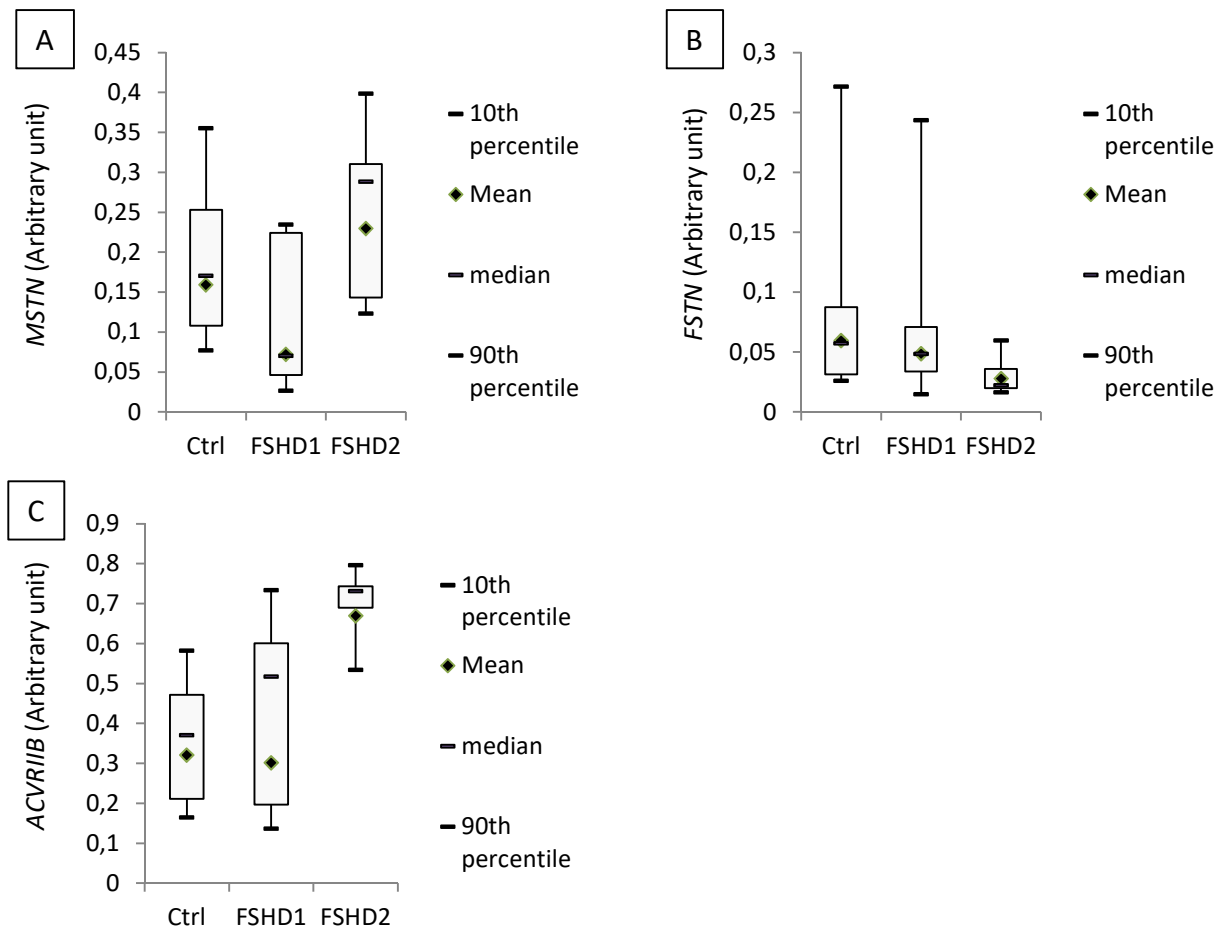


**Supplementary table 1** : Characteristics of LGMD's biopsies

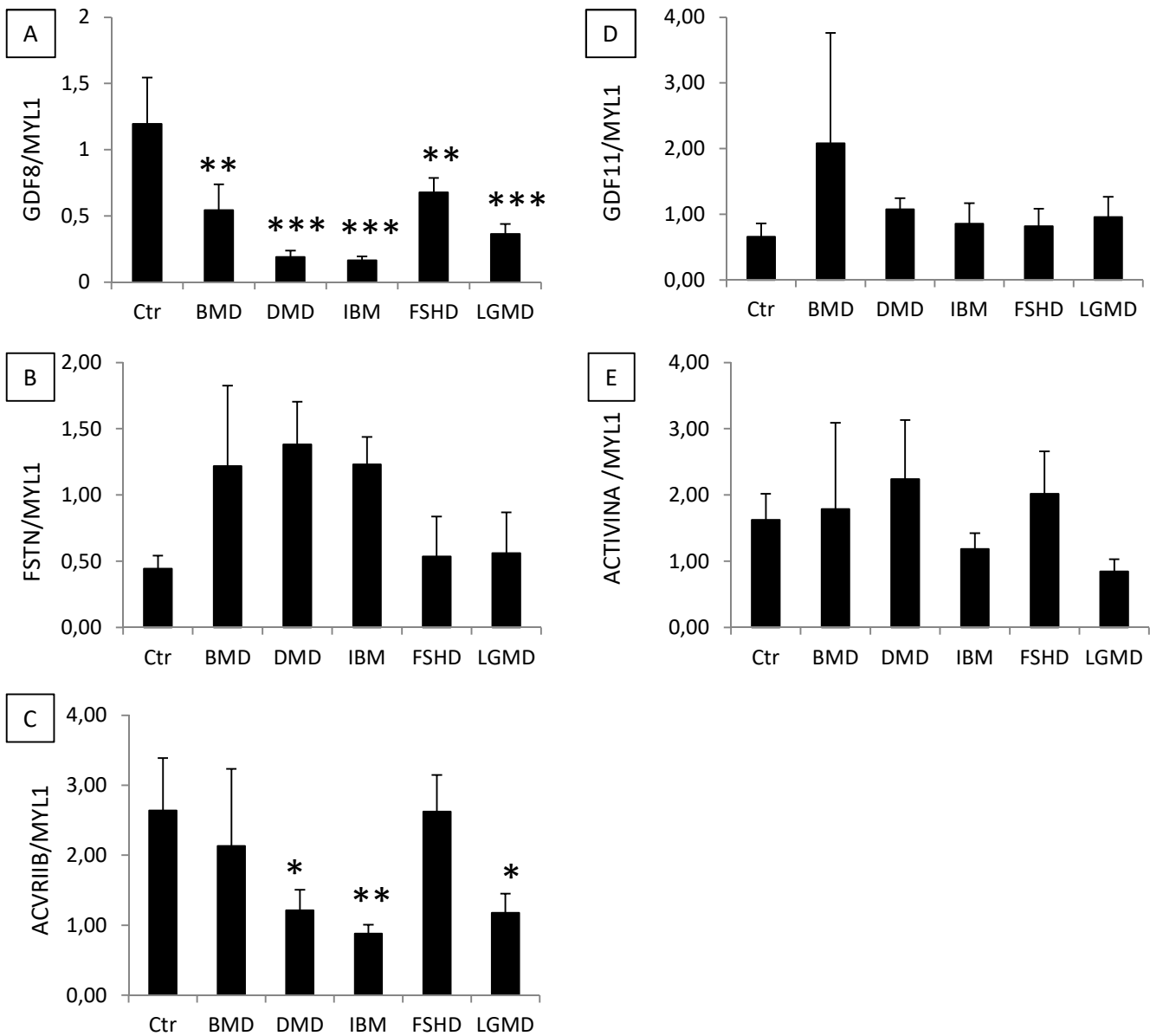
	<b>LGMD2A (n=1)</b>	<b>LGMD2B (n=4)</b>	<b>LGMD2C (n=2)</b>	<b>LGMD2D (n=2)</b>	<b>LGMD2L (n=2)</b>
<b>Age (years)</b>					
Mean	44,0	34,9	11,2	16,0	39,6
Range	-	24,2 - 50,1	8,6 - 13,8	12,1 - 20,0	21,3 - 57,9
<b>Gender</b>					
Female	-	3	1	1	-
Male	1	1	1	1	2
<b>Age of onset (years)</b>					
Mean	40,0	30,0	4,5	8,0	55,0
Range	-	25 - 35	4 - 5	6 - 10	-

**Supplementary table 2: Oligonucleotides used in this study**

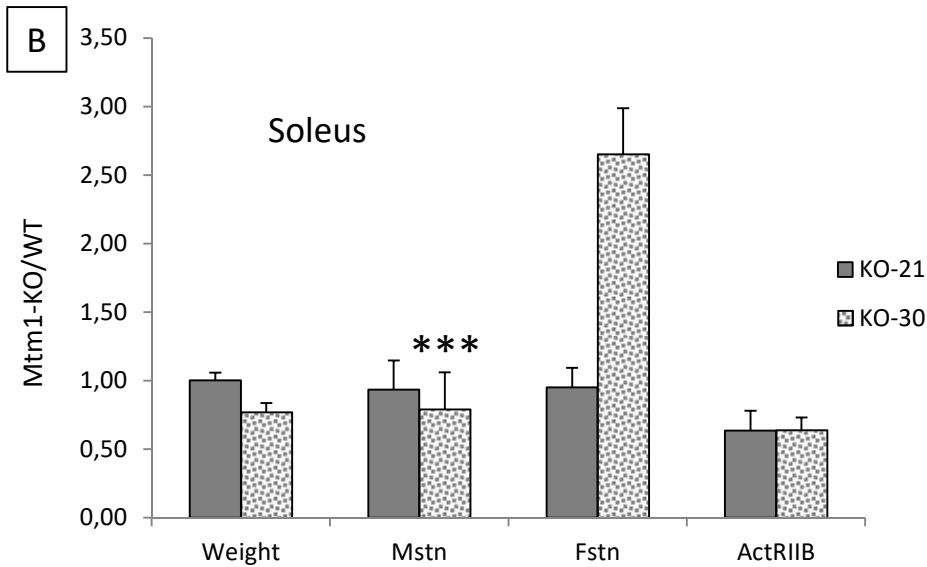
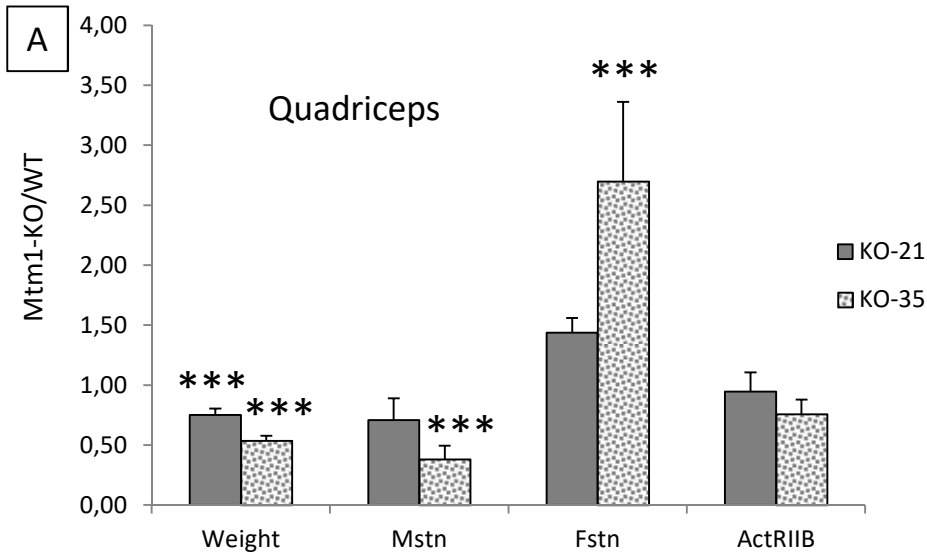
Gene symbol	Accession n°	Name	5'	3'	Amplicon length
<i>Actb</i>	NM_007393.5	b-actin_F	CTGGCTCCTAGCACCATGAA		123
		b-actin_R	CTGCTTGCTGATCCACATCT		
<i>Activin A</i>	NM_008380.2	Activin A -F	CACACTTCTGCACGCTCCAC		92
		Activin A -R	TTTGCCGAGTCAGGCACAG		
<i>Tubb5</i>	NM_011655.5	b-tubulin_F	CCTTCATTGGAAACAGCACA		222
		b-tubulin_R	CCTCCTCTCCGAAATCCTCT		
<i>Gapdh</i>	NM_001289726.1	Gapdh_F	TTGTGATGGGTGTGAACCAC		283
