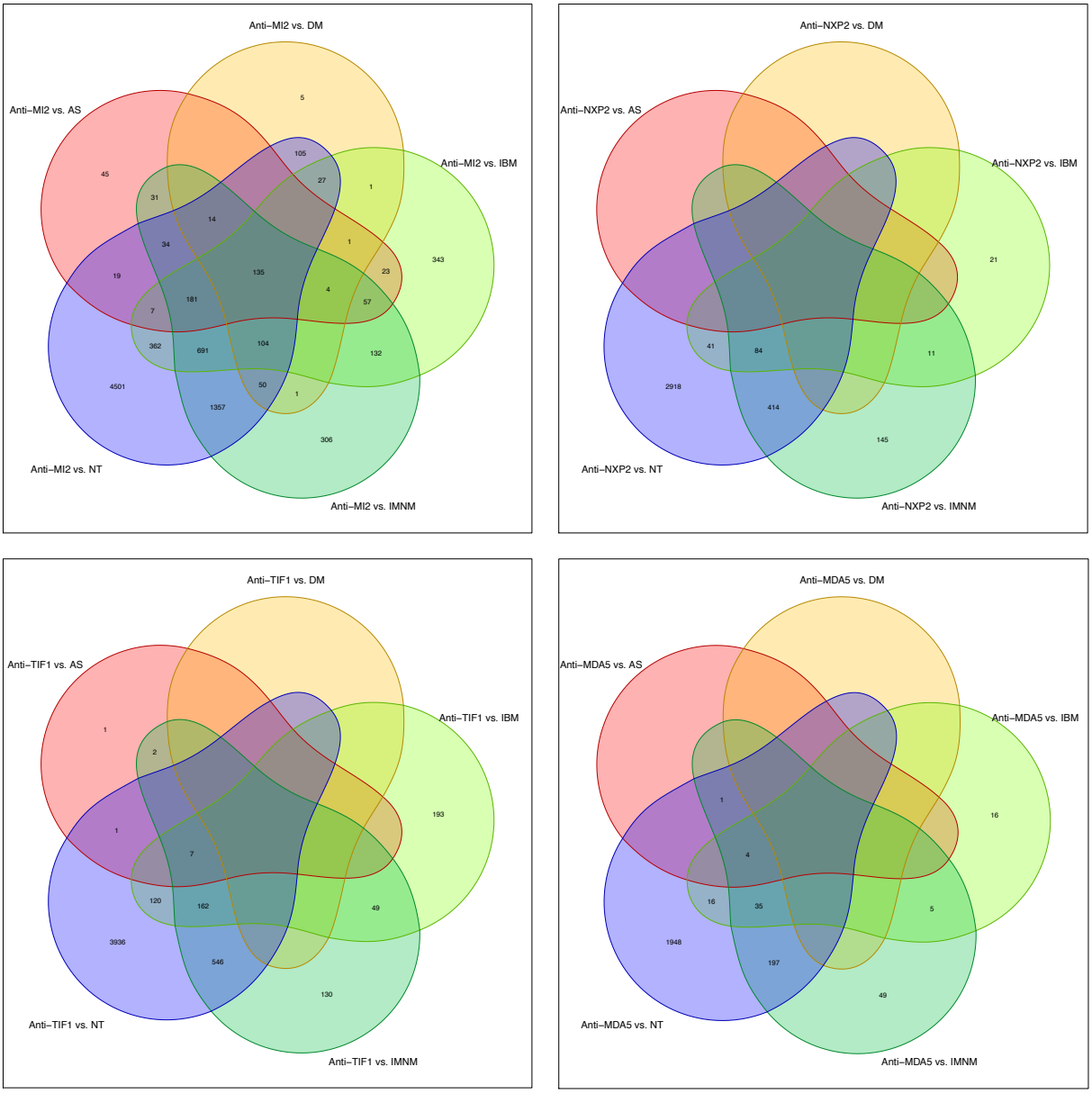
Supplementary Table 1. Differential expression of the set of 135 genes specifically overexpressed in anti-Mi2 dermatomyositis.

