

## 7 Literatur

- Abe, I., Okumoto, K., Tamura, S., and Fujiki, Y. 1998. Clofibrate-inducible, 28-kDa peroxisomal integral membrane protein is encoded by *PEX11*. *FEBS Lett.* 431:468-472.
- Albertini, M., Girzalsky, W., Veenhuis, M. and Kunau, W.H. 2001. Pex12p of *Saccharomyces cerevisiae* is a component of a multi-protein complex essential for peroxisomal matrix protein import. *Eur J Cell Biol.* 80:257-70.
- Albertini, M., P. Rehling, R. Erdmann, W. Girzalsky, J.A. Kiel, M. Veenhuis, and W.H. Kunau. 1997. Pex14p, a peroxisomal membrane protein binding both receptors of the two PTS-dependent import pathways. *Cell.* 89:83-92.
- Ausubel, F.J., Brent, R.; Kingston, R.E., Moore, D.D., Seidman, J.G., Smith, J.A., Struhl, K. 1989. Current Protocols in Molecular Biology. Greene Publishing Associates and Wiley Interscience, New York.
- Baerends, R.J.S., K.N. Faber, A.M. Kram, J.A.K.W. Kiel, I.J. van der Kleij, and M. Veenhuis. 2000. A stretch of positively charged amino acids at the N terminus of *Hansenula polymorpha* Pex3p is involved in incorporation of the protein into the peroxisomal membrane. *J Biol Chem.* 275:9986-95.
- 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.
- Baumgartner, U., B. Hamilton, M. Piskacek, H. Ruis, and H. Rottensteiner. 1999. Functional analysis of the Zn(2)Cys(6) transcription factors Oaf1p and Pip2p. Different roles in fatty acid induction of beta-oxidation in *Saccharomyces cerevisiae*. *J Biol Chem.* 274:22208-16.
- Bellion, E., and J.M. Goodman. 1987. Proton ionophores prevent assembly of a peroxisomal protein. *Cell.* 48:165-73.
- Biardi, L., and S.K. Krisans. 1996. Compartmentalization of cholesterol biosynthesis. Conversion of mevalonate to farnesyl diphosphate occurs in the peroxisomes. *J Biol Chem.* 271:1784-8.
- Blattner, J., B. Swinkels, H. Dorsam, T. Prospero, S. Subramani, and C. Clayton. 1992. Glycosome assembly in trypanosomes: variations in the acceptable degeneracy of a COOH-terminal microbody targeting signal. *J Cell Biol.* 119:1129-36.
- Bottger, G., P. Barnett, A.T. Klein, A. Kragt, H.F. Tabak, and B. Distel. 2000. *Saccharomyces cerevisiae* PTS1 receptor Pex5p interacts with the SH3 domain of the peroxisomal

- membrane protein Pex13p in an unconventional, non-PXXP-related manner. *Mol Biol Cell.* 11:3963-76.
- Braun, A., Kammerer, S., Weissenhorn, W., Weiss, E.H., and Cleve, H. 1994. Sequence of a putative human housekeeping gene (*HK33*) localized on chromosome 1. *Gene.* 146:291-295.
- Braverman, N., G. Dodt, S.J. Gould, and D. Valle. 1995. Disorders of peroxisome biogenesis. *Hum Mol Genet.* 4:1791-8.
- Braverman, N., G. Dodt, S.J. Gould, and D. Valle. 1998. An isoform of pex5p, the human PTS1 receptor, is required for the import of PTS2 proteins into peroxisomes. *Hum Mol Genet.* 7:1195-205.
- Braverman, N., G. Steel, C. Obie, A. Moser, H. Moser, S.J. Gould, and D. Valle. 1997. Human PEX7 encodes the peroxisomal PTS2 receptor and is responsible for rhizomelic chondrodysplasia punctata. *Nat Genet.* 15:369-76.
- Breidenbach, R.W., and H. Beevers. 1967. Association of the glyoxylate cycle enzymes in a novel subcellular particle from castor bean endosperm. *Biochem. Biophys. Res. Commun.* 27:462-469.
- Brocard, C., F. Kragler, M.M. Simon, T. Schuster, and A. Hartig. 1994. The tetratricopeptide repeat-domain of the PAS10 protein of *Saccharomyces cerevisiae* is essential for binding the peroxisomal targeting signal-SKL. *Biochem Biophys Res Commun.* 204:1016-22.
- Brocard, C., G. Lametschwandtner, R. Koudelka, and A. Hartig. 1997. Pex14p is a member of the protein linkage map of Pex5p. *Embo J.* 16:5491-500.
- Brosius, U., T. Dehmel, and J. Gartner. 2002. Two different targeting signals direct human peroxisomal membrane protein 22 to peroxisomes. *J Biol Chem.* 277:774-84.
- Brown, T.W., V.I. Titorenko, and R.A. Rachubinski. 2000. Mutants of the *Yarrowia lipolytica* PEX23 gene encoding an integral peroxisomal membrane peroxin mislocalize matrix proteins and accumulate vesicles containing peroxisomal matrix and membrane proteins. *Mol Biol Cell.* 11:141-52.
- Chang, C.C., W.H. Lee, H. Moser, D. Valle, and S.J. Gould. 1997. Isolation of the human PEX12 gene, mutated in group 3 of the peroxisome biogenesis disorders. *Nat Genet.* 15:385-8.
- Chang, C.C., D.S. Warren, K.A. Sacksteder, and S.J. Gould. 1999. PEX12 interacts with PEX5 and PEX10 and acts downstream of receptor docking in peroxisomal matrix protein import. *J Cell Biol.* 147:761-74.
- Collins, C.S., J.E. Kalish, J.C. Morrell, J.M. McCaffery, and S.J. Gould. 2000. The peroxisome biogenesis factors Pex4p, Pex22p, Pex1p und Pex6p act in the terminal steps of peroxisomal matrix protein import. *Mol Cell Bio.* 20:7516-26.