		Gapdh_R	TTCAGCTCTGGGATGACCTT		
<i>Gdf11</i>	NM_010272	Gdf11 - F	ATCAGCCGGGAGGTAGTGAA		159
		Gdf11 - R	CTGGGCCATGCTTATGACCGT		
<i>Hprt</i>	NM_013556.2	Hprt1_F	GCAAACCTTTGCTTTCCCTGG		85
		Hprt1_R	ACTTCGAGAGGTCCTTTTCACC		
<i>Rplp0</i>	NM_007475.5	P0_F	CTCCAAGCAGATGCAGCAGA		87
		P0_R	ATAGCCTTGCGCATCATGG		
<i>Acvr2b</i>	NM_007397.3	ActrIIB_F	GCTCAGCTCATGAACGACT		68
		ActrIIB_R	CTCTGCCACGACTGCTTGT		
<i>Fst</i>	NM_001301373.1	Fstn_F	CTCTCAAGTGGATGATTTTC		345
		Fstn_R	ACAGTAGGCATTATTGGTCTG		
<i>Mstn</i>	NM_010834.3	Mstn_F	GCACTGGTATTTGGCAGAGTA		345
		Mstn_R	CACACTCTCCTGAGCAGTAAT		
<i>ACTIVIN A</i>	ENST00000242208.4	F-ACTIVIN A	TTATGGAGCAGACCTCGGAG		75
		R-ACTIVIN A	AAATCTCGAAGTGCAGCGTC		
<i>B2M</i>	NM_004048	F_B2M	CTCTCTTTCTGGCCTGGAGG		67
		R_B2M	TGCTGGATGACGTGAGTAAACC		
<i>GAPDH</i>	ENST00000229239	F-GAPDH2	AAGGTGAAGGTCCGAGTCAACGG		199
		R-GAPDH2	TGACAAGCTTCCCGTTCTCAGCC		
<i>GUS</i>	ENST00000304895	F-GUS	CTCATTTGGAATTTTGCCGATT		81
		R-GUS	CCGAGTGAAGATCCCCTTTTA		
