

Figure Legends

Supplemental Figure S1: *Pex1*-Knock-in / Knock-out Construct Design. The construct was designed with several important features. Most importantly, a single nucleotide substitution was introduced in exon 15 to cause the amino acid substitution p.G844D. In addition, *loxP* sites were strategically introduced within introns 11 and 13 that would yield a null allele if *Pex1*-G844D carrier or homozygous mice were crossed with a *Cre* recombinase mouse. Thus, we have the opportunity to generate germ line null mice or tissue specific knock outs.

Supplemental Figure S2: *Pex1* Mouse and Human Protein Alignment. Alignment of the human PEX1 and murine *Pex1* amino acid sequences shows that the orthologous human PEX1-p.G843D and murine *Pex1*-p.G844D amino acids reside in a highly conserved sequence tract.

Supplemental Figure S3: PEX14 Immunostaining in *Pex1*-G844D and Wild Type Mice. The number and size of peroxisome structures are qualitatively similar in the cell lines derived from *Pex1*-G844D and wild type mouse pups.