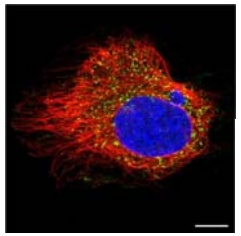
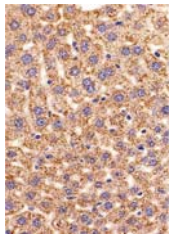
Peroxisomes

small, membrane-enclosed organelles that contain enzymes (50) involved in a variety of metabolic reaction, specialized for carrying out oxidative reactions.

Peroxisome in light microscopy

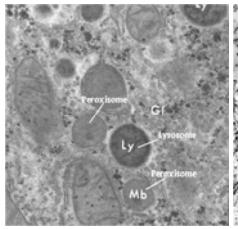


Peroxisomal trans-2-enoyl-CoA reductase
http://www.proteinatlas.org/dictionary/cell/vesicles+%28peroxisome%29+4

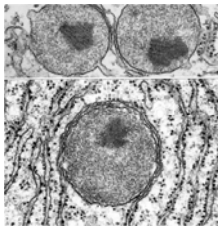


Peroxisome in hepatocytes
http://www.niehs.nih.gov/research/resources/liverpath/toxicologic/index.cfm

Peroxisome ultrastructure

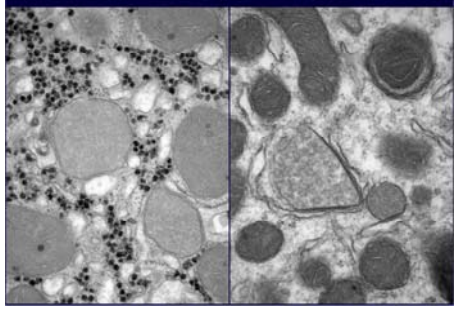


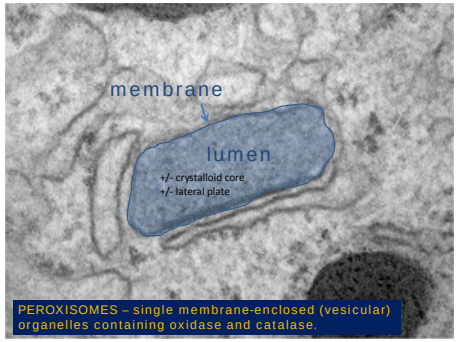
http://www.carolguze.com/text/442-11-clinical_genetics.shtml



http://www.visualphotos.com/photo/1x8466350/peroxisomes.jpg

Peroxisome ultrastructure

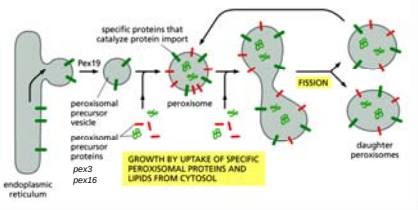




PEROXISOMES – single membrane-enclosed (vesicular) organelles containing oxidase and catalase.

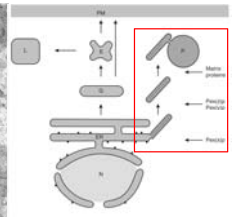
Peroxisome biogenesis

Peroxins (pex1 –pex24)



Peroxisome biogenesis

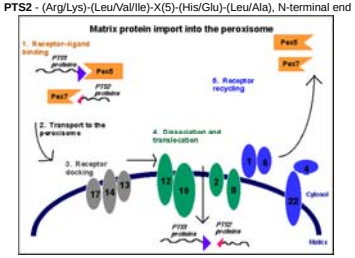
Adabella van der Zand et al. J Cell Sci 2006; 119: 989-994



Tabak HF, Murk JL, Braakman I, Geuze HJ. **Peroxisomes start their life in the endoplasmic reticulum.** Traffic. 2003; 4(8): 512-8.

Peroxisome biogenesis - matrix protein import

PTS1 - Ser-Lys-Leu-COOH, C-terminal end
PTS2 - (Arg/Lys)-(Leu/Val/Ile)-X(5)-(His/Glu)-(Leu/Ala), N-terminal end



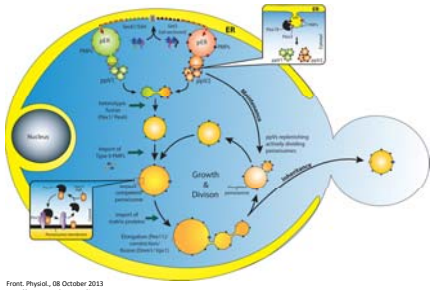
TRANSLOCATION

http://carolguze.com/images/cellorganelles/peroximport.gif

Peroxisomes
RER
ribosomes
lamellae

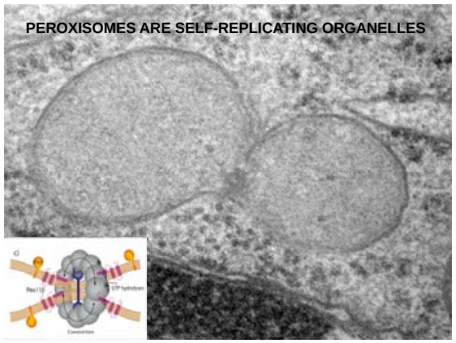
Tabak HF, Murk JL, Braakman I, Geuze HJ. **Peroxisomes start their life in the endoplasmic reticulum.** Traffic. 2003; 4(8): 512-8.