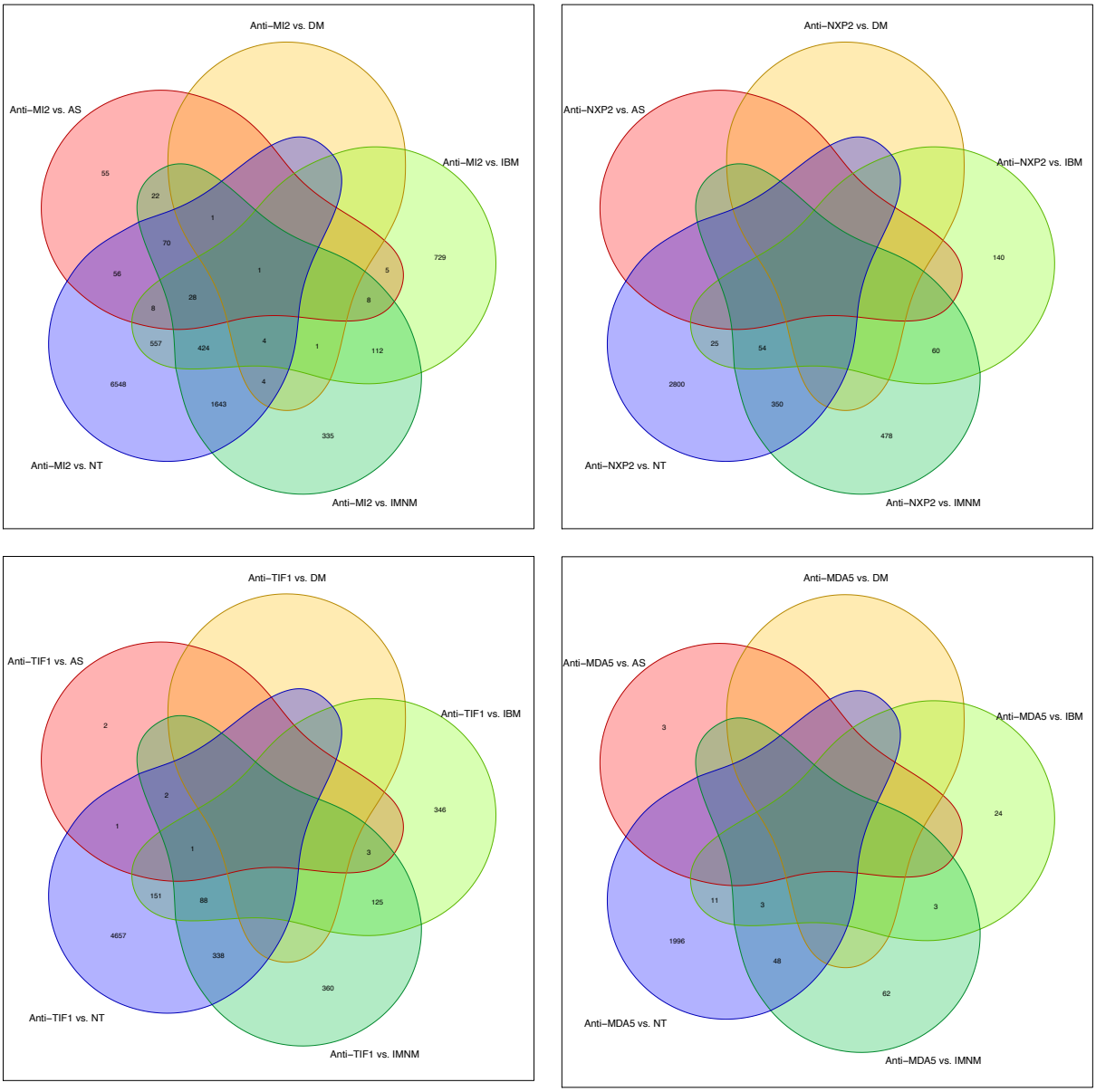
	Anti-Mi2 vs. ALL		Anti-Mi2 vs. NT		Anti-Mi2 vs. DM		Anti-Mi2 vs. AS		Anti-Mi2 vs. IMNM		Anti-Mi2 vs. IBM	
Gene	logFC	adj.P.Val	logFC	adj.P.Val	logFC	adj.P.Val	logFC	adj.P.Val	logFC	adj.P.Val	logFC	adj.P.Val
ABCG4	2.1	3e-07	2.3	4e-06	2.2	0.002	2.2	0.02	2.0	3e-05	1.5	0.03
ADAT3	1.2	7e-06	1.3	2e-05	1.2	0.04	1.3	0.04	1.2	8e-05	1.2	0.01
ALPG	3.0	3e-07	3.1	4e-06	3.1	0.009	3.1	0.02	2.9	6e-04	3.5	0.004
ANKRD20A19P	4.5	3e-22	4.3	2e-08	4.6	8e-06	4.6	7e-04	4.6	7e-11	4.3	0.002
APC2	2.9	2e-11	3.3	3e-07	2.6	0.009	3.1	0.005	2.8	1e-06	3.3	0.003
AQP5	2.7	2e-09	2.9	7e-09	2.6	0.003	2.8	0.002	2.4	2e-05	2.9	0.001
ARHGDIG	2.7	1e-09	3.4	7e-06	2.4	0.01	3.1	0.01	2.3	4e-05	2.6	0.02
B4GALNT4	3.2	2e-16	3.2	6e-07	2.8	3e-05	3.5	4e-04	3.3	2e-10	3.1	0.001
BAHD1	0.6	2e-06	0.4	0.01	0.7	0.01	0.7	0.007	0.7	7e-06	0.5	0.04
BAIAP3	1.5	2e-13	1.0	9e-05	1.7	3e-04	1.9	3e-04	1.5	5e-10	1.6	0.001
BANCR	2.8	1e-11	2.8	1e-08	2.6	0.003	2.7	0.003	3.0	2e-08	2.6	0.003
BEAN1	2.0	1e-05	3.1	1e-09	2.2	0.02	2.1	0.03	1.3	0.005	1.7	0.03
C1QTNF8	2.9	2e-11	2.7	9e-07	3.0	5e-04	3.2	6e-04	2.9	4e-07	3.1	0.001
CACNA1G	3.1	8e-15	3.4	5e-07	3.2	1e-04	4.0	6e-04	2.7	1e-07	2.9	0.004
CADM4	1.1	5e-10	0.8	4e-04	1.2	3e-04	1.4	2e-04	1.0	5e-08	1.4	2e-04
CAMKV	4.1	7e-21	4.0	1e-09	4.3	3e-07	4.0	3e-04	4.2	1e-12	3.6	0.002
CAMSAP3	3.0	8e-12	3.1	1e-06	2.7	1e-04	2.6	0.004	3.1	3e-08	2.9	0.004
CAPN12	3.0	8e-17	3.8	2e-07	2.8	6e-04	3.6	0.004	2.8	4e-09	2.1	0.03
CASKIN1	2.5	1e-14	2.8	1e-08	3.1	3e-05	2.3	0.003	2.0	3e-08	2.8	6e-04
CBARP	2.3	9e-15	2.8	3e-09	2.0	2e-04	2.1	0.006	2.0	7e-08	3.0	2e-04
CD164L2	1.8	9e-06	1.5	7e-04	2.1	0.005	1.7	0.05	1.7	0.001	1.7	0.03
CHGA	3.4	4e-14	3.4	1e-09	3.5	3e-05	2.8	0.009	3.5	1e-09	4.1	2e-05
CHRM4	5.0	3e-28	5.4	8e-10	4.8	8e-07	4.0	0.01	5.4	3e-14	4.3	0.004
COL11A2	2.0	2e-13	1.2	2e-04	2.4	1e-04	2.4	3e-04	2.0	4e-11	2.4	2e-04
COL26A1	2.7	3e-09	2.5	1e-05	2.6	0.002	2.9	0.004	2.7	1e-06	2.6	0.005
COL2A1	3.8	7e-16	3.7	3e-05	3.7	6e-05	4.0	0.007	3.7	2e-08	3.9	0.007
COL9A3	1.8	8e-08	1.9	2e-04	1.6	0.03	1.8	0.02	2.0	2e-06	1.6	0.03
COX6B2	4.5	2e-19	4.6	7e-09	4.5	1e-04	4.8	6e-04	4.4	3e-10	4.6	0.002
CPNE6	4.4	8e-21	4.4	5e-09	4.3	6e-06	4.7	3e-04	4.5	7e-11	3.7	0.005
CRB3	2.4	4e-08	2.5	4e-06	2.0	0.04	2.6	0.007	2.5	7e-06	2.6	0.003
CSDC2	1.9	1e-06	3.3	4e-09	2.2	0.002	1.7	0.04	1.1	0.005	1.4	0.04
DACT3	1.5	1e-10	2.0	9e-08	1.3	0.01	1.8	0.002	1.4	3e-06	1.4	0.01
