

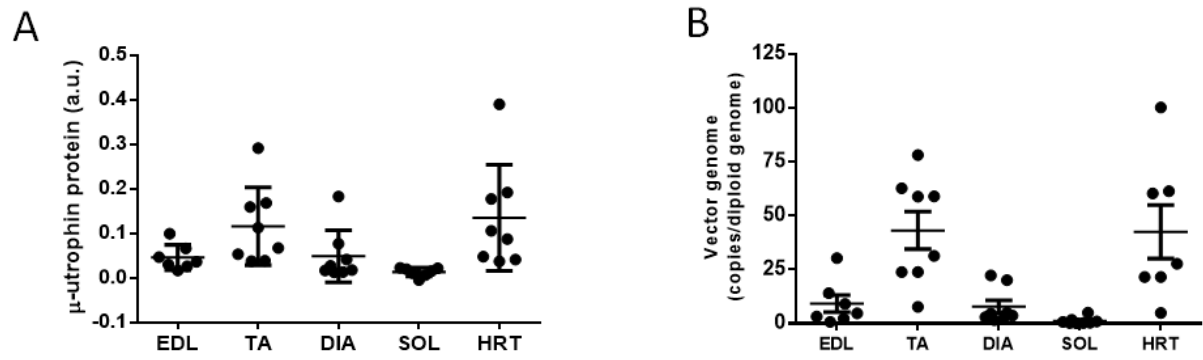
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Supplemental Information

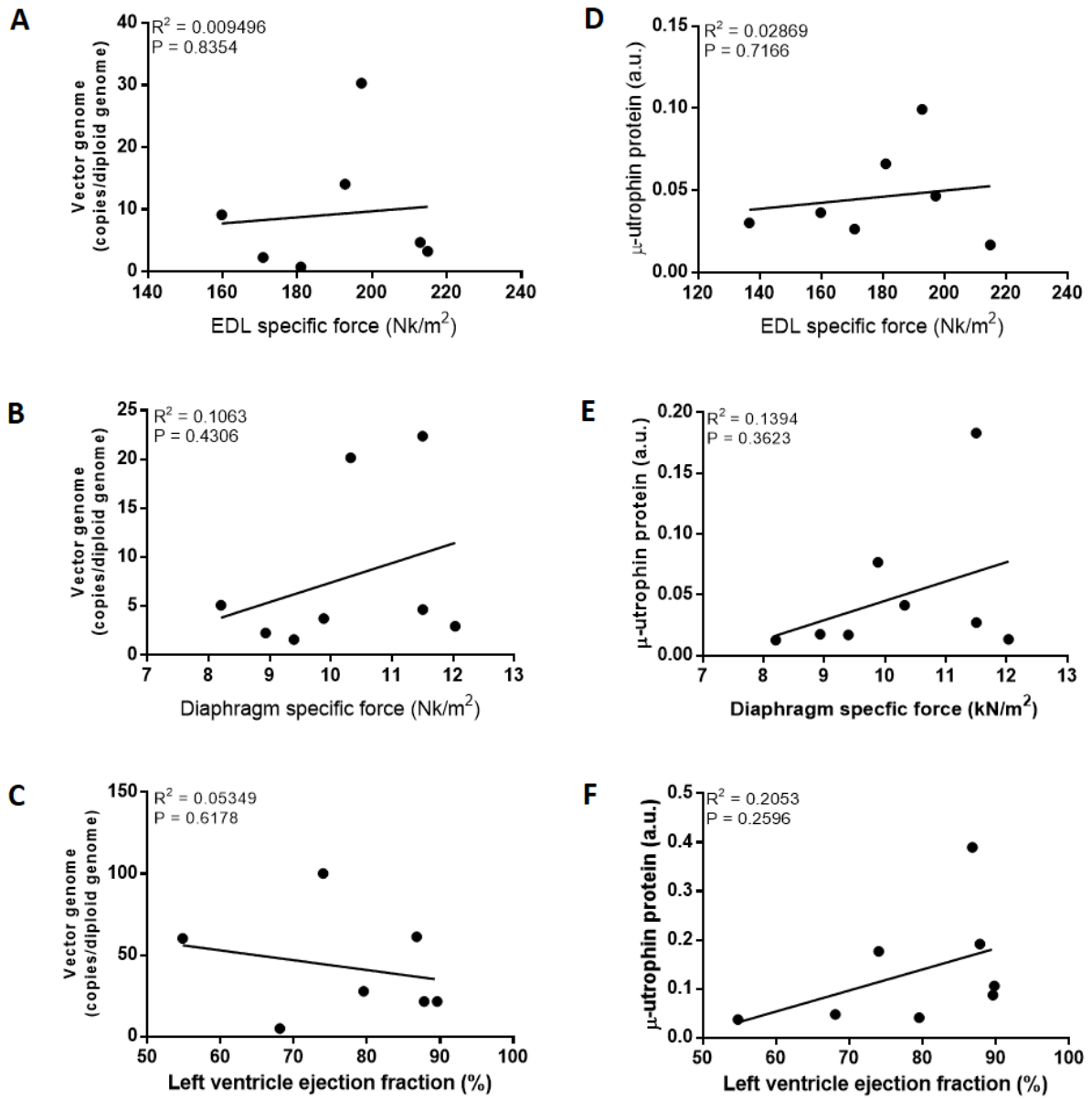
**Micro-utrophin Improves Cardiac
and Skeletal Muscle Function of Severely
Affected D2/*mdx* Mice**

