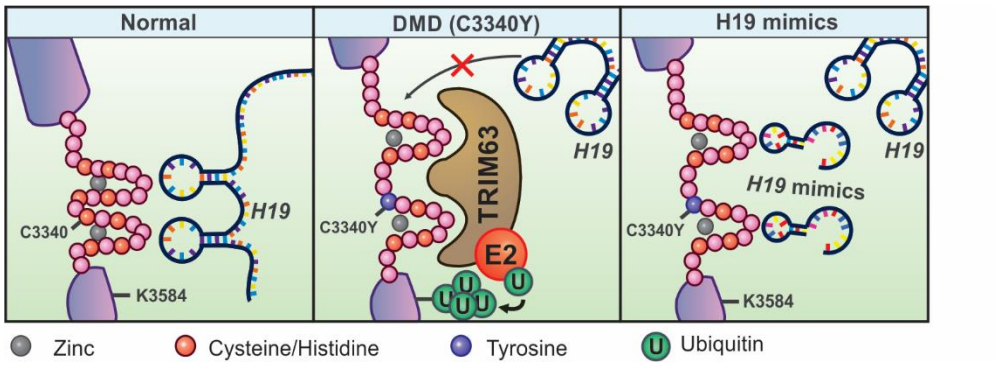
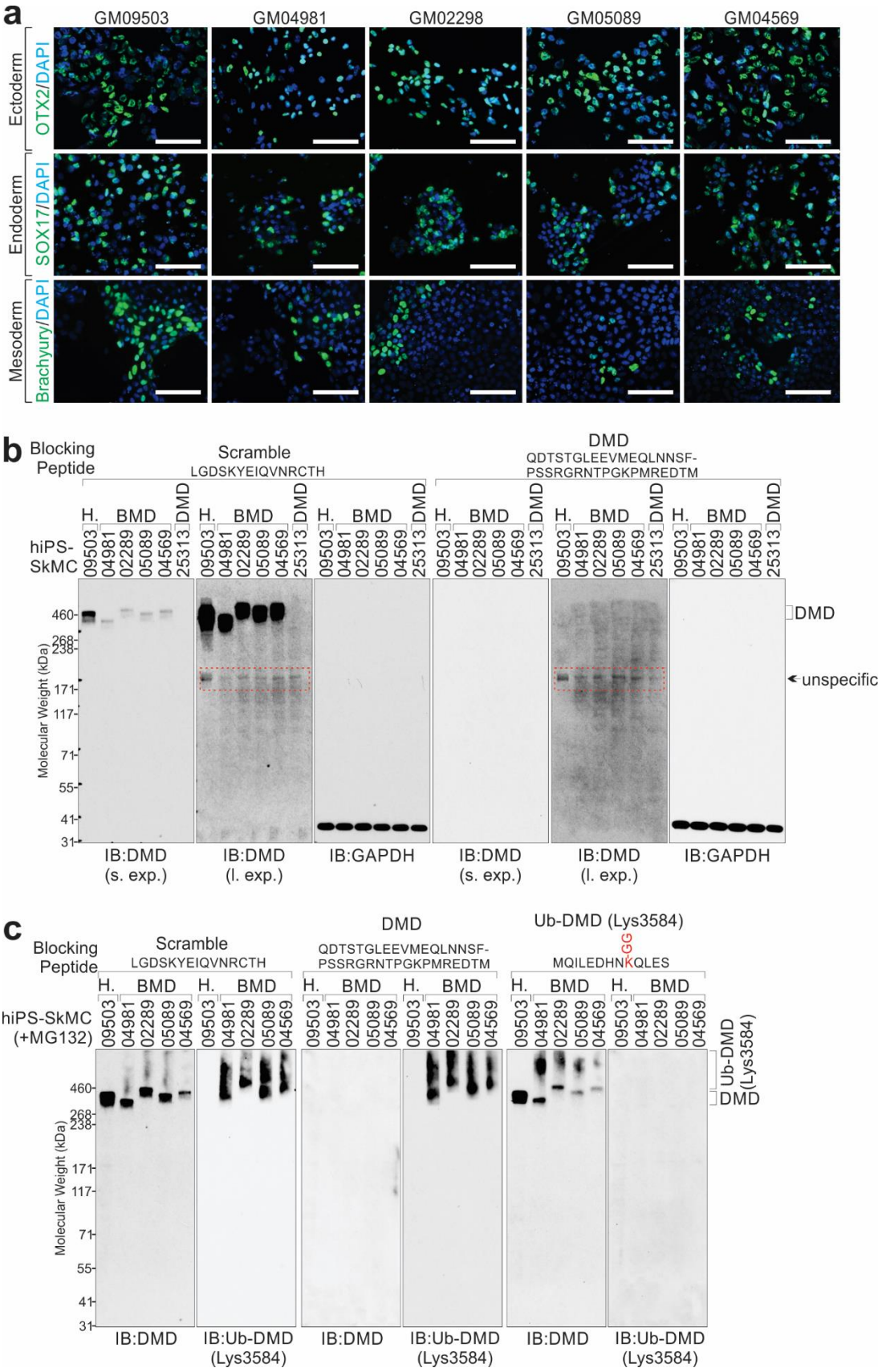
Supplementary Information

