# Supplemental data

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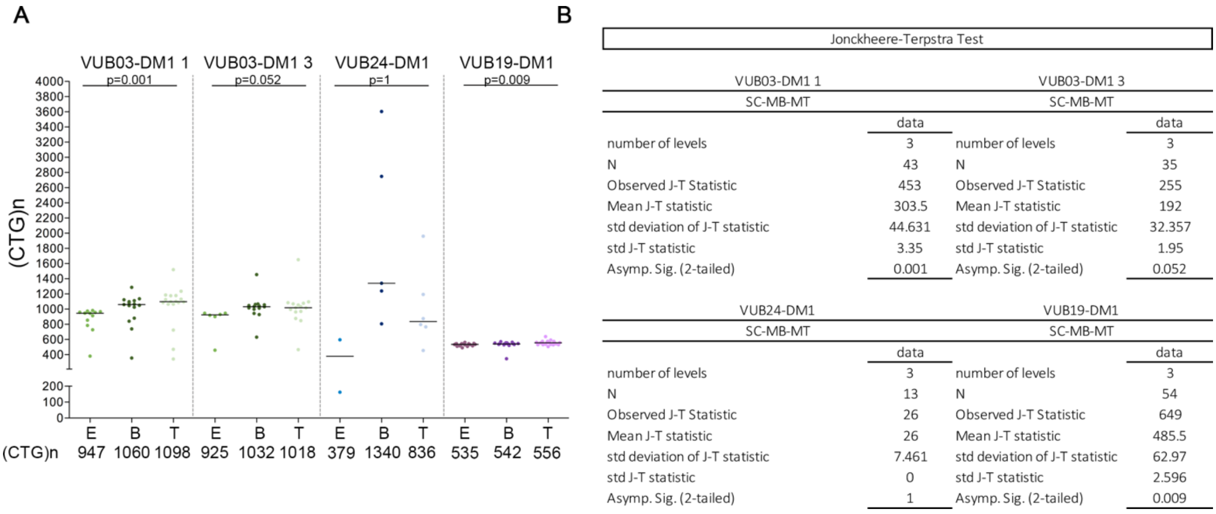


Jonckheere-Terpstra Test

| VUB03-DM1 1<br>SC-MB-MT | data | VUB03-DM1 3<br>SC-MB-MT | data |
|---|---|---|---|
| number of levels | 3 | number of levels | 3 |
| N | 43 | N | 35 |
| Observed J-T Statistic | 453 | Observed J-T Statistic | 255 |
| Mean J-T statistic | 303.5 | Mean J-T statistic | 192 |
| std deviation of J-T statistic | 44.631 | std deviation of J-T statistic | 32.357 |
| std J-T statistic | 3.35 | std J-T statistic | 1.95 |
| Asymp. Sig. (2-tailed) | 0.001 | Asymp. Sig. (2-tailed) | 0.052 |

| VUB24-DM1<br>SC-MB-MT | data | VUB19-DM1<br>SC-MB-MT | data |
|---|---|---|---|
| number of levels | 3 | number of levels | 3 |
| N | 13 | N | 54 |
| Observed J-T Statistic | 26 | Observed J-T Statistic | 649 |
| Mean J-T statistic | 26 | Mean J-T statistic | 485.5 |
| std deviation of J-T statistic | 7.461 | std deviation of J-T statistic | 62.97 |
| std J-T statistic | 0 | std J-T statistic | 2.596 |
| Asymp. Sig. (2-tailed) | 1 | Asymp. Sig. (2-tailed) | 0.009 |

**Fig. S1.** CTG repeat size in the four hESCs and their derived myoblasts and myotubes.

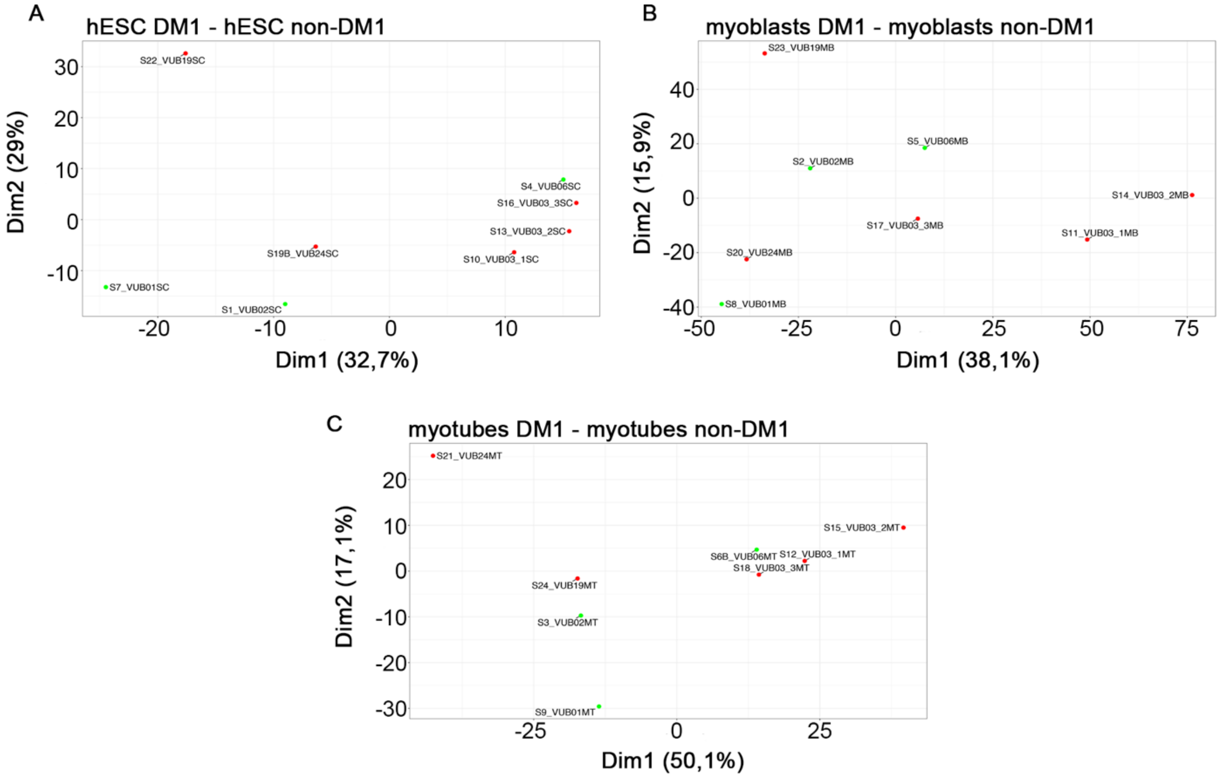
**(A)** The median CTG repeat size is shown for all 3 cell types (hESCs, myoblasts, myotubes) of VUB03-DM1, VUB24-DM1 and VUB19-DM1. Every dot represents the median repeat size of one low input LongAmp PCR reaction, and reflects the repeat size which was dominantly present in that PCR reaction. The horizontal line for every dot cluster represent the median for that cluster. Individual repeat sizes of every dot are presented in **Table S1**. **(B)** The Jonkheere-Terpstra test details. The Jonckheere-Terpstra test was performed on every hESC line separately, including the three different cell types. N is the number of data points.

Abbreviations: E and SC: human embryonic stem cells; B and MB: myoblasts; T and MT: myotubes.

**Table S1.** Individual CTG repeat values for the Fig. S1 and is presented in a separate excel sheet.

PacBio results are shown per individual LongAmp PCR reaction and per DM1 cell line for hESCs, myoblasts and myotubes. Number of reads, maximum and minimum repeat size, median and mean CTG repeat sizes are shown per PCR reaction. The values at the bottom of the rows with the mean and median per PCR reaction indicate the mean or median for the whole sample.

https://figshare.com/s/491658bfa49266b27e9a



**(A)**

**Fig. S2.** Samples cluster together according to cell type not according to DM1 vs non-DM1.

**(A)** Principle component analysis of DM1 hESCs versus non-DM1 hESCs, **(B)** Principle component analysis of DM1 myoblasts versus non-DM1 myoblasts, **(C)** Principle component analysis of DM1 myotubes versus non-DM1 myotubes. Abbreviations: SC: human embryonic stem cells; MB: myoblasts; MT: myotubes.

**Table S2. List of GO pathways related to muscle differentiation commonly or differentially regulated in DM1 or non-DM1 cell lines from human embryonic stem cells to myoblasts.**

Abbreviations: NES: normalized enrichment score

| common in DM1 and nonDM1 | | |
|---|---|---|
| **pathway** | **padj** | **NES** |
| GO_ACTIN_FILAMENT_BASED_PROCESS | 0.01265662 | 1.82755139 |
| GO_ACTIN_FILAMENT_BINDING | 0.01265662 | 1.65965972 |
| GO_ACTIN_FILAMENT_BUNDLE | 0.01265662 | 1.72006638 |
| GO_ACTIN_FILAMENT_ORGANIZATION | 0.01265662 | 1.80606287 |
| GO_ACTIN_MONOMER_BINDING | 0.01265662 | 1.92364276 |
| GO_ACTOMYOSIN | 0.01265662 | 1.92669322 |
| GO_MESODERM_DEVELOPMENT | 0.02718075 | 1.51100596 |
| GO_MESODERM_MORPHOGENESIS | 0.01265662 | 2.09475924 |
| GO_MESODERMAL_CELL_DIFFERENTIATION | 0.02718075 | 1.79060856 |
| GO_MUSCLE_CELL_DEVELOPMENT | 0.03352309 | 1.53029422 |
| GO_MUSCLE_CELL_DIFFERENTIATION | 0.01265662 | 2.00327926 |
| GO_MUSCLE_CELL_MIGRATION | 0.01265662 | 2.0880279 |
| GO_MUSCLE_STRUCTURE_DEVELOPMENT | 0.01265662 | 2.14812435 |
| GO_MUSCLE_TISSUE_DEVELOPMENT | 0.01265662 | 2.11831806 |
| GO_MYOBLAST_DIFFERENTIATION | 0.01265662 | 1.91262265 |
| GO_MYOBLAST_FUSION | 0.01265662 | 1.97777147 |
| GO_POSITIVE_REGULATION_OF_MUSCLE_CELL_DIFFERENTIATION | 0.01265662 | 1.88645592 |
| GO_POSITIVE_REGULATION_OF_MYOBLAST_DIFFERENTIATION | 0.01265662 | 1.95881106 |
| GO_POSITIVE_REGULATION_OF_MYOTUBE_DIFFERENTIATION | 0.02718075 | 1.70879793 |
| GO_POSITIVE_REGULATION_OF_SKELETAL_MUSCLE_TISSUE_DEVELOPMENT | 0.02047867 | 1.74902239 |
| GO_POSITIVE_REGULATION_OF_SMOOTH_MUSCLE_CELL_MIGRATION | 0.01265662 | 1.9709965 |
| GO_POSITIVE_REGULATION_OF_STRIATED_MUSCLE_CELL_DIFFERENTIATION | 0.01265662 | 1.75918725 |
| GO_REGULATION_OF_ACTIN_FILAMENT_BASED_PROCESS | 0.01265662 | 1.71984935 |