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Supplemental Figures



Supp. Figure 1.



Supp. Figure 2.

Supplemental Figure Legends

Supp. Figure 1. Quantification of μ -utrophin protein expression and vector genome copy number in cardiac and skeletal muscles of D2/*mdx* mice administered AAV- μ -Utro. Quantification of (A) μ -utrophin protein expression and (B) qPCR detection of the CMV promoter in cardiac and skeletal muscles from D2/*mdx* mice administered AAV- μ -Utro. Data represented as mean \pm SEM, $n = 8$ /group. EDL, extensor digitorum longus; TA, tibialis anterior; DIA, diaphragm; SOL, soleus; HRT, heart.

Supp. Figure 2. Assessment of the linear relationship between μ -utrophin protein expression or vector genome expression and functional parameters of cardiac and skeletal muscles from D2/*mdx* mice administered AAV- μ -Utro. Assessment of the linear relationship between μ -utrophin protein expression and (A) specific force extensor digitorum longus (EDL) muscles, (B) specific force of diaphragm muscle strips and (C) left ventricle ejection fraction of D2/*mdx* mice administered AAV- μ -Utro. Assessment of the linear relationship between vector genome expression and (D) specific force of EDL muscles, (E) specific force of diaphragm muscle strips and (F) left ventricle ejection fraction of D2/*mdx* mice administered AAV- μ -Utro. No statistically significant correlations was observed between parameters. $n = 8$ /group.

Supplementary Table 1. Body mass, kyphosis score and muscle mass of 14-, 21- and 28-week BL10/WT, BL10/*mdx*, D2/WT and D2/*mdx* mice

			14 weeks	21 weeks	28 weeks
Body mass (g)	BL10	WT	28 ± 1	30 ± 1	33 ± 2
		<i>mdx</i>	35 ± 1 *	34 ± 2	39 ± 1 *
	D2	WT	28 ± 1	33 ± 1	31 ± 2
		<i>mdx</i>	24 ± 1	27 ± 1 *	30 ± 2
Kyphosis score	BL10	WT	1 ± 0	1.1 ± 0.1	1 ± 0
		<i>mdx</i>	1 ± 0	1.3 ± 0.2	2.1 ± 0.2 *
	D2	WT	1.7 ± 0.1	2.8 ± 0.2	3.4 ± 0.1
		<i>mdx</i>	2.2 ± 0.2 *	2.2 ± 0.1	3.2 ± 0.2

