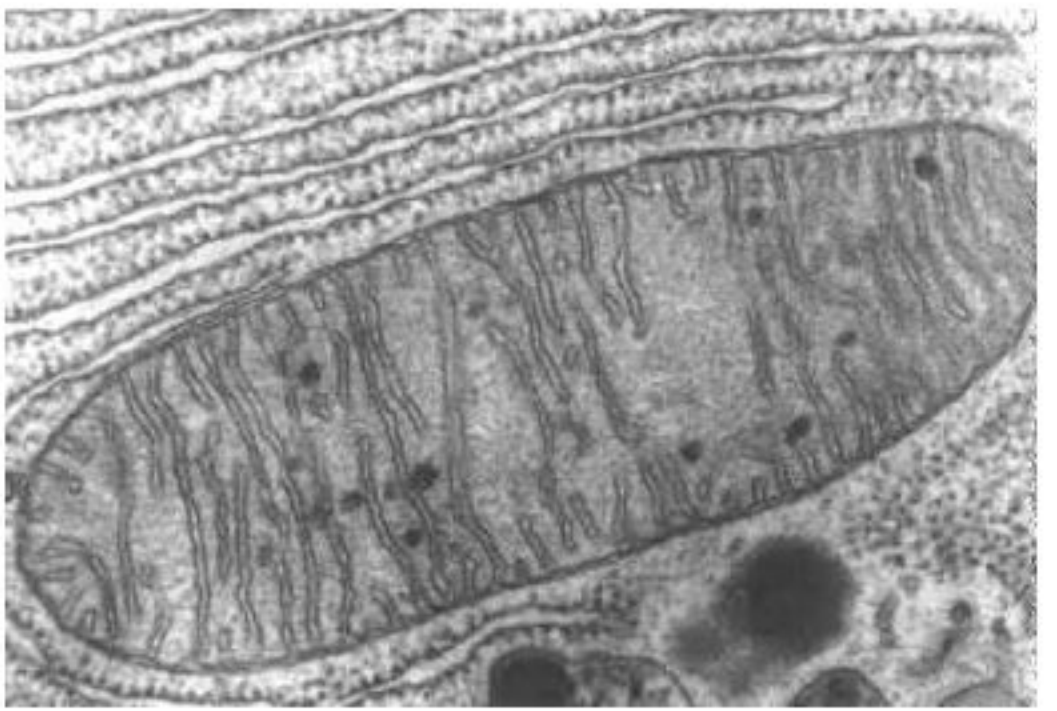
PLAN

MITOCHONDRIE

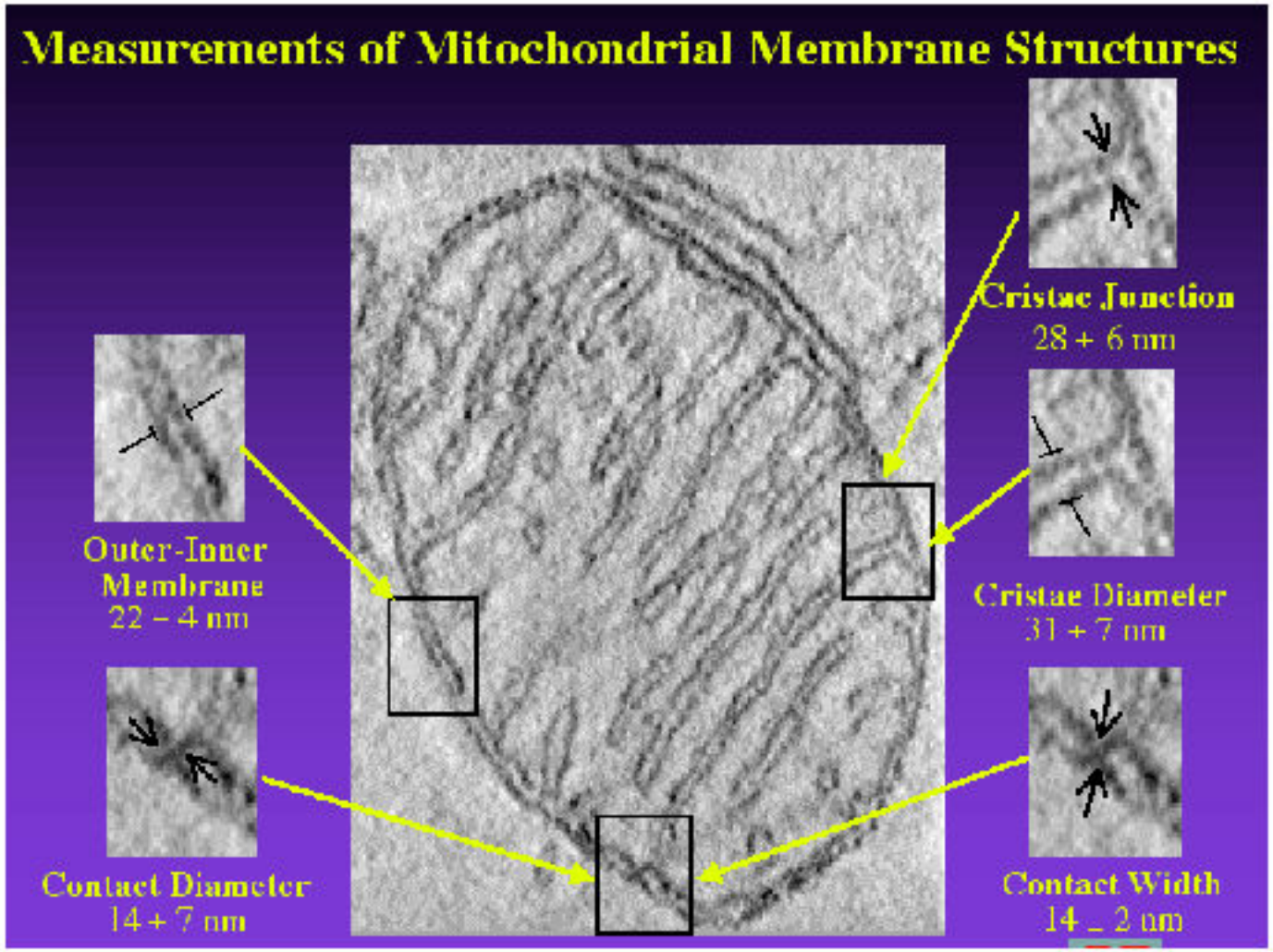


généralités

D'après A Textbook of Histology
by D Fawcett. Chapman and Hall, N.Y. 1994



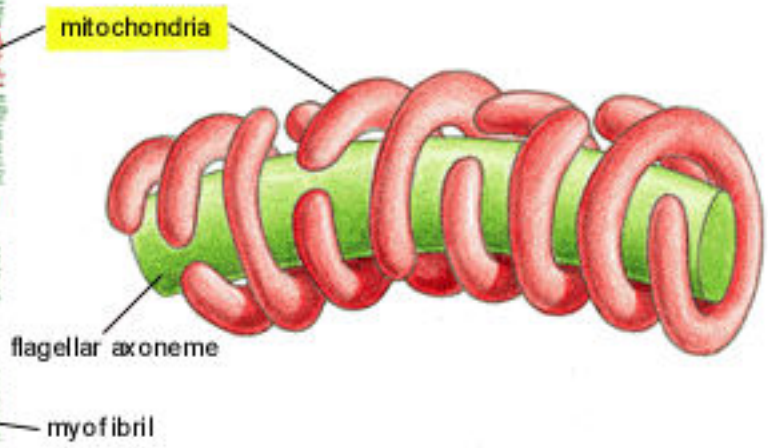
Morphologie