- 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.* 269:21835-44.
- Cregg, J.M., I.J. van der Klei, G.J. Sulter, M. Veenhuis, and W. Harder. 1990. Peroxisome-deficient mutants of *Hansenula polymorpha*. *Yeast*. 6:87-97.
- Crookes, W.K., and Olsen, L.J. 1999. Peroxin puzzles and folded freight: peroxisomal protein import in review. *Naturwissenschaften*. 86:51-61.
- Dammai, V., and S. Subramani. 2001. The human peroxisomal targeting signal receptor, Pex5p, is translocated into the peroxisomal matrix and recycled to the cytosol. *Cell*. 105:187-96.
- de Hoop, M.J., and G. Ab. 1992. Import of proteins into peroxisomes and other microbodies. *Biochem J*. 286:657-669.
- DeDuve, C., and P. Baudhuin. 1966. Peroxisomes (microbodies and related particles). *Physiol. Rev.* 46:323-357.
- Diestelkotter, P., and W.W. Just. 1993. In vitro insertion of the 22-kD peroxisomal membrane protein into isolated rat liver peroxisomes. *J Cell Biol.* 123:1717-25.
- Distel, B., R. Erdmann, S.J. Gould, G. Blobel, D.I. Crane, J.M. Cregg, G. Dodt, Y. Fujiki, J.M. Goodman, W.W. Just, J.A. Kiel, W.H. Kunau, P.B. Lazarow, G.P. Mannaerts, H.W. Moser, T. Osumi, R.A. Rachubinski, A. Roscher, S. Subramani, H.F. Tabak, T. Tsukamoto, D. Valle, I. van der Klei, P.P. van Veldhoven, and M. Veenhuis. 1996. A unified nomenclature for peroxisome biogenesis factors. *J Cell Biol.* 135:1-3.
- Dodt, 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. *Nat Genet.* 9:115-25.
- Dodt, G., and S.J. Gould. 1996. Multiple PEX genes are required for proper subcellular distribution and stability of Pex5p, the PTS1 receptor: evidence that PTS1 protein import is mediated by a cycling receptor. *J Cell Biol.* 135:1763-74.
- Einwächter, H., S. Sowinski, W.H. Kunau, and W. Schliebs. 2001. *Yarrowia lipolytica* Pex20p, *Saccharomyces cerevisiae* Pex18p/Pex21p and mammalian Pex5pL fulfil a common function in the early steps of the peroxisomal PTS2 import pathway. *EMBO Rep.* 2:1035-9.
- Eitzen, G.A., Aitchison, J.D., Szilard, R.K., Veenhuis, M., Nuttley, W.M., and Rachubinski, R.A. 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.

- Eitzen, G.A., Szilard, R.K., and Rachubinski, R.A. 1997. Enlarged peroxisomes are present in oleic acid-grown *Yarrowia lipolytica* overexpressing the *PEX16* gene encoding an intraperoxisomal peripheral membrane peroxin. *J. Cell Biol.* 137:1265-1278.
- Elgersma, Y., M. Elgersma-Hooisma, T. Wenzel, J.M. McCaffery, M.G. Farquhar, and S. Subramani. 1998. A mobile PTS2 receptor for peroxisomal protein import in *Pichia pastoris*. *J Cell Biol.* 140:807-20.
- Elgersma, Y., L. Kwast, A. Klein, T. Voorn-Brouwer, M. van den Berg, B. Metzig, T. America, H.F. Tabak, and B. Distel. 1996a. The SH3 domain of the *Saccharomyces cerevisiae* peroxisomal membrane protein Pex13p functions as a docking site for Pex5p, a mobile receptor for the import PTS1-containing proteins. *J Cell Biol.* 135:97-109.
- Elgersma, Y., L. Kwast, M. van den Berg, W.B. Snyder, B. Distel, S. Subramani, and H.F. Tabak. 1997. Overexpression of Pex15p, a phosphorylated peroxisomal integral membrane protein required for peroxisome assembly in *S.cerevisiae*, causes proliferation of the endoplasmic reticulum membrane. *Embo J.* 16:7326-41.
- 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*. 135:731-40.
- Elgersma, Y., A. Vos, M. van den Berg, C.W. van Roermund, P. van der Sluijs, B. Distel, and H.F. Tabak. 1996b. Analysis of the carboxyl-terminal peroxisomal targeting signal 1 in a homologous context in *Saccharomyces cerevisiae*. *J Biol Chem.* 271:26375-82.
- Erdmann, R., and G. Blobel. 1995. Giant peroxisomes in oleic acid-induced *Saccharomyces cerevisiae* lacking the peroxisomal membrane protein Pmp27p. *J Cell Biol.* 128:509-23.
- Erdmann, R., and G. Blobel. 1996. Identification of Pex13p a peroxisomal membrane receptor for the PTS1 recognition factor. *J Cell Biol.* 135:111-21.
- Erdmann, R., and W.H. Kunau. 1992. A genetic approach to the biogenesis of peroxisomes in the yeast *Saccharomyces cerevisiae*. *Cell Biochem Funct.* 10:167-74.
- Erdmann, R., and W.H. Kunau. 1994. Purification and immunolocalization of the peroxisomal 3-oxoacyl-CoA thiolase from *Saccharomyces cerevisiae*. *Yeast*. 10:1173-82.
- Erdmann, R., M. Veenhuis, and W.H. Kunau. 1997. Peroxisomes: organelles at the cross-roads. *Trends Cell Biol.* 7:400-407.
- Erdmann, R., M. Veenhuis, D. Mertens, and W.H. Kunau. 1989. Isolation of peroxisome-deficient mutants of *Saccharomyces cerevisiae*. *Proc Natl Acad Sci U S A.* 86:5419-23.
- 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.

- Feilotter, H.E., G.J. Hannon, C.J. Ruddell, and D. Beach. 1994. Construction of an improved host strain for two hybrid screening. *Nucleic Acids Res.* 22:1502-3.
- Fields, S., and O. Song. 1989. A novel genetic system to detect protein-protein interactions. *Nature.* 340:245-6.
- Frank, R. 1992. Spot synthesis: an easy technique for the positionally addressable, parallel chemical synthesis on a membrane support. *Tetrahedron.* 44:6031-6040.
- Fransen, M., C. Brees, E. Baumgart, J.C. Vanhooren, M. Baes, G.P. Mannaerts, and P.P.V. Veldhoven. 1995. Identification and characterization of the putative human peroxisomal C-terminal targeting signal import receptor. *J Biol Chem.* 270:7731-6.
- Fransen, M., T. Wylin, C. Brees, G.P. Mannaerts, and P.P.V. Veldhoven. 2001. Human pex19p binds peroxisomal integral membrane proteins at regions distinct from their sorting sequences. *Mol Cell Biol.* 21:4413-24.
- Fransen, M., S.R. Terlecky, and S. Subramani. 1998. Identification of a human PTS1 receptor docking protein directly required for peroxisomal protein import. *Proc Natl Acad Sci U S A.* 95:8087-92.
- Fujiki, Y., K. Okumoto, H. Otera, and S. Tamura. 2000. Peroxisome biogenesis and molecular defects in peroxisome assembly disorders. *Cell Biochem Biophys.* 32:155-64.
- Fukuda, S., H. Shimozawa, Y. Suzuki, Z. Zhang, S. Tomatsu, T. Tsukamoto, N. Hashiguchi, T. Osumi, M. Masuno, K. Imaizumi, Y. Kuroki, Y. Fujiki, T. Orii, and N. Kondo. 1996. Human peroxisome assembly factor-2 (PAF-2): A gene responsible for group C peroxisome biogenesis disorders in humans. *Am. J. Hum. Gen.* 59:1210-1220.
- Geisbrecht, B.V., K. Schulz, K. Nau, M.T. Geraghty, H. Schulz, R. Erdmann, and S.J. Gould. 1999. Preliminary characterization of Yor180Cp: identification of a novel peroxisomal protein of *Saccharomyces cerevisiae* involved in fatty acid metabolism. *Biochem Biophys Res Commun.* 260:28-34.
- Geisbrecht, B.V., D. Zhu, K. Schulz, K. Nau, J.C. Morrell, M. Geraghty, H. Schulz, R. Erdmann, and S.J. Gould. 1998. Molecular characterization of *Saccharomyces cerevisiae* Delta3, Delta2-enoyl-CoA isomerase. *J Biol Chem.* 273:33184-91.
- Geraghty, M.T., D. Bassett, J.C. Morrell, G.J. Gatto, J. Bai, B.V. Geisbrecht, P. Hieter, and S.J. Gould. 1999. Detecting patterns of protein distribution and gene expression in silico. *Proc Natl Acad Sci USA.* 96:2937-42.
- Gietl, C. 1990. Glyoxisomal malate dehydrogenase from watermelon is synthesized with an amino-terminal transit peptide. *Proc. Natl. Acad. Sci. U S A.* 87:33184-33191.
- Gietl, C., K.N. Faber, I.J. van der Klei, and M. Veenhuis. 1994. Mutational analysis of the N-terminal topogenic signal of watermelon glyoxysomal malate dehydrogenase using the heterologous host *Hansenula polymorpha*. *Proc. Natl. Acad. Sci. U S A.* 91:3151-3155.