| | | |
|---|---|---|
| GO_REGULATION_OF_ACTIN_FILAMENT_DEPOLYMERIZATION | 0.01265662 | 1.76999533 |
| GO_REGULATION_OF_ACTIN_FILAMENT_LENGTH | 0.01265662 | 1.87975112 |
| GO_REGULATION_OF_MYOBLAST_DIFFERENTIATION | 0.01265662 | 1.96570676 |
| GO_REGULATION_OF_SKELETAL_MUSCLE_TISSUE_DEVELOPMENT | 0.01265662 | 1.92787985 |
| GO_REGULATION_OF_SMOOTH_MUSCLE_CELL_PROLIFERATION | 0.01265662 | 1.64533319 |
| GO_SKELETAL_MUSCLE_CELL_DIFFERENTIATION | 0.01265662 | 1.8912858 |
| GO_SKELETAL_MUSCLE_ORGAN_DEVELOPMENT | 0.01265662 | 1.85674546 |
| GO_SMOOTH_MUSCLE_CELL_DIFFERENTIATION | 0.02047867 | 1.72172324 |
| GO_SMOOTH_MUSCLE_TISSUE_DEVELOPMENT | 0.01265662 | 2.10317899 |
| **only in nonDM1** | | |
| **pathway** | **padj** | **NES** |
| GO_MUSCLE_ORGAN_MORPHOGENESIS | 0.02063181 | 1.79042048 |
| GO_RESPONSE_TO_MUSCLE_STRETCH | 0.0271632 | 1.72427596 |
| GO_NEGATIVE_REGULATION_OF_MUSCLE_TISSUE_DEVELOPMENT | 0.0391335 | 1.6233145 |
| GO_POSITIVE_REGULATION_OF_MUSCLE_TISSUE_DEVELOPMENT | 0.0391335 | 1.60740162 |
| **only in DM1** | | |
| **pathway** | **padj** | **NES** |
| GO_MUSCLE_CELL_PROLIFERATION | 0.01265662 | 1.9935163 |
| GO_MUSCLE_CELL_CELLULAR_HOMEOSTASIS | 0.02047867 | 1.92889912 |
| GO_STRUCTURAL_CONSTITUENT_OF_MUSCLE | 0.01265662 | 1.87357571 |
| GO_MYOFILAMENT | 0.02718075 | 1.75936398 |
| GO_MYOFIBRIL_ASSEMBLY | 0.02047867 | 1.70005522 |
| GO_ACTIN_MYOSIN_FILAMENT_SLIDING | 0.04057227 | 1.66507122 |
| GO_POSITIVE_REGULATION_OF_SMOOTH_MUSCLE_CELL_PROLIFERATION | 0.04630565 | 1.56071946 |

## Table S3. List of GO pathways related to muscle differentiation commonly or differentially regulated in DM1 or non-DM1 cell lines from myoblasts to myotubes.

Abbreviations: NES: normalized enrichment score

| **common in DM1 and nonDM1** | | |
|---|---|---|
| **pathway** | **padj** | **NES** |
| GO_ACTIN_FILAMENT_BASED_MOVEMENT | 0.0182555 | 1.84590055 |
| GO_ACTIN_MEDIATED_CELL_CONTRACTION | 0.0181814 | 2.19563341 |
| GO_ACTIN_MYOSIN_FILAMENT_SLIDING | 0.0181814 | 2.17862925 |
| GO_ACTION_POTENTIAL | 0.0182555 | 1.80776418 |
| GO_ACTOMYOSIN_STRUCTURE_ORGANIZATION | 0.02939984 | 1.61938644 |
| GO_MUSCLE_CELL_DEVELOPMENT | 0.01858137 | 1.68050193 |
| GO_MUSCLE_CELL_DIFFERENTIATION | 0.01985494 | 1.63851458 |
| GO_MUSCLE_CONTRACTION | 0.01960765 | 1.8034354 |
| GO_MUSCLE_ORGAN_DEVELOPMENT | 0.02014791 | 1.74404638 |
| GO_MUSCLE_STRUCTURE_DEVELOPMENT | 0.02126039 | 1.68710183 |
| GO_MUSCLE_SYSTEM_PROCESS | 0.01959137 | 1.80356905 |
| GO_MUSCLE_TISSUE_DEVELOPMENT | 0.031786 | 1.53735186 |
| GO_MYOFIBRIL_ASSEMBLY | 0.0181814 | 1.99425034 |
| GO_MYOFILAMENT | 0.0181814 | 1.7783661 |
| GO_MYOSIN_II_COMPLEX | 0.0181814 | 1.85943125 |
| GO_REGULATION_OF_MUSCLE_SYSTEM_PROCESS | 0.0313491 | 1.38870282 |
| GO_SKELETAL_MUSCLE_ORGAN_DEVELOPMENT | 0.01858137 | 1.60400751 |
| GO_STRIATED_MUSCLE_CELL_DIFFERENTIATION | 0.0313491 | 1.54769956 |
| GO_STRIATED_MUSCLE_CONTRACTION | 0.04783482 | 1.51151411 |
| GO_STRUCTURAL_CONSTITUENT_OF_MUSCLE | 0.0181814 | 2.07551241 |
| **only in nonDM1** | | |
| **pathway** | **padj** | **NES** |
| GO_ACTIN_FILAMENT_BUNDLE_ORGANIZATION | 0.02500129 | -1.7082806 |
| GO_MESODERM_MORPHOGENESIS | 0.03669841 | -1.5419403 |
| GO_POSITIVE_REGULATION_OF_ACTIN_FILAMENT_POLYMERIZATION | 0.01180616 | -1.8566163 |
| GO_REGULATION_OF_ACTIN_FILAMENT_LENGTH | 0.01180616 | -1.6299536 |
| GO_ACTIN_FILAMENT_BASED_PROCESS | 0.01180616 | 1.52952967 |
| GO_ACTININ_BINDING | 0.04629302 | 1.63068784 |
| GO_MUSCLE_FIBER_DEVELOPMENT | 0.01180616 | 1.78597005 |
| GO_MUSCLE_HYPERTROPHY | 0.01180616 | 2.02477825 |

| | | |
|---|---|---|
| GO_MYOSIN_COMPLEX | 0.01180616 | 1.76727751 |
| GO_MYOSIN_FILAMENT | 0.01180616 | 2.10635835 |
| GO_POSITIVE_REGULATION_OF_SKELETAL_MUSCLE_TISSUE_DEVELOPMENT | 0.042022 | 1.6967193 |
| GO_REGULATION_OF_ACTIN_FILAMENT_BASED_MOVEMENT | 0.02468595 | 1.76604176 |
| GO_REGULATION_OF_MUSCLE_ADAPTATION | 0.01180616 | 1.75761499 |
| GO_REGULATION_OF_MUSCLE_CELL_DIFFERENTIATION | 0.04185 | 1.41027052 |
| GO_REGULATION_OF_MUSCLE_CONTRACTION | 0.01180616 | 1.826582 |
| GO_REGULATION_OF_MUSCLE_TISSUE_DEVELOPMENT | 0.01916667 | 1.60113238 |
| GO_REGULATION_OF_SKELETAL_MUSCLE_TISSUE_DEVELOPMENT | 0.04496701 | 1.54391244 |
| GO_REGULATION_OF_STRIATED_MUSCLE_CONTRACTION | 0.01180616 | 1.84471796 |
| GO_SKELETAL_MUSCLE_CELL_DIFFERENTIATION | 0.01180616 | 2.02984286 |
| GO_SKELETAL_MUSCLE_CONTRACTION | 0.01180616 | 1.99822543 |
| GO_STRIATED_MUSCLE_ADAPTATION | 0.02468595 | 1.88461344 |
| GO_ALPHA_ACTININ_BINDING | 0.04619133 | 1.64472534 |
| **only in DM1** | | |
| **pathway** | **padj** | **NES** |
| GO_MUSCLE_MYOSIN_COMPLEX | 0.0181814 | 2.03464647 |

## Table S4. Genes included in the heatmap in Figure 1E.

This heatmap includes the differential gene expression from hESC to Myoblast for the genes involved in the development of skeletal muscle, as listed in the Hallmark Myogenesis library of GSEA. The table and heatmap only include values for genes with an FDR<0.05. FC: log2 fold change, FDR: false discovery rate.

| | **Non DM1** | | **DM1** | |
|---|---|---|---|---|
| **Gene** | **Log2FC** | **FDR** | **Log2FC** | **FDR** |
| *PGAM2* | -7.6364474 | 0.00050996 | -4.1372501 | 0.00228038 |
| *ACTN3* | -7.1062215 | 0.00169109 | -4.9963056 | 0.00141402 |
| *VIPR1* | -6.684418 | 0.00011729 | -6.5755281 | 1.1054E-05 |
| *GNAO1* | -6.1910983 | 4.1641E-05 | -6.0808602 | 1.0679E-07 |
| *ACTA1* | -5.3121312 | 0.00459045 | | |
| *KCNH2* | -5.1236904 | 0.00183986 | -2.6026062 | 0.01207283 |
| *MYH2* | -5.0635294 | 0.00520164 | -2.0813847 | 0.02477974 |
| *FXYD1* | -4.8936168 | 0.00169415 | | |
| *PTGIS* | -4.5349603 | 0.00461408 | -3.3097771 | 0.00055805 |
| *SORBS1* | -4.204472 | 5.6523E-05 | -3.193996 | 1.3774E-06 |
| *ERBB3* | -3.8512355 | 0.00043801 | | |
| *HRC* | -3.6297947 | 0.01160065 | | |
| *MYOZ1* | -3.482335 | 0.00717859 | -2.6183037 | 0.02848058 |
| *CACNA1H* | -3.411508 | 0.00461981 | -2.6953797 | 0.00109621 |
| *PYGM* | -3.3522263 | 0.01775395 | | |
| *GJA5* | -3.0896497 | 0.00474154 | -1.8749708 | 0.04047632 |
| *CKB* | -2.8788679 | 0.00326882 | | |
| *FGF2* | -2.5262915 | 0.00955584 | -3.2596399 | 0.00104467 |
| *CLU* | -2.3593071 | 0.01343509 | -1.3162698 | 0.0061474 |
| *SOD3* | -2.2542128 | 0.02673005 | | |
| *PTP4A3* | -2.1849369 | 0.00643007 | -1.1815027 | 0.04094168 |
| *SLC6A8* | -2.0690044 | 0.0002861 | -1.6167478 | 1.3472E-05 |
| *FKBP1B* | -2.0080864 | 0.0059866 | -2.0506157 | 5.2646E-06 |
| *SH3BGR* | -1.8581192 | 0.01436419 | -1.7977259 | 0.00025212 |
| *GPX3* | -1.8481428 | 0.03745818 | -2.3940478 | 0.00012671 |
| *FOXO4* | -1.826665 | 0.0068506 | -1.6908778 | 0.00308057 |
| *CFD* | -1.7590589 | 0.03498819 | -2.2272838 | 0.00151544 |
| *LARGE1* | -1.3101683 | 0.00102905 | -0.9173283 | 0.00416748 |
| *NOS1* | -1.2629876 | 0.02469434 | | |
| *LAMA2* | -1.2051618 | 0.02739755 | | |
| *HDAC5* | -1.1630103 | 0.00354764 | -0.8288821 | 0.00606047 |
| *ABLIM1* | -1.1609756 | 0.0013776 | | |
| *MYH11* | -1.1157403 | 0.03127347 | | |
| *FABP3* | -1.0708169 | 0.04014091 | | |
| *OCEL1* | -0.9854368 | 0.02040891 | -0.942291 | 0.00134744 |
| *DTNA* | -0.9735365 | 0.01792914 | -0.7680321 | 0.02640046 |