DHRS2	5.1	2e-28	6.0	7e-12	4.1	3e-05	4.7	0.001	5.4	1e-14	5.2	6e-04
DLGAP3	2.4	3e-09	2.9	3e-07	2.3	0.002	2.3	0.02	2.2	9e-06	2.2	0.01
DPYSL4	1.4	3e-09	1.4	3e-06	1.3	0.02	1.4	0.02	1.2	3e-05	2.0	6e-04
ECE2	3.9	3e-17	3.6	7e-07	4.0	2e-04	4.0	0.002	4.0	3e-09	4.1	0.002
EFNA3	1.4	4e-07	1.4	2e-04	1.5	0.01	1.5	0.02	1.3	8e-05	1.4	0.02
EFNB3	1.5	1e-04	1.3	0.002	1.9	0.02	1.7	0.03	1.3	0.003	1.7	0.006
ELFN1	1.7	2e-08	1.5	2e-06	2.0	0.004	1.9	0.01	1.5	1e-06	1.6	0.004
ENSG00000142539.9	3.9	1e-13	3.7	6e-07	4.1	5e-05	3.5	0.01	4.2	5e-10	3.8	0.004
ENSG00000189093.16	1.1	2e-11	1.1	8e-07	1.3	8e-04	1.1	0.005	1.0	9e-07	0.9	0.009
ENSG00000218416.4	2.3	5e-07	2.2	2e-06	2.0	0.008	2.0	0.02	2.5	1e-06	2.6	3e-04
ENSG00000251511.1	2.6	8e-13	2.3	2e-07	2.9	6e-05	2.6	0.002	2.6	5e-08	2.7	0.001
ENSG00000255375.3	3.1	2e-11	3.1	2e-07	2.9	0.003	2.5	0.03	3.4	1e-08	3.5	5e-04
ENSG00000256481.1	2.3	3e-10	2.2	2e-08	2.6	7e-05	2.1	0.005	2.3	3e-06	2.5	7e-04
ENSG00000260293.2	2.1	6e-08	2.7	1e-07	1.9	0.04	2.4	0.009	1.9	6e-06	1.9	0.007
ENSG00000267110.1	2.3	2e-05	1.7	0.006	2.5	0.007	2.5	0.01	2.3	3e-04	3.0	7e-04
ENSG00000272512.1	2.9	8e-13	2.5	3e-06	1.7	0.02	3.1	7e-04	3.6	6e-13	3.3	2e-04
ENSG00000272884.1	2.1	8e-04	1.5	0.05	2.3	0.02	3.1	0.004	2.1	4e-04	1.8	0.05
ENSG00000275437.1	2.2	1e-09	2.2	5e-06	2.1	0.002	1.9	0.007	2.4	2e-08	2.1	0.001
ENSG00000286311.1	3.3	2e-14	3.2	4e-10	3.3	2e-05	2.9	4e-04	3.3	4e-11	3.6	2e-05
ENSG00000287069.1	1.7	5e-04	1.4	0.001	2.0	0.02	1.7	0.03	1.7	0.003	1.5	0.05
ENSG00000286766.1	1.8	8e-07	2.3	7e-07	2.2	3e-04	1.9	0.01	1.3	0.002	1.2	0.04
ENSG00000289332.1	4.3	1e-24	4.3	4e-10	4.4	1e-06	4.4	3e-04	4.2	1e-11	4.7	1e-04
ENTPD8	3.2	5e-13	3.5	4e-09	2.5	0.006	3.7	3e-04	3.3	2e-08	3.6	2e-04
EPPK1	2.7	2e-09	3.0	9e-05	2.9	8e-04	3.0	0.006	2.5	9e-07	2.4	0.02
ERF	1.3	2e-08	1.5	3e-07	1.4	0.004	1.1	0.03	1.1	1e-05	1.1	0.02
ESPNP	3.0	1e-11	3.2	9e-09	3.2	4e-04	2.4	0.02	3.1	4e-08	2.9	0.002
FAM131C	2.6	7e-08	3.1	6e-07	2.1	0.03	2.5	0.02	2.6	2e-05	2.3	0.02
FAM171A2	3.1	1e-15	3.7	6e-07	4.2	9e-07	2.6	0.02	2.3	2e-06	3.1	0.005
FBXL16	3.8	8e-17	4.8	1e-08	3.7	3e-05	3.5	0.006	3.7	3e-08	2.9	0.01
FGFRL1	0.9	7e-08	0.4	0.008	1.0	7e-04	1.2	2e-04	0.8	4e-06	1.1	5e-04
FIGNL2	1.8	6e-07	1.8	6e-05	1.9	0.01	1.8	0.01	1.7	2e-05	1.5	0.02
FLRT1	2.5	2e-09	3.0	7e-08	2.1	0.01	3.1	4e-04	2.2	5e-06	2.5	0.004
GATA5	2.3	6e-08	2.4	4e-06	2.2	0.007	2.1	0.02	2.3	1e-05	2.4	0.004
GCGR	4.2	1e-17	4.2	7e-08	4.2	2e-05	4.2	9e-04	4.5	2e-11	3.7	0.006