- Gietz, D., A. Jean, R.A. Woods, and R.H. Schiestl. 1992. Improved method for high efficiency transformation of intact yeast cells. *Nucleic Acids Res.* 20:1425.
- Gietz, R.D., and A. Sugino. 1988. New yeast-Escherichia coli shuttle vectors constructed with in vitro mutagenized yeast genes lacking six-base pair restriction sites. *Gene.* 74:527-34.
- Girzalsky, W., P. Rehling, K. Stein, J. Kipper, L. Blank, W.H. Kunau, and R. Erdmann. 1999. Involvement of Pex13p in Pex14p localization and peroxisomal targeting signal 2-dependent protein import into peroxisomes. *J Cell Biol.* 144:1151-62.
- Glover, J.R., D.W. Andrews, and R.A. Rachubinski. 1994. *Saccharomyces cerevisiae* peroxisomal thiolase is imported as a dimer. *Proc. Natl. Acad. Sci. USA.* 91.
- Goldfischer, S., C.L. Moor, A.B. Johnson, J.S. A, M.P. Valsamis, H.K. Wisniewski, R.H. Ritch, W.T. Norton, I. Rapin, and I.M. Garner. 1973. Peroxisomal and mitochondrial defects in cerebrohepatorenal syndrome. *Science.* 182:62-64.
- Götte, K., W. Girzalsky, M. Linkert, E. Baumgart, S. Kammerer, W.H. Kunau, and R. Erdmann. 1998. Pex19p, a farnesylated protein essential for peroxisome biogenesis. *Mol Cell Biol.* 18:616-28.
- Gould, S.J. 2001. The Peroxisome for The Scientist. In: <http://www.peroxisome.org/Scientist/Biogenesis/> (unveröffentlichte Beobachtungen).
- Gould, S.J., J.E. Kalish, J.C. Morrell, J. Bjorkman, A.J. Urquhart, and D.I. Crane. 1996. Pex13p is an SH3 protein of the peroxisome membrane and a docking factor for the predominantly cytoplasmic PTs1 receptor. *J Cell Biol.* 135:85-95.
- Gould, S.J., G.A. Keller, N. Hosken, J. Wilkinson, and S. Subramani. 1989. A conserved tripeptide sorts proteins to peroxisomes. *J Cell Biol.* 108:1657-64.
- Gould, S.J., G.A. Keller, M. Schneider, S.H. Howell, L.J. Garrard, J.M. Goodman, B. Distel, H. Tabak, and S. Subramani. 1990. Peroxisomal protein import is conserved between yeast, plants, insects and mammals. *Embo J.* 9:85-90.
- 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.* 8:613-28.
- Gould, S.J., and D. Valle. 2000. Peroxisome biogenesis disorders: genetics and cell biology. *Trends Genet.* 16:340-5.
- Güldener, U., S. Heck, T. Fielder, J. Beinhauer, and J.H. Hegemann. 1996. A new efficient gene disruption cassette for repeated use in budding yeast. *Nucleic Acids Res.* 24:2519-24.
- Hajra, A.K., and J.E. Bishop. 1982. Glycerolipid biosynthesis in peroxisomes via the acyldihydroxyacetone pathway. *Ann.N.Y.Acad.Sci.* 386:170-182.
- Heinemann, P., and W.W. Just. 1992. Peroxisomal protein import. *In vivo* evidence for a novel translocation competent compartment. *FEBS Lett.* 300:179-82.

- Henke, B., W. Girzalsky, V. Berteaux-Lecellier, and R. Erdmann. 1998. IDP3 encodes a peroxisomal NADP-dependent isocitrate dehydrogenase required for the beta-oxidation of unsaturated fatty acids. *J Biol Chem.* 273:3702-11.
- Hettema, E.H., W. Girzalsky, M. van Den Berg, R. Erdmann, and B. Distel. 2000. *Saccharomyces cerevisiae* Pex3p and Pex19p are required for proper localization and stability of peroxisomal membrane proteins. *Embo J.* 19:223-33.
- Hettema, E.H., C.C.M. Ruigrok, M.G. Koerkamp, M. van den Berg, H.F. Tabak, B. Distel, and I. Braakman. 1998. The cytosolic DnaJ-like protein djp1p is involved specifically in peroxisomal protein import. *J Cell Biol.* 142:421-34.
- Hettema, E.H., C.W. van Roermund, B. Distel, M. van den Berg, C. Vilela, C. Rodrigues-Pousada, R.J. Wanders, and H.F. Tabak. 1996. The ABC transporter proteins Pat1 and Pat2 are required for import of long-chain fatty acids into peroxisomes of *Saccharomyces cerevisiae*. *Embo J.* 15:3813-22.
- Heyman, F.A., E. Monosov, and S. Subramani. 1994. Role of the *PAS1* gene of *Pichia pastoris* in peroxisome biogenesis. *J Cell Biol.* 127:1259-1273.
- Hinnen, A., J.B. Hicks, and G.R. Fink. 1978. Transformation of yeast. *Proc Natl Acad Sci U S A.* 75:1929-33.
- 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.* 114:1167-78.
- Holroyd, C., and R. Erdmann. 2001. Protein translocation machineries of peroxisomes. *FEBS Lett.* 501:6-10.
- Honsho, M., and Y. Fujiki. 2001. Topogenesis of Peroxisomal Membrane Protein Requires a Short, Positively Charged Intervening-loop Sequence and Flanking Hydrophobic Segments. STUDY USING HUMAN MEMBRANE PROTEIN PMP34. *J. Biol. Chem.* 276:9375-9382.
- Huang, Y., R. Ito, T. Hashimoto, and M. Ito. 2000. A Missense Mutation in the RING finger Motif of PEX2 Protein Disturbs the Import of Peroxisome Targeting Signal 1 (PTS1)-Containing Protein but NOT the PTS2-Containing Protein. *Biochem. Biophys. res. Commun.* 270:717-721.
- Huhse, B., P. Rehling, M. Albertini, L. Blank, K. Meller, and W.H. Kunau. 1998. Pex17p of *Saccharomyces cerevisiae* is a novel peroxin and component of the peroxisomal protein translocation machinery. *J Cell Biol.* 140:49-60.
- Imanaka, T., Y. Shiina, T. Takano, T. Hashimoto, and T. Osumi. 1996. Insertion of the 70 kDa peroxisomal membrane protein into peroxisomal membranes *in vivo* and *in vitro*. *J. Biol. Chem.* 271:3706-3713.

