Correction

BIOCHEMISTRY

Correction for "Heptahelical protein PQLC2 is a lysosomal cationic amino acid exporter underlying the action of cysteamine in cystinosis therapy," by Adrien Jézégou, Elisa Llinares, Christine Anne, Sylvie Kieffer-Jaquinod, Seana O'Regan, Joëlle Aupetit, Allel Chabli, Corinne Sagné, Cécile Debacker, Bernadette Chadefaux-Vekemans, Agnès Journet, Bruno André, and Bruno Gasnier, which appeared in issue 50, December 11, 2012, of *Proc Natl Acad Sci USA* (109: E3434-E3443; first published November 20, 2012; 10.1073/pnas.1211198109).

The authors note that within Figure 6A, a structural formula of the mixed disulfide appeared incorrectly. The corrected Figure 6 and its legend appear below. This error does not affect the conclusions of the article.

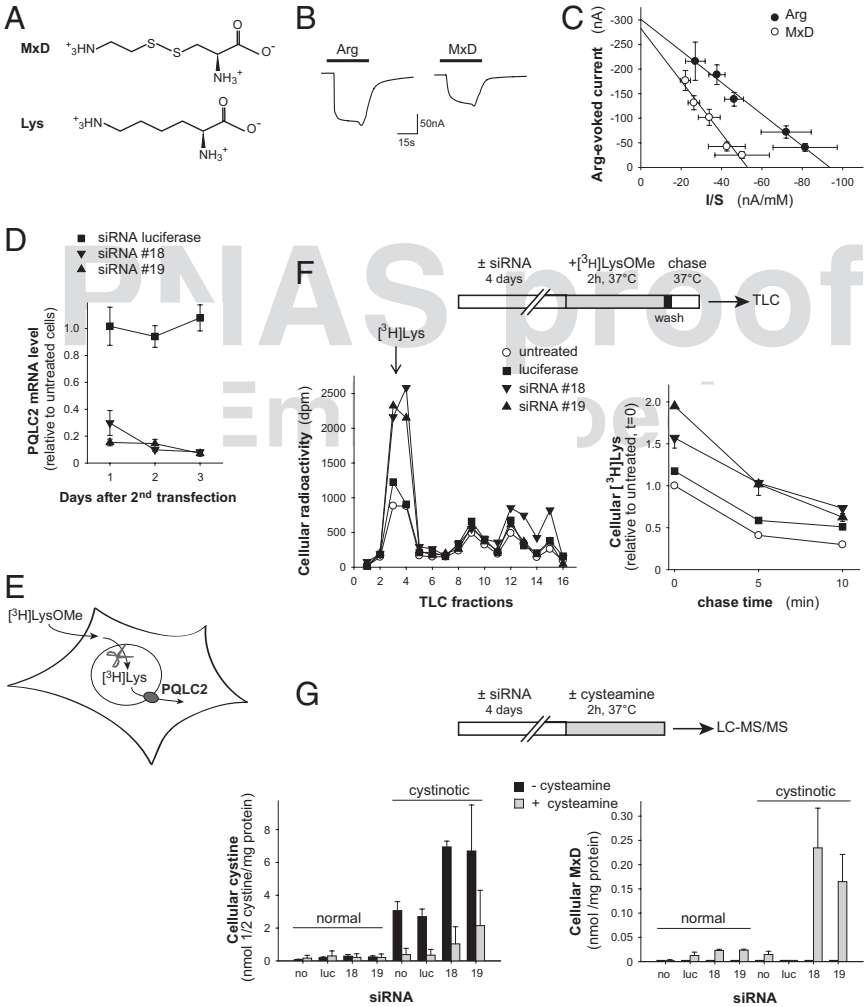

Fig. 6. PQLC2 exports a key chemical intermediate in cysteamine therapy of cystinosis. (*A*) Chemical structure of the MxD resembles that of lysine. (*B*) Current traces evoked by MxD and arginine (10 mM each) on a representative PQLC2-LL/AA-EGFP oocyte at −40 mV and pH 5.0. (*C*) Saturation kinetics of paired MxD and arginine responses (means ± SEMs of five oocytes from two batches). K_m and I_{max} values are reported in the main text. I/S, current/substrate concentration ratio. (*D*) Kinetics of PQLC2 mRNA knockdown in human cystinotic fibroblasts after two rounds of siRNA transfection. Two PQLC2-targeted siRNAs are compared with a luciferase-targeted negative control. Means ± SEMs of four measurements are shown. (*E* and *F*) PQLC2 gene silencing decreases the clearance of lysine from an intracellular compartment. (*E*) Scheme depicts how lysosomes are preferentially loaded with amino acids in whole cells using a methyl ester precursor. After loading human fibroblasts with [^{3}H]Lys*O*Me, the fate of the resulting intracellular [^{3}H]Lys pool was monitored by TLC. (*F*) Plots show representative chromatograms (*Left*) and representative [^{3}H]Lys clearance kinetics (*Right*), respectively. PQLC2 gene silencing increases the intracellular [^{3}H]Lys pool. (*G*) Effect of PQLC2 gene silencing on intracellular cystine and MxD levels. PQLC2 knockdown exacerbates cystine storage (*Left*) and dramatically increases the level of MxD induced by cysteamine (*Right*) in human cystinotic fibroblasts, as illustrated in this representative experiment (means ± SEMs of three measurements). luc, luciferase; no, untreated.

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