Supplementary Figure and Figure legend



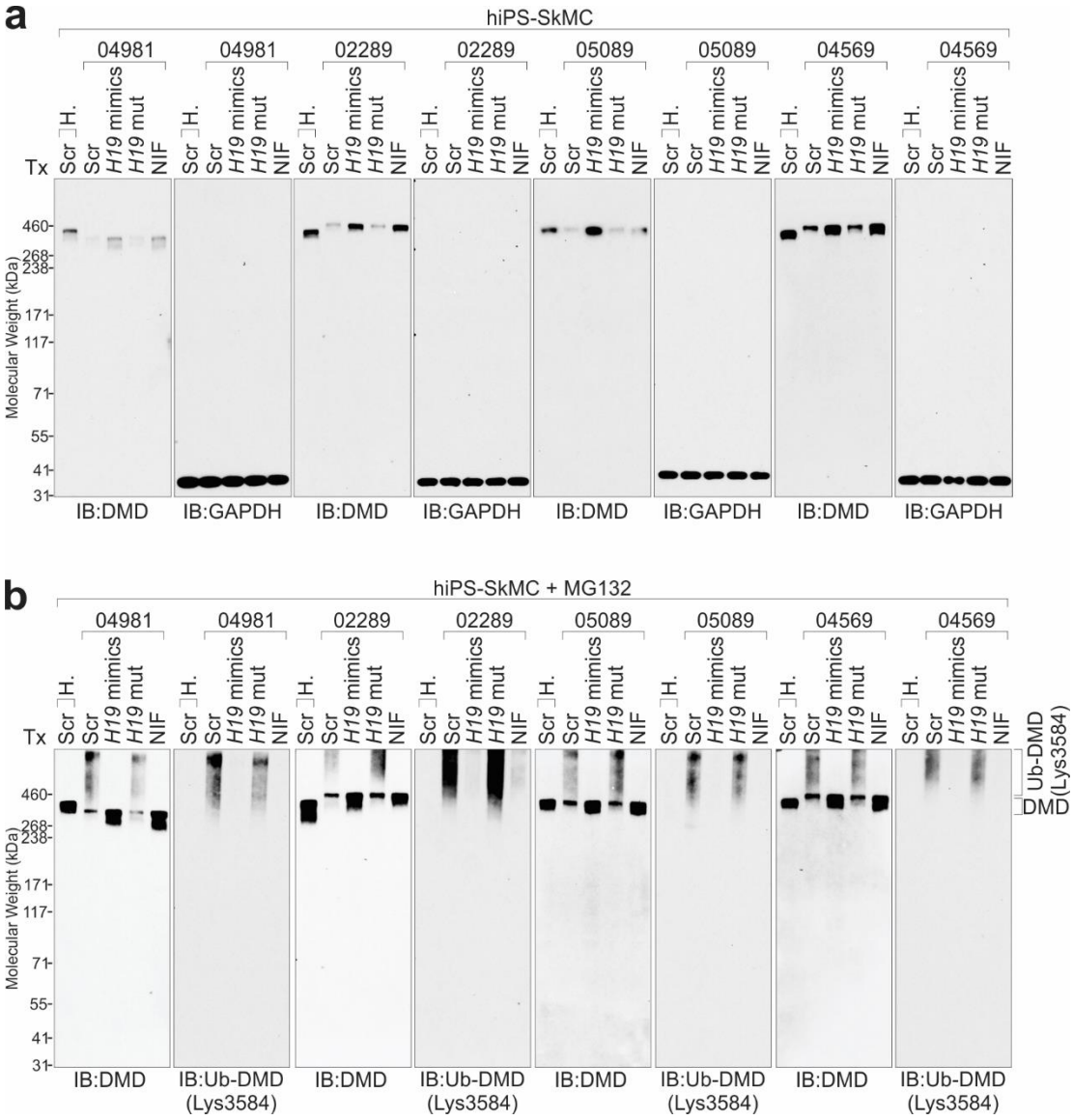
Supplementary Fig. 1 H19 mimics interfere with TRIM63-DMD interactions.