GIMD1	2.6	1e-12	2.3	3e-07	2.8	1e-04	2.4	0.006	2.6	4e-08	2.9	4e-04
GRB7	1.9	3e-06	2.4	6e-08	1.9	0.03	1.4	0.05	1.8	3e-05	1.8	0.001
GRIN2D	2.7	4e-13	3.9	3e-09	1.8	0.04	2.3	0.02	2.6	8e-08	2.9	0.003
GRIN3B	3.1	2e-12	3.4	5e-08	2.9	5e-04	2.8	0.02	3.3	2e-09	2.8	0.005
HCN2	3.5	1e-15	4.2	3e-07	3.3	5e-04	3.3	0.02	3.5	2e-08	2.9	0.02
HR	1.6	1e-07	2.1	5e-08	1.7	0.01	1.6	0.01	1.3	8e-05	1.3	0.02
IFITM5	5.4	2e-30	5.1	3e-09	5.5	5e-07	5.4	3e-04	5.4	2e-12	5.6	4e-04
IGLON5	3.6	3e-14	3.5	9e-08	3.6	1e-04	3.8	6e-04	3.6	2e-07	3.8	0.001
IL11	3.8	2e-13	4.3	6e-09	3.1	0.01	4.0	0.002	3.8	4e-08	3.9	0.003
JPH3	2.8	4e-11	3.2	6e-08	2.7	5e-04	3.2	6e-04	2.7	4e-08	2.4	0.004
KCNJ4	5.6	6e-34	5.6	5e-11	5.9	6e-09	4.8	0.001	5.7	3e-15	5.8	2e-04
KIF1A	4.7	3e-22	4.8	3e-07	3.7	2e-04	5.3	2e-04	5.5	2e-15	4.0	0.002
KLK13	2.5	4e-06	2.3	1e-05	2.6	0.01	2.6	0.004	2.6	1e-06	2.3	0.007
KRTAP2-3	1.9	4e-06	1.4	0.006	2.3	0.004	1.9	0.05	2.0	1e-04	2.1	0.02
LCAL1	2.0	4e-08	1.7	2e-05	2.4	4e-04	2.0	0.004	2.0	2e-05	2.0	0.004
LCN12	2.0	3e-11	2.2	2e-07	1.7	0.01	2.2	0.005	2.0	2e-08	2.5	7e-04
LINC00624	2.8	1e-11	2.7	8e-08	2.5	6e-04	3.0	3e-04	2.9	1e-08	3.4	3e-05
LKAAEAR1	3.0	6e-14	2.9	2e-08	3.5	3e-06	2.7	0.003	3.0	5e-09	2.9	7e-04
MADCAM1	2.5	4e-20	2.6	1e-07	2.6	5e-05	2.8	0.001	2.3	3e-10	2.7	0.001
MMEL1-AS1	2.0	2e-07	1.7	2e-05	2.2	0.007	2.0	0.007	2.0	1e-05	2.1	0.003
MMP24	1.8	4e-06	2.8	4e-07	2.1	0.004	2.1	0.01	1.1	0.005	1.4	0.04
MYRF	2.3	5e-09	2.9	3e-08	2.2	0.01	2.3	0.006	2.2	7e-09	1.5	0.02
NCKAP5L	0.8	2e-05	1.0	4e-07	0.8	0.005	0.9	0.02	0.4	0.03	0.8	0.007
NECAB2	2.8	1e-10	3.3	4e-08	2.4	0.006	3.1	0.01	2.8	7e-07	2.5	0.01
NKAIN4	4.2	2e-20	4.2	2e-09	4.2	5e-06	4.4	2e-04	4.5	2e-14	3.7	0.002
NLGN2	1.0	1e-12	1.3	1e-11	0.8	0.007	1.0	7e-04	0.9	2e-08	1.0	4e-04
NTN3	1.4	9e-04	0.9	0.04	1.6	0.03	1.6	0.02	1.5	0.002	1.4	0.04
OVOL1	3.1	7e-15	2.8	9e-08	3.4	1e-05	3.2	4e-04	3.2	8e-10	3.0	0.002
P2RX2	5.5	8e-35	5.5	2e-12	5.1	9e-08	5.0	3e-04	5.8	1e-17	5.8	2e-05
PALM3	3.0	4e-12	3.1	7e-08	3.3	2e-04	2.9	0.007	3.0	6e-08	2.7	0.005
PDIA2	2.5	8e-13	3.1	7e-08	2.0	0.009	2.8	0.002	2.5	4e-08	2.5	0.003
PHOX2A	2.2	8e-08	2.1	4e-06	2.3	0.009	2.4	0.002	2.2	1e-05	2.7	4e-04
PLEKHG4B	2.2	6e-06	1.7	0.002	2.3	0.004	2.5	0.009	2.5	1e-06	1.7	0.02
PLPPR3	4.3	1e-25	4.2	3e-11	4.5	1e-08	4.5	9e-06	4.2	1e-13	4.8	5e-06
PPP1R1B	3.9	2e-14	4.4	1e-07	3.5	4e-04	4.6	9e-04	3.9	6e-09	2.6	0.02
PRKCG	3.5	4e-14	3.4	2e-07	3.5	3e-04	3.6	0.002	3.7	6e-10	2.7	0.02
