

Fig. S1. qRT-PCR analysis of *mbl* silencing. cDNA was obtained from 50 flies of the indicated genotypes, and the levels of *mbl* expression were measured by quantitative real time PCR (qRT-PCR). *mbl* expression levels were normalized relative to the reference gene *tubulin 84C* and subsequently each genotype was divided by the value obtained for wild-type (*OrR*) control flies. *UAS-IR-mbl* flies, which carry an RNAi construct against *mbl* but no Gal4 driver, were used as an additional control. *mbl* silencing in *Act5C-Gal4 UAS-IR-mbl* flies was of approximately 50%. This value is in the range of the *mbl* expression reduction obtained with other classical mutant alleles of the gene. Three biological replicates and three technical replicates per biological sample were used. Graph bars represent mean values and their standard error.

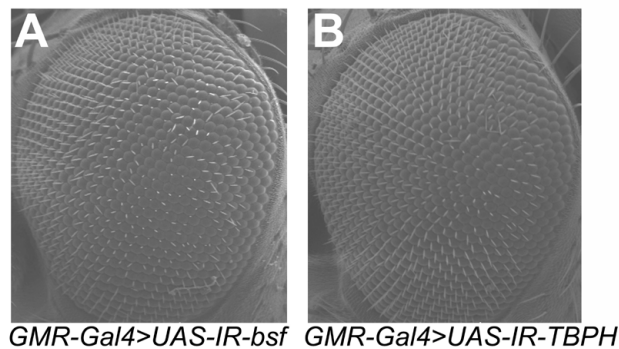


Fig. S2. Silencing of *bsf* or *TBPH* in the eye does not cause phenotypes. *GMR*-driven expression of *bsf* (A) or *TBPH* (B) RNAi transgenes did not affect eye morphology, ruling out that and additive phenomenon was responsible for their genetic interaction with i(CTG)480.

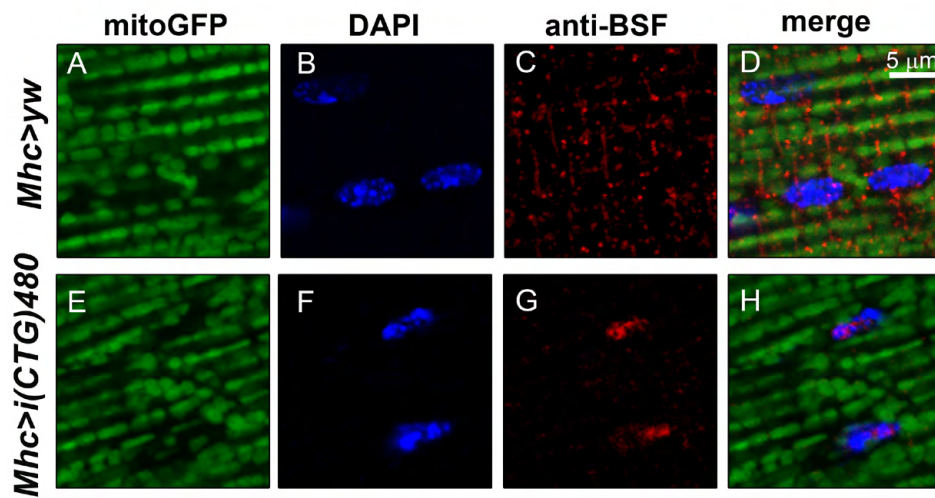


Fig. S3. bsf does not colocalize with mitochondria in the *Drosophila* muscle sarcomere. (A-D) Fluorescent confocal images of thorax cryosections from flies expressing a GFP-tagged mitochondrial marker (*Mhc-Gal4 UAS-mitoGFP*; green) and stained with an anti-BSF antibody (red). Nuclei are shown counterstained with DAPI (blue). The images confirm that BSF localizes preferentially in cytoplasmic bands, and to a lesser degree in the nuclei. No colocalization with the mitochondria was found. (H-K) Colocalization with DAPI confirmed that CTG-repeat expression (*Mhc-Gal4 UAS-i(CTG)480>UAS-mitoGFP*) abolished BSF cytoplasmic signal and caused BSF to re-locate into the nucleus. No mitochondrial signal was observed in this case either.