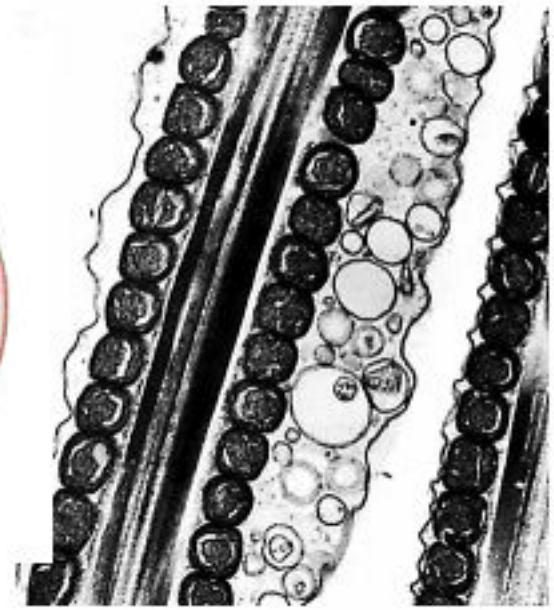


http://www.SCi.sdsu.edu/TFrey/MitoMovie.htm

Localisation

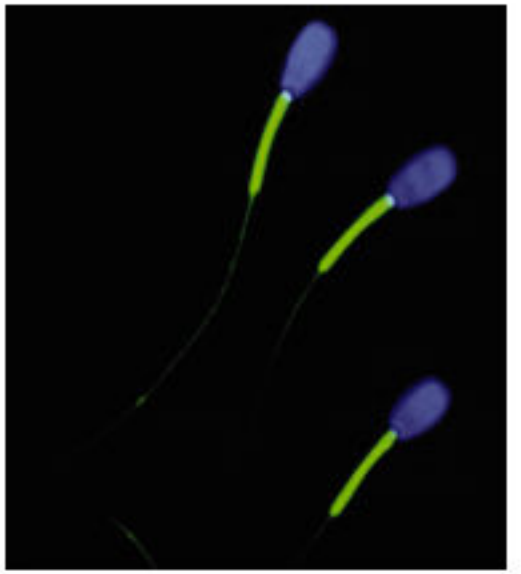
CARDIAC MUSCLE



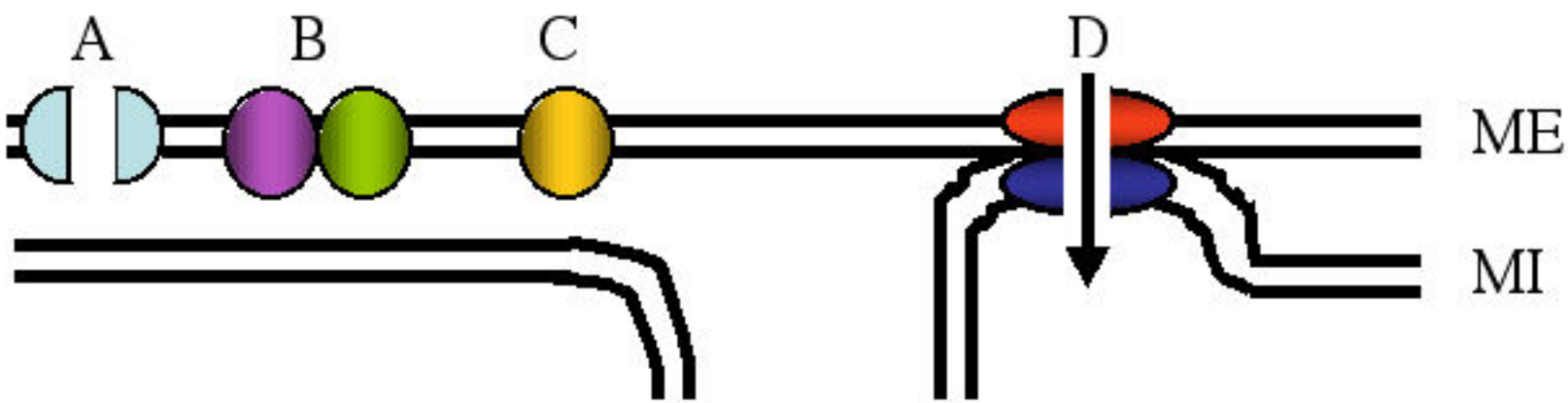


SPERM TAIL

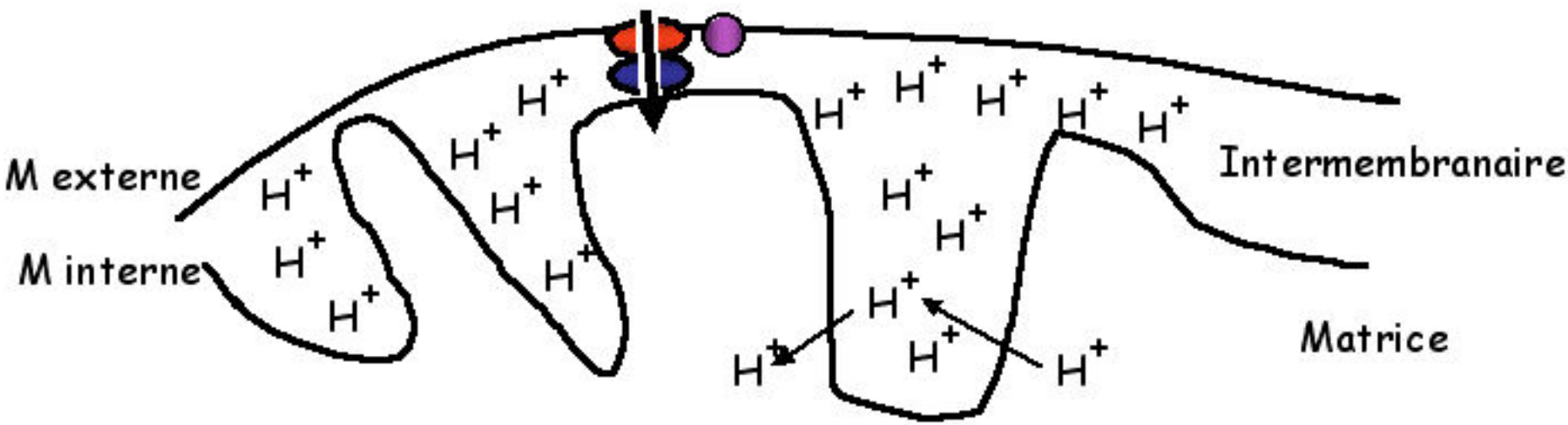