| | | | | |
|---|---|---|---|---|
| *AGL* | -0.6869365 | 0.03243653 | | |
| *PFKM* | -0.4816099 | 0.03997276 | | |
| *SCD* | -0.4237087 | 0.04357237 | | |
| *BAG1* | 0.52291356 | 0.03661361 | | |
| *TPM3* | 0.5531979 | 0.02759006 | | |
| *SH2B1* | 0.63269704 | 0.01812807 | 0.65558606 | 0.00456142 |
| *SPTAN1* | 0.6420548 | 0.02553393 | 0.8783119 | 0.00133428 |
| *GABARAPL2* | 0.76464736 | 0.00303357 | 0.79314506 | 0.0011554 |
| *ITGB5* | 0.84040954 | 0.0433597 | 0.64552128 | 0.00101551 |
| *DMPK* | 0.93892448 | 0.03801914 | 2.33839218 | 0.00481972 |
| *CNN3* | 0.95240685 | 0.00765872 | 0.8327663 | 0.00028287 |
| *SORBS3* | 0.96911095 | 0.00389133 | 0.70863702 | 0.00207322 |
| *MEF2A* | 1.03887422 | 0.00189322 | 1.67415001 | 0.00167855 |
| *HSPB8* | 1.06612798 | 0.0461395 | | |
| *MAPK12* | 1.1287699 | 0.00696582 | 0.71548276 | 0.00924735 |
| *MEF2D* | 1.21509755 | 0.00035714 | 1.45068928 | 0.00044345 |
| *CHRNB1* | 1.28042522 | 0.0279838 | 2.13804309 | 0.01104284 |
| *PKIA* | 1.30821332 | 0.00351101 | 1.57559488 | 6.2214E-05 |
| *MRAS* | 1.35782071 | 0.00285817 | 2.66211884 | 0.00114645 |
| *PLXNB2* | 1.42669337 | 0.00073714 | 1.30189246 | 1.2604E-05 |
| *RB1* | 1.50245783 | 0.00088596 | 1.37371906 | 6.5545E-05 |
| *CRAT* | 1.54450558 | 0.00122494 | 1.81393102 | 1.8562E-05 |
| *ACSL1* | 1.54936383 | 0.03491145 | 1.66672016 | 0.00069319 |
| *PPP1R3C* | 1.63281955 | 0.03958486 | 1.77194249 | 0.00447141 |
| *MYO1C* | 1.7080987 | 0.00099124 | 1.25387419 | 2.4556E-05 |
| *KLF5* | 1.74023751 | 0.00602375 | 2.91415737 | 0.00127289 |
| *GADD45B* | 1.74119308 | 0.00469833 | 1.49723194 | 0.01308782 |
| *AK1* | 1.78885198 | 0.00087464 | 1.99424536 | 0.00022705 |
| *BDKRB2* | 1.82705386 | 0.03652561 | 2.40704318 | 0.00141779 |
| *STC2* | 1.84520704 | 0.01910589 | 2.09760189 | 0.01258574 |
| *BIN1* | 1.88045433 | 0.00025973 | 2.67073101 | 0.0012023 |
| *AEBP1* | 1.88665576 | 0.00077659 | 1.4746003 | 0.00698089 |
| *IGFBP3* | 1.91990724 | 0.01449659 | 2.58270264 | 0.00037647 |
| *LPIN1* | 2.04999279 | 0.00011903 | 2.00339165 | 8.1572E-06 |
| *NCAM1* | 2.25890788 | 0.00478228 | 3.93859662 | 0.00028913 |
| *NQO1* | 2.30884621 | 0.00069818 | 2.07424127 | 0.00019916 |
| *SGCA* | 2.37896995 | 0.02383693 | 4.62772958 | 0.01354487 |
| *SIRT2* | 2.45797159 | 0.00028937 | 3.5285191 | 0.00035307 |
| *MYH3* | 2.50843605 | 0.02332573 | 6.95308417 | 0.00703814 |
| *PDLIM7* | 2.72154165 | 7.5496E-05 | 2.64355517 | 1.7823E-06 |
| *TPM2* | 2.80871992 | 0.00078469 | 2.93465456 | 8.1347E-06 |
| *MYLK* | 2.83151781 | 0.00443069 | 2.76635452 | 1.8483E-05 |
| *CTF1* | 2.87495473 | 0.00077659 | 2.47213046 | 0.00018835 |

| | | | | |
|---|---|---|---|---|
| *MYH9* | 2.94513142 | 0.00015746 | 2.49330023 | 4.8667E-06 |
| *SVIL* | 3.01375511 | 0.00048534 | 2.95019569 | 7.4847E-08 |
| *COL4A2* | 3.04453224 | 0.00090295 | 2.78856241 | 5.8933E-05 |
| *TAGLN* | 3.06344378 | 0.00107551 | 1.87855956 | 0.00392026 |
| *ITGB1* | 3.0831708 | 0.00017315 | 2.60753692 | 7.4703E-06 |
| *GSN* | 3.09622146 | 0.0006844 | 3.52197322 | 3.2212E-05 |
| *SPARC* | 3.13473222 | 0.00019542 | 2.77060242 | 8.6243E-06 |
| *COL6A2* | 3.13819281 | 0.00077529 | 2.44537108 | 1.3031E-06 |
| *COL15A1* | 3.16355938 | 0.01475994 | 5.30371238 | 0.00108984 |
| *COL1A1* | 3.18915404 | 0.00104841 | 3.20779302 | 0.00025212 |
| *EPHB3* | 3.20310055 | 0.01904683 | 4.43428353 | 0.00073895 |
| *SMTN* | 3.27096114 | 0.00020537 | 2.63538877 | 1.0895E-05 |
| *MYLPF* | 3.31537411 | 0.01292004 | 7.21912669 | 0.00660378 |
| *SSPN* | 3.49300238 | 0.00029642 | 4.18483126 | 7.3464E-05 |
| *TGFB1* | 3.61486094 | 0.00163264 | 3.59273433 | 2.0195E-06 |
| *WWTR1* | 3.67345661 | 5.9126E-05 | 3.61272202 | 1.0047E-06 |
| *MYBPH* | 3.77887647 | 0.02689955 | 9.05159429 | 0.00545013 |
| *PRNP* | 3.96818459 | 0.00032957 | 3.45581746 | 1.0663E-05 |
| *KCNH1* | 3.97792156 | 0.00032767 | 4.612112 | 1.0941E-05 |
| *MEF2C* | 4.23754976 | 0.00011903 | 7.50750712 | 0.00047727 |
| *KIFC3* | 4.46366974 | 0.00106394 | 4.0230589 | 4.1261E-07 |
| *SPHK1* | 4.50026891 | 0.00011983 | 3.76449516 | 0.00044345 |
| *CDKN1A* | 4.51078872 | 0.00093463 | 3.45718467 | 0.00192736 |
| *HBEGF* | 4.53786655 | 0.00278658 | 3.48187621 | 0.00141428 |
| *LSP1* | 4.83654505 | 0.00989165 | 5.39217829 | 0.00094403 |
| *BHLHE40* | 4.86953754 | 0.00055626 | 3.84464037 | 2.5057E-05 |
| *MYOG* | 5.60690782 | 0.01467014 | 9.67484023 | 0.00227968 |
| *COL6A3* | 6.37157841 | 0.00048213 | 6.10605019 | 1.3089E-07 |
| *ADAM12* | 6.50025487 | 0.00017569 | 6.86283676 | 2.0486E-07 |
| *CDH13* | 6.52977417 | 0.00034417 | 6.0534943 | 1.6907E-06 |
| *SGCD* | 6.60504238 | 9.2577E-05 | 8.31569881 | 0.00036429 |
| *IGFBP7* | 7.86491015 | 0.00040475 | 7.40743712 | 1.35E-06 |
| *COL3A1* | 8.18258098 | 0.0037429 | 8.7579506 | 3.229E-06 |
| *MYL4* | 9.10699754 | 0.00855629 | 7.5034369 | 0.00370619 |
| *CHRNA1* | 9.43800137 | 9.337E-05 | 10.3710038 | 0.00016395 |

## Table S5. Genes included in the heatmap in Figure 2D.

This heatmap includes the differential gene expression from Myoblast to Myotube for the genes involved in the development of skeletal muscle, as listed in the Hallmark Myogenesis library of GSEA. The table and heatmap only include values for genes with an FDR<0.05. FC: log2 fold change, FDR: false discovery rate.

