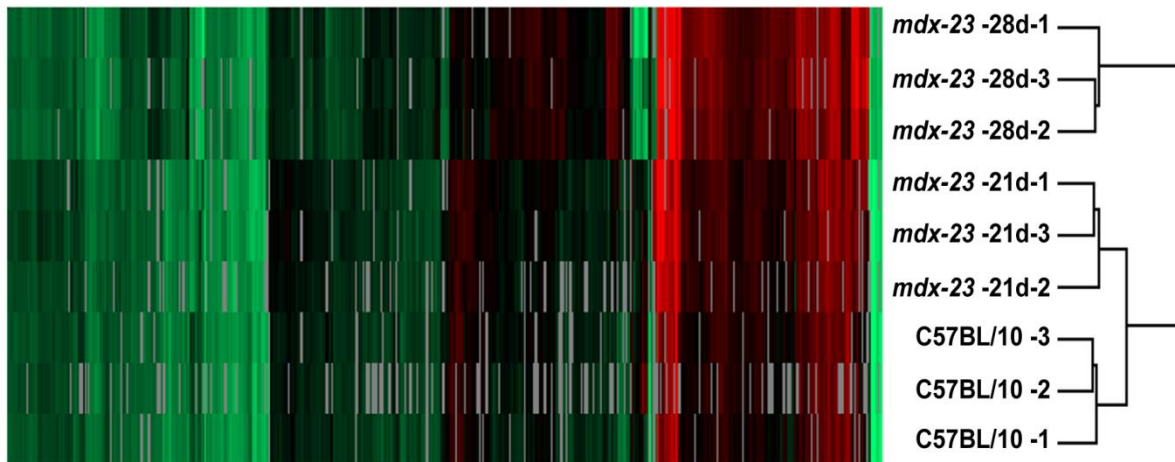
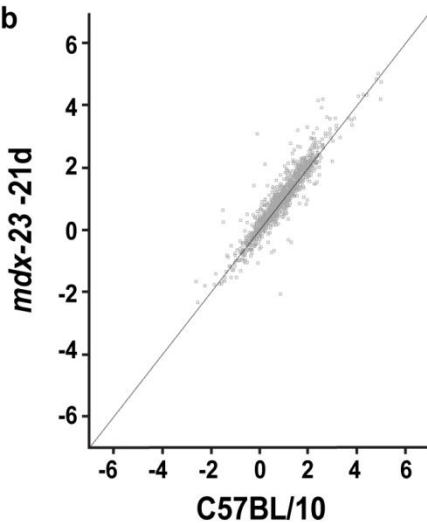


