

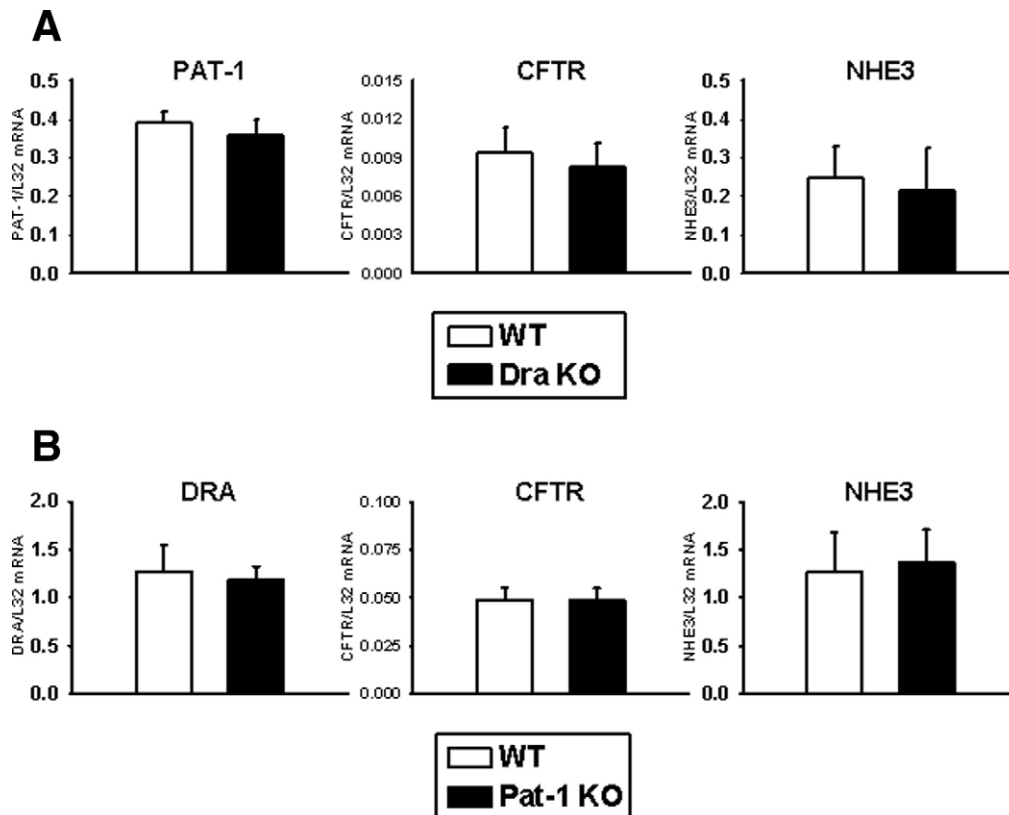
**Supplementary Table 1.** Blood Hematocrit, pCO<sub>2</sub>, HCO<sub>3</sub><sup>-</sup>, and pH of WT and Dra KO Mice

	Hct	pH	HCO <sub>3</sub> <sup>-</sup> (mmol/L)	pCO <sub>2</sub> (mm Hg)	No.
WT	40.4 ± 1.5	7.17 ± 0.03	17.0 ± 1.8	46.1 ± 3.2	6
Dra KO	44.9 ± 2.1	7.26 ± 0.06	29.0 ± 2.3 <sup>a</sup>	66.3 ± 6.3 <sup>a</sup>	6

NOTE. Blood analysis. Dra WT and KO mice were anesthetized with an intraperitoneal injection of avertin (Sigma, St. Louis, MO). Blood was drawn from a left-side cardiac puncture into heparin-treated syringes and immediately analyzed for gases and pH using an IRMA Blood Analysis System (Diametrics Medical, St. Paul, MN).

Hct, blood hematocrit.

<sup>a</sup>Significantly different from WT, *P* < .05.



**Supplementary Figure 1.** (A) mRNA expression of Pat-1, Cfr, and Nhe3 is unchanged in the Dra KO jejunum. (B) mRNA expression of Dra, Cfr, and Nhe3 is unchanged in the Pat-1 KO jejunum. mRNA expression was measured by quantitative real-time PCR using jejunal preparations from WT (open bars) and KO (solid bars) littermate mice (*n* = 6 mice). The mRNA concentration for a specific gene was normalized to the concentration of ribosomal protein L32 mRNA to control for differences in cell number and the quality of the RNA preparation. Method: Full-thickness intestinal samples were collected for measurement of mRNA expression using real-time quantitative PCR analysis, as previously described.<sup>1</sup> Total RNA was extracted using Tri-Reagent (Molecular Research Center, Cincinnati, OH), and 1.6  $\mu$ g were reverse transcribed using SuperScriptIII First-Strand Synthesis System for RT-PCR (Invitrogen, Carlsbad, CA). The relative mRNA concentration in the samples (0.08  $\mu$ g complementary DNA [cDNA]) was estimated using a LightCycler FastStart DNA Master SYBR Green I (Roche, Indianapolis, IN), according to the manufacturer's directions. Real-time PCR was performed with the following primers: Cfr-forward (F) 5'-CAAACAAGTGAATCTGAAGGC-3' and Cfr-reverse (R) 5'-GCTCAGATCGCATCAAGC-3' for Cfr; Dra-F 5'-GGCGGCAAGTGTAGCATTCAACT-3' and Dra-R 5'-GTGGGCTAAAGCCAAACAGCATCAA-3' for Dra; Nhe3\_5'a\_mr (forward) 5'-GGCCTTCATTGCTCCCAAG-3' and Nhe3\_3'a\_mrh (reverse) 5'-ATGCTTGTACTCTGCCGAGG-3' for Nhe3; Pat-1 F 5'-CCAGCAATCAAGAAGATGCC-3' and Pat-1 R 5'-ACACTTCCACTTCAATCTCCC-3' for Pat-1, and L32-F 5'-CATCTGTTTACGGCATCATG-3' and L32-R 5'-CGCTCCCATACCGATGTTGG-3' for L32. A melting curve analysis was performed after each PCR to ensure the specificity of quantification. The relative concentration of cDNA in each sample was determined from the threshold of linear amplification. The threshold was converted to relative concentration based on a formula that was generated by measuring the exponential phase of serially diluted standards, according to the manufacturer's instructions. Reference: 1. Clarke LL, Gawenis LR, Bradford EM, et al. Abnormal Paneth cell granule dissolution and compromised resistance to bacterial colonization in the intestine of CF mice. *Am J Physiol* 2004;286:G1050-G1058.