- Imanaka, T., G.M. Small, and P.B. Lazarow. 1987. Translocation of acyl-CoA oxidase into peroxisomes requires ATP hydrolysis but not a membran potential. *J. Cell. Biol.* 140:2915-2922.
- James, P., J. Halladay, and E.A. Craig. 1996. Genomic libraries and a host strain designed for highly efficient two-hybrid selection in yeast. *Genetics*. 144:1425-36.
- Jansen, G.A., R. Ofman, S. Ferdinandusse, L. Ijlst, A.O. Muijsers, O.H. Skjeldal, O. Stokke, C. Jakobs, G.T. Besley, J.E. Wraith, and R.J. Wanders. 1997. Refsum disease is caused by mutations in the phytanoyl-CoA hydroxylase gene. *Nat Genet*. 17:190-3.
- Jones, J.M., J.C. Morrell, and S.J. Gould. 2001. Multiple distinct targeting signals in integral peroxisomal membrane proteins. *J Cell Biol*. 153:1141-50.
- Jones, J.M., K. Nau, M.T. Geraghty, R. Erdmann, and S.J. Gould. 1999. Identification of peroxisomal acyl-CoA thioesterases in yeast and humans. *J Biol Chem*. 274:9216-23.
- 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 J*. 15:3275-85.
- 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*. 15:6406-19.
- Kammerer, S., Holzinger, A., Welsch, U., and Roscher, A.A. 1998. Cloning and characterization of the gene encoding the human peroxisomal assembly protein Pex3p. *FEBS Lett*. 429:53-60.
- Kinoshita, N., K. Ghaedi, N. Shimozawa, R.J.A. Wanders, Y. Matsuzono, T. Imanaka, K. Okumoto, Y. Suzuki, N. Kondo, and Y. Fujiki. 1998. Newly identified Chinese hamster ovary cell mutants are defective in biogenesis of peroxisomal membrane vesicles (Peroxisomal ghosts), representing a novel complementation group in mammals. *J Biol Chem*. 273:24122-30.
- Koller, A., W.B. Snyder, K.N. Faber, T.J. Wenzel, L. Rangell, G.A. Keller, and S. Subramani. 1999. Pex22p of *Pichia pastoris*, essential for peroxisomal matrix protein import, anchors the ubiquitin-conjugating enzyme, Pex4p, on the peroxisomal membrane. *J Cell Biol*. 146:99-112.
- Komori, M., Rasmussen, S.W., Kiel, J.A.K.W., Baerends, R.J.S., Cregg, J.M., van der Klei, I.J., and Veenhuis, M. 1997. The *Hansenula polymorpha* PEX14 gene encodes a novel peroxisomal membrane protein essential for peroxisome biogenesis. *EMBO J*. 16.
- Kragler, F., Lageder, A., Raupachova, J., Binder, M., and Hartig, A. 1993. Two independent peroxisomal targeting signals in catalase A of *Saccharomyces cerevisiae*. *J. Cell Biol*. 120:665-673.

- Kramer, A., Schuster, A., Reineke, U., Malin, R., Volkmer-Engert, R., Landgraf, C., and Schneider-Mergener, J. 1994. Combinatorial Cellulose-Bound Peptide Libraries: Screening tools for the Identification of Peptides That Bind Ligands with Predefined Specificity. *Methods:A Companion to Methods in Enzymology.* 6:388-395.
- Kramer, A., and J. Schneider-Mergener. 1998. Synthesis of Peptide libraries on continuous cellulose membranes. *Meth.Mol.Biol.* 87:25-39.
- Krisans, S.K. 1996. Cell compartmentalization of cholesterol biosynthesis. *Ann N Y Acad Sci.* 804:142-64.
- Laemmli, U.K. 1970. Cleavage of structural proteins during the assembly of the head of bacteriophage T4. *Nature.* 227:680-5.
- Lametschwandtner, G., C. Brocard, M. Fransen, P. Van Veldhoven, J. Berger, and A. Hartig. 1998. The difference in recognition of terminal tripeptides as peroxisomal targeting signal 1 between yeast and human is due to different affinities of their receptor Pex5p to the cognate signal and to residues adjacent to it. *J Biol Chem.* 273:33635-43.
- Lazarow, P.B., and Fujiki, Y. 1985. Biogenesis of peroxisomes. *Annu. Rev. Cell biol.* 1:489-530.
- Lazarow, P.B., and H.W. Moser. 1995. Disorders in peroxisome biogenesis. In The metabolic and molecular bases of inherited disease. Vol. 2. C.R. Scriver, Beaudet, C.R., Sly, W.S., Valle, D., editor. McGraw-Hill Book Co., New York, N.Y. 2287-2324.
- Lee, M.S., and R.T. Mullen, und Trelease, R.N. 1997. Oilseed isocitrate lyases lacking their essential type 1 peroxisomal targeting signal are piggybacked into glyoxysomes. *Plant Cell.* 9:185-197.
- Li, X., E. Baumgart, J.C. Morrell, G. Jimenez-Sanchez, D. Valle, and S.J. Gould. 2002. PEX11{beta} Deficiency Is Lethal and Impairs Neuronal Migration but Does Not Abrogate Peroxisome Function. *Mol. Cell. Biol.* 22:4358-4365.
- Liu, H., X. Tan, K. A. Russel, 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.
- Maniatis, T., E.F. Fritsch, and J. Sambrook. 1982. Molecular cloning: A laboratory manual. Cold Spring Harbor Laboratoy Press, New York.
- Marshall, P.A., Y.I. Krimkevich, R.H. Lark, J.M. Dyer, M. Veenhuis, and J.M. Goodman. 1995. Pmp27 promotes peroxisomal proliferation. *J Cell Biol.* 129:345-55.
- 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 J.* 13:4908-18.