Supplementary Materials

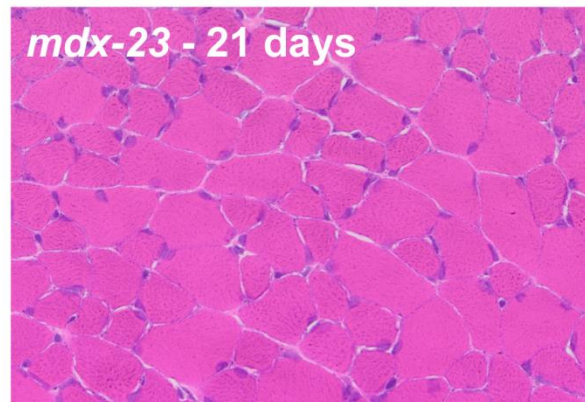
a



b

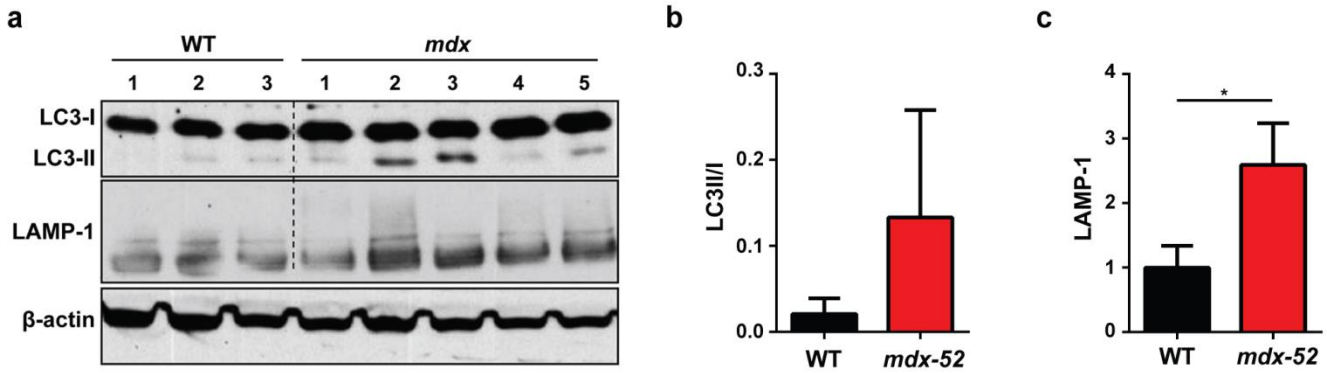


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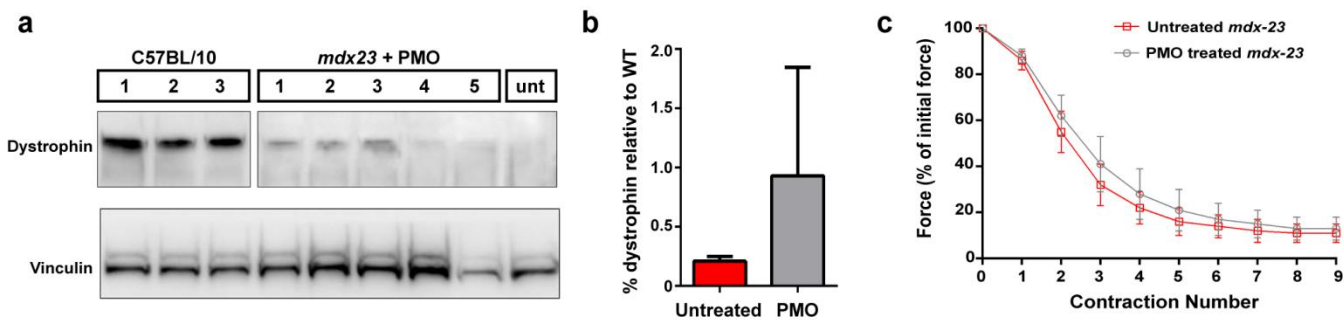


Supplemental Figure 1: Analysis of protein changes at disease onset in *mdx* mice. (a)

Hierarchical cluster analysis of the 1138 proteins identified in *mdx*-23 at 21 and 28 days of age and the parental WT strain (C57BL/10). (b) Pearson correlation analysis of proteins at 21 days of age *mdx*-23 and parental WT strain. (c) Images showing histological features of gastrocnemius muscle cross-sections from 21 day old *mdx*-23. Note the absence of small, centrally nucleated, regenerating fibers in the muscles at 21 days.



Supplemental Figure 2: (a) Western Blot analysis of *mdx-52* proteins at 28 days of age from five independent mice. **(b)** Densitometric analysis of LC3I and LC3II. Graph shows LC3II-to-LC3I ratio. **(c)** Densitometric analysis of LAMP-1 (normalized to the loading control – β -actin). (*) $p \leq 0.05$ by Student's *t* test.



Supplemental Figure 3: Effect of exon skipping on *mdx* sarcolemmal repair. (a) Western Blot of dystrophin protein from EDL of five *mdx-23* animals one month after one systemic (intravenous) injection of PMO. **(b)** Dystrophin level was quantified by normalizing the protein levels to loading control and presented as a percentage of the total WT dystrophin level. **(c)** Analysis of loss in EDL muscle contractile force following 10% LC-injuries.

Supplemental Table 2: Mitochondrial proteins with altered expression in 28 days old *mdx* muscle

Entry	Entry name
O35459	Delta(3,5)-Delta(2,4)-dienoyl-CoA isomerase, mitochondrial
Q9CPQ1	Cytochrome c oxidase subunit 6C
P56392	Cytochrome c oxidase subunit 7A1, mitochondrial
P43023	Cytochrome c oxidase subunit 6A2, mitochondrial
P51174	Long-chain specific acyl-CoA dehydrogenase, mitochondrial
P19536	Cytochrome c oxidase subunit 5B, mitochondrial
Q8BMF4	Dihydrolipoyllysine-residue acetyltransferase component of pyruvate dehydrogenase complex, mitochondrial (EC 2.3.1.12)
Q9CZU6	Citrate synthase, mitochondrial
P19783	Cytochrome c oxidase subunit 4 isoform 1, mitochondrial
P56391	Cytochrome c oxidase subunit 6B1
Q9EQ20	Methylmalonate-semialdehyde dehydrogenase [acylating]
Q64521	Glycerol-3-phosphate dehydrogenase, mitochondrial
P45952	Medium-chain specific acyl-CoA dehydrogenase, mitochondrial
Q8BH59	Calcium-binding mitochondrial carrier protein Aralar1
Q8K1Z0	Ubiquinone biosynthesis protein COQ9, mitochondrial
Q06185	ATP synthase subunit e, mitochondrial (ATPase subunit e)
Q9DCW4	Electron transfer flavoprotein subunit beta (Beta-ETF)
Q9DB20	ATP synthase subunit O, mitochondrial
Q9CQX8	28S ribosomal protein S36, mitochondrial
Q91WD5	NADH dehydrogenase [ubiquinone] iron-sulfur protein 2, mitochondrial
P70404	Isocitrate dehydrogenase [NAD] subunit gamma 1, mitochondrial
P50544	Very long-chain specific acyl-CoA dehydrogenase, mitochondrial
Q9WTP6	Adenylate kinase 2, mitochondrial (AK 2)
Q8BK30	NADH dehydrogenase [ubiquinone] flavoprotein 3, mitochondrial (Complex I-9kD)
Q6P8J7	Creatine kinase S-type, mitochondrial
P99029	Peroxiredoxin-5, mitochondrial
Q91VR2	ATP synthase subunit gamma, mitochondrial (F-ATPase gamma subunit)
P47791	Glutathione reductase, mitochondrial (GR)
Q91WS0	CDGSH iron-sulfur domain-containing protein 1 (MitoNEET)
Q9D0K2	Succinyl-CoA:3-ketoacid coenzyme A transferase 1, mitochondrial

Mitochondrial proteins whose level was significantly altered in 28 days old *mdx* mice compared to the age-matched control are listed such that black font represent proteins with lower expression in *mdx23* mice. The proteins listed in black font showed reduced level, while those in white font showed increased expression.