

Table S1. Dystrophin antibodies.

name	clone	immunogen (human dystrophin)	epitope			vendor (catalogue number)	references for characterization of antibodies
			exons	region	amino acids		
MANEX1A	4C7	recombinant aa1-68		amino terminus	includes aa 2-4	DSHB	6, 11
MANEX7B	8E11	recombinant fragment encoded by exons 4-16	7-8	actin binding domain		DSHB	2
MANHINGE1B	10F9	peptide aa255-279 conjugated to albumin	8	hinge 1	255-270	DSHB	1
DYSB	34C5	recombinant aa321-494	10-12	R1-2	321-494	Leica (NCL-DYSB)	
MANDYS19	8F6	recombinant aa816-1749	20-21	R5	856-915	DSHB	8, 9
MANDYS16	1B12	recombinant aa816-1749	27	R8	1226-1245	DSHB	8, 9
MANDYS1	3B7	recombinant dystrophin fragment	31-32	R10-11	1431-1505	DSHB	8, 9
MANDYS110	3H10	recombinant aa1749-2248 as TrpE fusion protein	38-39	R14	1774-1856	DSHB	9, 10
MANDYS102	7D2	recombinant aa1749-2248 as TrpE fusion protein	43	R16	2047-2105	DSHB	9, 10

MANEX45A	8F10	recombinant fragment encoded by exons 45-46	45	R17		DSHB	11
MANEX46B	7G1	recombinant fragment encoded by exons 45-46	46	R18		DSHB	11
MANEX50	6A9	recombinant fragment encoded by exons 45-50	50	hinge 3		DSHB	11
MANDRA1	7A10	recombinant dystrophin fragment		carboxy terminus	3667-3671	DSHB	3, 4, 5, 7, 10
rabbit polyclonal		synthetic peptide, aa3661-3677		carboxy terminus	3661-3677	abcam [®] (ab15277)	

abcam[®] = <http://www.abcam.com/>

DSHB = Developmental Studies Hybridoma Bank, The University of Iowa (<http://dshb.biology.uiowa.edu/>)

Leica = Leica Microsystems (<http://www.leica-microsystems.com/products/total-histology/novocastra-reagents/>)

References

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Legends to supplementary figures.

Figure S1. Complete primary sequence of the region of dystrophin coded by exons 6 to 14. Domains of the protein are marked at the beginning of the lines. Exons were coloured sequentially to highlight their boundaries and phase with the protein domains. The number of the residue beginning an exon is noted under the sequence. The sequence recognized by the antibody MANHINGE1B is underlined in exon 8.

Figure S2. Superposition of the ^1H NMR spectra of R12 WT and R12 L427P. The spectra were acquired at 500 MHz on a Bruker Avance with water presaturation. Several resonances at high field and the dispersion of the amide protons are indicative of a folded wild type protein. In contrast, the spectrum of the mutated protein rather corresponds to an only partly folded protein as indicated by the broad NH resonances and the absence of high field signals.

Figure S3. (A) The three first principal components (PC) of motion measured from the R1-2 WT MD trajectory. Motion arrays are centred on $\text{C}\alpha$ atoms, sized and coloured depending on their amplitude from blue (low) to red (high). (B) Overlap of covariance matrices of the ten first principal modes is presented at bottom right. Second principal mode seems to remain unaffected by the L427P mutation. The first and the third most contributing components to the global motion (sum over 35% of the total) are the most strongly modified by the single mutation. Only 34% similarity has been measured between WT and mutant for PC1 and PC3.