- Matsuzono, Y., N. Kinoshita, S. Tamura, N. Shimozawa, M. Hamasaki, K. Ghaedi, R.J. Wanders, Y. Suzuki, N. Kondo, and Y. Fujiki. 1999. Human PEX19: cDNA cloning by functional complementation, mutation analysis in a patient with Zellweger syndrome, and potential role in peroxisomal membrane assembly. *Proc Natl Acad Sci U S A.* 96:2116-21.
- McCammon, M.T., J.A. McNew, P.J. Willy, and J.M. Goodman. 1994. An internal region of the peroxisomal membrane protein PMP47 is essential for sorting to peroxisomes. *J Cell Biol.* 124:915-25.
- 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 [published erratum appears in *J Cell Biol* 1993 Sep;122(5):following 1143]. *J Cell Biol.* 121:761-74.
- McNew, J.A., and J.M. Goodman. 1994. An oligomeric protein is imported into peroxisomes *in vivo*. *J Cell Biol.* 127:1245-57.
- McNew, J.A., and J.M. Goodman. 1996. The targeting and assembly of peroxisomal proteins: some old rules do not apply. *Trends Biochem. Sci.* 21:54-58.
- Mihalik, S.J., J.C. Morrell, D. Kim, K.A. Sacksteder, P.A. Watkins, and S.J. Gould. 1997. Identification of PAHX, a Refsum disease gene. *Nat Genet.* 17:185-9.
- Moser, A.B., M. Rasmussen, S. Naidu, P.A. Watkins, M. McGuiness, A.K. Hajra, G. Chen, G. Raymond, A. Liu, D. Gordon, K. Garnaas, D.S. Walton, O.H. Skjeldal, M.A. Guggenheim, L.G. Jackson, E.R. Elias, and H.W. Moser. 1995. Phenotype of patients with peroxisomal disorders subdivided into sixteen complementation groups. *J. Pediatr.* 127:13-22.
- Motley, A., E. Hettema, B. Distel, and H. Tabak. 1994. Differential protein import deficiencies in human peroxisome assembly disorders. *J Cell Biol.* 125:755-67.
- Motley, A., M.J. Lumb, P.B. Oatey, P.R. Jennings, P.A. De Zoysa, R.J. Wanders, H.F. Tabak, and C.J. Danpure. 1995. Mammalian alanine/glyoxylate aminotransferase 1 is imported into peroxisomes via the PTS1 translocation pathway. Increased degeneracy and context specificity of the mammalian PTS1 motif and implications for the peroxisome-to-mitochondrion mistargeting of AGT in primary hyperoxaluria type 1. *J Cell Biol.* 131:95-109.
- Motley, A.M., E.H. Hettema, E.M. Hogenhout, P. Brites, A.L. ten Asbroek, F.A. Wijburg, F. Baas, H.S. Heijmans, H.F. Tabak, R.J. Wanders, and B. Distel. 1997. Rhizomelic chondrodysplasia punctata is a peroxisomal protein targeting disease caused by a non-functional PTS2 receptor. *Nat Genet.* 15:377-80.

- Müller, W.H., van der Krift, T.P., Krouwer, A.J.J., Wosten, H.A.B. and van der Voort, L.H.M. 1991. Localisation of the pathway of the penicillin biosynthesis in *Penicillium chrysogenum*. *EMBO J.* 10:489-496.
- Mumberg, D., R. Muller, and M. Funk. 1994. Regulatable promoters of *Saccharomyces cerevisiae*: comparison of transcriptional activity and their use for heterologous expression. *Nucleic Acids Res.* 22:5767-8.
- Nakagawa, T., T. Imanaka, M. Morita, K. Ishiguro, H. Yurimoto, A. Yamashita, N. Kato, and Y. Sakai. 2000. Peroxisomal membrane protein Pmp47 is essential in the metabolism of middle-chain fatty acid in yeast peroxisomes and is associated with peroxisome proliferation. *J Biol Chem.* 275:3455-61.
- Neer, E.J., C.J. Schmidt, R. Nambudripad, and T.F. Smith. 1994. The ancient regulatory-protein family of WD-repeat proteins. *Nature*. 371:297-300.
- Novikoff, A.B., and W.Y. Shin. 1964. The endoplasmatic reticulum in the golgi zone and its relations to microbodies, golgiapparatus and autophagic vacuoles in rat liver cells. *J. Mikros. Oxford*. 3:187-206.
- Nuttley, W.M., Brade, A.M., Eitzen, G.A., Veenhuis, M., Aitchison, J.D., Szilard, R.K., Glover, J.R., and Rachubinski, R.A. 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., Szilard, R.K., Smith, J.J., Veenhuis, M. and Rachubinski, R.A. 1995. The *PAH2* gene is required for peroxisome assembly in the methylotrophic yeast *Hansenula polymorpha* and encodes a member of the tetratricopeptide repeat family of proteins. *Gene*. 160:33-39.
- Nuttley, W.M., A.M. Brade, C. Gaillardin, G.A. Eitzen, J.R. Glover, J.D. Aitchison, and R.A. Rachubinski. 1993. Rapid identification and characterization of peroxisomal assembly mutants in *Yarrowia lipolytica*. *Yeast*. 9:507-517.
- Okumoto, K., I. Abe, and Y. Fujiki. 2000. Molecular anatomy of the peroxin Pex12p: ring finger domain is essential for Pex12p function and interacts with the peroxisome-targeting signal type 1-receptor Pex5p and a ring peroxin, Pex10p. *J Biol Chem.* 275:25700-10.
- Okumoto, K., N. Shimozawa, A. Kawai, S. Tamura, T. Tsukamoto, T. Osumi, H. Moser, R.J. Wanders, Y. Suzuki, N. Kondo, and Y. Fujiki. 1998. PEX12, the pathogenic gene of group III Zellweger syndrome: cDNA cloning by functional complementation on a CHO cell mutant, patient analysis, and characterization of PEX12p. *Mol Cell Biol.* 18:4324-36.
- Opperdoes, F.R., and Borst, P. 1977. Localization of nine glycolytic enzymes in a microbody-like organelle in *Trypanosoma Brucei*: the glycosome. *FEBS letters*. 80:360-364.