Molecular Biology of the Cell, 3rd edn



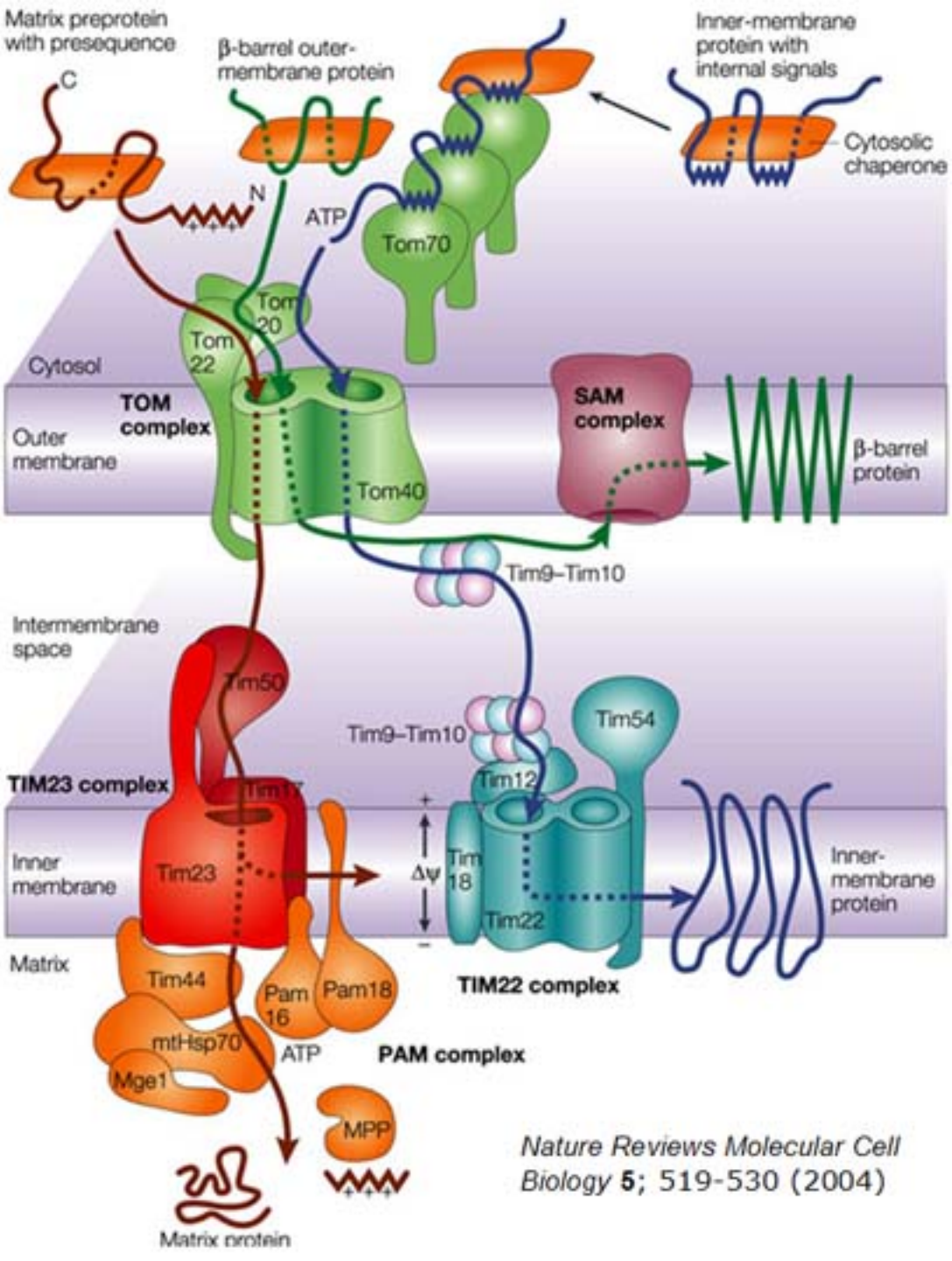
Membrane externe



Espace intermembranaire

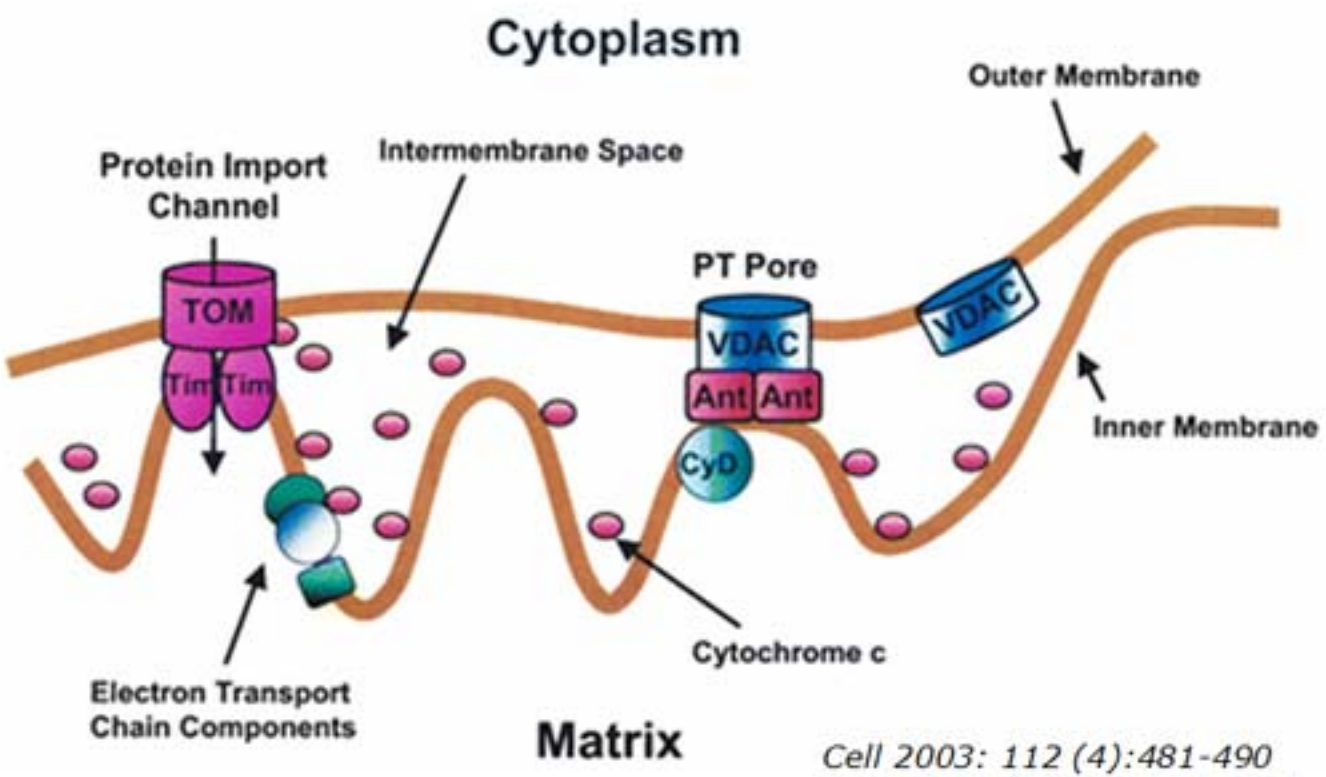


Importation de protéines cytosoliques



Nature Reviews Molecular Cell Biology **5**; 519-530 (2004)