<i>RPLP0</i>	ENSG00000089157	F-P0	TCCAGGCTTTAGGTATCACCAC		94
		R-P0	GCTCCCACTTTGTCTCCAGTC		
<i>PPIA</i>	ENST00000355968	F-PPIA	CCTAAAGCATAACGGGTCCTG		133
		R-PPIA	TTTCACTTTGCCAAACACCA		
<i>ACVR2B</i>	ENST00000352511	F52-AcvRIIb	CTCCTCTGGGGATCGCTGT		84
		R135-AcvRIIb	CTCCAGTTGGCGTTGTAGT		
<i>FSTL</i>	ENST00000256759	F1-FSTL	CGGCTGAGCACCTCGTG		155
		R1-FSTL	TTCTTGTTTCATTCGGCATT		
<i>GDF11</i>	ENST00000257868.9	F-GDF11	ATTGGCAGAGCATCGACTTC		182
		R-GDF11	TTTTGTGTTCTCTAGGACTCG		
<i>MSTN</i>	NM_005259	F-972	TTTTACCCAAAGCTCCTCCA		258
		R-3017	GAGTCTCGACGGGTCTCAAA		
<i>MYL1</i>	ENST00000352451	F-MYL1	GCAATGAAGAGCTGAATGCCA		126
		R-MYL1	TGCAAAAGACACGCAGACCCT		



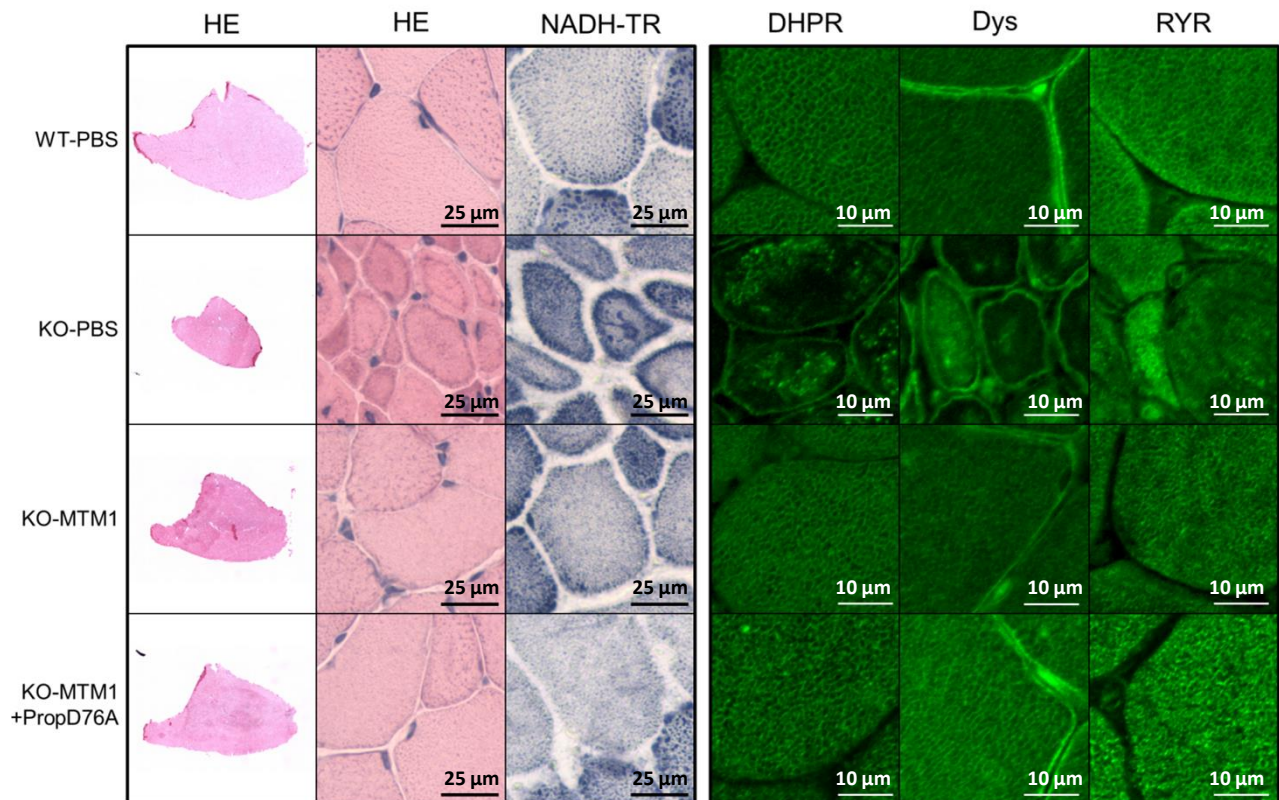
**Supplementary figure 1:** mRNA levels of either *MSTN* (A), *FSTN* (B), or *ACVR1B* (C) were measured by RT-qPCR in healthy controls (Ctrl, N=9), FSHD1 (N=8), FSHD2 (N=5) patients. Horizontal lines are medians, the extremities of the boxes are delimited by the first and third quartile, and the whiskers correspond to the 10th and 90th percentile. A one-way ANOVA, followed by the Fisher's Least Significant Difference multiple comparison test was performed.