- Opperdoes, F.R. 1988. Glycosomes may provide clues to the import of peroxisomal proteins. *Trends Biochem Sci.* 13:255-60.
- Osumi, T., T. Tsukamoto, S. Hata, S. Yokota, S. Miura, Y. Fujiki, M. Hijikata, S. Miyazawa, and T. Hashimoto. 1991. Amino-terminal presequence of the precursor of peroxisomal 3-ketoacyl-CoA thiolase is a cleavable signal peptide for peroxisomal targeting. *Biochem. Biophys. Res. Commun.* 181:947-954.
- Otera, H., T. Harano, M. Honsho, K. Ghaedi, S. Mukai, A. Tanaka, A. Kawai, N. Shimizu, and Y. Fujiki. 2000. The mammalian peroxin Pex5pL, the longer isoform of the mobile peroxisome targeting signal (PTS) type 1 transporter, translocates the Pex7p.PTS2 protein complex into peroxisomes via its initial docking site, Pex14p. *J Biol Chem.* 275:21703-14.
- Otera, H., K. Okumoto, K. Tateishi, Y. Ikoma, E. Matsuda, M. Nishimura, T. Tsukamoto, T. Osumi, K. Ohashi, O. Higuchi, and Y. Fujiki. 1998. Peroxisome targeting signal type 1 (PTS1) receptor is involved in import of both PTS1 and PTS2: studies with PEX5-defective CHO cell mutants. *Mol Cell Biol.* 18:388-99.
- Passreiter, M., M. Anton, D. Lay, R. Frank, C. Harter, F.T. Wieland, K. Gorgas, and W.W. Just. 1998. Peroxisome biogenesis: involvement of ARF and coatomer. *J Cell Biol.* 141:373-383.
- Patel, S., Latterich, M. 1998. The AAA team: related ATPases with diverse functions. *Trends Cell Bio.* 8:65-71.
- Pause, B., Saffrich, R., Hunziker, A., Ansorge, W. and Just, W.W. 2000. Targeting of the 22 kDa integral peroxisomal membrane protein. *FEBS Lett.* 471:23-28.
- Portsteffen, H., Beyer, A., Becker, E., Epplen, C., Pawlak, A., Kunau, W.-H. and Dodt, G. 1997. Human *PEX1* is mutated in complementation group 1 of the peroxisome biogenesis disorders. *Nature Genet.* 17:449-452.
- Powers, J.M., and H.W. Moser. 1998. Peroxisomal disorders: genotype, phenotype, major neuropathologic lesions, and pathogenesis. *Brain Pathol.* 8:101-120.
- Purdue, P.E., and P.B. Lazarow. 2001a. Peroxisome biogenesis. *Annu Rev Cell Dev Biol.* 17:701-52.
- Purdue, P.E., and P.B. Lazarow. 2001b. Pex18p is constitutively degraded during peroxisome biogenesis. *J Biol Chem.* 276:47684-9.
- Purdue, P.E., X. Yang, and P.B. Lazarow. 1998. Pex18p and Pex21p, a novel pair of related peroxins essential for peroxisomal targeting by the PTS2 pathway. *J Cell Biol.* 143:1859-69.
- Purdue, P.E., J.W. Zhang, M. Skoneczny, and P.B. Lazarow. 1997. Rhizomelic chondrodysplasia punctata is caused by deficiency of human PEX7, a homologue of the yeast PTS2 receptor. *Nat Genet.* 15:381-4.

- Reguenga, C., M.E. Oliveira, A.M. Gouveia, C. Sa-Miranda, and J.E. Azevedo. 2001. Characterization of the mammalian peroxisomal import machinery: Pex2p, Pex5p, Pex12p, and Pex14p are subunits of the same protein assembly. *J Biol Chem.* 276:29935-42.
- Rehling, P., M. Albertini, and W.-H. Kunau. 1996a. Protein Import into Peroxisomes: New Developments. *Ann. NY Acad. of Sci.* 804:34-36.
- Rehling, P., M. Marzioch, F. Niesen, E. Wittke, M. Veenhuis, and W.H. Kunau. 1996b. The import receptor for the peroxisomal targeting signal 2 (PTS2) in *Saccharomyces cerevisiae* is encoded by the PAS7 gene. *Embo J.* 15:2901-13. abs.html.
- Rehling, P., A. Skaletz-Rorowski, W. Girzalsky, T. Voorn-Brouwer, M.M. Franse, B. Distel, M. Veenhuis, W.H. Kunau, and R. Erdmann. 2000. Pex8p, an intraperoxisomal peroxin of *Saccharomyces cerevisiae* required for protein transport into peroxisomes binds the PTS1 receptor pex5p. *J Biol Chem.* 275:3593-602.
- Reuber, B.E., E. Germain-Lee, C.S. Collins, J.C. Morrell, R. Ameritunga, H.W. Moser, D. Valle, and S.J. Gould. 1997. Mutations in PEX1 are the most common cause of peroxisome biogenesis disorders. *Nat Genet.* 17:445-8.
- Rottensteiner, H., A.J. Kal, B. Hamilton, H. Ruis, and H.F. Tabak. 1997. A heterodimer of the Zn2Cys6 transcription factors Pip2p and Oaf1p controls induction of genes encoding peroxisomal proteins in *Saccharomyces cerevisiae*. *Eur J Biochem.* 247:776-83.
- Sacksteder, K.A., J.M. Jones, S.T. South, X. Li, Y. Liu, and S.J. Gould. 2000. PEX19 binds multiple peroxisomal membrane proteins, is predominantly cytoplasmic, and is required for peroxisome membrane synthesis. *J Cell Biol.* 148:931-44.
- Sacksteder, K.A., J.C. Morrell, R.J. Wanders, R. Matalon, and S.J. Gould. 1999. MCD encodes peroxisomal and cytoplasmic forms of malonyl-CoA decarboxylase and is mutated in malonyl-CoA decarboxylase deficiency. *J Biol Chem.* 274:24461-8.
- Saidowsky, J., G. Dodt, K. Kirchberg, A. Wegner, W. Nastainczyk, W.H. Kunau, and W. Schliebs. 2001. The di-aromatic pentapeptide repeats of the human peroxisome import receptor PEX5 are separate high affinity binding sites for the peroxisomal membrane protein PEX14. *J Biol Chem.* 276:34524-9.
- Salomons, F.A., J.A. Kiel, K.N. Faber, M. Veenhuis, and I.J.v.d. Klei. 2000. Overproduction of Pex5p stimulates import of alcohol oxidase and dihydroxyacetone synthase in a *Hansenula polymorpha* Pex14 null mutant. *J Biol Chem.* 275:12603-11.
- Schäfer, H., K. Nau, A. Sickmann, R. Erdmann, and H.E. Meyer. 2001. Identification of peroxisomal membrane proteins of *Saccharomyces cerevisiae* by mass spectrometry. *Electrophoresis.* 22:2955-68.
- Schepers, L., M. Casteels, J. Vamecq, G. Parmentier, P.P. Van Veldhoven, and G.P. Mannerts. 1988.  $\beta$ -Oxidation of the carboxyl side chain of prostaglandin E2 in rat liver peroxisomes and mitochondria. *J. Biol. Chem.* 263:2724-2731.