Composition: résumé



Cell 2003: 112 (4):481-490
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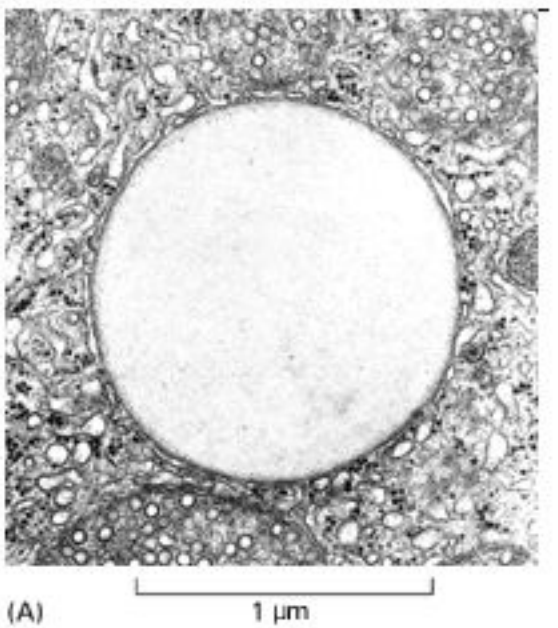
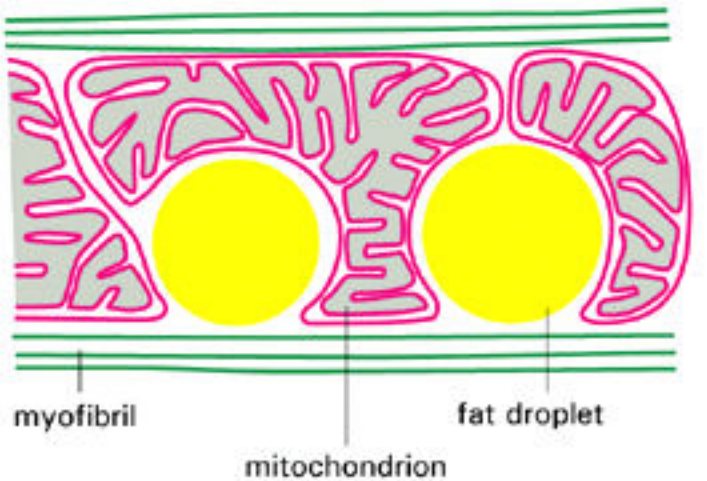
Fonctions