Graphic illustration of the H19-dependent competition with TRIM63 in interacting with dystrophin.


a
GM09503
GM04981
GM02298
GM05089
GM04569
Ectoderm
OTX2/DAPI
Endoderm
SOX17/DAPI
Mesoderm
Brachyury/DAPI
b
Blocking Peptide
Scramble
LGDSKYEIQVNRCTH
DMD
QDTSTGLEEVMEQLNNSF-PSSRGRNTPGKPMREDTM
H.
BMD
DMD
hiPS-SkMC
09503
04981
02289
05089
04569
25313
Molecular Weight (kDa)
460
268
238
171
117
71
55
41
31
DMD
unspecific
IB:DMD (s. exp.)
IB:DMD (l. exp.)
IB:GAPDH
IB:DMD (s. exp.)
IB:DMD (l. exp.)
IB:GAPDH
c
Blocking Peptide
Scramble
LGDSKYEIQVNRCTH
DMD
QDTSTGLEEVMEQLNNSF-PSSRGRNTPGKPMREDTM
Ub-DMD (Lys3584)
MQILEDHNKGGQLES
hiPS-SkMC (+MG132)
Ub-DMD (Lys3584)
DMD
IB:DMD
IB:Ub-DMD (Lys3584)
IB:DMD
IB:Ub-DMD (Lys3584)
IB:DMD
IB:Ub-DMD (Lys3584)

Supplementary Fig. 2 *Dmd* C3333Y mutation facilitates poly-ubiquitination and protein degradation of DMD.

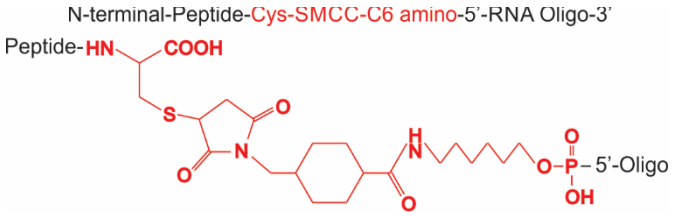
a, Pluripotency determination using indicated antibodies of hiPS cells derived from indicated donors. Scale bars, 100 μm. **b,** IB detection using indicated antibodies and blocking peptides using cell lysates extracted from indicated hiPS-SkMCs. c, Blocking Peptide Competition Assay using cell lysates extracted from indicated iPS-SkMCs and blocking peptides as shown, followed with IB detection using indicated antibodies. MG132 was used as proteasome inhibitor.