| | **Non DM1** | | **DM1** | |
|---|---|---|---|---|
| **Gene** | **Log2FC** | **FDR** | **Log2FC** | **FDR** |
| *LAMA2* | 3.434466542 | 0.004964119 | 2.558175579 | 0.002277686 |
| *COL3A1* | 2.969467392 | 0.042640266 | 2.955398545 | 0.003068552 |
| *DMD* | 2.767419811 | 0.009233199 | 2.785701929 | 0.003537978 |
| *SORBS1* | 3.356207514 | 0.010852453 | 2.837099843 | 0.004705953 |
| *IGF1* | 7.961961829 | 0.021266006 | 6.601104416 | 0.005055313 |
| *ABLIM1* | 2.032069952 | 0.007201063 | 1.303012396 | 0.006641053 |
| *GJA5* | 7.350950766 | 0.002335051 | 5.903106902 | 0.011650128 |
| *TSC2* | 0.588021565 | 0.022460709 | 0.513039783 | 0.011790923 |
| *ITGB5* | -1.029597026 | 0.026519573 | -1.095285801 | 0.011876489 |
| *CRYAB* | 4.338175471 | 0.034820232 | 4.160120198 | 0.012695223 |
| *KCNH2* | 6.514672136 | 0.005838995 | 4.339128578 | 0.012740782 |
| *FABP3* | 3.917470453 | 0.018246247 | 2.844873071 | 0.014690511 |
| *NQO1* | -1.585215391 | 0.011538682 | -1.701832158 | 0.016059109 |
| *SSPN* | 2.325759034 | 0.008582274 | 1.854418399 | 0.016346349 |
| *HSPB2* | 7.605691691 | 0.007093457 | 5.701880592 | 0.016384447 |
| *AGL* | 2.19265002 | 0.010213886 | 2.202678156 | 0.01644433 |
| *TGFB1* | -1.4248959 | 0.01054286 | -1.499853944 | 0.016523106 |
| *CKMT2* | 5.510813574 | 0.007047628 | 4.359050833 | 0.019705103 |
| *CD36* | 6.080426272 | 0.00316201 | 4.824471292 | 0.019878718 |
| *TCAP* | 3.558564383 | 0.032768447 | 3.69826496 | 0.021562269 |
| *ACHE* | 2.587647573 | 0.012991014 | 2.852586981 | 0.023164772 |
| *IGFBP3* | 2.158494815 | 0.038977168 | 2.034892733 | 0.024755399 |
| *SGCG* | 5.431689296 | 0.008587939 | 3.852361426 | 0.025090027 |
| *SPTAN1* | 0.891044981 | 0.026626872 | 1.02850752 | 0.025130091 |
| *MYL6B* | 2.271145922 | 0.005809485 | 2.011751398 | 0.025749356 |
| *FST* | 2.397183778 | 0.027555253 | 2.57317549 | 0.02608245 |
| *NAV2* | 0.932944466 | 0.043370045 | 1.452889679 | 0.02704499 |
| *FOXO4* | 2.35168128 | 0.01166004 | 2.003659929 | 0.027181795 |
| *PSEN2* | 2.464107869 | 0.010811335 | 2.214830878 | 0.02744485 |
| *SVIL* | 0.90121126 | 0.047568627 | 0.983028954 | 0.029391722 |
| *SPHK1* | -2.711792186 | 0.014526251 | -2.363562269 | 0.033438218 |
| *SLN* | 6.689599383 | 0.020011912 | 5.239655295 | 0.034400081 |
| *STC2* | -1.878642266 | 0.021167168 | -1.401806338 | 0.034646023 |
| *TNNC2* | 9.265908784 | 0.003587066 | 5.654457864 | 0.036055991 |
| *CACNG1* | 12.8305577 | 0.002038081 | 5.851439514 | 0.036566693 |
| *KLF5* | 3.317568511 | 0.005502216 | 2.267084435 | 0.036849485 |

| | | | | |
|---|---|---|---|---|
| *MAPRE3* | 2.703345609 | 0.008206323 | 1.800803074 | 0.037299158 |
| *EIF4A2* | 0.538257268 | 0.015001838 | 0.523123553 | 0.03787702 |
| *COX6A2* | 7.460777533 | 0.026418018 | 4.278824389 | 0.038200662 |
| *PKIA* | 1.719028816 | 0.011076449 | 1.523168325 | 0.038819486 |
| *LDB3* | 6.076404982 | 0.004860348 | 3.719347159 | 0.039278995 |
| *SORBS3* | -1.001467484 | 0.02041369 | -0.550195828 | 0.041575078 |
| *SPEG* | 1.56457793 | 0.047736298 | 1.266826569 | 0.042457936 |
| *CAMK2B* | 3.697435383 | 0.021131387 | 2.620925713 | 0.04257657 |
| *TPM3* | -1.116836487 | 0.007923713 | -0.806040209 | 0.043181949 |
| *PRNP* | -0.958295281 | 0.043240439 | -0.911032826 | 0.04653724 |
| *CSRP3* | 5.176373525 | 0.015988941 | 3.822859929 | 0.049102734 |
| *MYL1* | 8.690728906 | 0.005249948 | 4.997857263 | |
| *ITGB1* | -1.32925361 | 0.012021653 | -0.956007405 | |
| *MYOM1* | 8.406506963 | 0.00437319 | 4.64701646 | |
| *MYOM2* | 6.149523336 | 0.009035857 | 3.951858259 | |
| *FXYD1* | 5.844446055 | 0.015558367 | 3.8650738 | |
| *TNNI2* | 8.721228713 | 0.00383752 | 4.527387147 | |
| *MRAS* | 2.93920966 | 0.002788993 | 1.814355448 | |
| *PFKM* | 0.53224965 | 0.029222214 | 0.651726107 | |
| *MYH7* | 7.370984742 | 0.010366123 | 4.306603954 | |
| *NCAM1* | 3.693855646 | 0.008582274 | 2.021400055 | |
| *DTNA* | 1.390336879 | 0.008571628 | 1.230408857 | |
| *DES* | 7.034624445 | 0.004698949 | 3.991609377 | |
| *CASQ2* | 8.629082168 | 0.005614601 | 4.269249435 | |
| *REEP1* | 4.211069025 | 0.021167168 | 2.801647477 | |
| *TNNT3* | 9.110154294 | 0.006431548 | 4.377343417 | |
| *MYL4* | 6.992537157 | 0.006402841 | 3.650003756 | |
| *MYH3* | 9.621307216 | 0.006432159 | 4.201878419 | |
| *ATP2A1* | 4.708730253 | 0.016644763 | 3.365619922 | |
| *ERBB3* | 5.219661863 | 0.003163749 | 2.760341145 | |
| *HRC* | 8.424311718 | 0.004502011 | 3.651198099 | |
| *CKM* | 10.24258556 | 0.004458363 | 4.58463117 | |
| *MEF2C* | 5.093576545 | 0.003993906 | 2.832725766 | |
| *ACTA1* | 8.031125266 | 0.002788993 | 3.735337947 | |
| *PTP4A3* | 1.812624089 | 0.038026673 | 1.23971904 | |
| *PC* | 1.488430115 | 0.039138238 | 1.195889434 | |
| *MYLPF* | 8.283638421 | 0.00437319 | 3.739768021 | |
| *CHRNG* | 5.137513774 | 0.007488854 | 2.445679116 | |
| *ACTN2* | 7.826633701 | 0.003473675 | 3.651630753 | |
| *MYH9* | -1.273563434 | 0.016945518 | -0.851558215 | |
| *MAPK12* | -0.482987897 | 0.033912329 | -0.528816105 | |
| *CAV3* | 7.610257638 | 0.003427943 | 3.225843372 | |
| *RYR1* | 6.04432954 | 0.014192291 | 3.044566438 | |

| | | | |
|---|---|---|---|
| *BIN1* | 1.795943956 | 0.008310537 | 1.383843173 |
| *SGCD* | 3.14043155 | 0.00437319 | 2.166361257 |
| *SGCA* | 4.713782962 | 0.00494004 | 2.798394202 |
| *SIRT2* | 1.737372248 | 0.006447267 | 1.332688915 |
| *PPFIA4* | 3.497203613 | 0.01497645 | 1.830181151 |
| *ENO3* | 4.48638605 | 0.013809202 | 2.260413772 |
| *LPIN1* | -0.542167579 | 0.019825178 | -0.382264166 |
| *MYH8* | 10.04747762 | 0.007033353 | 3.832054535 |
| *COL15A1* | 4.448857219 | 0.006614878 | 1.525656537 |
| *TNNT1* | 3.566188914 | 0.031091592 | 1.998472503 |
| *TNNT2* | 5.315065188 | 0.026132879 | 2.616352844 |
| *DMPK* | 1.680564158 | 0.005490022 | 1.129093845 |
| *TNNC1* | 7.914029201 | 0.002599395 | 2.770542097 |
| *ACTC1* | 6.249879394 | 0.011480179 | 2.758916827 |
| *TNNI1* | 6.175935646 | 0.016910263 | 2.458983265 |
| *CKB* | 3.613770376 | 0.008629059 | 1.788170616 |
| *MYBPH* | 8.177178805 | 0.009033672 | 2.769047414 |
| *CHRNB1* | 1.533927426 | 0.03392957 | 1.168247921 |
| *MYOG* | 6.047326723 | 0.018906755 | 2.385511018 |
| *MEF2D* | 0.522128005 | 0.015614808 | 0.391114879 |
| *SMTN* | -0.985283507 | 0.03666777 | -0.18946943 |

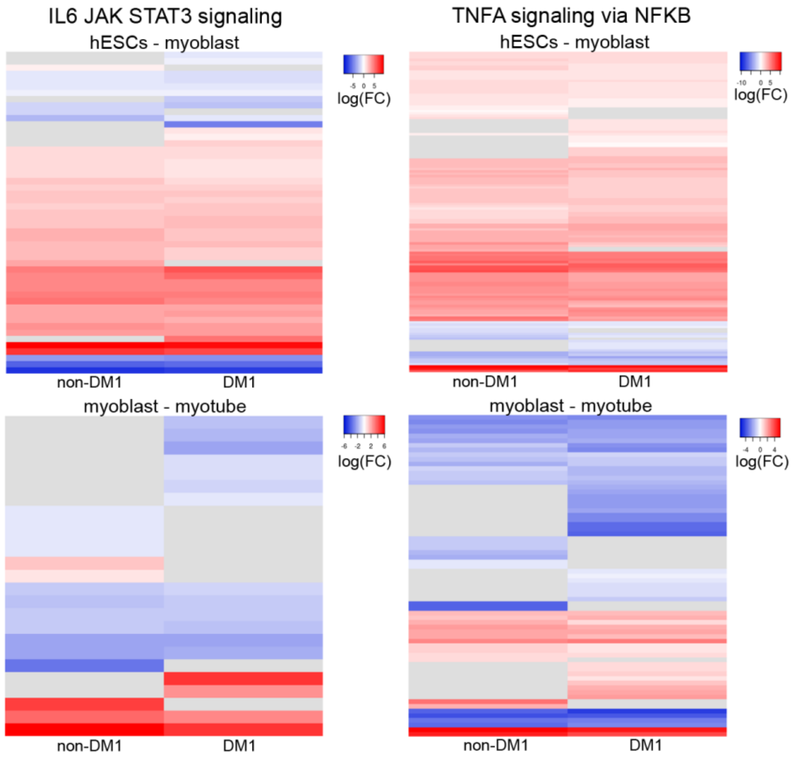


**Fig. S3.** Heatmaps representing the log fold change of genes belonging to IL6 JAK STAT3 signaling, and TNFA signaling via NFKB pathways.

Grey lines indicate genes with FDR>0.05. Abbreviation: NES: normalized enrichment score; FC: fold change.