/6 6/7
 <CH2>lqqtNSEKILLSWVRQSTRNYPQVNVINFTTSWSDGLALNALIHSHRPDLFDWNSVVCQGSAT
 131 178

7/8
 QRLEHAFNIARYQLGIE KLDPEDVDTTYPDKKSILMYITSLFQVLPQQVSIE
 217

8/9
 <HINGE1>AIQEVEMLPRPPKVTKEEHFQLHHQMHSQQITVSLAGGYERTSSPKPRFKSYAYTQAAY
 MANHINGE1B 278

9/10
 VTTSDPTRSPFPSQHLEAPEDKSFSSLMES
 321

10/11
 <R1>EVNLDRYQTALEEVLSWLLSAEDTLQAGGEISNDVEVVKDQFHTHEGYMMDLTAHQGRVGNILQ
 384

* 11/12
 LGSKLIGTGKLSDEETEVEQEQMNLNSRWECLRVASMEKQSNLH
 445

12/13
 <R2>RVLMDLQNGKLKELNDWLTKTEERTRKMEEEPLGPDLEDLKRQVQGHKVLQEDLEGEQVRVNSL
 495

13/14
 THMVVVDESSGDHATAALEEQLKVLGDRWANICRWTEDRVLLQDI
 535

Fig. S1

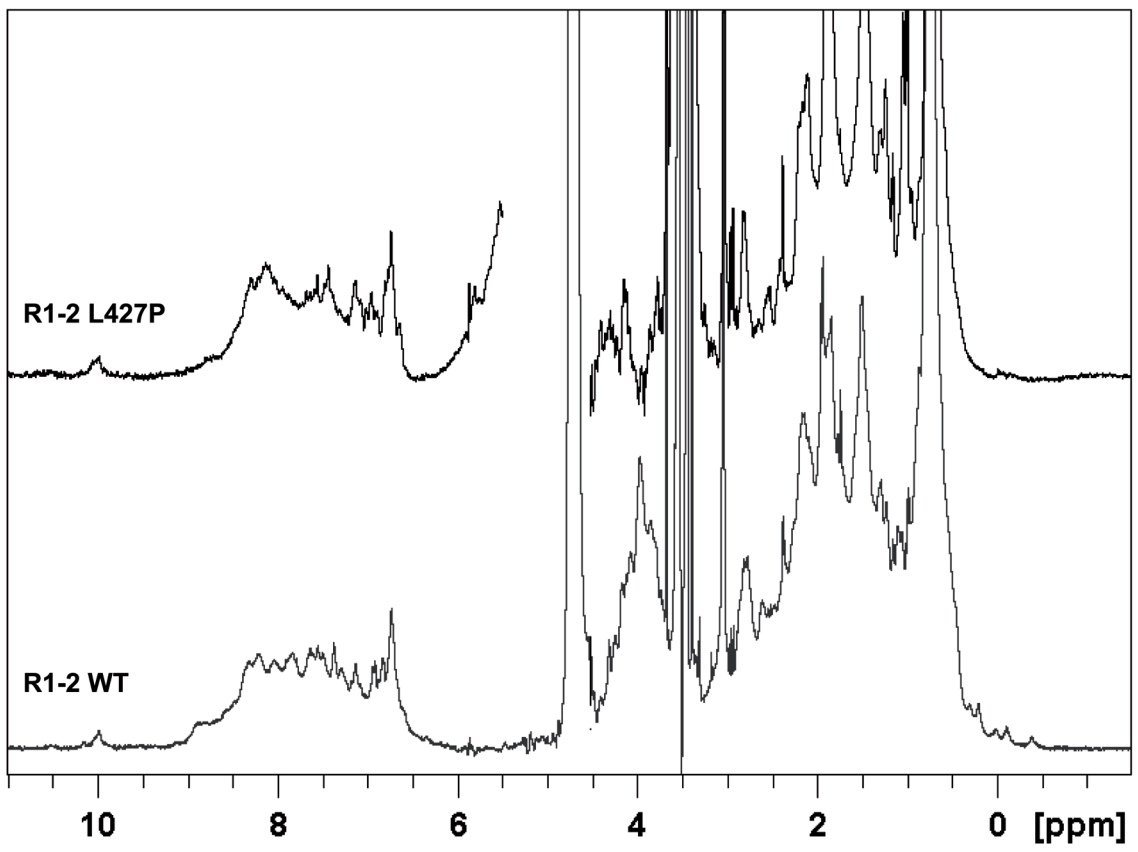


Fig. S2

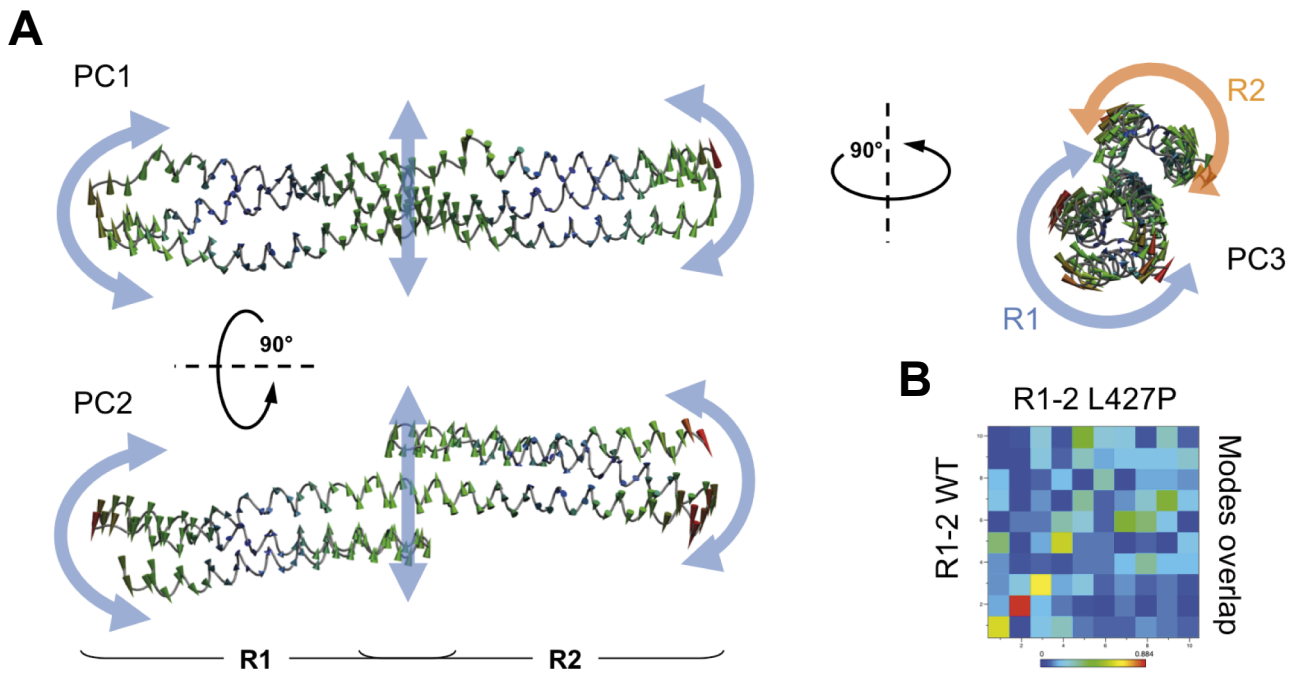


Fig. S3