- Schliebs, W., J. Saidowsky, B. Agianian, G. Dodt, F.W. Herberg, and W.H. Kunau. 1999. Recombinant human peroxisomal targeting signal receptor PEX5. Structural basis for interaction of PEX5 with PEX14. *J Biol Chem.* 274:5666-73.
- Schrader, M., B.E. Reuber, J.C. Morrell, G. Jimenez-Sanchez, C. Obie, T.A. Stroh, D. Valle, T.A. Schroer, and S.J. Gould. 1998. Expression of PEX11beta mediates peroxisome proliferation in the absence of extracellular stimuli. *J Biol Chem.* 273:29607-14.
- Shimizu, N., R. Itoh, Y. Hirono, H. Otera, and K. Ghaedi. 1999. The peroxin Pex14p-cDNA cloning by functional complementation on a Chinese hamster ovary cell mutant, characterization, and functional analysis. *J. Biol. Chem.* 274:12593-604.
- Shimozawa, N., Y. Suzuki, Z. Zhang, A. Imamura, N. Kondo, N. Kinoshita, Y. Fujiki, T. Tsukamoto, T. Osumi, T. Imanaka, T. Orii, F. Beemer, P. Mooijer, C. Dekker, and R.J. Wanders. 1998. Genetic basis of peroxisome-assembly mutants of humans, Chinese hamster ovary cells, and yeast: identification of a new complementation group of peroxisome-biogenesis disorders apparently lacking peroxisomal-membrane ghosts [letter]. *Am J Hum Genet.* 63:1898-903.
- 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.* 255:1132-1134.
- Sikorski, R.S., and P. Hieter. 1989. A system of shuttle vectors and yeast host strains designed for efficient manipulation of DNA in *Saccharomyces cerevisiae*. *Genetics.* 122:19-27.
- Small, G.M., Y. Luo, T. Wang, and I.V. Karpichev. 1996. Molecular regulation of peroxisomal acyl-CoA oxidase in yeast. *Ann N Y Acad Sci.* 804:362-72.
- Smith, J.J., and R.A. Rachubinski. 2001. A role for the peroxin Pex8p in Pex20p-dependent thiolase import into peroxisomes of the yeast *Yarrowia lipolytica*. *J Biol Chem.* 276:1618-25.
- Snyder, W.B., K.N. Faber, T.J. Wenzel, A. Koller, G.H. Luers, L. Rangell, G.A. Keller, and S. Subramani. 1999a. Pex19p interacts with Pex3p and Pex10p and is essential for peroxisome biogenesis in *Pichia pastoris*. *Mol Biol Cell.* 10:1745-61.
- Snyder, W.B., A. Koller, A.J. Choy, M.A. Johnson, J.M. Cregg, L. Rangell, G.A. Keller, and S. Subramani. 1999b. Pex17p is required for import of both peroxisome membrane and luminal proteins and interacts with Pex19p and the peroxisome targeting signal-receptor docking complex in *Pichia pastoris*. *Mol Biol Cell.* 10:4005-19.
- Snyder, W.B., A. Koller, A.J. Choy, and S. Subramani. 2000. The peroxin Pex19p interacts with multiple, integral membrane proteins at the peroxisomal membrane. *J Cell Biol.* 149:1171-8.
- Soukupova, M., C. Sprenger, K. Gorgas, W.H. Kunau, and G. Dodt. 1999. Identification and characterization of the human peroxin PEX3. *Eur J Cell Biol.* 78:357-74.

- South, S.T., and S.J. Gould. 1999. Peroxisome synthesis in the absence of preexisting peroxisomes. *J Cell Biol.* 144:255-66.
- South, S.T., K.A. Sacksteder, X. Li, Y. Liu, and S.J. Gould. 2000. Inhibitors of COPI and COPII do not block PEX3-mediated peroxisome synthesis. *J Cell Biol.* 149:1345-60.
- Spong, A.P., and S. Subramani. 1993. Cloning and characterization of PAS5: a gene required for peroxisome biogenesis in the methylotrophic yeast *Pichia pastoris*. *J Cell Biol.* 123:535-48.
- Stein, K., A. Schell-Steven, R. Erdmann, and H. Rottensteiner. 2002. Interactions of Pex7p and Pex18p/Pex21p with the peroxisomal docking machinery: implications for the first steps in PTS2 protein import. *Mol Cell Biol.* in press
- Subramani, S. 1993. Protein import into peroxisomes and biogenesis of the organelle. *Annu Rev Cell Biol.* 9:445-78.
- Subramani, S. 1996. Protein translocation into peroxisomes. *J Biol Chem.* 271:32483-6.
- Subramani, S. 1998. Components involved in peroxisome import, biogenesis, proliferation, turnover, and movement. *Physiol Rev.* 78:171-88.
- Subramani, S., A. Koller, and W.B. Snyder. 2000. Import of peroxisomal matrix and membrane proteins. *Annu Rev Biochem.* 69:399-418.
- Swinkels, B.W., S.J. Gould, A.G. Bodnar, R.A. Rachubinski, and S. Subramani. 1991. A novel, cleavable peroxisomal targeting signal at the amino-terminus of the rat 3-ketoacyl-CoA thiolase. *Embo J.* 10:3255-62.
- 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.* 131:1453-69.
- Takada, Y., and Noguchi, T. (1986). 1986. Ureidoglycolate lyase, a new metalloenzyme of peroxisomal urate degradation in marine fish liver. *Biochem. J.* 235:391-397.
- Tan, X., Waterham, H.R., Veenhuis, M., and Cregg, J.M. 1995. The *Hansenula polymorpha* PER8 gene encodes a novel peroxisomal integral membrane protein involved in proliferation. *J. Cell Biol.* 128:307-319.
- Terlecky, S.R., W.M. Nuttley, D. McCollum, E. Sock, and S. Subramani. 1995. The *Pichia pastoris* peroxisomal protein PAS8p is the receptor for the C-terminal tripeptide peroxisomal targeting signal. *Embo J.* 14:3627-34.
- Terlecky, S.R., E.A. Wiemer, W.M. Nuttley, P.A. Walton, and S. Subramani. 1996. Signals, receptors, and cytosolic factors involved in peroxisomal protein import. *Ann N Y Acad Sci.* 804:11-20.
- Titorenko, V.I., and R.A. Rachubinski. 2001. The life cycle of the peroxisome. *Nat Rev Mol Cell Biol.* 2:357-68.

