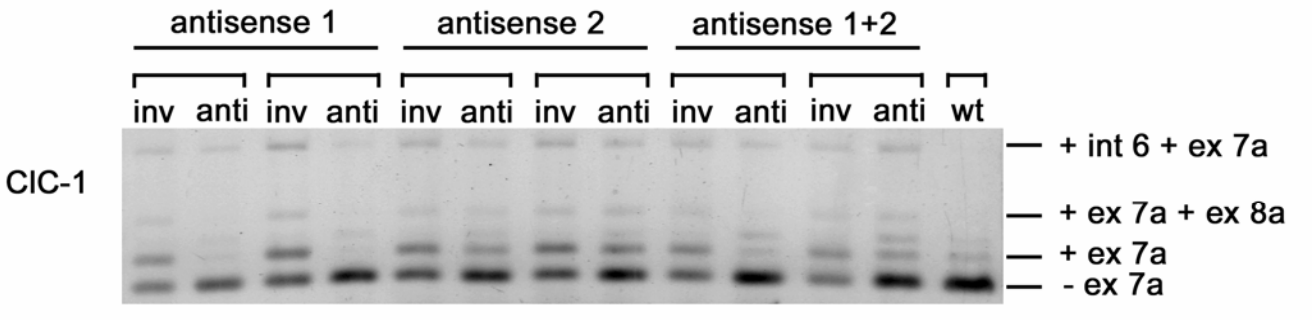
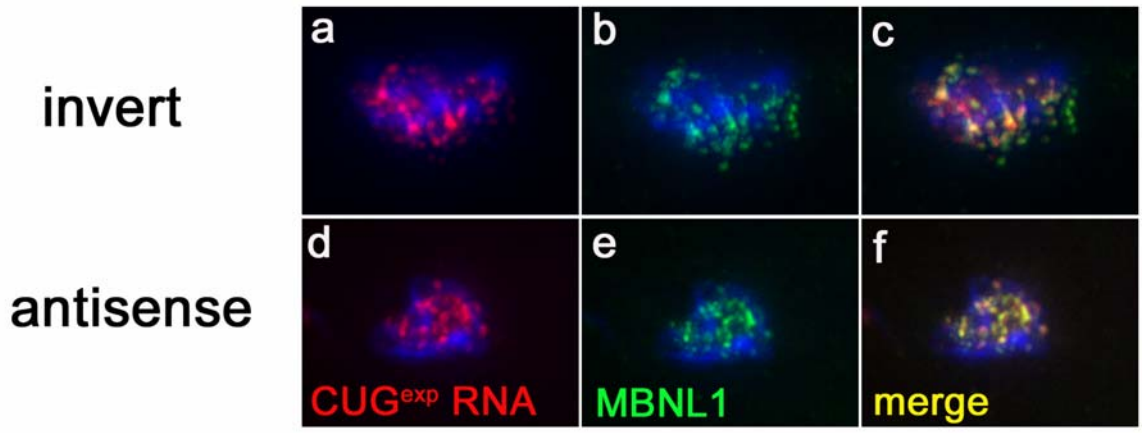


Supplementary Fig 1. Comparison of antisense morpholinos targeting ClC-1 exon 7a. Antisense oligo was injected into tibialis anterior (TA) muscle of *HSA*^{LR} mice and invert oligo (inv) (20 μg) was injected into the contralateral TA. Tissue was obtained 3 weeks later for analysis of ClC-1 splicing by RT-PCR. Antisense morpholino targeting the 3' splice site (antisense 1; 20 μg) induced a higher level of exon 7a skipping than antisense morpholino directed against the 5' splice site (antisense 2; 20 μg). Effects of antisense 1 alone (20 μg) were similar to co-injection of antisense 1 and 2 (10 μg each) (n = 3 each group; 2 from each group are shown).



Supplementary Fig 2. Antisense morpholino had no effect on the formation of ribonuclear inclusions. Fluorescence *in situ* hybridization and immunofluorescence demonstrate co-localization of CUG^{exp} RNA (red) and MBNL1 protein (green) in muscle nuclei (blue) 3 weeks after injection of HSA^{LR} TA muscle with invert (a-c) and antisense (d-f) morpholino.

Supplementary methods Fluorescence *in situ* hybridization and immunofluorescence analysis for MBNL1 protein was performed as described previously[1].

Reference

1. Lin, X. et al. Failure of MBNL1-dependent post-natal splicing transitions in myotonic dystrophy. *Hum Mol Genet* **15**, 2087-97 (2006).