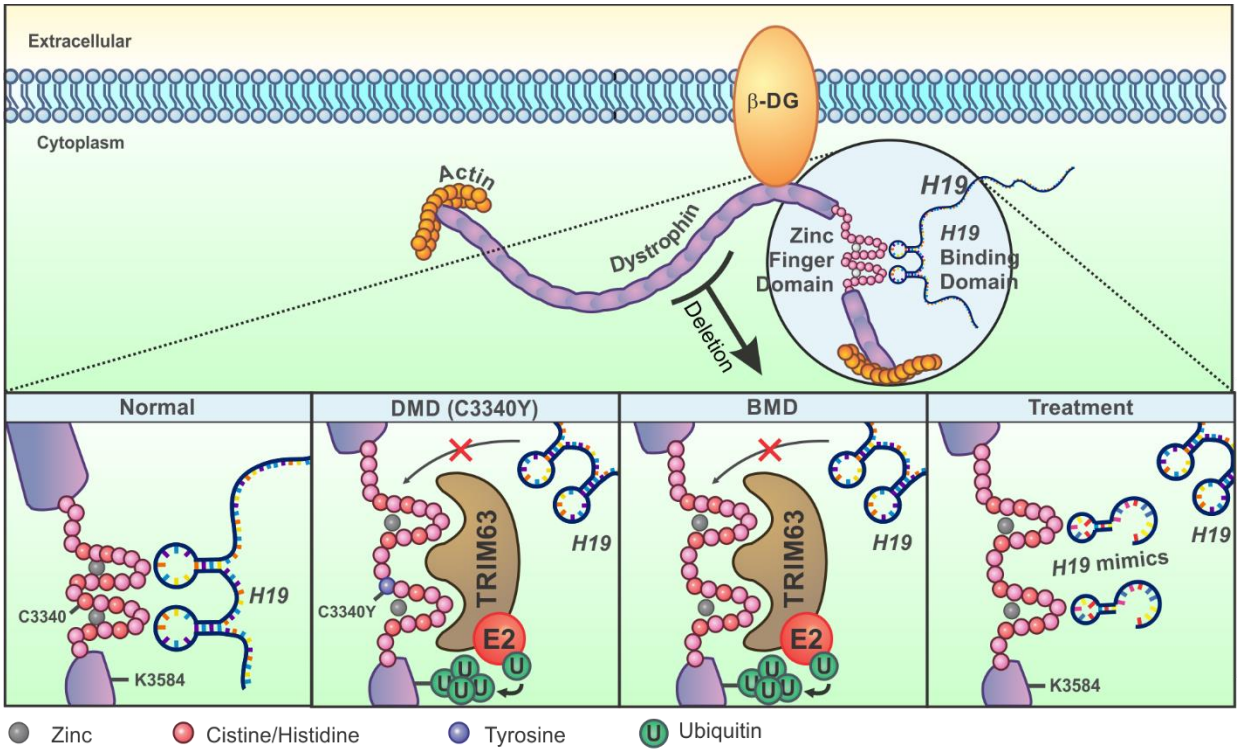
Supplementary Fig. 3 *H19* and Nifenazone extend the half-life of DMD protein.

a, IB detection using indicated antibodies of hiPS-SkMCs derived from healthy donor (09503) or BMD patients in the presence of scramble (Scr), AGR-*H19*, AGR-*H19* mutant, or NIF treatment.

b, IB detection using indicated antibodies of hiPS-SkMCs derived from healthy donor (09503) or BMD patients as indicated, in the presence of scramble (Scr), AGR-*H19*, AGR-*H19* mutant or NIF treatment. MG132 was used as proteasome inhibitor.



Supplementary Fig. 4 Illustration of SMCC linker used to conjugate peptide and RNA oligonucleotides used for designing of AGR-*H19*.



Supplementary Figure 5. *H19* competes with TRIM63 in associating with dystrophin. Graphic illustration of the biological importance of *H19* in interacting with dystrophin to inhibit the TRIM63-medicated, K48-linked poly-ubiquitination of dystrophin at K3584.