# Table S6. Genes included in the heatmaps in Figure 3B and Fig. S3.

Genes in the heatmap including the differential gene expression from stem cell to myoblast for the genes involved in the interferon alfa response, as listed in the Hallmark Interferon Alfa Response library of GSEA. The table and heatmap only include values for genes with an FDR<0.05. FC: log2 fold change, FDR: false discovery rate.

| | **Non DM1** | | **DM1** | |
|---|---|---|---|---|
| **Gene** | **Log2FC** | **FDR** | **Log2FC** | **FDR** |
| *RTP4* | -9.8710063 | 0.00293264 | -9.1145961 | 6.7332E-06 |
| *BST2* | -5.6689928 | 0.00255575 | -3.711485 | 0.00012479 |
| *C1S* | -3.6145526 | 0.00392271 | -3.0921577 | 0.00013651 |
| *IFITM1* | -3.783782 | 0.00072326 | -2.6419353 | 9.8759E-05 |
| *DHX58* | -2.6405175 | 0.00193729 | -2.6029477 | 0.00179974 |
| *BATF2* | -3.673551 | 0.0026175 | -2.3881501 | 0.00069974 |
| *IFI30* | -2.6087761 | 0.00148313 | -2.033113 | 0.00088395 |
| *MX1* | | | -1.2735789 | 0.01345829 |
| *OAS1* | -0.9674436 | 0.00665148 | -1.1838165 | 0.00029696 |
| *CD74* | -1.9637571 | 0.00468261 | -1.1216428 | 0.0034503 |
| *CCRL2* | | | -0.9619376 | 0.03405482 |
| *TRIM14* | -1.0517142 | 0.01177066 | -0.838942 | 0.00210212 |
| *ADAR* | -0.4538553 | 0.04326151 | -0.5490263 | 0.02580798 |
| *PNPT1* | | | -0.5315989 | 0.00552356 |
| *NUB1* | 0.52746854 | 0.0226403 | 0.32253053 | 0.01901764 |
| *CMTR1* | 0.31552143 | 0.0556944 | 0.35378361 | 0.00860065 |
| *LAP3* | 0.5442846 | 0.01821382 | 0.39143634 | 0.01018341 |
| *SLC25A28* | 0.66935904 | 0.03862344 | 0.50214236 | 0.00673699 |
| *OGFR* | | | 0.54021449 | 0.00991492 |
| *HLA-C* | | | 0.63838056 | 0.04410707 |
| *PARP12* | 0.89486011 | 0.01799961 | 0.69650345 | 0.00106813 |
| *TRIM5* | 0.86991497 | 0.00539115 | 0.69817181 | 0.01045006 |
| *TRIM25* | 0.72164805 | 0.00253848 | 0.76537427 | 0.00103142 |
| *NCOA7* | | | 0.8193937 | 0.0001103 |
| *CNP* | 0.91720551 | 0.00075643 | 0.93802208 | 9.2195E-05 |
| *TDRD7* | 0.97659574 | 0.03256812 | 0.98949348 | 0.00395936 |
| *MOV10* | | | 0.99001671 | 0.00089653 |
| *PARP14* | | | 1.03639898 | 0.00016168 |
| *IFIH1* | | | 1.07481521 | 0.04631453 |
| *PSME2* | 1.03072437 | 0.00182484 | 1.16555153 | 0.00026052 |
| *CASP8* | 1.68294852 | 0.00618372 | 1.18602132 | 0.00456825 |
| *TAP1* | 1.02090412 | 0.01523837 | 1.21394533 | 0.00147742 |
| *ELF1* | 0.95741278 | 0.03258727 | 1.3127996 | 4.2495E-06 |
| *IRF2* | 1.2394834 | 0.00328061 | 1.35173854 | 0.000708 |
| *STAT2* | 1.31630803 | 0.00361701 | 1.48633341 | 1.6408E-05 |
| *PSME1* | 1.04515988 | 0.00389609 | 1.52652453 | 0.00044223 |

| | | | | |
|---|---|---|---|---|
| *CD47* | 1.69648427 | 0.00107538 | 1.60142131 | 0.00011415 |
| *LAMP3* | 1.90135588 | 0.00602392 | 1.60559618 | 9.7329E-05 |
| *EPSTI1* | | | 1.64657152 | 0.00390111 |
| *IL4R* | 1.69524039 | 0.00088818 | 1.65641998 | 5.886E-05 |
| *LGALS3BP* | 1.81575474 | 0.00880171 | 1.84981875 | 0.00021108 |
| *LPAR6* | -2.1040376 | 0.00247152 | 1.8846873 | 0.00049018 |
| *DDX60* | 1.21449143 | 0.02815044 | 1.92595807 | 0.0004263 |
| *PSMB8* | 1.45747843 | 0.00678832 | 1.97870711 | 7.9371E-05 |
| *SP110* | 1.90136616 | 0.00273328 | 2.01496921 | 3.2379E-06 |
| *OASL* | | | 2.02315681 | 0.00013915 |
| *CMPK2* | 1.69994872 | 0.02669673 | 2.13970382 | 0.00022211 |
| *IFI35* | 1.66901342 | 0.02967756 | 2.29758269 | 0.00054942 |
| *IRF9* | 1.95929158 | 0.00064738 | 2.40028118 | 1.0354E-05 |
| *IL15* | 2.77078982 | 0.01500403 | 2.47188007 | 2.5073E-05 |
| *PSMB9* | | | 2.48831084 | 0.0018983 |
| *PARP9* | 2.17773954 | 0.00223631 | 2.82072021 | 1.4696E-05 |
| *RSAD2* | | | 2.87013131 | 0.00081155 |
| *CSF1* | 3.27242899 | 0.00057967 | 2.89144771 | 9.9354E-05 |
| *TENT5A* | 3.28138347 | 0.00159613 | 3.39027052 | 1.0692E-05 |
| *CXCL10* | 4.05629843 | 0.01356744 | 3.45304179 | 0.00127289 |
| *CXCL11* | 4.05629843 | 0.01356744 | 3.45304179 | 0.00127289 |
| *IRF1* | 3.11318206 | 0.00071388 | 3.51620023 | 2.9883E-07 |
| *B2M* | 3.41653854 | 0.00029518 | 3.79500874 | 4.6206E-06 |
| *IL7* | 4.26428611 | 4.1641E-05 | 4.14466523 | 3.2372E-07 |
| *IFIT3* | 3.44806214 | 0.00396568 | 4.14775246 | 2.8538E-06 |
| *IFIT2* | 3.24152066 | 0.01379642 | 4.24433006 | 4.4988E-06 |
| *GMPR* | 2.95956246 | 0.0048119 | 4.43908047 | 7.0662E-05 |
| *SAMD9* | 5.00413302 | 0.00100066 | 5.33463933 | 9.1734E-05 |
| *IFI27* | 4.68177109 | 0.00025745 | 5.34058862 | 8.8438E-07 |
| *UBA7* | 4.7775481 | 0.00262178 | 6.19323937 | 4.1436E-06 |
| *IFI44L* | 7.74855836 | 0.00460375 | 7.66551219 | 5.7307E-05 |
| *SAMD9L* | 8.38014829 | 0.00112005 | 7.69210855 | 2.9942E-05 |
| *GBP2* | 6.29738047 | 0.00556511 | 8.21587074 | 1.6461E-05 |
| *GBP4* | 8.02976632 | 4.1641E-05 | 8.87883336 | 3.3853E-08 |
| *IFI44* | 10.4740058 | 0.00215472 | 10.6670916 | 6.1887E-05 |
| *SELL* | -1.086175 | 0.00286981 | | |
| *TRIM21* | 0.84855874 | 0.01841346 | | |
| *USP18* | 1.68356502 | 0.01309787 | | |

Genes in the heatmap including the differential gene expression from myoblast to myotube for the genes involved in the interferon alfa response, as listed in the Hallmark Interferon Alfa Response library of GSEA. The table and heatmap only include values for genes with an FDR<0.05. FC: log2 fold change, FDR: false discovery rate.

| | **Non DM1** | | **DM1** | |
|---|---|---|---|---|
| **Gene** | **Log2FC** | **FDR** | **Log2FC** | **FDR** |
| *TRIM25* | -1.0161334 | 0.00407008 | -1.2705931 | 0.00127928 |
| *PSMA3* | -0.5968589 | 0.01195124 | -0.6457465 | 0.00145744 |
| *CD47* | -0.8292759 | 0.01368849 | -0.8668103 | 0.03670892 |
| *GMPR* | 2.0767409 | 0.01461171 | | |
| *USP18* | 1.02658003 | 0.0431299 | | |
| *TRIM21* | -0.5586301 | 0.04649472 | | |
| *IL15* | | | -1.2110939 | 0.00660677 |
| *IRF1* | | | -2.2835597 | 0.00754558 |
| *PARP12* | | | -1.0440708 | 0.00886797 |
| *CASP8* | | | -0.8250892 | 0.00955576 |
| *PSME2* | | | -0.9304172 | 0.01070195 |
| *IL4R* | | | -0.8773788 | 0.01497536 |
| *OGFR* | | | -0.5435969 | 0.02437785 |
| *IFI35* | | | -1.6916088 | 0.02590576 |
| *PSMB8* | | | -1.3500904 | 0.02745125 |
| *IFIT3* | | | -1.4702349 | 0.0295467 |
| *MVB12A* | | | -0.3519241 | 0.0317893 |
| *PLSCR1* | | | -0.8445372 | 0.03327363 |
| *LGALS3BP* | | | -1.3929268 | 0.03612207 |
| *EPSTI1* | | | 1.54308177 | 0.03908688 |
| *SP110* | | | -0.6331772 | 0.04014417 |
| *CMPK2* | | | -1.0556658 | 0.04543234 |
| *C1S* | | | 2.65304697 | 0.04649819 |
| *MOV10* | | | -0.6852287 | 0.0495637 |

Genes in the heatmap including the differential gene expression from myoblast to myotube for the genes involved in the mTORC1 signaling, as listed in the Hallmark mTORC1 - Genes up-regulated through activation of mTORC1 complex- library of GSEA. The table and heatmap only include values for genes with an FDR<0.05. FC: log2 fold change, FDR: false discovery rate.

| | **Non DM1** | | **DM1** | |
|---|---|---|---|---|
| **Genes** | **Log2FC** | **FDR** | **Log2FC** | **FDR** |
| *GGA2* | 1.39395718 | 0.00390196 | | |
| *ERO1A* | -1.3435659 | 0.00418508 | -1.474942 | 0.0015409 |
| *CD9* | -1.3961864 | 0.00437319 | | |
| *ENO1* | -1.6348037 | 0.0056941 | -1.677491 | 0.00096568 |
| *EDEM1* | -1.1586112 | 0.00640284 | | |
| *EGLN3* | 5.14064024 | 0.00640284 | | |
| *GAPDH* | -1.2550856 | 0.00644727 | | |
| *VLDLR* | 1.20587612 | 0.00663228 | | |
| *AURKA* | -1.7566402 | 0.00667111 | -1.7725795 | 0.00289283 |
| *LDHA* | -2.0607803 | 0.00744482 | -2.1336723 | 0.0015409 |
| *TXNRD1* | -0.7779018 | 0.00765844 | | |
| *SRD5A1* | -1.129161 | 0.00831054 | | |
| *NUP205* | -0.7874342 | 0.00831054 | | |
| *SLC2A1* | -1.7736938 | 0.00858794 | | |
| *TFRC* | -1.7180821 | 0.00874769 | -1.3687902 | 0.00145744 |
| *SLC9A3R1* | -1.034294 | 0.00900165 | | |
| *GBE1* | -1.2782428 | 0.00952393 | | |
| *RRM2* | -1.6230648 | 0.01001437 | -1.8757533 | 0.00354383 |
| *PSMA3* | -0.5968589 | 0.01195124 | -0.6457465 | 0.00145744 |
| *FGL2* | 3.1913025 | 0.01259737 | | |
| *BUB1* | -1.7073953 | 0.01281787 | -2.2026589 | 0.00129302 |
| *PLOD2* | -1.3974453 | 0.01290488 | | |
| *ACTR2* | -0.7889837 | 0.01293796 | | |
| *IDH1* | -0.5842155 | 0.01375289 | | |
| *BTG2* | 1.32461259 | 0.01461171 | | |
| *PPIA* | -0.7345603 | 0.01461171 | | |
| *POLR3G* | -1.3856651 | 0.01473787 | | |
| *PNP* | -2.150089 | 0.0157362 | | |
| *EBP* | -0.9706267 | 0.01614303 | | |
| *DDX39A* | -0.8585984 | 0.01625575 | | |
| *MCM4* | -0.9702213 | 0.01827781 | | |
| *PSMD12* | -0.6742016 | 0.01914544 | | |
| *PSMD14* | -0.7025409 | 0.01961858 | | |
| *PSMC2* | -0.4749448 | 0.0197571 | | |
| *ACTR3* | -0.8638794 | 0.02035067 | | |
| *SLC6A6* | -1.5638856 | 0.02065512 | | |
| *PSME3* | -0.5587381 | 0.02083138 | | |