PRMT8	2.2	9e-06	2.2	1e-05	1.9	0.05	2.4	0.006	2.3	1e-05	1.7	0.04
PRR35	5.5	1e-31	5.4	6e-10	5.7	5e-08	5.4	4e-04	5.5	2e-13	5.7	3e-04
PRX	1.1	3e-07	0.8	0.01	1.1	0.01	1.3	0.003	1.2	5e-07	1.0	0.01
PTH2	1.7	9e-06	1.4	5e-04	1.8	0.02	1.7	0.04	1.7	7e-04	2.0	0.006
PTPRN	3.5	6e-13	4.1	2e-09	2.4	0.04	3.4	0.007	3.8	2e-09	3.6	0.002
RAB26	1.9	6e-08	1.0	0.006	2.4	7e-04	2.6	7e-04	1.9	9e-08	1.7	0.005
RAB3B	4.0	1e-16	3.6	5e-07	3.9	1e-04	3.9	4e-04	4.7	1e-15	3.5	0.002
RAC3	2.3	8e-09	2.8	2e-06	1.9	0.007	2.1	0.01	2.4	3e-08	2.0	0.01
RAP1GAP	2.4	5e-09	2.8	4e-06	2.0	0.02	2.5	0.006	2.7	2e-08	1.8	0.02
RTN4RL2	1.2	4e-08	1.2	2e-05	1.1	0.05	1.3	0.02	1.1	7e-06	1.4	0.002
SCRT1	6.3	2e-39	6.3	2e-11	6.3	1e-08	6.0	3e-04	6.3	4e-15	6.9	9e-05
SEMA4C	0.7	5e-06	0.7	8e-05	0.8	0.004	0.8	0.004	0.5	0.002	0.6	0.005
SEMA4G	0.9	9e-04	0.6	0.008	1.6	0.008	1.0	0.03	0.6	0.009	0.8	0.04
SHISA7	3.4	3e-14	3.0	4e-07	3.6	2e-05	3.6	4e-04	3.7	1e-11	3.2	0.002
SLC25A29	0.8	6e-07	0.7	4e-05	0.7	0.04	0.9	0.007	0.8	2e-07	0.7	0.002
SLC30A3	2.9	1e-11	3.8	2e-07	3.1	0.002	2.5	0.04	2.6	7e-07	2.6	0.02
SMPD4P1	2.1	6e-05	1.5	0.004	2.0	0.01	2.3	0.005	2.2	1e-04	3.0	9e-05
SOX15	1.3	1e-04	1.0	0.002	1.8	0.02	1.9	0.01	0.9	9e-04	1.2	0.02
SRRM3	2.2	1e-11	2.8	5e-08	1.8	0.03	1.9	0.02	2.1	6e-07	2.4	0.004
SSUT2P8	2.0	5e-07	1.7	9e-05	2.3	0.003	2.0	0.02	2.0	1e-04	2.3	0.004
STAC2	2.4	4e-09	2.8	1e-06	2.3	0.008	2.2	0.02	2.1	4e-05	2.7	0.003
TEX19	3.3	2e-13	4.0	3e-11	3.5	1e-05	2.6	0.02	3.1	3e-07	3.3	0.002
TH	3.0	3e-12	2.8	4e-07	3.3	7e-05	2.8	0.004	3.0	5e-08	3.3	3e-04
TMEM151A	4.1	1e-22	4.1	5e-10	4.0	7e-06	3.8	8e-04	4.1	2e-11	4.3	2e-04
TMEM151B	3.6	1e-15	3.3	2e-07	3.4	8e-05	3.8	2e-04	3.8	3e-11	3.6	3e-04
TMPRSS9	3.1	5e-11	2.8	2e-07	3.3	2e-04	2.3	0.02	3.3	9e-11	3.3	3e-04
TULP1	2.7	6e-10	3.2	3e-09	2.7	0.002	2.5	0.02	2.7	2e-06	2.4	0.01
UTF1	2.4	3e-07	2.2	7e-05	2.7	0.003	2.6	0.008	2.4	8e-05	2.1	0.03
VWA5B2	2.3	8e-08	2.3	5e-06	2.7	3e-05	2.7	2e-04	1.9	6e-05	2.5	4e-04
WFDC2	4.2	1e-24	5.1	2e-09	4.4	5e-07	4.1	0.003	3.6	3e-09	4.5	0.001
WNT16	2.9	8e-10	3.0	3e-08	2.4	0.02	3.0	0.004	3.0	7e-07	3.3	8e-04
YBX2	1.6	3e-13	2.0	2e-08	1.5	8e-04	1.6	0.004	1.6	5e-09	1.4	0.01
ZNF467	1.2	3e-08	1.3	4e-06	1.3	0.006	1.3	0.007	1.0	1e-04	1.1	0.01

ALL: all the other muscle biopsies, NT: histologically normal biopsies, DM: dermatomyositis, AS: antisynthetase syndrome, IBM: inclusion body myositis, IMNM: immune-mediated necrotizing myositis.

