

Supplemental Information

**Promoting readthrough of nonsense mutations
in CF mouse model: Biodistribution and efficacy
of NV848 in rescuing CFTR protein expression**

Ignazio Fiduccia, Federica Corrao, Maria Grazia Zizzo, Riccardo Perriera, Francesco Genovese, Emanuele Vitale, Davide Ricci, Raffaella Melfi, Marco Tutone, Andrea Pace, Laura Lentini, and Ivana Pibiri

Table S1. Mice body weight at T^0 and T^{15} days.

		CFTR ^{WT}		CFTR ^{G542X/G542X}	
		g	± SEM	g	± SEM
Weight treatment	t^0	11.8	0.8	8.2	1.0
Weight treatment	t^{15}	17.4	0.9	14.0	0.8

Table S2. Sequence of primers used for DNA amplification to detect CFTR WT (primers P3 and P2) and CFTR G542X (primers P1 and P2) alleles.

Name	Nucleotide Sequence	Type of primer	Allele identified
P 1	5' – ACA AGA CAA CAC AGT TCT CT – 3'	Forward	CFTR G542X
P 2	5' – TCC ATG CAC CAT AAC AAC AAG T – 3'	Reverse	CFTR (Common)
P 3	5' – ACA AGA CAA CAC AGT TCT TG – 3'	Forward	CFTR WT

Table S3. Primer sequences for real-time RT PCR to calculate relative gene expression for the CFTR gene.

Gene		Nucleotide Sequence	
CFTR mouse	FORWARD	5' – CTACATGGAACACATACCTTCG - 3'	Divangahi M., et al., 2009
CFTR mouse	REVERSE	5' – GGTGATAATCACTGCATAGC – 3'	Divangahi M., et al., 2009
ACTIN mouse	FORWARD	5' – ACCGTCAAAAGATGACCCAGA- 3'	Designed with Primer Express software (Applied Biosystems)
ACTIN mouse	REVERSE	5' – GAGGCATACAGGGACAGCACA – 3'	Designed with Primer Express software (Applied Biosystems)