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## A Unified Nomenclature for Peroxisome Biogenesis Factors

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For the past 10 years, there has been substantial progress in the field of peroxisome biogenesis. One key to this progress has been the use of genetic approaches in a wide variety of experimental organisms (4, 8, 11, 15, 18, 21, 26, 35, 38, 47, 48). To date, these systems have been used to identify thirteen proteins required for peroxisome biogenesis, three of which have also been shown to be defective in the lethal peroxisome biogenesis disorders. However, the diversity of experimental systems has also led to a profusion of names for peroxisome assembly genes and proteins. These include the acronyms PAS, PAF, PER, PAY, PEB, and PMP and span an even greater array of numbering systems. At the request of the Editors of *The Journal of Cell Biology* and for the benefit of all concerned, we considered several options for gene and protein names, numbering systems, and possible definitions for the types of genes and protein to be included. We propose here a unified protein and gene nomenclature for peroxisome biogenesis factors.

Proteins involved in peroxisome (microbody) biogenesis (inclusive of peroxisomal matrix protein import, membrane biogenesis, peroxisome proliferation, and peroxi-

some inheritance) will be designated peroxins, with PEX representing the gene acronym. However, even though defects in peroxisomal metabolic enzymes or transcription factors may affect peroxisome proliferation and/or morphology, such proteins shall not be included in this group. The proteins and genes will be numbered by date of published characterization, both for known factors and for those identified in the future. When necessary, species of origin may be specified by one letter abbreviations for genus and species (e.g., *ScPEX1*<sup>l</sup> for the *Saccharomyces cerevisiae* *PEX1* gene). To minimize ambiguities in naming additional proteins that may be identified in the future, we urge authors before publication to contact an ad hoc nomenclature committee (see below) who will be responsible for numbering new peroxins.

The new nomenclature for peroxisome assembly genes and proteins is outlined in Table I. Questions should be addressed to the first three authors of this letter, who organized the nomenclature revision and who will comprise the nomenclature committee for the next 12 months. We thank *The Journal of Cell Biology* for stimulating the unification of our nomenclature and for providing the opportunity to present our resolution. We hope that these

Please address all correspondence to Dr. Stephen J. Gould, Departments of Biological Chemistry and Cell Biology and Anatomy, The Johns Hopkins University School of Medicine, 725 North Wolfe Street, Baltimore, MD 21205. Tel: (410) 955-3085. Fax: (410) 955-0215. E-Mail: Stephen.Gould@qmail.bs.jhu.edu

1. Abbreviations used in this paper: Sc, *Saccharomyces cerevisiae*; Pp, *Pichia pastoris*; Rn, *Rattus norvegicus*; Hs, *Homo sapiens*; Pa, *Podospora anserina*; Hp, *Hansenula polymorpha*; Yl, *Yarrowia lipolytica*; Cb, *Candida boidinii*; PBD, peroxisome biogenesis disorder.

