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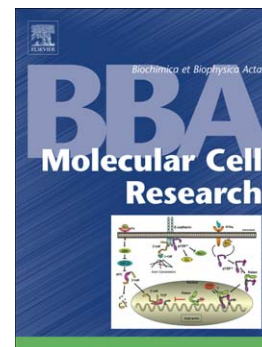
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Peroxisomal Protein Import Pores

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Abstract

Peroxisomal protein import is essentially different to the translocation of proteins into other organelles. The molecular mechanisms by which completely folded or even oligomerized proteins cross the peroxisomal membrane remain to be disclosed. The identification of a water-filled pore that is mainly constituted by Pex5 and Pex14 led to the assumption that proteins are translocated through a large, probably transient, protein-conducting channel. Here, we will review the work that led to the identification of this translocation pore. In addition, we will discuss the main biophysical features of the pore and compare it with other protein-translocation channels. This article is part of a special issue entitled "Assembly, Maintenance and Dynamics of Peroxisomes"

Introduction

Peroxisomes are ubiquitous organelles of eukaryotic cells. As their name implies peroxisomes harbour enzymes important for the oxidation of organic compounds and consequently one of their main functions is the reduction of reactive oxygen species [1]. Peroxisomes also play a major role in the metabolism of fatty acids, cholesterol and D-amino acids [2]. Importantly, mammalian peroxisomes harbour the first reactions to synthesize plasmalogens, which represent the most abundant class of phospholipids in the myelin sheath that is wrapped around axons. Since a lack of plasmalogen leads to severe abnormalities in the myelination of neurons, peroxisomal disorders often lead to neurological disease patterns [3]. Proteins that are involved in the biogenesis, division and inheritance of peroxisomes are called peroxins (encoded by PEX genes) and so far 34 different PEX genes have been identified [4-7]. Several inherited diseases including the Zellweger syndrome spectrum (ZSS) disorders and rhizomelic chondrodysplasia punctata (RCDP) type 1, caused by defects of peroxisomal biogenesis factors have been identified [8-12].

As peroxisomes carry out a large variety of metabolic tasks a constant and probably highly regulated exchange of metabolites between the organelle and the cytosol has to be assured. The permeability of the peroxisomal membrane is a matter of debate and is discussed in excellent reviews elsewhere [13, 14]. Altogether, surprisingly little is known about the molecular identity of transporters and channels for metabolites but also about translocation sites for peroxisomal proteins within the peroxisomal membrane.

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