

SUPPLEMENTS

Supplementary table 1. Blood values in C57Bl/6 mice and *Atp6v1b1*^{-/-} mice under normal diet and HCl load for 4 days. Data are presented as mean ± SEM, n = 5-9 animals/ group, * p<0.05 vs baseline period in same genotype, # p<0.05 vs C57BL6 mice during the same period.

Supplementary table 2. Body weight, food intake, and urinary values of C57BL6 mice and *Atp6v1b1*^{-/-} mice under normal diet and during an HCl load for 4 days. Data are presented as mean ± SEM, n = 5-9 animals/ group, * p<0.05 vs baseline period in same genotype, # p<0.05 vs C57BL6 mice during the same period.

Supplementary table 3. Sequences of the primers and probes used for RT-qPCR

Supplementary figure 1. Unchanged mRNA expression of the B1, B2 and a4 subunits of the H⁺-ATPase in *Rhcg*^{+/+} and *Rhcg*^{-/-} mice.

RT-qPCR was performed on total kidney to determine relative mRNA levels of several H⁺-ATPase relevant for intercalated cell H⁺-ATPase function. None of the mRNA values measured of the B1 (**A**), B2 (**B**) and a4 (**C**) isoforms were different between the 2 genotypes. Values are mean ± SEM (n= 5 mice).

Supplementary data

Supplementary table 1. Blood values in *Atp6v1b1*^{+/+} mice and *Atp6v1b1*^{-/-} mice under normal diet and HCl load for 4 days

	Basal status		2 days HCl		4 days HCl	
	<i>C57BL6</i> (n=9)	<i>Atp6v1b1</i> ^{-/-} (n=7)	<i>C57BL6</i> (n=7)	<i>Atp6v1b1</i> ^{-/-} (n=6)	<i>C57BL6</i> (n=7)	<i>Atp6v1b1</i> ^{-/-} (n=5)
pH	7.35 ± 0.01	7.40 ± 0.01	7.11 ± 0.02*	7.15 ± 0.02*	7.24 ± 0.02	6.87 ± 0.12 ^{*#}
pCO ₂ (mmHg)	40.0 ± 0.6	40.6 ± 1.2	37.3 ± 1.2	39.3 ± 1.5	39.4 ± 1.5	45.4 ± 2.5 [#]
HCO ₃ (mM)	21.6 ± 0.7	24.8 ± 1.2	11.5 ± 0.8*	12.8 ± 0.6*	16.4 ± 0.9*	8.7 ± 1.8 ^{*#}
pO ₂	52.1 ± 2.2	48.8 ± 2.0	60.4 ± 2.4	62.6 ± 3.4	58.4 ± 2.5	83.8 ± 8.7 ^{*#}
Na ⁺ (mM)	144.6 ± 0.5	145.4 ± 0.3	148.0 ± 1.1*	147.8 ± 1.5	151.4 ± 0.5*	150.8 ± 2.4*
Cl ⁻ (mM)	109.7 ± 0.6	108.3 ± 0.7	123.4 ± 1.6*	121.0 ± 1.5*	120.0 ± 1.6*	133.6 ± 2.8 ^{*#}
Ca ²⁺ (mM)	1.26 ± 0.01	1.23 ± 0.01	1.34 ± 0.01*	1.33 ± 0.02*	1.36 ± 0.02*	1.60 ± 0.1 ^{*#}
Hb (g/dl)	15.1 ± 0.2	15.7 ± 0.2	15.6 ± 0.3	16.7 ± 0.2 ^{*#}	15.3 ± 0.2	18.8 ± 1.2 ^{*#}

* p<0.05 vs baseline period in same genotype, # p<0.05 vs *Atp6v1b1*^{+/+} mice during the same period.

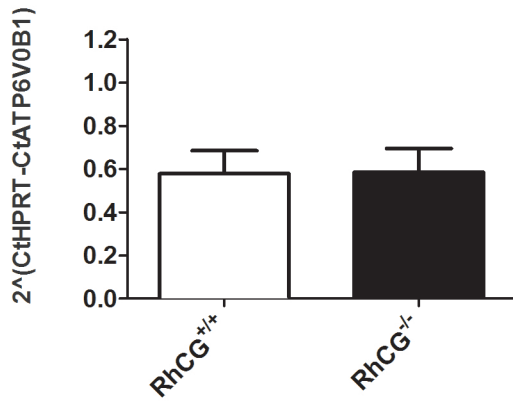
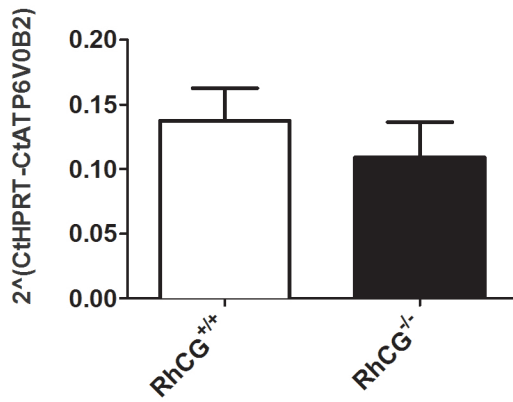
Supplementary table 2. Weight, food intake and urinary values in *Atp6v1b1*^{+/+} mice and *Atp6v1b1*^{-/-} mice under normal diet and during an HCl load for 4 days

