Supplemental Material:



Figure S1. Urinary creatinine concentration and osmolality during NH₄CI
Ioading. Time 0 represents baseline (prior to treatment). Data are means ± SEM; n =
8. No significant differences compared to baseline were observed.







Figure S3. Urinary creatinine concentration and osmolality during NH₄CI
Ioading. Time 0 represents baseline (prior to treatment). Data are means ± SEM; n =
8. No significant differences compared to baseline were observed.



Figure S4. Effect of NaHCO₃ **loading on B subunit abundance in urinary exosomes.** Immunoblots of urinary exosomes isolated from 7 different participants, probed with B1 (upper panel), B2 (middle panel) and alix (lower panel) antibodies.



Figure S5. Urinary Na (A), K (B), CI (C), NH₄ (D), creatinine (E) and osmolality (F) during NH₄Cl loading in two dRTA patients. Time 0 represents baseline (prior to treatment).