- Titorenko, V.I., J.J. Smith, R.K. Szilard, and R.A. Rachubinski. 1998. Pex20p of the yeast *Yarrowia lipolytica* is required for the oligomerization of thiolase in the cytosol and for its targeting to the peroxisome. *J Cell Biol.* 142:403-20.
- Titorenko, V.I., J.J. Smith, R.K. Szilard, and R.A. Rachubinski. 2000. Peroxisome biogenesis in the yeast *Yarrowia lipolytica*. *Cell Biochem Biophys.* 32:21-6.
- Tsukamoto, T., Miura, S. and Fujiki, Y. 1991. Restoration by a 35K membrane protein of peroxisome assembly in a peroxisome-deficient mammalian cell mutant. *Nature.* 350:77-81.
- Tsukamoto, T., S. Miura, T. Nakai, S. Yokota, N. Shimozawa, Y. Suzuki, T. Orii, Y. Fujiki, F. Sakai, A. Bogaki, H. Yasumo, and T. Osumi. 1995. Peroxisome assembly factor-2, a putative ATPase cloned by functional complementation of a peroxisome-deficient mammalian cell mutant. *Nature Genet.* 11:395-401.
- 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.
- Urquhart, A.J., D. Kennedy, S.J. Gould, and D.I. Crane. 2000. Interaction of Pex5p, the type 1 peroxisome targeting signal receptor, with the peroxisomal membrane proteins Pex14p and Pex13p. *J Biol Chem.* 275:4127-36.
- van der Klei, I.J., Hilbrands, R.E., Swaving, G.J., Waterham, H.R., Vrielink, E.G., Titorenko, V.I., Cregg, J.M., Harder, W., and Veenhuis, M. 1995. The *Hansenula polymorpha* PER3 gene is essential for the import of PTS1 proteins into the peroxisomal matrix. *J. Biol. Chem.* 270:17229-17236.
- van der Klei, I.J., and Veenhuis, M. 1996. Peroxisome biogenesis in the yeast *Hansenula polymorpha*: a structural and functional analysis. *Ann. New York Acad. Sci.* 804:47-59.
- van der Klei, I.J., R.E. Hilbrands, J.A. Kiel, S.W. Rasmussen, J.M. Cregg, and M. Veenhuis. 1998. The ubiquitin-conjugating enzyme Pex4p of *Hansenula polymorpha* is required for efficient functioning of the PTS1 import machinery. *Embo J.* 17:3608-18.
- 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* [published erratum appears in Proc Natl Acad Sci U S A 1995 Jun 6;92(12):5759]. *Proc Natl Acad Sci U S A.* 90:11782-6.
- 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.* 119:153-62.
- van der Voorn, L., Ploegh, H.L. 1992. The WD-40 repeat. *FEBS Lett.* 307:131-134.

- Veenhuis, M., M. Mateblowski, W.H. Kunau, and W. Harder. 1987. Proliferation of microbodies in *Saccharomyces cerevisiae*. *Yeast*. 3:77-84.
- 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*. 1216:325-8.
- Walton, P.A., P.E. Hill, and S. Subramani. 1995. Import of stably folded proteins into peroxisomes. *Mol Biol Cell*. 6:675-83.
- Walton, P.A., M. Wendland, S. Subramani, R.A. Rachubinski, and W.J. Welch. 1994. Involvement of 70-kD heat-shock proteins in peroxisomal import. *J Cell Biol*. 125:1037-46.
- Wanders, R.J., R.B. Schutgens, and P.G. Barth. 1995. Peroxisomal disorders: a review. *J Neuropathol Exp Neurol*. 54:726-39.
- Wanders, R.J., and J.M. Tager. 1998. Lipid metabolism in peroxisomes in relation to human disease. *Mol Aspects Med*. 19:69-154.
- Warren, D.S., J.C. Morrell, H.W. Moser, D. Valle, and S.J. Gould. 1998. Identification of PEX10, the gene defective in complementation group 7 of the peroxisome-biogenesis disorders. *Am J Hum Genet*. 63:347-59.
- Waterham, H.R., de Vries, Y., Russel, K.A., Xie, W., Veenhuis, M., and Cregg, J.M. 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.
- Waterham, H.R., and J.M. Cregg. 1997. Peroxisome biogenesis. *Bioessays*. 19:57-66.
- 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 peroxisome biogenesis and encodes a peroxisomal matrix protein with both carboxy- and amino-terminal targeting signals. *J. Cell Biol*. 127:737-749.
- Wendland, M., and S. Subramani. 1993. Cytosol-dependent peroxisomal protein import in a permeabilized cell system. *J Cell Biol*. 120:675-85.
- Wiebel, F.F., and Kunau, W.-H. 1992. The PAS2 protein essential for peroxisome biogenesis is related to ubiquitin-conjugating enzymes. *Nature*. 359:73-76.
- Wiemer, E.A., W.M. Nuttley, B.L. Bertolaet, X. Li, U. Francke, M.J. Wheelock, U.K. Anne, K.R. Johnson, and S. Subramani. 1995a. Human peroxisomal targeting signal-1 receptor restores peroxisomal protein import in cells from patients with fatal peroxisomal disorders. *J Cell Biol*. 130:51-65.
- Wiemer, E.A., S.R. Terlecky, W.M. Nuttley, and S. Subramani. 1995b. Characterization of the yeast and human receptors for the carboxy-terminal tripeptide peroxisomal targeting signal. *Cold Spring Harb Symp Quant Biol*. 60:637-48.

- 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.* 271:18973-80.
- Will, G.K., M. Soukupova, X. Hong, K.S. Erdmann, J.A. Kiel, G. Dodt, W.H. Kunau, and R. Erdmann. 1999. Identification and characterization of the human orthologue of yeast Pex14p. *Mol Cell Biol.* 19:2265-77.
- 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, PXAAA1, encodes a cytoplasmic ATPase required for stability of the PTS1 receptor. *Embo J.* 15:2914-23.
- Zhang, B., S.L. Marcus, K.S. Miyata, S. Subramani, J.P. Capone, and R.A. Rachubinski. 1993. Characterization of protein-DNA interactions within the peroxisome proliferator-responsive element of the rat hydratase-dehydrogenase gene. *J Biol Chem.* 268:12939-45.
- Zhang, J.W., and P.B. Lazarow. 1995. *PEB1 (PAS7)* 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.
- Zhang, J.W., and P.B. Lazarow. 1996. Peb1p (Pas7p) is an intraperoxisomal receptor for the NH<sub>2</sub>-terminal, type 2, peroxisomal targeting sequence of thiolase: Peb1p itself is targeted to peroxisomes by an NH<sub>2</sub>-terminal peptide. *J Cell Biol.* 132:325-34.
- Zhang, Z., Y. Suzuki, N. Shimozawa, S. Fukuda, A. Imamura, T. Tsukamoto, T. Osumi, Y. Fujiki, T. Orii, R.J. Wanders, P.G. Barth, H.W. Moser, B.C. Paton, G.T. Besley, and N. Kondo. 1999. Genomic structure and identification of 11 novel mutations of the PEX6 (peroxisome assembly factor-2) gene in patients with peroxisome biogenesis disorders. *Hum Mutat.* 13:487-96.
- Zoeller, R.A., A. Morand, M.J. Santos, P.B. Lazarow, T. Hashimoto, A.M. Tarakoff, and C.R.H. Raetz. 1989. Chinese hamster ovary cell mutants defective in peroxisome biogenesis. *J. Biol. Chem.* 264:21872-21878.
- Zoeller, R.A., and C.R. Raetz. 1986. Isolation of animal cell mutants deficient in plasmalogen biosynthesis and peroxisome assembly. *Proc Natl Acad Sci U S A.* 83:5170-4.