Table I. Description of Peroxins and PEX Genes

PEX gene	Peroxin characteristics	Former name
PEX 1	117–127 kD AAA ATPase; subcellular distribution is unknown.	<i>ScPASI</i> (12) <i>PpPASI</i> (16)
PEX 2	C <sub>3</sub> HC <sub>4</sub> zinc-binding integral peroxisomal membrane protein; 35–52 kD; mutations responsible for complementation group 10 of the PBD.	<i>RnPAF1</i> (33) <i>HsPAF1</i> (29) <i>PaCARI</i> (2) <i>PpPER6</i> (40)
PEX 3	51–52-kD integral peroxisomal membrane protein lacking similarity to other proteins.	<i>ScPAS3</i> (17) <i>HpPER9</i> (1) <i>PpPAS2</i> (43)
PEX 4	21–24-kD peroxisome-associated ubiquitin-conjugating enzyme.	<i>ScPAS2</i> (42) <i>PpPAS4</i> (3)
PEX 5	PTS1 receptor; 64–69-kD protein containing 8–7 tetratricopeptide repeats; localized to the cytoplasm and peroxisome; mutations responsible for complementation group 2 of the PBD.	<i>PpPAS8</i> (24) <i>ScPAS10</i> (37) <i>HsPXR1</i> (5) <i>HsPTS1R</i> (13, 44) <i>HpPER3</i> (36) <i>HpPAH2</i> (27) <i>YIPAY32</i> (31)
PEX 6	Belongs to the AAA family of ATPases; 112–127 kD; localized to cytoplasm and peroxisome; mutations responsible for complementation group 4 of the PBD.	<i>PpPAS5</i> (30) <i>ScPAS8</i> (39) <i>YIPAY4</i> (25) <i>RnPAF2</i> (34) <i>HsPAAA1</i> (45)
PEX 7	PTS2 receptor; 42-kD protein containing 6 WD40 repeats localized to the cytoplasm and peroxisome.	<i>ScPAS7</i> (23) <i>ScPEB1</i> (46)
PEX 8	71–81-kD peroxisome-associated protein containing a PTS1 signal.	<i>HpPER1</i> (41) <i>PpPER3</i> (20)
PEX 9	42-kD integral peroxisomal membrane protein lacking similarity to other proteins.	<i>YIPAY2</i> (6)
PEX 10	C <sub>3</sub> HC <sub>4</sub> zinc-binding integral peroxisomal membrane protein; 34–48 kD.	<i>HpPER8</i> (32) <i>PpPAS7</i> (19)
PEX 11	27–32-kD peroxisome-associated protein involved in peroxisome proliferation.	<i>ScPMP27</i> (9, 22) <i>CbPMP30</i> (28)
PEX 12	48-kD C <sub>3</sub> HC <sub>4</sub> zinc-binding integral peroxisomal membrane protein.	<i>PpPAS10</i> (18)
PEX 13	SH3-containing, 40–43-kD integral peroxisomal membrane protein; binds the PTS1 receptor (7, 10, 14).	

changes will make it easier for the general scientific community, as well as ourselves, to follow the interesting and exciting research on peroxisome biogenesis.