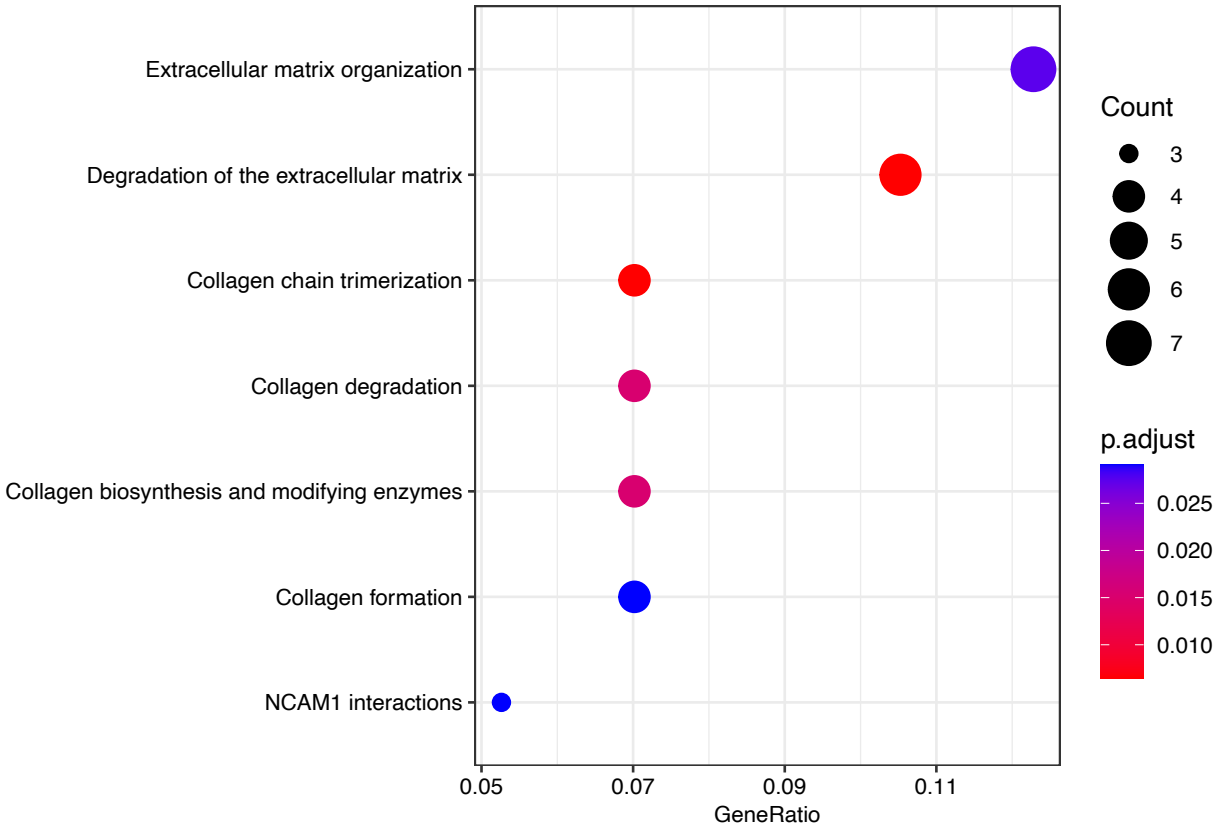
Supplementary Figure 1. Venn diagram showing the number of genes that were differentially overexpressed (q-value < 0.05) in each dermatomyositis subgroup compared to other myositis patients and normal muscle biopsies. DM: dermatomyositis, AS: antisynthetase syndrome, IBM: inclusion body myositis, IMNM: immune-mediated necrotizing myositis, NT: histologically normal biopsies.