Peroxisome biogenesis

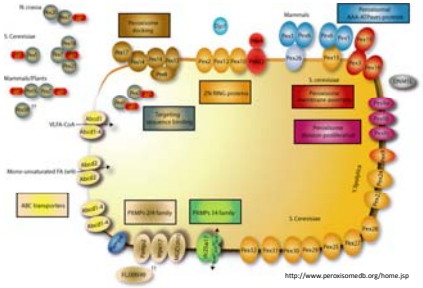


Front. Physiol., 08 October 2013
http://dx.doi.org/10.3389/fphys.2013.00286

PEROXISOMES ARE SELF-REPLICATING ORGANELLES



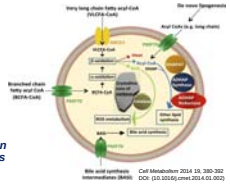
Peroxins and other PMPs



http://www.peroxisomedb.org/home.jsp

Peroxisome functions

- **β-oxidation** of long and very long fatty acids
- auxiliary β-oxidation, for the unsaturated fatty acids carrying double bonds at odd carbons
- **α-oxidation** of fatty acids with lateral methyl groups *(prenyl or isoprenyl structures)*
- **oxidative metabolism** of amino acids and purines
- **biosynthesis** of:
  - cholesterol,
  - biliary acids, and
  - plasmalogens

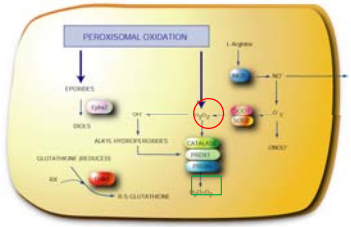


Cell Metabolism 2014 19, 380-392
DOI: (10.1016/j.cmet.2014.01.002)

N.B. It seems that ***all biochemical events in peroxisome are oxidative processes***

Catalase (CAT−**40%)** allows the degradation of the harmful hydrogen peroxide, produced in the peroxisome by several hydrogen-peroxide-producing oxidases, yielding water and oxygen.

Superoxide dismutases [SOD1 (Cu/ZnSOD) and SOD2 (MnSOD)] convert harmful superoxide radicals to molecular oxygen and hydrogen peroxide.

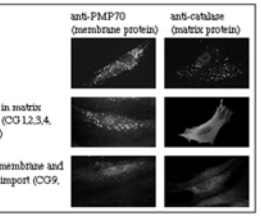


ANTIOXIDANTS

http://www.peroxisomedb.org/home.jsp

Peroxisome biogenesis

Experimental evidences about mechanisms



CG – complementation group

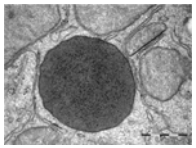
Peroxisomal diseases

TABLE 1. The Peroxisomal Disorders

Disorder	Abbreviation	Protein involved	Gene	Chromosome	MIM
Disorders of peroxisome biogenesis					
Zellweger syndrome	ZS	Peroxins	*PEX*-genes	Multiple loci	214100
Neonatal adrenoleukodystrophy	NALD	Peroxins	*PEX*-genes	Multiple loci	214110
Infantile Refsum disease	IRD	Peroxins	*PEX*-genes	Multiple loci	202370
Hyperpipecolic acidaemia*	HPA	Peroxins	*PEX*-genes	Multiple loci	266510
Rhizomelic chondrodysplasia punctata type 1	RCDP type 1	PexTp	*PEX7*	6q21-q22.2	215100
Single peroxisomal enzyme/transporter defects					
X-linked adrenoleukodystrophy	X-ALD	ALDP	*ABCD1*	Xq28	300100
Acyl-CoA oxidase deficiency (pseudo-neonatal ALD)	ACOX1-deficiency	Straight-chain acyl-CoA oxidase (ACOX; ACOX1)	*ACOX1*	17q25.1	264470
D-bifunctional protein deficiency	D-BP deficiency	D-BP/MFE/MFE1/ D-PBE	*HSD17B4*	5q2	261515
2-methylacyl-CoA racemase deficiency	Racemase deficiency	AMACR	*AMACR*	5p13.3-p12	604489
Rhizomelic chondrodysplasia punctata type 2 (DHAPAT deficiency)	RCDP type 2	DHAPAT	*GNPAT*	1q42.1-42.3	222765
Rhizomelic chondrodysplasia punctata type 3 (alkylDHAP synthase deficiency)	RCDP type 3	ADHAPS	*AGPS*	2q33	600121
Refsum disease (phytanoyl-CoA hydroxylase deficiency)	ARD	Phytanoyl-CoA hydroxylase (PhyH)	*PHYH/PAHX*	10p15-p14	266500
Hyperoxaluria type 1	PH1	Alanine glyoxylate aminotransferase (AGT)	*AGXT*	2q37.3	259900
Glutaric acidemia type 3	GA3	?	?	?	231690
Mevalonate kinase deficiency	MK-deficiency	Mevalonate kinase	*MVK*	12q24.1	251170, 260920
Catalasemia		Catalase	*CAT*	11p13	115500
Mulibrey nanism	MUL	Trim37p	*TRIM*	17q22-23	253250

*At least the four original HPA patients described by Gatfield et al. (1969), Burton et al. (1981), and Thomas et al. (1975) are now known to be affected by a disorder of peroxisome biogenesis. As described in the text, additional HPA patients have been described in literature in whom peroxisome biogenesis is normal. The primary defect in these patients remains to be identified.

http://www.peroxisomedb.org/home.jsp

Peroxisomes Summary

- Peroxisome is a membrane bound organelle with oxidative function using hydrogen peroxide
- Indirectly involved in energetic metabolism of the cells due to oxidation of fatty acids