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|---|---|---|---|---|
| *ACLY* | -0.6157864 | 0.02083138 | | |
| *TPI1* | -0.7977651 | 0.02314094 | | |
| *RPA1* | -0.6340378 | 0.02484571 | -0.7649368 | 0.00384886 |
| *FDXR* | -0.903531 | 0.02518585 | | |
| *MAP2K3* | -1.334313 | 0.02557279 | -1.1479065 | 0.00314594 |
| *CCT6A* | -0.5660775 | 0.02613288 | | |
| *MCM2* | -1.0112654 | 0.02698042 | | |
| *GLA* | -1.1385406 | 0.02843371 | | |
| *PSMD13* | -0.5794091 | 0.02853326 | | |
| *PSMG1* | -0.6666482 | 0.02879422 | | |
| *ADIPOR2* | -0.4734363 | 0.0290815 | | |
| *DHFR* | -0.9707819 | 0.02926847 | | |
| *PPA1* | -0.7761836 | 0.0321806 | | |
| *TUBG1* | -0.9423582 | 0.03292739 | | |
| *GSR* | -0.7397882 | 0.0345282 | -0.9562429 | 0.00246353 |
| *CCNF* | -0.8882368 | 0.03460799 | -1.3355966 | 0.00156703 |
| *GPI* | -0.5971081 | 0.0347557 | | |
| *ATP6V1D* | -0.5788874 | 0.03599709 | | |
| *CACYBP* | -0.6562446 | 0.03629176 | | |
| *UNG* | -0.6598043 | 0.03632507 | | |
| *PLK1* | -1.8293413 | 0.03674075 | -2.1954151 | 0.00204983 |
| *SORD* | -0.510324 | 0.03801432 | | |
| *INSIG1* | -1.2541102 | 0.0389763 | | |
| *GLRX* | -0.9993069 | 0.03945555 | | |
| *SLA* | 4.00791479 | 0.04056427 | | |
| *CYP51A1* | 0.58406845 | 0.04186838 | 0.84543848 | 0.00175946 |
| *HSPE1* | -0.6618585 | 0.04619077 | | |
| *ETF1* | -0.5282685 | 0.04631241 | | |
| *CYB5B* | -0.4931647 | 0.04649472 | | |
| *AK4* | -1.2301485 | 0.04649472 | | |
| *CDC25A* | -0.8583768 | 0.04848467 | | |
| *EEF1E1* | -0.6903684 | 0.04991954 | | |
| *NAMPT* | | | -1.2050141 | 0.00396463 |

Genes in the heatmap including the differential gene expression from myoblast to myotube for the genes involved in the canonical WNT signaling, as listed in the Hallmark WNT beta catenin signaling (Genes up-regulated by activation of WNT signaling through accumulation of beta catenin CTNNB1) library of GSEA. The table and heatmap only include values for genes with an FDR<0.05. FC: log2 fold change, FDR: false discovery rate.

| | **Non DM1** | | **DM1** | |
|---|---|---|---|---|
| **Gene** | **Log2FC** | **FDR** | **Log2FC** | **FDR** |
| *DLL1* | 3.02490657 | 0.00357914 | | |
| *PSEN2* | 2.46410787 | 0.01081134 | 2.21483088 | 0.02744485 |
| *NKD1* | 4.15693784 | 0.01546835 | 2.71488895 | 0.01281312 |
| *AXIN2* | 2.25386868 | 0.0160369 | 1.65954167 | 0.00394503 |
| *LEF1* | 2.33688891 | 0.01660013 | | |
| *DKK1* | -3.5601985 | 0.01701892 | | |
| *SKP2* | 1.40983119 | 0.0191727 | | |
| *WNT5B* | -2.184473 | 0.02528114 | | |
| *KAT2A* | 0.48746473 | 0.02801512 | | |
| *JAG2* | 1.80956951 | 0.03902908 | | |
| *GNAI1* | | | 0.4562088 | 0.02530757 |
| *TP53* | | | -0.3964804 | 0.03756924 |
| *NUMB* | | | -0.4112431 | 0.04817564 |

Genes in the heatmap including the differential gene expression from stem cell to myoblast for the genes involved in the IL6-JAK-STAT3 signaling, as listed in the Hallmark IL6-JAK-STAT3 signaling (Genes up-regulated by IL6 [GeneID=3569] via STAT3 [GeneID=6774], e.g., during acute phase response) library of GSEA. The table and heatmap only include values for genes with an FDR<0.05. FC: log2 fold change, FDR: false discovery rate.

| | **Non DM1** | | **DM1** | |
|---|---|---|---|---|
| **Gene** | **Log2FC** | **FDR** | **Log2FC** | **FDR** |
| *IL6ST* | 4.26428611 | 4.1641E-05 | 4.14466523 | 3.2372E-07 |
| *OSMR* | 4.50750983 | 4.4035E-05 | 4.64246571 | 1.6061E-07 |
| *CD44* | 7.1533028 | 5.6523E-05 | 6.32644941 | 1.0077E-07 |
| *ITGA4* | 4.75482789 | 9.337E-05 | 5.98071831 | 3.5189E-07 |
| *PDGFC* | 5.0575846 | 9.8128E-05 | 4.33901364 | 1.0601E-07 |
| *IFNAR1* | 1.97760937 | 9.8973E-05 | 1.92092762 | 4.7556E-07 |
| *STAM2* | 1.92235282 | 0.00017136 | 1.83483037 | 5.6503E-07 |
| *PIK3R5* | -7.9610759 | 0.00020256 | -7.7903413 | 5.7319E-06 |
| *JUN* | 2.69348552 | 0.00021404 | 2.02752476 | 0.00287086 |
| *TYK2* | 1.80504095 | 0.00026724 | 1.86369814 | 9.7896E-08 |
| *IL1R1* | 4.41550369 | 0.00029969 | 5.28856073 | 5.135E-06 |
| *TNFRSF12A* | 2.33354197 | 0.00030757 | 1.71676477 | 0.00063839 |
| *ITGB3* | 9.29609163 | 0.00034365 | 8.69967984 | 1.0041E-05 |
| *CSF1* | 3.27242899 | 0.00057967 | 2.89144771 | 9.9354E-05 |
| *PTPN1* | 1.13157544 | 0.00061288 | 0.88953856 | 0.00071451 |
| *IRF9* | 1.95929158 | 0.00064738 | 2.40028118 | 1.0354E-05 |
| *IL10RB* | 2.54786608 | 0.00069408 | 2.35882848 | 1.5019E-05 |
| *IRF1* | 3.11318206 | 0.00071388 | 3.51620023 | 2.9883E-07 |
| *IL4R* | 1.69524039 | 0.00088818 | 1.65641998 | 5.886E-05 |
| *INHBE* | -6.1033354 | 0.00111856 | -5.6992855 | 0.00109621 |
| *SOCS3* | 1.88891489 | 0.00120972 | 2.59961308 | 0.00022229 |
| *TNFRSF1A* | 2.01843338 | 0.00144618 | 2.14672085 | 2.0286E-06 |
| *LEPR* | 1.23861536 | 0.00155782 | 1.23600981 | 0.0031101 |
| *TGFB1* | 3.61486094 | 0.00163264 | 3.59273433 | 2.0195E-06 |
| *IFNGR2* | 1.1425538 | 0.00211175 | 0.76948797 | 0.00090272 |
| *FAS* | 2.49815015 | 0.00237573 | 1.82850815 | 0.00316376 |
| *IL17RA* | 1.97799425 | 0.00313285 | 1.45886921 | 1.7864E-05 |
| *STAT2* | 1.31630803 | 0.00361701 | 1.48633341 | 1.6408E-05 |
| *CBL* | -0.8496222 | 0.00384032 | -1.0970443 | 5.335E-05 |
| *PTPN2* | -0.7237833 | 0.0057656 | -0.7274825 | 0.00011301 |
| *IL13RA1* | 3.12011256 | 0.00614545 | 3.26150887 | 9.4861E-07 |
| *CNTFR* | -3.7908074 | 0.00658418 | -4.4382957 | 0.00077272 |
| *TNFRSF1B* | -2.9195845 | 0.00874089 | -1.0890794 | 0.03153736 |
| *PIM1* | -0.9760434 | 0.00962678 | -1.3917178 | 0.00263493 |
| *TLR2* | 2.75568692 | 0.01338685 | 2.07934841 | 0.00118947 |
| *CXCL1* | 4.05629843 | 0.01356744 | 3.45304179 | 0.00127289 |

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|---|---|---|---|---|
| *CXCL3* | -1.5796414 | 0.01695678 | -2.3876273 | 0.00531439 |
| *LTB* | 4.21479792 | 0.01856928 | 4.16486875 | 0.00101537 |
| *LTBR* | -1.7181058 | 0.02377898 | | |
| *IL17RB* | -0.9959749 | 0.03588731 | -1.2219837 | 0.00261062 |
| *PTPN11* | 0.45438024 | 0.04112286 | | |
| *STAT1* | 1.26168843 | 0.04498209 | 1.01012492 | 0.00016363 |
| *TNFRSF21* | | | 0.71504354 | 0.00097337 |
| *BAK1* | | | -0.62338 | 0.00460355 |
| *CSF2RA* | | | -2.0893389 | 0.00515515 |
| *A2M* | | | 5.17346952 | 0.00537684 |
| *MYD88* | | | 0.86705336 | 0.01076212 |
| *CD9* | | | -0.7455295 | 0.01546379 |

Genes in the heatmap including the differential gene expression from myoblast to myotube for the genes involved in the IL6-JAK-STAT3 signaling, as listed in the Hallmark IL6-JAK-STAT3 signaling (Genes up-regulated by IL6 [GeneID=3569] via STAT3 [GeneID=6774], e.g., during acute phase response) library of GSEA. The table and heatmap only include values for genes with an FDR<0.05. FC: log2 fold change, FDR: false discovery rate.

