Figure 1. Urea metabolism is impaired in patient-derived human-induced pluripotent stem cells (hiPSCs) differentiated into hepatocytes (hiPSC-Heps). (A) Scheme showing the urea cycle detoxifying ammonia (NH4+) into nontoxic urea in five consecutive urea cycle enzyme (UCE)—mediated reactions in mitochondria in hepatocytes. (B) Urea secretion in four control hiPSC-Hep lines (Ctrl_1-4) as compared with PHHs. Data represent the average of 4 to 12 independent biological samples. (C,D) Relative mRNA and protein expression of UCEs analyzed by real-time quantitative PCR (C) and western blot (D) in Ctrl_1 hiPSCs and hiPSC-Heps compared with primary human hepatocytes (PHHs). All samples shown were loaded on the same gel. The line indicates that the image was cropped. Data represent the average of two to four independent biological samples. One-way ANOVA; *p
