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Supplemental Figure 8

Polycystin 1 isoform 2 precursor [Homo sapiens]-4302aa

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optimal cleavage motifs for TACE and ADAM10. The N-terminal site (PRFSHSFPR) is predicted to generate a 259,000 MW C-terminal PC1 fragment; the next downstream site (PLTQSIQAN) is predicted to generate a 225,000 MW C-terminal PC1 fragment. The predicted molecular weights of these C-terminal fragments correlate with the 230-260 kDa bands that are recognized by NM002 and NM005 via immunoblot in the absence of EDTA containing buffer (see Supplemental Figure 3F).
Supplemental Text

CD16.7-PC1 WT localizes to primary cilia of 3T3 cells, but truncation mutants do not. The integral membrane CD16.7-PC1-WT fusion protein encoding the last 112 aa of the conserved human PC1 tail (termed CD16.7-PC1-WT) localizes to the primary cilia and Golgi of mouse Swiss 3T3 cells (Supplemental Figure 1). Truncation of the last 20 or 40 amino acids of the PC1 C-terminus, CD16.7-PC1-359 and CD16.7-PC1-339 respectively, abolishes transport to cilia and results in intracellular protein accumulation in the Golgi and possibly the ER suggested by rimming of nuclear envelope (Supplemental Figure 1, yellow arrows).

C-terminally FLAG-tagged PC2 binds Arf4
Cells expressing C-terminally FLAG-tagged PC2 (amino acids 1-219) were used to further confirm Arf4 binding specificity. FLAG-PC2 precipitated purified Arf4 above the background levels seen with FLAG-BAP control protein (Supplemental Figure 2). Arf4 was minimally precipitated with FLAG-PC2-ΔRVxPx deletion mutant and could not be distinguished from FLAG-BAP negative control levels. Immunoblotting for the immunoprecipitated FLAG proteins confirmed their equal precipitation.

Specific antibodies against Polycystin-1 recognize protein in Golgi
Antibodies directed against a peptide epitope in the third cytoplasmic loop region of PC1 (NM002 and NM032) and remote from the C-terminal ciliary targeting sequence were generated in two different rabbits. By immunofluorescence and immuno-EM NM002 showed PC1 enriched in the Golgi and cilia of cultured kidney epithelial cells (MDCKII and RCTE) (Supplemental Figure 3A-C and main text Figure 3A). Acetylated α-tubulin served as a ciliary marker and α-2,6 sialyltransferase or Golgi lights served as independent Golgi markers (Supplemental Figure 3A-B and main text Figure 3A). The peptide antigen used to generate NM002 and NM032 was originally used to generate an antibody for the first definitive immunolocalization of PC1 in tissues though the subcellular localization of PC1 was not described (Van Adelsberg et al., 1997). Antibody specificity of NM002 pAb was further confirmed by peptide-blocking and siRNA depletion experiments (Supplemental Figure 3D-E). Both immunostaining and immunoblot reactivity were decreased by nearly 70% following siRNA depletion of PC1. Comparative evaluation of NM002 and NM0032 against our well-characterized PC1 specific antibody (NM005) raised against a pMAL-PC1 C-terminus fusion protein (Ward et al., 1996; Roitbak et al., 2004; Xu et al., 2007) showed all the antibodies have similar immunoreactivities by Western blot (Supplemental Figure 3F).

Control immunoprecipitations for Arf4, ASAP1, Rab5, Rab7, Rab8 and Rab11.

Polycystin-1 accumulated with mutant Arf4 and Rab11 isoforms.
Arf4-GFP-WT localizes in the Golgi (refer to Figure 3). To determine analyze the function of Arf4 and Rab11 in Golgi export, we analyzed the expression of mutant proteins Arf4 I46D and Rab11 S25N. We used the Arf4 I46D mutant, which blocks ASAP1 interaction, causes morphological defects, including Golgi collapse, and results in retinal degeneration in transgenic animals (Deretic et al., 2005). Accordingly, Arf4-GFP-I46D accumulated in regions predicted to be collapsed Golgi in BHK and MDCK cells (Supplemental Figure 6, top and middle panels). PC1 was found to overlap with these Arf4-GFP-I46D accumulations. The Rab11 S25N dominant
negative mutant causes Golgi accumulation and has been shown to reduce the rate of transferrin recycling, as the Golgi serves as a hub for Rab11 recruitment and activation (Chen et al., 1998; Chen and Wandinger-Ness, 2001). PC1 signals strongly overlapped with GFP-Rab11 S25N, suggesting that this mutant causes PC1 retention in the Golgi (Supplemental Figure 6).

**Characterization of shRNA mediated depletion of Arf4.**
Arf4 shRNA (OligoIDs: V3LHS-410096, V2LMM-62890, V2LHS-271123, V3LHS-371138, V2LHS-92212) and Rab8 shRNA (OligoID: V3LHS-359728) clones obtained in pGIPZ-GFP vectors (ThermoScientific Open Biosystems, Huntsville, AL) were transfected into HeLa and RCTE cells using Lipofectamine 2000 and knockdown analyzed after 48 h by immunoblotting for Arf4 protein (Supplemental Figure 4).

**Metalloprotease cleavage sites in ectodomain of polycystin-1.**
Several potential metalloprotease recognition sequences were identified in the ectodomain of polycystin-1 based on reported consensus sequences (Caescu et al., 2009) (Supplemental Figure 5). Metalloprotease cleavage at these sites is expected to result in 230-250 kDa C-terminal cleavage fragments, which are in fact observed when polycystin immunoprecipitations are performed in the presence of magnesium and absence of EDTA (Supplemental Figure 3F). Similar degradation fragments were first reported in fetal kidney extracts (Van Adelsberg et al., 1997).

**Supplemental Figure Legends**

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Supplemental Movie 1. PC1 and Arf4-GFP colocalized to Golgi associated discrete orange punctae in MDCKII cells. Image was analyzed by confocal microscopy using a Zeiss LSM 510. MDCKII cells expressing Arf4-GFP (green) were transfected for 24 h, fixed and immunostained for PC1 with NM002 (red).

Supplemental Movie 2. Rab11, PC1 and ASAP1 colocalize in NK1 cells. Three dimensional confocal Z-stack of cultured normal human kidney (NK1) cells shows co-localization of Rab11 (red), PC1 (green), and ASAP1 (blue) in regions of the Golgi (pink punctae).
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