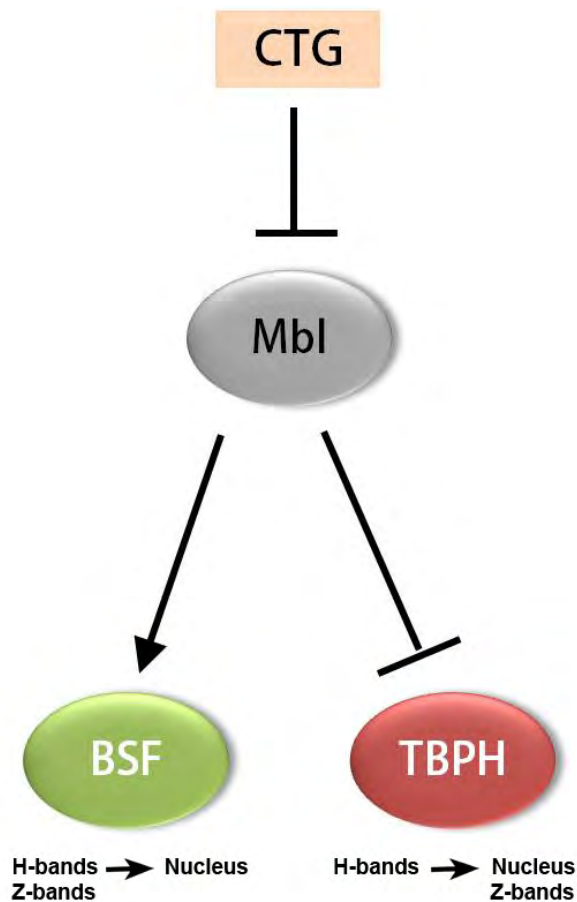


Fig. S4. Model of the interaction with BSF and TBPH. Mbl sequestration by CTG repeats would trigger mis-localization of TBPH, changing from predominantly cytoplasmic (sarcomeric H-bands) to predominantly nuclear with an aberrant presence in sarcomeric Z-bands. Mbl sequestration by CTG repeats would also induce a re-distribution of the BSF protein, from the cytoplasm to the nucleus.

[Click here to download Tables S1.](#)

Supplementary Table S2. DNA primer sequences and PCR conditions.

<i>Nested PCR⁽¹⁾</i>		
Primer	Tm	Sequence
IR-mbl-nested1f / IR-mbl-nested2r	65°C	TCAGCGGAAAAGGTGAATTATACC GCGGACTTGAAGAAGTCGGGGATCCCGTACTTTTACGCCGAT GG
IR-mbl-nested1f / IR-mbl-nested3r	65°C	TCAGCGGAAAAGGTGAATTATACC CAACATCGAGGACGGCAGCGTGCAGTACTTTTACGCCGATGG
IR-mbl-nested4f / GFPfrg2r	65°C	GTGAATTATACCAGAGCCTGC CTGCACGCTGCCGTCCTCGATGTTG
IR-mbl	65°C	GAAGATCTTCTACCAGAGCCTGAA
GFPfrgf / GFPfrgr	55°C	CGGGATCCCCGACTTCTTCAAGTCCGCC CGGGATCCCCGAGCTGCACGCTGCCGTCCT
<i>Quantitative Real Time PCR (qRT-PCR)</i>		
Primer	[nM]	Sequence
qPCRmbld / qPCRmblr	300	TTGAATCAAAATTATAGCCCAAGCT CGATTTTGCTCGTTAGCGTTT
TBPH-TranscriptLevelsf / TBPH-TranscriptLevelsr	250	TGCAGCGGCCACGAACAC CGGTCGCTGGTGCGAGGTTA
TBPH-AlternativeExonf / TBPH-AlternativeExonr	250	CGGTTGCTGTTCCCGCCACT GGCTACGGCATGGGTGGCAA
TBPH-UTR1f / TBPH-UTR1r	250	ATCCTGTCGTCTGTGTGTGAGTGC CCGCCAAACTGCCTCCGTCT
TBPH-UTR2f / TBPH-UTR2r	250	CGCACGCTCTCTCACACGAAT AGAACCGAGCGTGGAACGTACA
qTub84Bd / qTub84Br	900	CACTACACCATCGGCAAGGA GCAGACCGGTGCACTGATC
Rp49f / Rp49r	250	GGATCGATATGCTAAGCTGTCGCACA GGTGCCTTGTTCGATCCGTAACC

⁽¹⁾ The magnesium concentration used was 2 mM, except for the IR-mbl primer (3 mM). The number of PCR cycles was 30 in all cases.