| | **Non DM1** | | **DM1** | |
|---|---|---|---|---|
| **Gene** | **Log2FC** | **FDR** | **Log2FC** | **FDR** |
| *CD44* | -2.431373 | 0.00233505 | -2.1887476 | 0.00332717 |
| *CD36* | 6.08042627 | 0.00316201 | 4.82447129 | 0.01987872 |
| *CD9* | -1.3961864 | 0.00437319 | -1.3202866 | 0.00490647 |
| *TGFB1* | -1.4248959 | 0.01054286 | -1.4998539 | 0.01652311 |
| *PTPN1* | -0.5990345 | 0.01400909 | | |
| *CBL* | -0.5498962 | 0.02630241 | | |
| *HMOX1* | -1.4525025 | 0.0280504 | -1.010035 | 0.04107062 |
| *INHBE* | 3.42769504 | 0.03045296 | 2.91778296 | 0.01524444 |
| *A2M* | 4.59053439 | 0.03329534 | | |
| *IL6ST* | -0.6436064 | 0.03440211 | | |
| *LEPR* | -0.7105066 | 0.03478801 | | |
| *TNFRSF12A* | -1.6472052 | 0.03877374 | -1.2299748 | 0.04556517 |
| *ITGA4* | 1.29781713 | 0.03945555 | | |
| *STAT3* | 0.66499655 | 0.04262235 | | |
| *ITGB3* | -2.2464195 | 0.04905305 | -2.4031601 | 0.04865699 |
| *ACVRL1* | | | 2.5921333 | 0.00238614 |
| *IL17RA* | | | -0.8701443 | 0.00391008 |
| *IRF1* | | | -2.2835597 | 0.00754558 |
| *TNFRSF21* | | | -0.6900455 | 0.0103455 |
| *IL4R* | | | -0.8773788 | 0.01497536 |
| *TNFRSF1A* | | | -1.0208199 | 0.03128259 |

Genes in the heatmap including the differential gene expression from stem cell to myoblast for the genes involved in the TNFA signaling via NFKB, as listed in the Hallmark TNFA signaling via NFKB (Genes regulated by NF-kB in response to TNF [GeneID=7124].) library of GSEA. The table and heatmap only include values for genes with an FDR<0.05. FC: log2 fold change, FDR: false discovery rate.

| | **Non DM1** | | **DM1** | |
|---|---|---|---|---|
| **Gene** | **Log2FC** | **FDR** | **Log2FC** | **FDR** |
| *IL6ST* | 4.26428611 | 4.1641E-05 | 4.14466523 | 3.2372E-07 |
| *PLAU* | 5.88192357 | 4.1641E-05 | 5.54780578 | 1.9938E-06 |
| *CD44* | 7.1533028 | 5.6523E-05 | 6.32644941 | 1.0077E-07 |
| *LIF* | 4.49126759 | 8.1067E-05 | 2.90652773 | 0.00127148 |
| *FOSL2* | 5.60709275 | 8.9611E-05 | 5.15794233 | 3.3853E-08 |
| *RELA* | 1.95923029 | 0.00010117 | 1.81292324 | 1.4451E-07 |
| *SPHK1* | 4.50026891 | 0.00011983 | 3.76449516 | 0.00044345 |
| *SERPINB8* | 4.668887 | 0.00013613 | 4.1179032 | 9.738E-07 |
| *SMAD3* | 3.01940803 | 0.00015057 | 3.22507329 | 3.5407E-07 |
| *TNC* | 7.19549432 | 0.00017773 | 6.53578839 | 7.8018E-07 |
| *B4GALT1* | 2.34716809 | 0.00018416 | 2.29763423 | 5.1062E-07 |
| *KLF2* | 5.86774385 | 0.00021367 | 3.44550138 | 0.00489474 |
| *CLCF1* | 4.95462355 | 0.00021385 | 4.50462716 | 3.0993E-05 |
| *JUN* | 2.69348552 | 0.00021404 | 2.02752476 | 0.00287086 |
| *IER3* | 4.61430028 | 0.00021548 | 4.16879302 | 7.4331E-06 |
| *GADD45A* | 2.37615749 | 0.00022834 | 1.77177622 | 0.00586905 |
| *INHBA* | 5.90445041 | 0.00025618 | 5.4155506 | 0.00139389 |
| *SPSB1* | 2.67608397 | 0.00025973 | 2.5137276 | 1.1426E-06 |
| *EGR2* | -3.567657 | 0.00026586 | -2.3488436 | 0.00229201 |
| *CCND1* | 3.62372926 | 0.00027331 | 1.99576619 | 0.00036472 |
| *DUSP4* | 4.99023601 | 0.00028991 | 6.56130903 | 1.3912E-05 |
| *PLAUR* | 4.37883786 | 0.00032423 | 3.75679352 | 4.6629E-05 |
| *PFKFB3* | 1.64385068 | 0.00040511 | 1.82839884 | 2.2004E-06 |
| *MAFF* | 3.04888309 | 0.00042883 | 2.47066841 | 0.00046907 |
| *TANK* | 1.47074501 | 0.0004501 | 1.51923592 | 2.3746E-06 |
| *PDLIM5* | 1.56274579 | 0.00053927 | 1.35287403 | 0.00049762 |
| *BTG3* | 1.34733034 | 0.0005397 | 0.93795034 | 0.0002928 |
| *BHLHE40* | 4.86953754 | 0.00055626 | 3.84464037 | 2.5057E-05 |
| *NR4A1* | -3.5040709 | 0.00056571 | -2.8485536 | 0.00020533 |
| *CSF1* | 3.27242899 | 0.00057967 | 2.89144771 | 9.9354E-05 |
| *FJX1* | 2.82362985 | 0.0006258 | 1.95049402 | 2.9199E-05 |
| *LDLR* | 1.40863979 | 0.00067307 | 0.93166494 | 0.00474641 |
| *PHLDA1* | 3.15350847 | 0.00068365 | 3.00703719 | 2.1122E-05 |
| *TIPARP* | 2.65146779 | 0.00068652 | 2.77018488 | 2.7692E-06 |
| *PLK2* | 4.18741038 | 0.00070396 | 4.16711182 | 3.1759E-06 |
| *IRF1* | 3.11318206 | 0.00071388 | 3.51620023 | 2.9883E-07 |
| *SERPINE1* | 7.33181838 | 0.00072915 | 5.88437571 | 0.00082336 |

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|---|---|---|---|---|
| *SNN* | -1.3941582 | 0.00075036 | -1.2038017 | 8.5995E-05 |
| *PTX3* | 6.60548669 | 0.00081759 | 5.42035491 | 1.9576E-06 |
| *PTGS2* | 3.23038972 | 0.00081858 | 2.97712131 | 0.02046786 |
| *EHD1* | 2.15142068 | 0.00082951 | 1.8946288 | 4.3527E-05 |
| *FOSL1* | 5.48607244 | 0.00088633 | 3.8891711 | 0.00082335 |
| *CDKN1A* | 4.51078872 | 0.00093463 | 3.45718467 | 0.00192736 |
| *TNFAIP3* | 3.35810143 | 0.00095142 | 3.27851577 | 7.3386E-05 |
| *IER5* | 3.03155773 | 0.0009862 | 2.30612188 | 1.0126E-05 |
| *KLF6* | 2.59934461 | 0.00105975 | 2.33854264 | 1.8973E-05 |
| *RCAN1* | 3.84643829 | 0.00107611 | 3.13346514 | 0.00115921 |
| *BIRC3* | 4.56988422 | 0.00109641 | 4.81220036 | 4.2402E-05 |
| *PHLDA2* | 4.59568862 | 0.00114657 | 4.34233954 | 5.8592E-06 |
| *TGIF1* | -1.2380713 | 0.00120375 | -0.8684406 | 0.00210157 |
| *SOCS3* | 1.88891489 | 0.00120972 | 2.59961308 | 0.00022229 |
| *TNIP1* | 2.51333976 | 0.00121207 | 2.14218623 | 1.9077E-06 |
| *KLF10* | 1.53785774 | 0.00128274 | 1.43648325 | 0.00111541 |
| *LAMB3* | 4.56557751 | 0.00147924 | 2.17779783 | 0.00883152 |
| *NFKB1* | 2.91851395 | 0.00149282 | 2.20798068 | 9.4429E-07 |
| *PTGER4* | 2.98924487 | 0.0018416 | 3.64082741 | 2.8794E-05 |
| *BIRC2* | 1.19462475 | 0.00185609 | 0.96830537 | 9.1092E-05 |
| *LITAF* | -2.1644054 | 0.00195738 | -2.8840659 | 1.2199E-05 |
| *IFNGR2* | 1.1425538 | 0.00211175 | 0.76948797 | 0.00090272 |
| *MSC* | 3.58561922 | 0.00246516 | 4.85471623 | 1.9163E-05 |
| *TNFRSF9* | 9.73702815 | 0.00259861 | 7.74972642 | 2.2307E-06 |
| *KLF9* | 2.54511022 | 0.00262398 | 3.5627719 | 8.8011E-05 |
| *PMEPA1* | 2.34737165 | 0.00266264 | 2.0197655 | 0.00074986 |
| *ATP2B1* | 1.24963088 | 0.00268751 | 1.98921288 | 0.00381694 |
| *DUSP2* | -3.3430403 | 0.00277126 | -3.8870379 | 1.3395E-05 |
| *HBEGF* | 4.53786655 | 0.00278658 | 3.48187621 | 0.00141428 |
| *CEBPD* | 1.98691503 | 0.00282013 | 2.17173985 | 0.00025627 |
| *AREG* | 5.378497 | 0.00297827 | 5.40119768 | 8.2429E-05 |
| *BCL3* | 3.88778452 | 0.00318883 | 4.43261698 | 2.3507E-06 |
| *JUNB* | 2.49081017 | 0.0033155 | 2.99464106 | 4.8681E-07 |
| *NFIL3* | 1.3318753 | 0.00361713 | 1.14509689 | 4.451E-05 |
| *RHOB* | 1.99315782 | 0.00392704 | 1.25866895 | 0.02448467 |
| *MARCKS* | 1.4017005 | 0.00445957 | 1.27343242 | 6.8011E-06 |
| *GADD45B* | 1.74119308 | 0.00469833 | 1.49723194 | 0.01308782 |
| *IL7R* | 6.87695247 | 0.0047553 | 8.76481041 | 7.3636E-05 |
| *HES1* | 1.6312793 | 0.00484687 | 2.2504904 | 2.9199E-05 |
| *DENND5A* | 0.7961755 | 0.00494793 | 0.64816417 | 0.00124312 |
| *G0S2* | 3.73971023 | 0.00504929 | | |
| *ICOSLG* | -1.4611358 | 0.00516786 | | |
| *IL1B* | 3.17661945 | 0.00604946 | | |