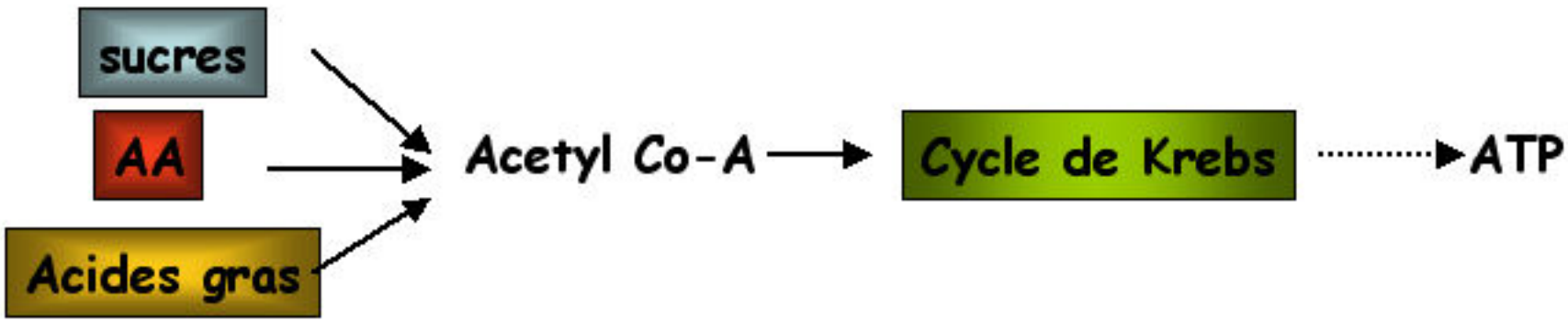
- **β-oxydation des acides gras:**



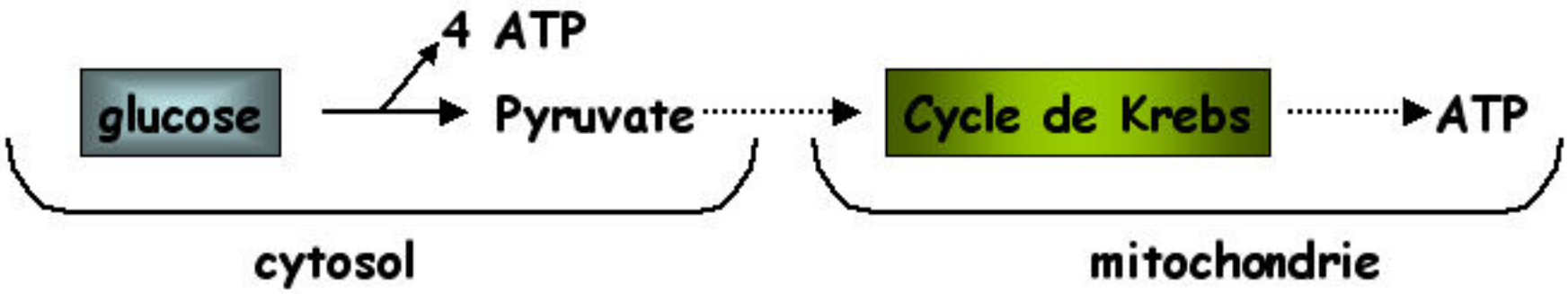


Molecular Biology of the Cell, 3rd edn

Production d'énergie: organisation générale



Cycle de Krebs



Vocabulaire

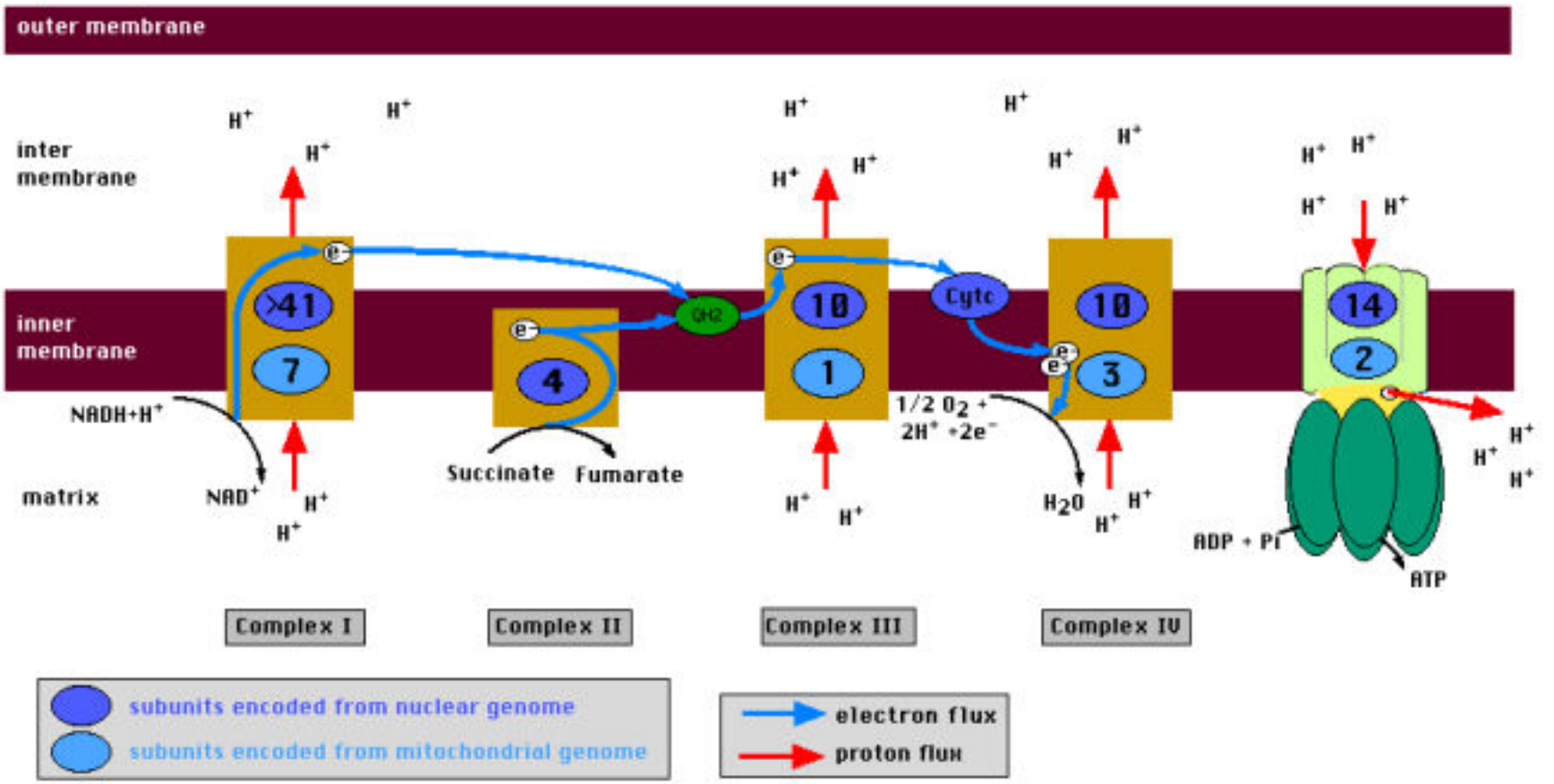
- **Le NADH.**

NADH = Nicotinamide Adénine Dinucléotide réduit.
Le NAD+ est converti en NADH au cours de l'oxydation d'une molécule.
Le NAD+ est capable d'accepter deux électrons et un proton pour donner un NADH.
Le NADH est produit au cours de nombreuses réactions : glycolyse, hélice de Lynen et cycle de Krebs .

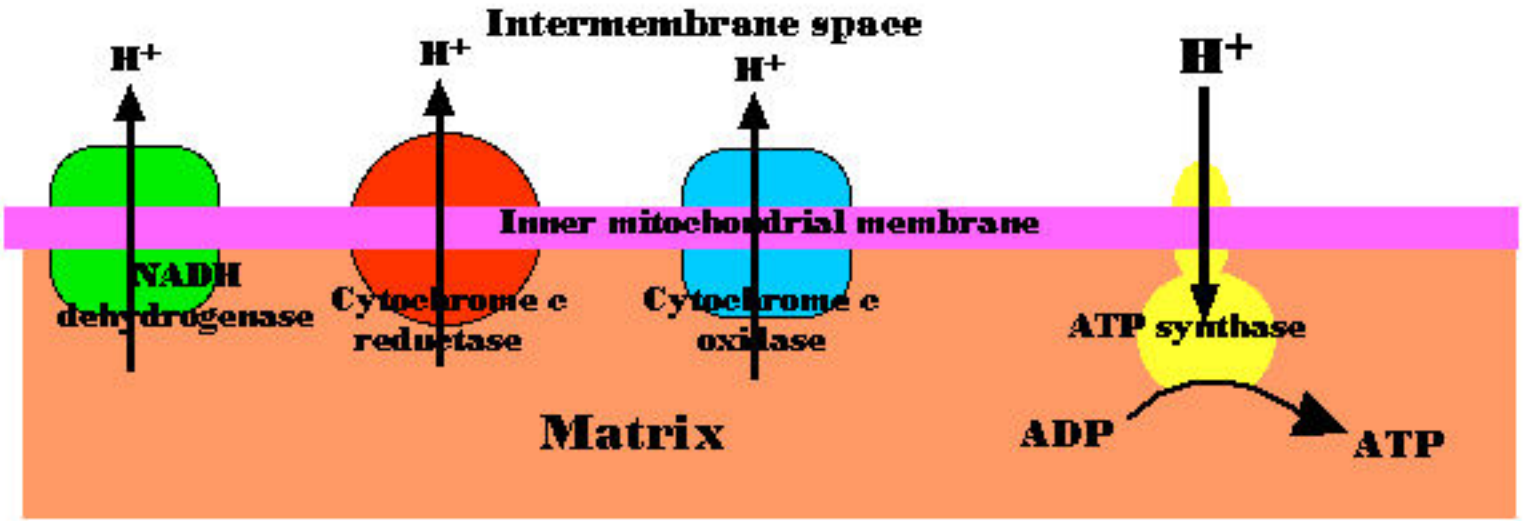
- **Le FADH2.**

FADH2 = Flavine Adénosine Dinucléotide réduit.
Le FAD est converti en FADH2 au cours de l'oxydation d'une molécule.
Le FAD est capable d'accepter deux électrons et deux protons pour former le FADH2.
La principale réaction mitochondriale qui produit du FADH2 est l'oxydation du succinate en fumarate au cours du cycle de Krebs .
L'enzyme qui catalyse cette réaction (succinate déshydrogénase) est un des complexes de la chaine respiratoire.
Le FADH2 est également produit au cours de la transformation d'acides gras en acétylCoA .

