

Pratt et al., <http://www.jgp.org/cgi/content/full/jgp.201010557/DC1>

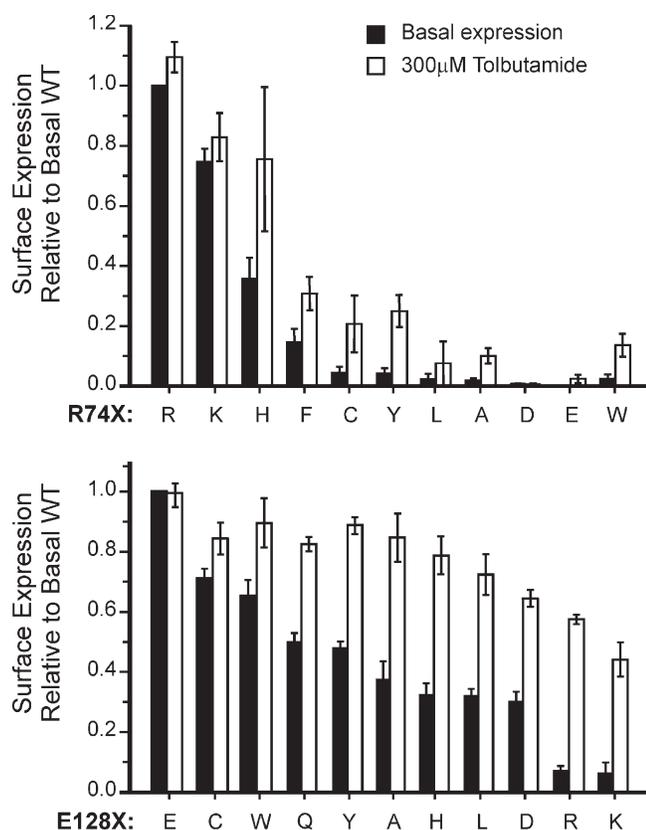
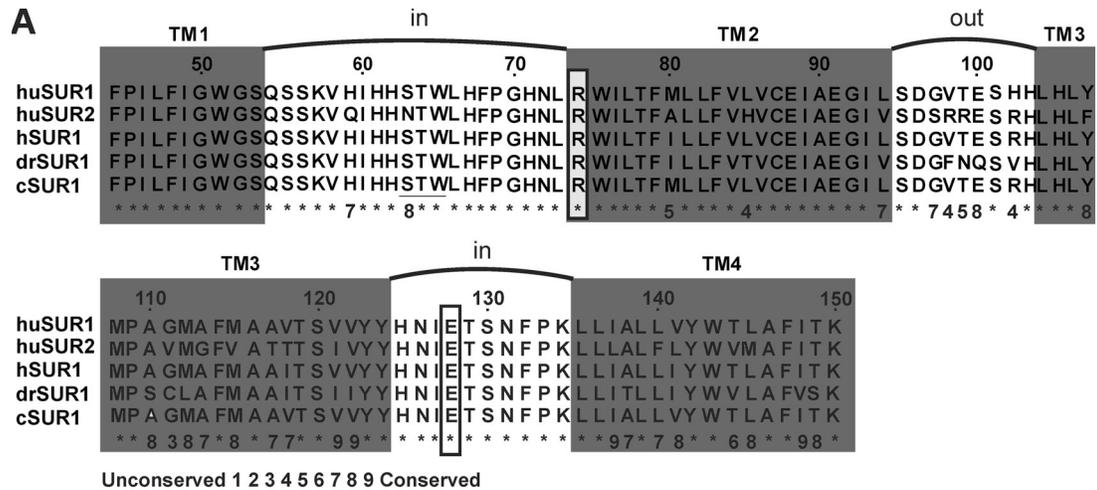


Figure S1. Quantification of the effect of tolbutamide on surface expression of R74X- and E128X-mutant channels. COSm6 cells cotransfected with WT Kir6.2 and WT, R74X (top), or E128X (bottom) fSUR1 were treated overnight with 300 μM tolbutamide before chemiluminescence assays. Black bars, basal surface expression (from Fig. 1); white bars, tolbutamide rescue. Error bars represent SEM; $n \geq 3$ for each condition.



B
TOPCONS Transmembrane Segment Predictions

	TM1	TM2	TM3	TM4	TM5
WT	33-53	74-94	104-124	135-155	167-187
E128X	--	--	--	--	--
R74K	--	--	--	--	--
R74H	31-51	72-92	--	--	--
R74F	31-51	72-92	--	--	--
R74C	31-51	72-92	--	--	--
R74Y	31-51	72-92	--	--	--
R74L	31-51	72-92	--	--	--
R74A	31-51	72-92	--	--	--
R74W	31-51	72-92	--	--	--
R74D	31-51	73-93	103-123	--	--
R74E	31-51	73-93	103-123	--	--

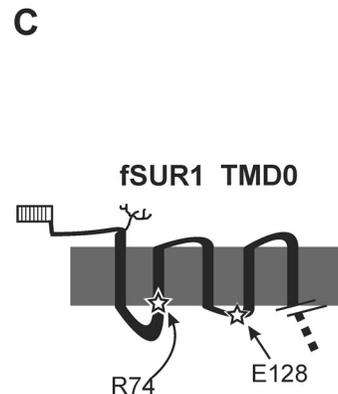


Figure S2. TMD0 is highly conserved between human SUR1 and SUR2, and among species. (A) The alignment was performed using PRALINE; residues 44–150 of TMD0 are shown. R74 and E128 (black boxes) and predicted TM segments (gray boxes, predictions by TOPCONS) are also shown. Species include: hu, human; h, hamster; dr, *Danio rerio*/zebra fish; c, canine. The degree of conservation between sequences is calculated on a scale of 1–9 below the sequences. Cytoplasmic (in) or extracellular (out) loops are also indicated. (B) TOPCONS prediction software was used to determine the possible effects of R74X mutations on TM segment topology of TMD0. Deviations from hypothetical WT TM segment spans are indicated in the table. (C) A schematic of the membrane topology of WT SUR1 TMD0 (amino acids 1–198, end noted by break) is shown, highlighting R74 and E128 (stars), FLAG epitope (flag), and glycosylation site (branch).