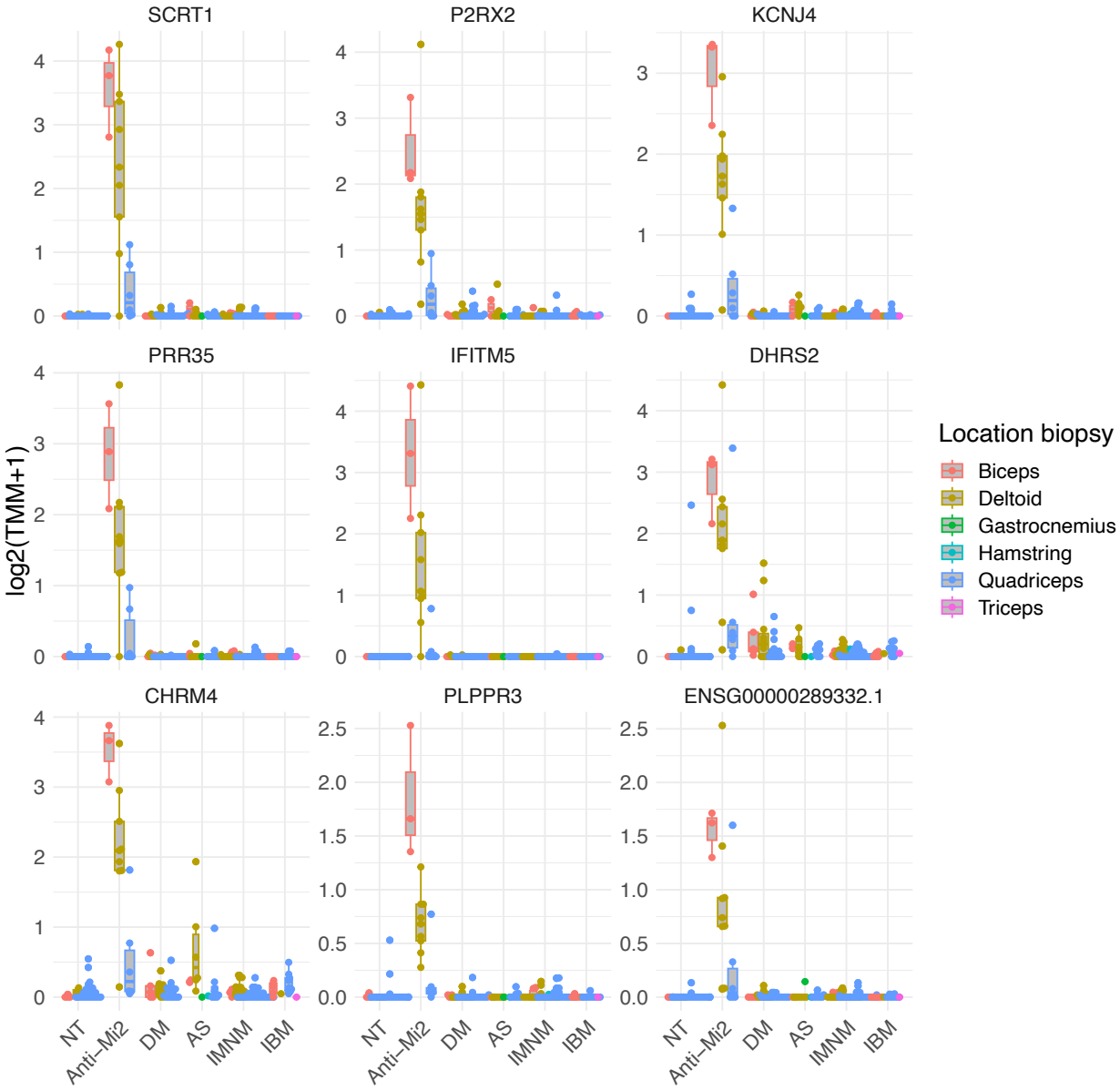
Supplementary Figure 2. Venn diagram showing the number of genes that were differentially underexpressed (q-value < 0.05) in each dermatomyositis subgroup compared to other myositis patients and normal muscle biopsies. DM: dermatomyositis, AS: antisynthetase syndrome, IBM: inclusion body myositis, IMNM: immune-mediated necrotizing myositis, NT: histologically normal biopsies.



Supplementary Figure 3. Pathway enrichment analysis of the set of genes specifically overexpressed in anti-Mi2 using the Reactome database.



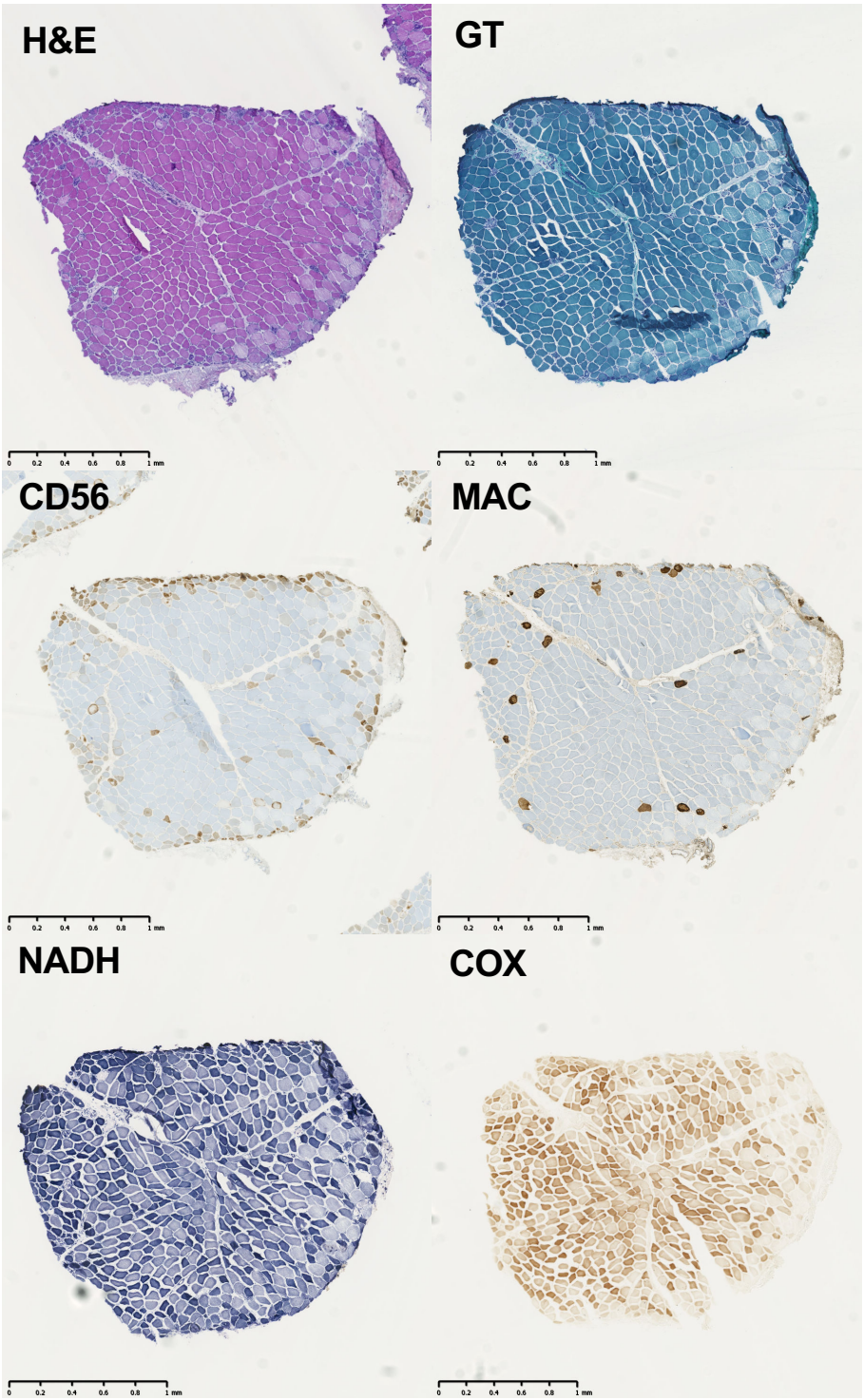
Supplementary Figure 4. Most differentially overexpressed genes in anti-Mi2 dermatomyositis muscle compared to all the other muscle biopsies included in the study according to the location of the muscle biopsy (histologically normal muscle biopsies [NT], non-anti-Mi2 dermatomyositis [DM], antisynthetase syndrome [AS], immune-mediated necrotizing myositis [IMNM], and inclusion body myositis [IBM]).



Supplementary Table 2. Association between the set of anti-Mi2-specific genes and genes previously proposed to be regulated by the CHD4/NuRD complex in skeletal or cardiac muscle using five different publicly available datasets from different animal models deficient in CHD4. CHD4-mck, CHD4-myh6, CHD4-corin, and CHD4-nkx_E10.5 were obtained from PMID 27166947, whereas CHD4-nkx_E9.5, and CHD4-nkx_E10 were obtained from PMID 29891665 (GSE109012). The list of mouse genes was restricted to those having a human homolog and the mouse homologs of the anti-Mi2 specific genes (n=114) were used for the analysis. The composite analysis of upregulated and downregulated genes contains more genes than the global composite analysis because some genes were underexpressed in one dataset and overexpressed in another.