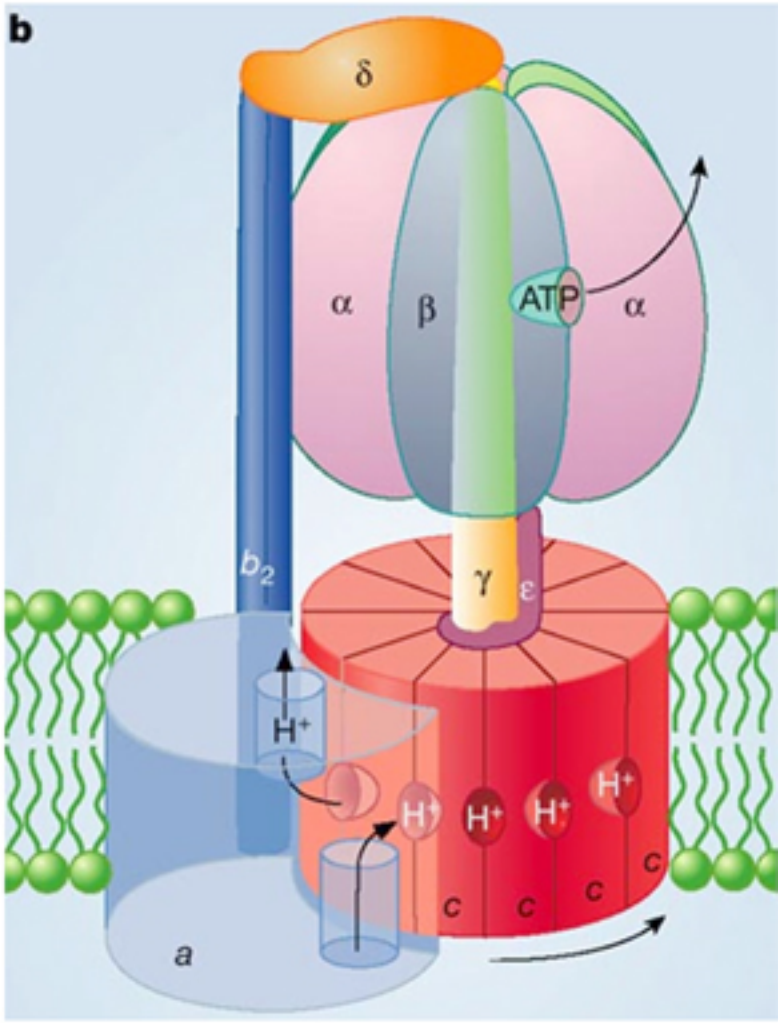
Chaine respiratoire



Gradient électrochimique

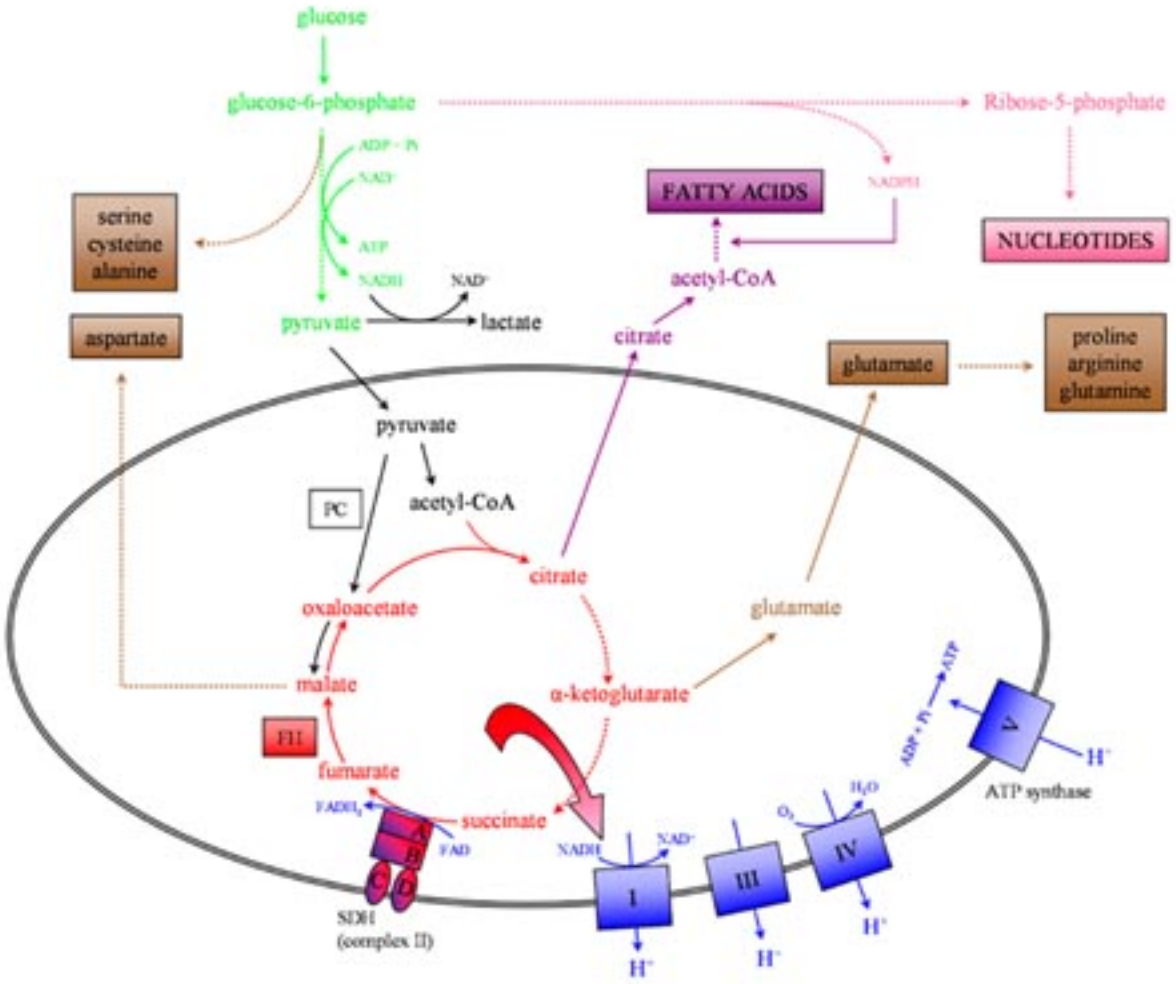


ATP synthase



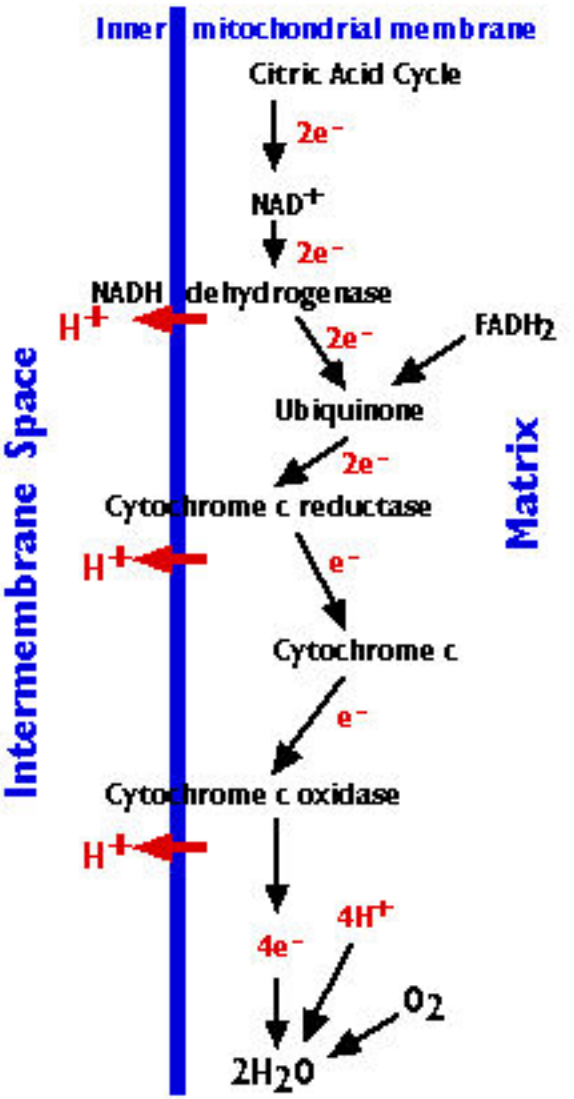
Nature **427**, 407 - 408 (29 January 2004)