## References

- Baerends, R.J.S., S.W. Rasmussen, R.E. Hilbrands, M. van der Heide, K.N. Faber, P.T.W. Reuvekamp, J.A.K.W. Kiel, J.M. Cregg, I.J. van der Klei, and M. Veenhuis. 1996. The *Hansenula polymorpha* *PER9* gene encodes a peroxisomal membrane protein essential for peroxisome assembly and integrity. *J. Biol. Chem.* 271:8887–8894.
- Berteaux-Lecellier, V., M. Picard, C. Thompson-Coffe, D. Zickler, A. Panyvier-Adoutte, and J.-M. Simonet. 1995. A nonmammalian homolog of the *PAF1* gene (Zellweger syndrome) discovered as a gene involved in caryogamy in the fungus *Podospora anserina*. *Cell* 81:1043–1051.
- Crane, D.I., J.E. Kalish, and S.J. Gould. 1994. The *Pichia pastoris* *PAS4* gene encodes a ubiquitin-conjugating enzyme required for peroxisome assembly. *J. Biol. Chem.* 269:21835–21844.
- Cregg, J.M., I.J. Vankiel, G.J. Sulter, M. Veenhuis, and W. Harder. 1990. Peroxisome-deficient mutants of *Hansenula polymorpha*. *Yeast* 6:87–97.
- Dodd, G., N. Braverman, C. Wong, A. Moser, H.W. Moser, P. Watkins, D. Valle, and S.J. Gould. 1995. Mutations in the PTS1 receptor gene, *PXR1*, define complementation group 2 of the peroxisome biogenesis disorders. *Nature Genet.* 9:115–124.
- Eitzen, G.A., J.D. Aitchison, R.K. Szilard, M. Veenhuis, M.W. Nuttley, and R.A. Rachubinski. 1995. The *Yarrowia lipolytica* gene *PAY2* encodes a 42-kDa peroxisomal integral membrane protein essential for matrix protein import and peroxisome enlargement but not for peroxisome membrane proliferation. *J. Biol. Chem.* 270:1429–1436.
- Elgersma, Y., L. Kwast, A. Klein, T. Voorn-Brouwer, M. van den Berg, B. Metzig, T. America, H.F. Tabak, and B. Distel. 1996. The SH3 domain of the *Saccharomyces cerevisiae* peroxisomal membrane protein Pex13p functions as a docking site for Pex5p, a mobile receptor for the import of PTS1 containing proteins. *J. Cell Biol.* 135:97–109.
- Elgersma, Y., M. van den Berg, H.F. Tabak, and B. Distel. 1993. An efficient positive selection procedure for the isolation of peroxisomal import and peroxisome assembly mutants of *Saccharomyces cerevisiae*. *Genetics* 135:731–740.
- Erdmann, R., and G. Blobel. 1995. Giant peroxisomes in oleic acid-induced *Saccharomyces cerevisiae* lacking the peroxisomal membrane protein Pmp27p. *J. Cell Biol.* 128:509–523.
- Erdmann, R., and G. Blobel. 1996. Identification of Pex13p, a peroxisomal membrane receptor for the PTS1 recognition factor. *J. Cell Biol.* 135:111–121.
- Erdmann, R., D. Veenhuis, D. Mertens, and W.-H. Kunau. 1989. Isolation of peroxisome-deficient mutants of *Saccharomyces cerevisiae*. *Proc. Natl. Acad. Sci. USA* 86:5419–5423.
- Erdmann, R., F.F. Wiebel, A. Flessau, J. Rytka, A. Beyer, K.U. Frohlich, and W.-H. Kunau. 1991. *PAS1*, a yeast gene required for peroxisome biogenesis, encodes a member of a novel family of putative ATPases. *Cell* 64:499–510.
- Fransen, M., C. Brees, E. Baumgart, J.C.T. Vanhooren, M. Baes, G.P. Mannaerts, and P.P. Van Veldhoven. 1995. Identification and characterization of the putative human peroxisomal C-terminal targeting signal import receptor. *J. Biol. Chem.* 270:7731–7736.
