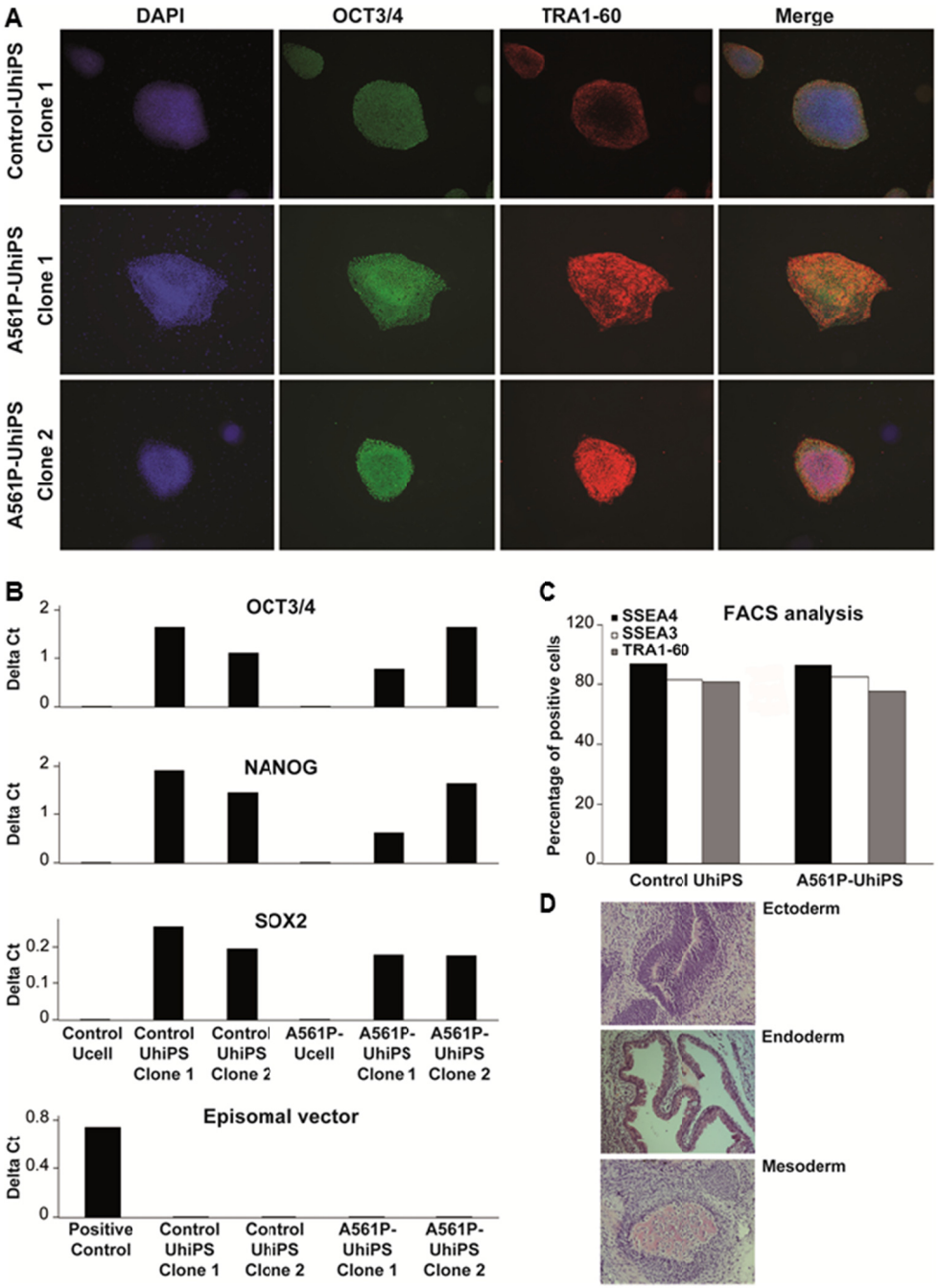
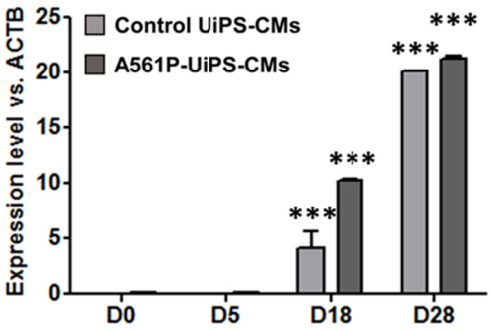
Supplemental data