**Supplementary figure 2:** mRNA levels of either *MSTN* (A), *FSTN* (B), *ACVR1B* (C), *GDF11* (D) and *ACTIVIN A* (E) were measured by RT-qPCR and normalized by the expression of the MLC-3fast myosin heavy chain (*MYL1*), in healthy controls (Ctrl, N=9), BMD (N=6), DMD (N=17), IBM (N=17), FSHD (N=13) and LGMD (N=11) patients. All graphs represent mean  $\pm$  SD. A one-way ANOVA, followed by the Fisher's Least Significant Difference multiple comparison test was performed.  $p < 0.05$ ; \*\* $<0.01$ ; \*\*\* $<0.001$



**Supplementary figure 3:** weight and mRNA levels of either *Mstn*, *Fstn*, *Acvr11b* were measured by RT-qPCR in the quadriceps (A) or the soleus (B) of either 21, 30 or 25 day old Mtm1-KO mice. n, number of mice used for each analysis: for the quadriceps, n = 9 and 8 at day 21, 8 and 8 at day 35 for WT and KO-Mtm1 mice respectively; for the soleus, n = 8 and 7 at day 21, 9 and 9 at day 35 for WT and KO-Mtm1 mice respectively. All graphs represent mean  $\pm$  SD. A one-way ANOVA, followed by the Fisher's Least Significant Difference multiple comparison test was performed. p \* < 0.05; \*\* < 0.01; \*\*\* < 0.001



**Supplementary figure 4:** Effect of Mtm1-rescue and mSeAP-PropD76A co-injection on muscle morphological features after 2 weeks of rAAV transduction in the Tibialis anterior (TA) in knockout mouse model. Histological aspect of PBS-treated WT and KO muscle and after injection of a rAAV2/1-Des-Mtm1 and rAAV2/1-CMV-mSeAP-PropD76A. Muscular fiber of TA cross-sections were stained on left panel with hematoxilin and eosin (HE) (magnification  $\times 40$  and  $\times 100$ ), mitochondria and endoplasmic reticulum were stained with nicotimanide adenine dinucleotide tetrazolium reductase (NADH-TR) (magnification  $\times 100$ ), and on right panel dihydropyridine receptor (DHPR), dysferlin (Dys) and ryanodine receptor (RYR) were detected by immunofluorescence (magnification  $\times 1200$ ).