- Gould, S.J., J.E. Kalish, J.C. Morrell, J. Bjorkman, A.J. Urquhart, and D.I. Crane. 1996. An SH3 protein in the peroxisome membrane is a docking factor for the PTS1 receptor. *J. Cell Biol.* 135:85–95.
- Gould, S.J., D. McCollum, A.P. Spong, J.A. Heyman, and S. Subramani. 1992. Development of the yeast *Pichia pastoris* as a model organism for a genetic and molecular analysis of peroxisome assembly. *Yeast* 8:613–628.
- Heyman, J.A., E. Monosov, and S. Subramani. 1994. Role of the *PAS1* gene of *Pichia pastoris* in peroxisome biogenesis. *J. Cell Biol.* 127:1259–1273.
- Höfeld, J., M. Veenhuis, and W.H. Kunau. 1991. *PAS3*, a *Saccharomyces cerevisiae* gene encoding a peroxisomal integral membrane protein essential for peroxisome biogenesis. *J. Cell Biol.* 114:1167–1178.
- Kalish, J.E., G.A. Keller, J.C. Morrell, S.J. Mihalik, B. Smith, J.M. Cregg, and S.J. Gould. 1996. Characterization of a novel component of the peroxisomal protein import apparatus using fluorescent peroxisomal proteins. *EMBO (Eur. Mol. Biol. Organ.) J.* 15:3275–3285.
- Kalish, J.E., C. Theda, J.C. Morrell, J.M. Berg, and S.J. Gould. 1995. Formation of the peroxisome lumen is abolished by loss of *Pichia pastoris* *Pas7p*, a zinc-binding integral membrane protein of the peroxisome. *Mol. Cell. Biol.* 15:6406–6419.
- Liu, H., X. Tan, K.A. Russell, M. Veenhuis, and J.M. Cregg. 1995. *PER3*, a gene required for peroxisome biogenesis in *Pichia pastoris*, encodes a peroxisomal membrane protein involved in protein import. *J. Biol. Chem.* 270:10940–10951.
- Liu, H., X. Tan, M. Veenhuis, D. McCollum, and J.M. Cregg. 1992. An efficient screen for peroxisome-deficient mutants of *Pichia pastoris*. *J. Bacteriol.* 174:4943–4951.
- Marshall, P., Y. Krimkevich, R. Lark, J. Dyer, M. Veenhuis, and J. Goodman. 1995. Pmp27 promotes peroxisomal proliferation. *J. Cell Biol.* 129:345–355.
- Marzioch, M., R. Erdmann, M. Veenhuis, and W.-H. Kunau. 1994. *PAS7* encodes a novel yeast member of the WD-40 protein family essential for import of 3-oxoacyl-CoA thiolase, a PTS2-containing protein, into peroxisomes. *EMBO (Eur. Mol. Biol. Organ.) J.* 13:4908–4918.
- McCollum, D., E. Monosov, and S. Subramani. 1993. The *pas8* mutant of *Pichia pastoris* exhibits the peroxisomal protein import deficiencies of Zellweger syndrome cells. The *PAS8* protein binds to the COOH-terminal tripeptide peroxisomal targeting signal and is a member of the TPR protein family. *J. Cell Biol.* 121:761–774.
- Nuttley, W.M., A.M. Brade, G.A. Eitzen, M. Veenhuis, J.D. Aitchison, R.K. Szilard, J.R. Glover, and R.A. Rachubinski. 1994. *PAY4*, a gene required for peroxisome assembly in the yeast *Yarrowia lipolytica*, encodes a novel member of a family of putative ATPases. *J. Biol. Chem.* 269:556–566.
- Nuttley, W.M., A.M. Brade, C. Gaillardin, G.A. Eitzen, J.R. Glover, J.D. Aitchinson, and R.A. Rachubinski. 1993. Rapid identification and characterization of peroxisomal assembly mutants in *Yarrowia lipolytica*. *Yeast* 9:507–517.
- Nuttley, W.M., R.K. Szilard, J.J. Smith, M. Veenhuis, and R.A. Rachubinski. 1995. The *PAH2* gene is required for peroxisome assembly in the methyotrophic yeast *Hansenula polymorpha* and encodes a member of the tetratricopeptide repeat family of proteins. *Gene (Amst.)* 160:33–39.

