

**ADDITIONAL FILE 3**

*Table S1. Quantitative analyses of fibers containing areas of structural disarray (unstructured cores and contracture cores) reveals that alterations are far more frequent in CASQ1-null fibers.*

<b>EDL</b> <b>(n=2)</b>	<b>Age</b> <b>(months)</b>	<b>Total No. of</b> <b>analyzed</b> <b>fibers (No. of</b> <b>mice)</b>	<b>No. of fiber with alterations</b>		<b>C</b> <b>% Total fiber</b> <b>area altered</b>
			<b>A</b> <b>Unstructured</b> <b>Cores</b>	<b>B</b> <b>Contracture</b> <b>Cores</b>	
<b>WT</b>	4	34 (2)	0 (0%)	0 (0%)	<b>0</b>
<b>WT</b>	14	58 (2)	2 (3%)	0 (0%)	<b>3</b>
<b>WT</b>	20	56 (2)	0 (0%)	1 (2%)	<b>2</b>
<b>WT</b>	24	65 (2)	1 (2%)	2 (3%)	<b>5</b>
<b>WT</b>	27	63 (2)	4 (6%)	1 (2%)	<b>8</b>
<b>CASQ1-null</b>	4	41 (2)	1 (2%)	0 (0%)	<b>2<sup>&amp;</sup></b>
<b>CASQ1-null</b>	14	62 (2)	10 (17%)	4 (6%)	<b>23**</b>
<b>CASQ1-null</b>	20	83 (2)	13 (16%)	5 (6%)	<b>22**</b>
<b>CASQ1-null</b>	24	85 (2)	14 (16%)	7 (8%)	<b>24**</b>
<b>CASQ1-null</b>	27	81 (2)	9 (11%)	10 (13%)	<b>24*</b>

Columns A and B) Fibers exhibiting unstructured and contracture cores are frequent in EDL muscles of CASQ1-null mice starting at 14 months, but rare in EDL muscles of age-matched WT mice. There it seems to be trend for an increased percentage of contracture cores (from 6 to 13% between 14 and 27 months of age), which suggests that probably unstructured cores may be an early-point in formation of contracture cores. Column C) The total percentage of fiber area exhibiting ultrastructural decay (unstructured cores + contraction cores) increases significantly from 4 to 14 months of age in CASQ1-null muscles, but remains unchanged at later stages. \*p < 0.05 and \*\*p < 0.01 compared to age matched WT; &p < 0.05 compared to 14-month-old CASQ1-null.