	All the genes			**Upregulated genes**			**Downregulated genes**		
	Mi2	**No Mi2**	**p-value**	**Mi2**	**No Mi2**	**p-value**	**Mi2**	**No Mi2**	**p-value**
Composite	54% (61)	26% (4804)	6e-10	49% (56)	15% (2828)	5e-17	7% (8)	12% (2161)	1
CHD4-mck	18% (20)	7% (1352)	3e-04	17% (19)	5% (827)	1e-06	1% (1)	3% (525)	1
CHD4-myh6	11% (12)	3% (571)	3e-04	9% (10)	2% (351)	8e-05	2% (2)	1% (220)	0.4
CHD4-corin	12% (14)	3% (635)	5e-05	11% (13)	3% (479)	1e-05	1% (1)	1% (156)	0.6
CHD4-nkx_E9.5	32% (37)	15% (2833)	5e-06	28% (32)	8% (1468)	3e-10	4% (5)	7% (1365)	0.9
CHD4-nkx_E10	21% (24)	7% (1254)	8e-07	21% (24)	5% (861)	7e-10	0% (0)	2% (393)	1
CHD4-nkx_E10.5	4% (5)	1% (238)	0.02	4% (5)	1% (217)	0.01	0% (0)	0% (21)	1

Supplementary Figure 5. Representative stainings of the muscle biopsy sections used for Figure 7 and panel B of Figure 6 including H&E (hematoxylin and eosin) for general morphology, Gomori trichrome (GT), NADH (nicotinamide adenine dinucleotide) and COX (cytochrome c oxidase), as well as immunohistochemical stains for CD56 and membrane attack complex (MAC).



Supplementary Figure 6. Result of the confocal immunofluorescence analysis, showing the separate fluorescent channels of the regions of interest depicted in Figure 7. MAdCAM-1 and SCRT1 were visualized in the red channel, while human IgG (IGG) was detected in the green channel. The blue channel corresponds to Hoechst staining, which was used to visualize nuclei, and the magenta channel corresponds to laminin (LMN) staining.