28. Sakai, Y., P.A. Marshall, A. Saiganji, K. Takabe, H. Saiki, N. Kato, and J.M. Goodman. 1995. The *Candida boidinii* peroxisomal membrane protein Pmp30 has a role in peroxisomal proliferation and is functionally homologous to Pmp27 from *Saccharomyces cerevisiae*. *J. Bacteriol.* 177: 6773–6781.
29. Shimozawa, N., T. Tsukamoto, Y. Suzuki, T. Orii, Y. Shirayoshi, T. Mori, and Y. Fujiki. 1992. A human gene responsible for Zellweger syndrome that affects peroxisome assembly. *Science (Wash. DC)*. 255:1132–1134.
30. Spong, A.P., and S. Subramani. 1993. Cloning and characterization of PASS: a gene required for peroxisome biogenesis in the methylotrophic yeast *Pichia pastoris*. *J. Cell Biol.* 123:535–548.
31. Szilard, R.K., V.I. Titorenko, M. Veenhuis, and R.A. Rachubinski. 1995. Pay32p of the yeast *Yarrowia lipolytica* is an intraperoxisomal component of the matrix protein translocation machinery. *J. Cell Biol.* 131: 1453–1469.
32. Tan, X., H.R. Waterham, M. Veenhuis, and J.M. Cregg. 1995. The *Hansenula polymorpha* PER8 gene encodes a novel peroxisomal integral membrane protein involved in proliferation. *J. Cell Biol.* 128:307–319.
33. Tsukamoto, T., S. Miura, and Y. Fujiki. 1991. Restoration by a 35K membrane protein of peroxisome assembly in a peroxisome-deficient mammalian cell mutant. *Nature (Lond.)*. 350:77–81.
34. Tsukamoto, T., S. Miura, T. Nakai, S. Yokota, N. Shimozawa, Y. Suzuki, T. Orii, Y. Fujiki, F. Sakai, A. Bogaki, et al. 1995. Peroxisome assembly factor-2, a putative ATPase cloned by functional complementation on a peroxisome-deficient mammalian cell mutant. *Nature Genet.* 11:395–401.
35. Tsukamoto, T., S. Yokota, and Y. Fujiki. 1990. Isolation and characterization of Chinese hamster ovary cell mutants defective in assembly of peroxisomes. *J. Cell Biol.* 110:651–660.
36. van der Klei, I.J., R.E. Hibbrands, G.J. Swaving, H.R. Waterham, E.G. Vrieling, V.I. Titorenko, C.J.M.W. Harder, and M. Veenhuis. 1995. The *Hansenula polymorpha* PER3 gene is essential for the import of PTS1 proteins into the peroxisome matrix. *J. Biol. Chem.* 270:17229–17236.
37. van der Leij, I., M.M. Franse, Y. Elgersma, B. Distel, and H.F. Tabak. 1993. PAS10 is a tetratricopeptide-repeat protein that is essential for the import of most matrix proteins into peroxisomes of *Saccharomyces cerevisiae*. *Proc. Natl. Acad. Sci. USA*. 90:11782–11786.
38. van der Leij, I., M. van den Berg, R. Boot, M. Franse, B. Distel, and H.F. Tabak. 1992. Isolation of peroxisome assembly mutants from *Saccharomyces cerevisiae* with different morphologies using a novel positive selection procedure. *J. Cell Biol.* 119:153–162.
39. Voorn-Brouwer, T., I. van der Leij, W. Hemrika, B. Distel, and H.F. Tabak. 1993. Sequence of the PAS8 gene, the product of which is essential for biogenesis of peroxisomes in *Saccharomyces cerevisiae*. *Biochim. Biophys. Acta*. 1216:325–328.
40. Waterham, H.R., Y. de Vries, K.A. Russel, W. Xie, M. Veenhuis, and J.M. Cregg. 1996. The *Pichia pastoris* PER6 gene product is a peroxisomal integral membrane protein essential for peroxisome biogenesis and has sequence similarity to the Zellweger syndrome protein PAF-1. *Mol. Cell. Biol.* 16:2527–2536.
41. Waterham, H.R., V.I. Titorenko, P. Haima, J.M. Cregg, W. Harder, and M. Veenhuis. 1994. The *Hansenula polymorpha* PER1 gene is essential for peroxisomal matrix protein with both carboxy-and amino-terminal targeting signals. *J. Cell Biol.* 127:737–749.
42. Wiebel, F.F., and W.-H. Kunau. 1992. The Pas2 protein essential for peroxisome biogenesis is related to ubiquitin-conjugating enzymes. *Nature (Lond.)*. 359:73–76.
43. Wiemer, E.A.C., G.H. Luers, K.N. Faber, T. Wenzel, M. Veenhuis, and S. Subramani. 1996. Isolation and characterization of Pas2p, a peroxisomal membrane protein essential for peroxisome biogenesis in the methylotrophic yeast *Pichia pastoris*. *J. Biol. Chem.* 271:18973–18980.
44. Wiemer, E.A.C., W.M. Nuttley, B.L. Bertolaet, X. Li, U. Franke, M.J. Wheelock, W.K. Anne, K.R. Johnson, and S. Subramani. 1995. Human peroxisomal targeting signal-1 receptor restores peroxisomal protein import in cells from patients with fatal peroxisomal disorders. *J. Cell Biol.* 130:51–65.
45. Yahraus, T., N. Braverman, G. Dodt, J.E. Kalish, J.C. Morrell, H.W. Moser, D. Valle, and S.J. Gould. 1996. The peroxisome biogenesis disorder group 4 gene, *PXAAAI*, encodes a cytoplasmic ATPase required for stability of the PTS1 receptor. *EMBO (Eur. Mol. Biol. Organ.) J.* 15: 2914–2923.
46. Zhang, J.W., and P.B. Lazarow. 1995. *PEB1 (PAST7)* in *Saccharomyces cerevisiae* encodes a hydrophilic, intra-peroxisomal protein that is a member of the WD repeat family and is essential for the import of thiolase into peroxisomes. *J. Cell Biol.* 129:65–80.
47. Zhang, J.W., Y. Han, and P.B. Lazarow. 1993. Novel peroxisome clustering mutants and peroxisome biogenesis mutants of *Saccharomyces cerevisiae*. *J. Cell Biol.* 123:1133–1147.
48. Zoeller, R.A., and C.R.H. Raetz. 1986. Isolation of animal cell mutants deficient in plasmalogens biosynthesis and peroxisome assembly. *Proc. Natl. Acad. Sci. USA*. 83:5170–5174.