**Supplementary Table 2.** Unidirectional and Net Flux of Na<sup>+</sup> and Cl<sup>-</sup>, I<sub>sc</sub>, and G<sub>t</sub> of Jejunal Preparations From WT and Dra KO Mice During cAMP Stimulation

	J <sup>Na+</sup>			J <sup>Cl-</sup>			I <sub>sc</sub>	G <sub>t</sub>	No.
	M-S	S-M	Net	M-S	S-M	Net			
WT	19.5 ± 1.5	19.1 ± 1.3	0.4 ± 1.2	18.3 ± 2.3	32.7 ± 2.4	-14.4 ± 1.5	-9.1 ± 0.3	34.6 ± 0.3	9
Dra KO	19.7 ± 1.6	18.8 ± 1.3	0.9 ± 1.5	13.1 ± 1.1 <sup>a</sup>	27.3 ± 1.4	-14.2 ± 1.4	-8.6 ± 0.3	34.9 ± 1.0	8

NOTE. J and I<sub>sc</sub> in μEq/cm<sup>2</sup> · h; G<sub>t</sub> in mS/cm<sup>2</sup>.

J, ion flux; I<sub>sc</sub>, short-circuit current; G<sub>t</sub>, transepithelial conductance; S-M, serosal-to-mucosal; M-S, mucosal-to-serosal.

<sup>a</sup>Significantly different from WT, *P* < .05.

**Supplementary Table 3.** Unidirectional Mucosal-to-Serosal Flux of Cl<sup>-</sup>, I<sub>sc</sub>, and G<sub>t</sub> of Jejunal Preparations From WT and Pat-1 KO Mice in the Nominal Absence of Luminal HCO<sub>3</sub><sup>-</sup>

	J <sup>Cl-</sup>			No.
	M-S	I <sub>sc</sub>	G <sub>t</sub>	
WT	20.4 ± 0.3	0.1 ± 0.2	38.6 ± 2.2	8
Pat-1 KO	18.3 ± 0.7 <sup>a</sup>	0.1 ± 0.2	37.7 ± 0.9	7

NOTE. J and I<sub>sc</sub> in μEq/cm<sup>2</sup> · h; G<sub>t</sub> in mS/cm<sup>2</sup>.

J<sup>Cl<sup>-</sup><sub>m-s</sub></sup>, mucosal-to-serosal flux of <sup>36</sup>Cl; J, ion flux; I<sub>sc</sub>, short-circuit current; G<sub>t</sub>, transepithelial conductance.

<sup>a</sup>Significantly different from WT, *P* < .05.

**Supplementary Table 4.** Unidirectional and Net Flux of  $^{22}\text{Na}^+$  and  $^{36}\text{Cl}^-$ ,  $I_{\text{sc}}$ , and  $G_t$  of Jejunal Preparations From WT and Pat-1 KO Mice During cAMP Stimulation

	$J_{\text{Na}^+}$			$J_{\text{Cl}^-}$			$I_{\text{sc}}$	$G_t$	No.
	M-S	S-M	Net	M-S	S-M	Net			
WT	19.1 ± 0.9	21.1 ± 1.5	-2.0 ± 1.7	13.4 ± 1.2	28.2 ± 1.7	-14.8 ± 1.5	-9.4 ± 0.4	37.8 ± 1.5	7
Pat-1 KO	20.2 ± 0.8	22.7 ± 1.0	-2.5 ± 1.0	13.2 ± 1.2	24.4 ± 1.2 <sup>a</sup>	-11.2 ± 1.5	-8.9 ± 0.3	36.0 ± 0.7	10

NOTE. J and  $I_{\text{sc}}$  in  $\mu\text{Eq}/\text{cm}^2 \cdot \text{h}$ ;  $G_t$  in  $\text{mS}/\text{cm}^2$ .

J, ion flux;  $I_{\text{sc}}$ , short-circuit current;  $G_t$ , transepithelial conductance; S-M, serosal-to-mucosal; M-S, mucosal-to-serosal.

<sup>a</sup>Significantly different from WT,  $P < .05$ .