	Basal status		2 days HCl		4 days HCl	
	<i>C57BL6</i> (n=7)	<i>Atp6v1b1</i> ^{-/-} (n=7)	<i>C57BL6</i> (n=7)	<i>Atp6v1b1</i> ^{-/-} (n=7)	<i>C57BL6</i> (n=7)	<i>Atp6v1b1</i> ^{-/-} (n=5)
Weight (g)	22.7 ± 0.28	28.1 ± 0.5 [#]	20.4 ± 0.2*	24.6 ± 0.3 ^{*#}	20.5 ± 0.3*	23.0 ± 0.3 ^{*#}
Weight lose in % of body weight under basal status	0.1 ± 0.6	0.4 ± 0.9	10.3 ± 0.5*	13.0 ± 1.6 ^{*#}	11.9 ± 3.4*	18.8 ± 2.7 ^{*#}
Food intake (g/24hrs/body weight)	0.36 ± 0.01	0.36 ± 0.02	0.28 ± 0.01*	0.22 ± 0.03*	0.34 ± 0.04	0.26 ± 0.01 ^{*#}
Water intake (ml/24hrs/body weight)	0.17 ± 0.05	0.05 ± 0.01 [#]	0.20 ± 0.05	0.20 ± 0.04*	0.18 ± 0.03	0.23 ± 0.06*
Urine values						
volume (ml/24 h)	2.6 ± 0.4	3.0 ± 0.1	2.1 ± 0.2	4.7 ± 0.8 [#]	3.1 ± 0.2	3.9 ± 1.2
creatinine excretion (μmol/24 h)	3.7 ± 0.6	3.4 ± 0.2	3.4 ± 0.4	1.8 ± 0.3 ^{*#}	1.9 ± 0.2*	1.9 ± 0.7*
Urinary pH	6.2 ± 0.0	6.5 ± 0.1 [#]	5.4 ± 0.1*	6.0 ± 0.0 ^{*#}	5.7 ± 0.1*	6.0 ± 0.1 ^{*#}
UNH ₄ /UCr (mEq/mmol)	3.4 ± 0.3	11.3 ± 1.2 [#]	51.0 ± 2.0*	56.7 ± 13.8*	88.9 ± 6.2*	76.6 ± 7.2 ^{*#}
UPi/UCr (mEq/mmol)	13.1 ± 1.1	11.6 ± 0.7	14.3 ± 2.4	18.8 ± 3.2	16.4 ± 2.6	22.7 ± 5.2

* p<0.05 vs baseline period in same genotype, # p<0.05 vs *Atp6v1b1*^{+/+} mice during the same period.

Supplementary table 3. Primers and probes sequences of the gene expressions evaluated by RT-qPCR

Gene	Accession number	Forward primer	Reverse primer	Probe
Mouse <i>Atp6v1b1</i>	NM_134157	5'- CCCAGTATGCTGAGATTGTCAACTTTAC CCTCC -3'	5'- CCTGAACAATGGCCTTGGTC-3'	5'- CCCAGTATGCTGAGATTGTCAACTTTAC CCTCC-3'
Mouse <i>Atp6v1b2</i>	NM_007509	5'- GCTGGTGATCCTGACGGACA TGAG-3'	5'- CGTGCCGGTCGAGTGGAAGGTAGAAAC GG-3'	5'- CTGCGAGAGGTTTCAGCTG-3'
Mouse <i>Atp6V0a4</i>	NM_080467	5'- GACGGTGTCTCATCTATGTTTTGA-3'	5'- TTTGCCCTGCATGGTCTTGTC-3'	5'- TCTCTCCATTATCAAGAGCTCATGCCA - 3'
Mouse <i>Hprt</i>	NM_013556	5'- TTATCAGACTGAAGAGCTACTGTAAGAT C-3'	5'- TTACCAGTGTCAATTATATCTTCAACAAT C-3'	5'- TGAGAGATCATCTCCACCAATAACTTTT ATGTCCC-3'
Human <i>RhCG</i>	NM_016321	5'- CTACCGACGCAACCTAGAGC-3'	5'-GCTGTCCCCATGGTAGGATA-3	5'-CTTTGCCATGATTGGCACCC-3'
Human <i>ATP6V1B1</i>	NM_001692	5'-GAGATGATTCAGACGGGCATTT-3'	5'-CTGGTGAAGAAGTCCAAGGC-3'	5'-AATGAGATTGCCGCTCAGAT-3'
Human <i>ATP6V1B2</i>	NM_001693	5''-TGAAGGGACTTCAGGTATAGATGC-3'	5'-GGAAGTCTTCGGCCAGTACA-3'	5'-TGGTCGGGTATTCAATGGAT-3'
Human <i>ATP6V0A1</i>	NM_001130020	5'-TTCCTGGAACCTGACCGAATTA-3'	5'-AGGAGTGCCTCTCCCATCT-3'	5'-GAGGCTGAATTGCATCATCA-3'
Human <i>HPRT</i>	NM_000185	5'-AAGGGTGTTACGTTTAT-3'	5'-TATTTCTGTGCGTGAT-3'	5'- ACCTCCTTGGGATCTATTTACCG -3'

A**B****C**