HEART

Heart mass (mg)	BL10	WT	189.6 ± 23.3	138.1 ± 2.8	146.8 ± 7.2
		<i>mdx</i>	227.1 ± 7.2	152.3 ± 3.2 *	155.1 ± 3.0
	D2	WT	155.3 ± 18.6	155.9 ± 3.4	153.3 ± 4.9
		<i>mdx</i>	182.1 ± 13.5	153.0 ± 4.7	162.7 ± 8.1
Heart/body mass (mg/g)	BL10	WT	6.7 ± 0.7	4.7 ± 0.2	4.4 ± 0.2
		<i>mdx</i>	6.5 ± 0.3	4.3 ± 0.2	4.0 ± 0.1
	D2	WT	5.8 ± 0.9	4.6 ± 0.1	5.1 ± 0.3
		<i>mdx</i>	7.6 ± 0.2	5.8 ± 0.3 *	5.5 ± 0.2

SKELETAL MUSCLE

TA/body mass (mg/g)	BL10	WT	1.57 ± 0.38	1.45 ± 0.05	1.30 ± 0.60
		<i>mdx</i>	2.20 ± 0.08 *	2.29 ± 0.10 *	2.17 ± 0.06 *
	D2	WT	1.39 ± 0.90	1.13 ± 0.05	1.19 ± 0.07
		<i>mdx</i>	1.52 ± 0.14	1.31 ± 0.09	1.19 ± 0.08
EDL/body mass (mg/g)	BL10	WT	0.27 ± 0.026	0.32 ± .01	0.30 ± 0.01
		<i>mdx</i>	0.45 ± 0.02 *	0.47 ± 0.02 *	0.45 ± 0.01 *
	D2	WT	0.33 ± 0.06	0.26 ± 0.01	0.29 ± 0.01
		<i>mdx</i>	0.23 ± 0.26	0.31 ± 0.06	0.26 ± 0.03
Quad/body mass (mg/g)	BL10	WT	5.18 ± 0.73	6.19 ± 0.15	5.43 ± 0.51
		<i>mdx</i>	9.05 ± 0.31 *	9.77 ± 0.27 *	8.97 ± 0.21 *
	D2	WT	5.12 ± 0.34	4.99 ± 0.32	5.58 ± 0.24
		<i>mdx</i>	4.29 ± 0.26	3.96 ± 0.14 *	3.54 ± 0.22 *

TA, tibialis anterior; EDL, extensor digitorum longus; Quad, quadriceps. Data represented as mean \pm SEM, * $P < 0.05$ vs age-matched WT control, $n = 6$ /group.

Supplementary Table 2. Body mass, kyphosis score and muscle mass of 21-week-old D2/*mdx* mice administered AAV- μ -Utro compared to vehicle control mice.

	Untreated	AAV-μ-Utro
Body mass (g)	28 \pm 1	26 \pm 1
Kyphosis score	2.5 \pm 0.2	2.3 \pm 0.2
HEART		
Heart mass (mg)	165.9 \pm 7.7	156.0 \pm 8.3
Heart mass/body mass (mg/g)	6.0 \pm 0.3	6.0 \pm 0.2
SKELETAL MUSCLE		
TA/body mass (mg/g)	2.0 \pm 0.1	1.8 \pm 0.8
EDL/body mass (mg/g)	0.3 \pm 0.0	0.3 \pm 0.0
Quad/body mass (mg/g)	4.6 \pm 0.3	6.2 \pm 0.3
Sol/body mass (mg/g)	0.6 \pm 0.1	0.3 \pm 0.0

Data represented as mean \pm SEM, $n = 8$ /group. No statistical differences were observed between groups. Tibialis anterior, TA; extensor digitorum longus, EDL; quadriceps, Quad; soleus, Sol.