Résumé du métabolisme énergétique

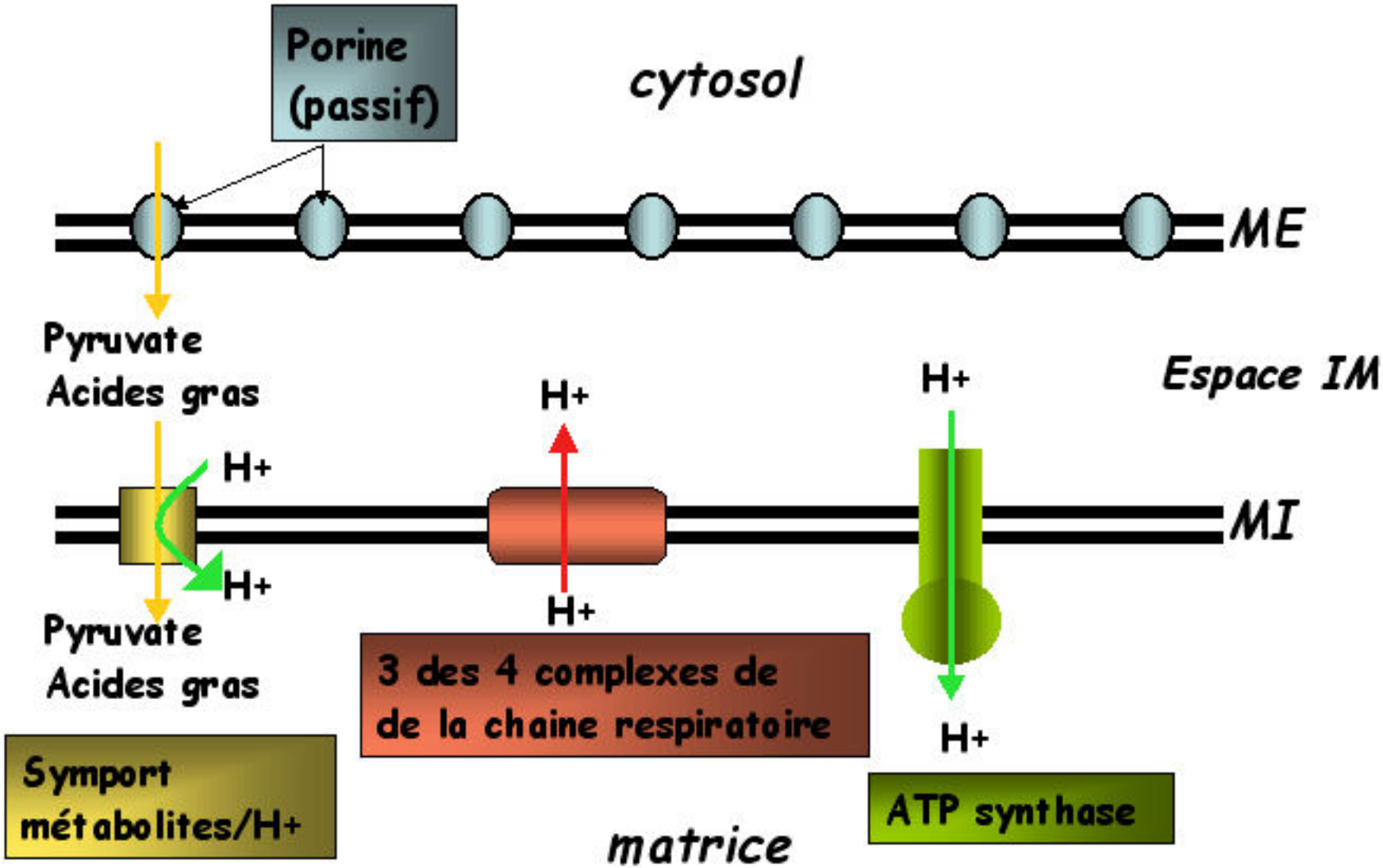


Oncogene (2006) 25, 4675–4682

Résumé

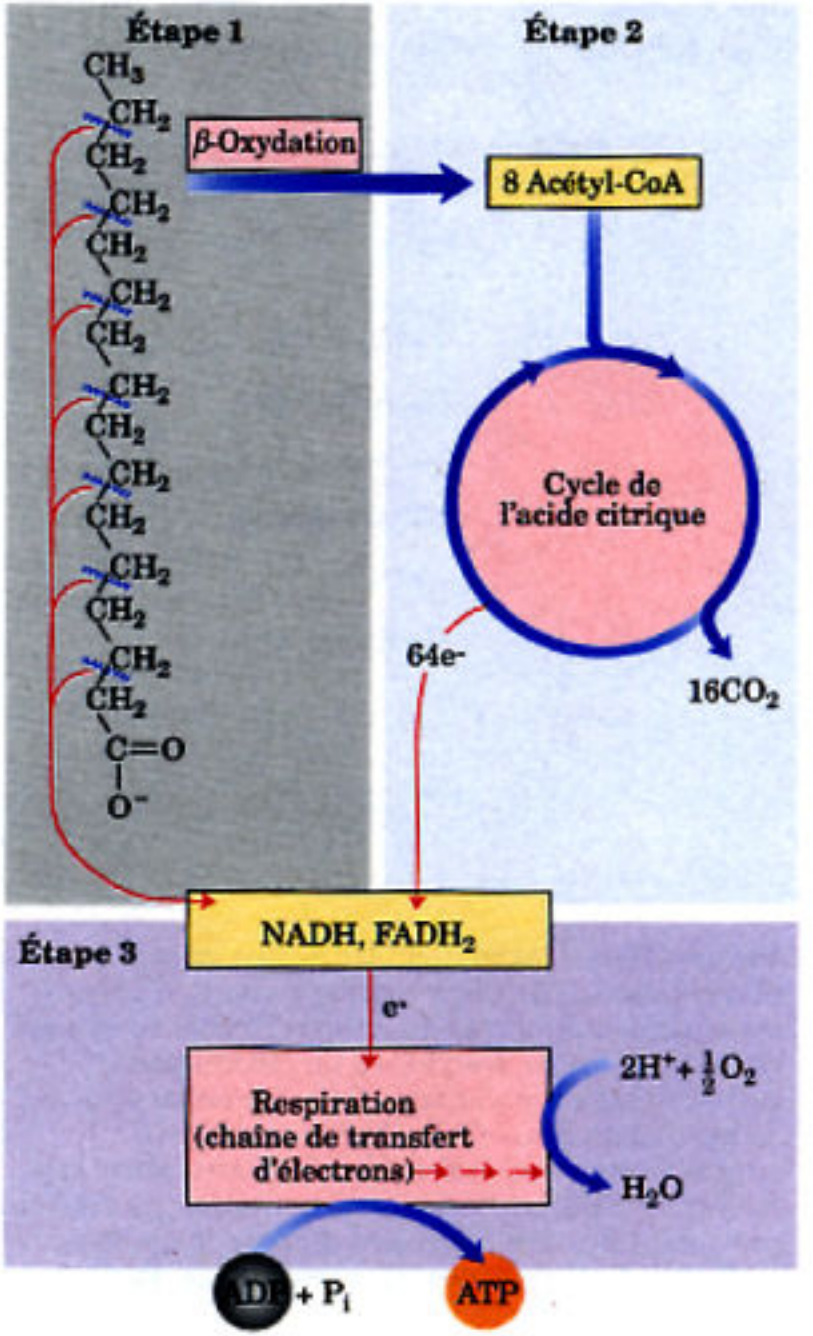

Inner mitochondrial membrane
Citric Acid Cycle
2e⁻
NAD+
2e⁻
NADH dehydrogenase
H+
2e⁻
FADH2
Ubiquinone
2e⁻
Cytochrome c reductase
H+
e⁻
Cytochrome c
e⁻
Cytochrome c oxidase
H+
4e⁻
4H+
O2
2H2O
Intermembrane Space
Matrix