Jouni et al.: **Towards Personalized Medicine:**

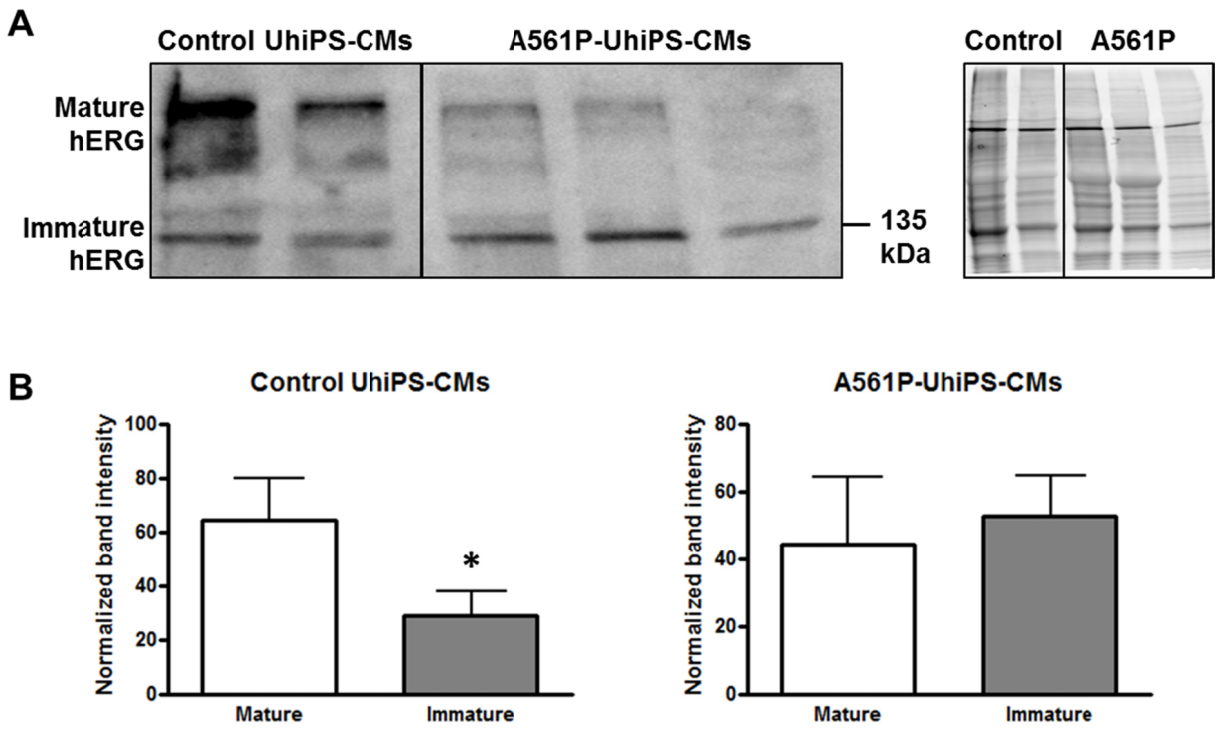
Using Cardiomyocytes Differentiated From Urine-Derived Pluripotent Stem Cells to Recapitulate Electrophysiological Characteristics of Type 2 Long QT Syndrome



Supplemental Figure 1: Characterization of hiPS cells from urine samples (UhiPS). (A) Endogenous pluripotent stem cell marker (OCT3/4 and TRA1-60) visualization by immunofluorescence staining of one Control UhiPS clones and two A561P-UhiPS clones. (B) Expression level of additional endogenous pluripotent stem cell marker (*OCT3/4*, *NANOG* and *SOX2)* genes by qRT-PCR in control and A561P-UhiPS cells as compared to their corresponding urine cells, and expression level of the episomal vector in the same hiPS clones, at passage 10. (C) Percentage of control and A561P-UhiPS cells expressing the pluripotency genes SSEA4, SSEA3 and TRA1-60 measured by flow cytometry. (D) Teratoma formation following injection of undifferentiated UhiPS cells in NOD/SCID mice. The presence of neural tissue (ectoderm, top), intestinal epithelium (endoderm, middle) and immature bone tissue and cartilage (mesoderm, bottom) is shown.



Supplemental Figure 2: Analysis of hERG gene expression. hERG encoding gene expression analysis, during cardiac differentiation (at day 0, 5, 18 and 28) of control UhiPS and of A561P-UhiPS derived cardiomyocytes, evaluated by RT-PCR. *** P<0.001 *versus* D0, no difference observed between control and mutated cells at each time-point.



Supplemental Figure 3: Analysis of hERG protein expression. (A) Left: Western blot analysis of hERG protein expression in control UhiPS-CMs and A561P-UhiPS-CMs (same blot). Two hERG specific bands were revealed (mature and immature subunits). Right: Stain-free expression served as internal control of gel loading. (B) Normalized intensity quantification of mature and immature bands in control UhiPS-CMs (n=3) and A561P-UhiPS-CMs (n=4).

Supplemental Tables

Supplemental Table 1. TaqMan probes and SYBR Green primers. Applied Biosystems references and gene names are listed.

TaqMan Probes	
Gene	**Reference**
ACTB	Hs99999903_m1
SOX2	Hs01053049_s1
NANOG	Hs02387400_g1
POU5F1	Hs04260367_g1
NKX2-5	Hs00231763-m1
GJA1	Hs00748445_s1
GJA5	Hs00270952_s1
RYR2	Hs00892883_m1
KCNJ2	Hs00265315_m1
KCNQ1	Hs00923522-m1
KCNH2	Hs04234270-g1
KCND3	Hs00542597_m1
CACNA1C	Hs00167681_m1
CACNA1G	Hs00367969_m1
SCN5A	Hs00165693_m1

Episomal detection primers	
Orientation	**Sequence**
Forward	GGCTCTCCCATGCATTCAAA
Reverse	GGCCCTCACATTGCCAAA

Supplemental Table 2. Quantitative parameters used for classification of action potentials obtained from patch-clamp experiments on control cells.

APs (n=41)	APD_{90} (ms)	Amplitude (mV)	dV/dt_{max} (V/s)	MDP (mV)	Peak to peak duration (s)
Nodal-like (n=5)	103.1±14.9	73.3±4.2	4.9±0.5	-49.6±4.7	0.4±0.1
Atrial-like (n=19)	185.8±15.8	81.8±1.9	6.9±0.3	-51.2±1.4	0.7±0.07
Ventricular-like (n=17)	444.4±54.3	102.9±1.8	20.3±3.6	-56.2±1.7	0.8±0.1