Résumé: transport des protons

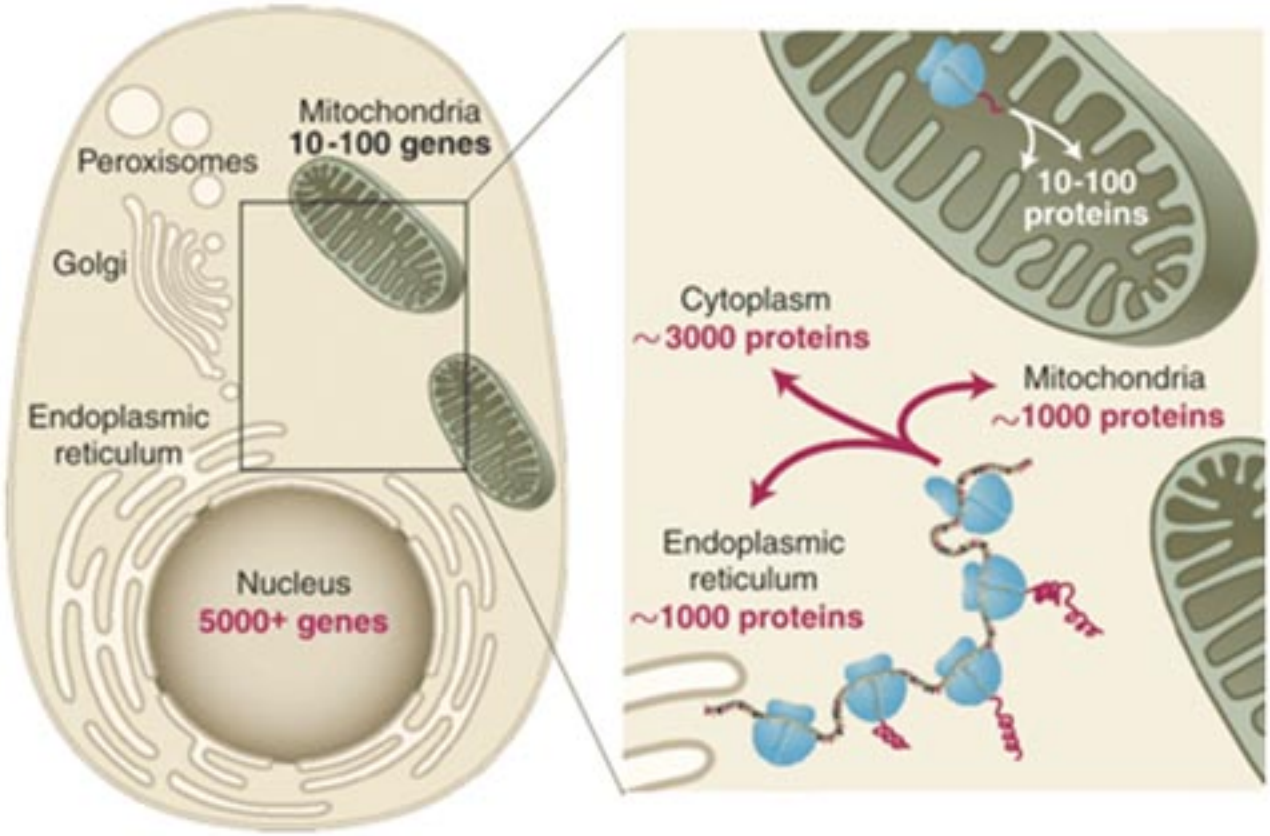


P. Lustenberger. Reproduction interdite

Résumé: lipides



Génome de la mitochondrie



Science (2006) 313; 5785: 314 - 318
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Maladies génétiques

The Spectrum of Mitochondrial Disease.

This table presents just a sample of the many genetic and acquired disorders that can result in primary or secondary disturbances of mitochondrial function. It demonstrates the importance of the intergenomic dialogue in contributing to the wide variety of disorders associated with mitochondrial dysfunction.

Some disorders known to be associated with mtDNA mutations	Some mendelian (nDNA) disorders of mitochondrial function involving regulation of fuel homeostasis	Some other primary disorders of intramitochondrial enzymes
MELAS	Luft disease	Methylmalonic acidemia
MERRF	Leigh syndrome (Complex I, COX, PDH)	Erythropoietic porphyria
NARP	Alpers Disease	Propionic acidemia
Myoneurogastrointestinal disorder and encephalopathy (MNGIE)	MCAD, SCAD, SCHAD, VLCAD, LCHAD	Acute intermittent porphyria
Pearson Marrow syndrome	Glutaric aciduria II	Variegate porphyria
Kearns-Sayre-CPEO	Lethal infantile cardiomyopathy	Maple syrup urine disease
Leber hereditary optic neuropathy (LHON)	Friedreich ataxia	Nonketotic hyperglycinemia
Aminoglycoside-associated deafness	Maturity onset diabetes of young	Hereditary sideroblastic anemia
Diabetes with deafness	Malignant hyperthermia	OTC Deficiency
	Disorders of ketone utilization	CPS Deficiency
	mtDNA depletion syndrome	
	Reversible COX deficiency of infancy	
	Various defects of the Krebs Cycle	
	Pyruvate dehydrogenase deficiency	
	Pyruvate carboxylase deficiency	
	Fumarase deficiency	
	Carnitine palmitoyl transferase deficiency	

http://www.mitoresearch.org/

Maladies avec dysfonctionnement mitochondrial non génétique

Disorders Sometimes Associated with Mitochondrial Dysfunction.

Some mendelian (nDNA) disorders with secondary disturbances in mitochondrial function†	*Polygenic and Genetotrophic*	*Autoimmune*	*Environmental*
Hemochromatosis	Aging	Multiple sclerosis	AZT toxicity
Wilson disease	Type II diabetes mellitus	Systemic lupus erythematosis	FIAU toxicity
Batten disease	Atherosclerotic heart disease	Rheumatoid arthritis	Lead, cyanide and mercury poisoning
Wolff-Parkinson-White††	Parkinson disease	Thyrotoxicosis	Ackee fruit toxic hypoglycemia
Huntington disease	Alzheimer dementia	Primary biliary cirrhosis	Doxorubicin cardiotoxicity
Menkes disease	Congestive heart failure	Procainamide lupus	Aminoglycoside ototoxicity and nephrotoxicity
Lesch-Nyhan syndrome	Maternally inherited migraine	Guillain-Barré syndrome	Amytal poisoning
	Niacin-responsive hypercholesterolemia		Carbon monoxide poisoning
† nDNA mutations affecting proteins that are not located in the mitochondria but which alter mitochondrial function.	Postpartum cardiomyopathy		Amphotericin nephrotoxicity
†† WPW generally has a non-mitochondrial cause but is sometimes seen in conjunction with cases of Complex I deficiency.	Alcoholic myopathy		MPTP Parkinsonism
	Wernicke encephalopathy		Vitamin deficiencies such as pellagra, beriberi, rickets, and ICU axonal neuropathy and pernicious anemia
	Reye syndrome		
	Burkitt lymphoma (BCL2)		
	Cancer metastasis (NM23)		
	Irritable bowel syndrome		
	Gastroparesis-GI dysmotility		

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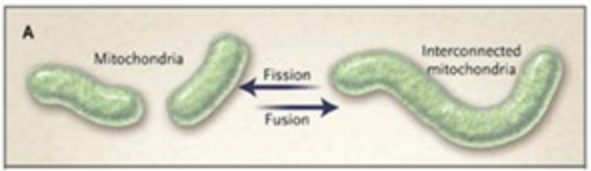
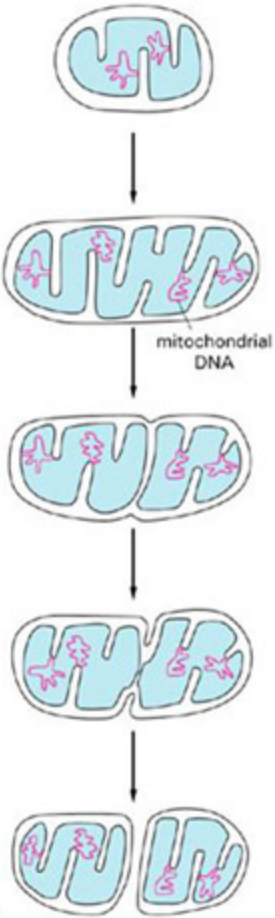
Division





Chan DC. N Engl J Med 2007;356(17):1707-9
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