| | | | | |
|---|---|---|---|---|
| *DUSP1* | 2.85932912 | 0.00639778 | 2.66007103 | 0.00044989 |
| *DNAJB4* | 1.66752872 | 0.00646698 | 1.23238173 | 0.00204993 |
| *SLC2A3* | -2.490632 | 0.00671404 | -2.4354326 | 8.9969E-06 |
| *CCL2* | 3.52144505 | 0.00721261 | 4.27255331 | 6.1722E-05 |
| *DUSP5* | 2.04034973 | 0.00858352 | 1.13728097 | 0.01513775 |
| *VEGFA* | 2.34009207 | 0.00964687 | 2.04636415 | 0.00904804 |
| *SDC4* | 1.27702129 | 0.00983729 | | |
| *PER1* | -0.9487304 | 0.01014042 | -0.5395033 | 0.01491049 |
| *NFE2L2* | 0.88435854 | 0.0105677 | 1.10888878 | 0.00011561 |
| *NFKBIA* | 1.12103951 | 0.01200224 | 0.74911493 | 0.008937 |
| *RELB* | 1.46356803 | 0.0120915 | 0.97011518 | 0.00947672 |
| *DRAM1* | 2.26758363 | 0.01284686 | 1.7168651 | 0.00818384 |
| *SQSTM1* | 1.19398864 | 0.01312401 | 1.23781692 | 0.00105948 |
| *TLR2* | 2.75568692 | 0.01338685 | 2.07934841 | 0.00118947 |
| *CXCL1* | 4.05629843 | 0.01356744 | 3.45304179 | 0.00127289 |
| *BTG2* | 1.63590986 | 0.01361685 | 1.50688623 | 0.0119254 |
| *IFIT2* | 3.24152066 | 0.01379642 | 4.24433006 | 4.4988E-06 |
| *GCH1* | -1.9681908 | 0.01384497 | -1.4944872 | 0.00673605 |
| *PPP1R15A* | 1.17026443 | 0.01386943 | 1.14940237 | 0.00805603 |
| *NFAT5* | 0.57650618 | 0.0146681 | 0.99085339 | 1.3572E-05 |
| *TAP1* | 1.02090412 | 0.01523837 | 1.21394533 | 0.00147742 |
| *CXCL3* | -1.5796414 | 0.01695678 | -2.3876273 | 0.00531439 |
| *SERPINB2* | 10.6075033 | 0.01786321 | 9.81919692 | 0.00049908 |
| *MAP2K3* | 0.94836477 | 0.01820229 | 0.80021588 | 0.00079607 |
| *IRS2* | 0.66352452 | 0.0193025 | | |
| *GFPT2* | -2.1920174 | 0.02142384 | -1.0625651 | 0.02509286 |
| *TNFAIP8* | 1.54523973 | 0.02259913 | 0.99360976 | 0.01111727 |
| *SGK1* | 1.35289701 | 0.02295154 | 1.99890101 | 0.0004821 |
| *PTPRE* | 1.53770403 | 0.02818597 | 2.54723116 | 0.00010847 |
| *NFKB2* | 0.89019259 | 0.02965579 | | |
| *SOD2* | 0.39954455 | 0.03543663 | | |
| *MXD1* | 1.22740039 | 0.03689353 | 1.088996 | 0.01695058 |
| *EGR3* | -2.5848334 | 0.04389306 | | |
| *SIK1* | 1.8359086 | 0.04523988 | 2.79649665 | 0.01681219 |
| *EGR1* | -1.4830135 | 0.04526919 | | |
| *GEM* | 1.40099197 | 0.04571177 | 2.23244479 | 7.5245E-05 |
| *EIF1* | 0.47845916 | 0.04798672 | | |
| *KDM6B* | | | 1.78397311 | 0.00029758 |
| *SAT1* | | | 1.19718371 | 0.00030512 |
| *PDE4B* | | | -1.7775686 | 0.0005334 |
| *BTG1* | | | 1.89339298 | 0.00088351 |
| *TUBB2A* | | | -0.866104 | 0.00658129 |
| *ZC3H12A* | | | 1.10377688 | 0.00917347 |

| | | |
|---|---|---|
| *ICAM1* | 1.7576845 | 0.00997947 |
| *CD83* | -1.1433299 | 0.01071153 |
| *TRIB1* | -1.2813529 | 0.01164771 |
| *SLC2A6* | 0.66936924 | 0.0125713 |
| *TSC22D1* | 0.36656198 | 0.01316564 |
| *F3* | 1.34449689 | 0.02047407 |
| *ABCA1* | 1.11670153 | 0.02159121 |
| *PLPP3* | -0.9462838 | 0.0289258 |
| *DDX58* | 0.63964827 | 0.02983679 |
| *CCNL1* | 0.30104418 | 0.03352216 |
| *CFLAR* | 1.144527 | 0.03471332 |
| *ZFP36* | 0.77305938 | 0.03667679 |
| *IFIH1* | 1.07481521 | 0.04631453 |

Genes in the heatmap including the differential gene expression from myoblast to myotube for the genes involved in the TNFA signaling via NFKB, as listed in the Hallmark TNFA signaling via NFKB (Genes regulated by NF-kB in response to TNF [GeneID=7124].) library of GSEA. The table and heatmap only include values for genes with an FDR<0.05. FC: log2 fold change, FDR: false discovery rate.

| | **Non DM1** | | **DM1** | |
|---|---|---|---|---|
| **Gene** | **Log2FC** | **FDR** | **Log2FC** | **FDR** |
| *PLAU* | -3.7253126 | 0.00233505 | -4.4394853 | 0.00125701 |
| *CD44* | -2.431373 | 0.00233505 | -2.1887476 | 0.00332717 |
| *CCND1* | -3.3758515 | 0.00256561 | -2.854504 | 0.00652244 |
| *KDM6B* | 1.68461199 | 0.00278899 | 0.83548083 | 0.01987352 |
| *ICOSLG* | 3.06923682 | 0.00316375 | | |
| *CEBPD* | 1.94083309 | 0.0045315 | 1.3928021 | 0.00705977 |
| *IL1B* | -3.6097374 | 0.00472061 | -3.4161109 | 0.03199129 |
| *EGR2* | 5.52392201 | 0.00500721 | 4.88603149 | 0.00088082 |
| *IER3* | -1.9627713 | 0.00643155 | | |
| *SERPINB8* | -2.0400797 | 0.00644727 | -1.8445558 | 0.01123286 |
| *PLAUR* | -3.2580499 | 0.00728704 | -3.0088861 | 0.00459787 |
| *PFKFB3* | -1.2937208 | 0.00774095 | -1.5452674 | 0.00185298 |
| *FOSL1* | -4.1635059 | 0.00782501 | -3.9771904 | 0.00085706 |
| *SPSB1* | -1.2978482 | 0.00889246 | | |
| *PER1* | 0.93214356 | 0.00941864 | 0.64116797 | 0.02365818 |
| *ATP2B1* | 1.71999824 | 0.01161296 | | |
| *ZBTB10* | 0.99628161 | 0.01183729 | 0.65840089 | 0.02068093 |
| *PHLDA2* | -2.2567037 | 0.01220746 | -2.3755433 | 0.0089609 |
| *ETS2* | -0.977517 | 0.01231359 | -1.5892265 | 0.00404488 |
| *VEGFA* | -2.5958891 | 0.01331465 | -2.7326907 | 0.0098362 |
| *SPHK1* | -2.7117922 | 0.01452625 | -2.3635623 | 0.03343822 |
| *BTG2* | 1.32461259 | 0.01461171 | 1.57064958 | 0.01837678 |
| *MSC* | 2.13075635 | 0.0198375 | 1.73099421 | 0.0266424 |
| *KLF9* | -1.7512155 | 0.0200161 | -2.7091698 | 0.00120837 |
| *IRS2* | -1.297685 | 0.02116717 | -1.0150547 | 0.01926929 |
| *MAP2K3* | -1.334313 | 0.02557279 | -1.1479065 | 0.00314594 |
| *EGR1* | 2.4955646 | 0.02913575 | 2.70121992 | 0.0043867 |
| *PHLDA1* | -1.1342375 | 0.02995247 | -1.7543035 | 0.00278674 |
| *DENND5A* | -0.5718393 | 0.03011756 | | |
| *TNC* | 1.32976083 | 0.03011893 | 1.34942421 | 0.03199123 |
| *LIF* | -1.8109141 | 0.03025927 | -2.0833327 | 0.02430623 |
| *EGR3* | 4.79107308 | 0.03266606 | 3.77781214 | 0.00320201 |
| *SAT1* | -1.4657417 | 0.03412257 | -1.6196244 | 0.02090836 |
| *IL6ST* | -0.6436064 | 0.03440211 | | |
| *SNN* | 0.70054093 | 0.0362185 | 0.68996913 | 0.04236406 |
| *BCL6* | 0.75612844 | 0.03683152 | | |
| *ID2* | -1.1352427 | 0.03709973 | | |

|  |  |  |  |  |
|---|---|---|---|---|
| *SERPINE1* | -3.6072948 | 0.03771482 |  |  |
| *MAFF* | -1.8013523 | 0.03798174 | -1.2231861 | 0.04833985 |
| *SDC4* | -1.2388338 | 0.04039827 |  |  |
| *INHBA* | -1.5555937 | 0.04132073 |  |  |
| *DUSP4* | -1.1872072 | 0.04208609 | -2.4941912 | 0.00384886 |
| *NR4A1* | 1.92101262 | 0.04405169 | 1.87336331 | 0.01163682 |
| *G0S2* |  |  | -3.5975766 | 0.00363902 |
| *NAMPT* |  |  | -1.2050141 | 0.00396463 |
| *IRF1* |  |  | -2.2835597 | 0.00754558 |
| *DUSP2* |  |  | 2.13306166 | 0.00955576 |
| *FJX1* |  |  | 0.73479925 | 0.01074221 |
| *ABCA1* |  |  | -2.1740011 | 0.01153193 |
| *BCL3* |  |  | -1.9951603 | 0.01227016 |
| *TNIP1* |  |  | -0.7465809 | 0.0151081 |
| *B4GALT5* |  |  | -0.61299 | 0.01746993 |
| *AREG* |  |  | -3.4324499 | 0.01751988 |
| *SQSTM1* |  |  | -0.8575789 | 0.01933262 |
| *LAMB3* |  |  | -2.2413078 | 0.02108977 |
| *SLC2A6* |  |  | -0.8473012 | 0.02108977 |
| *NR4A3* |  |  | 1.86056543 | 0.02689223 |
| *KLF2* |  |  | 1.22503553 | 0.02836673 |
| *TRIP10* |  |  | -0.3918087 | 0.0300163 |
| *TNF* |  |  | 1.34942421 | 0.03199123 |
| *TUBB2A* |  |  | 0.78802077 | 0.03437131 |
| *F2RL1* |  |  | -2.8280786 | 0.03625759 |
| *NFAT5* |  |  | -0.4541473 | 0.03787702 |
| *GFPT2* |  |  | -2.2206588 | 0.04171543 |
| *CDKN1A* |  |  | 1.68918886 | 0.04173948 |
| *RHOB* |  |  | 0.52301439 | 0.04269738 |
| *PDLIM5* |  |  | 0.90131606 | 0.04302633 |

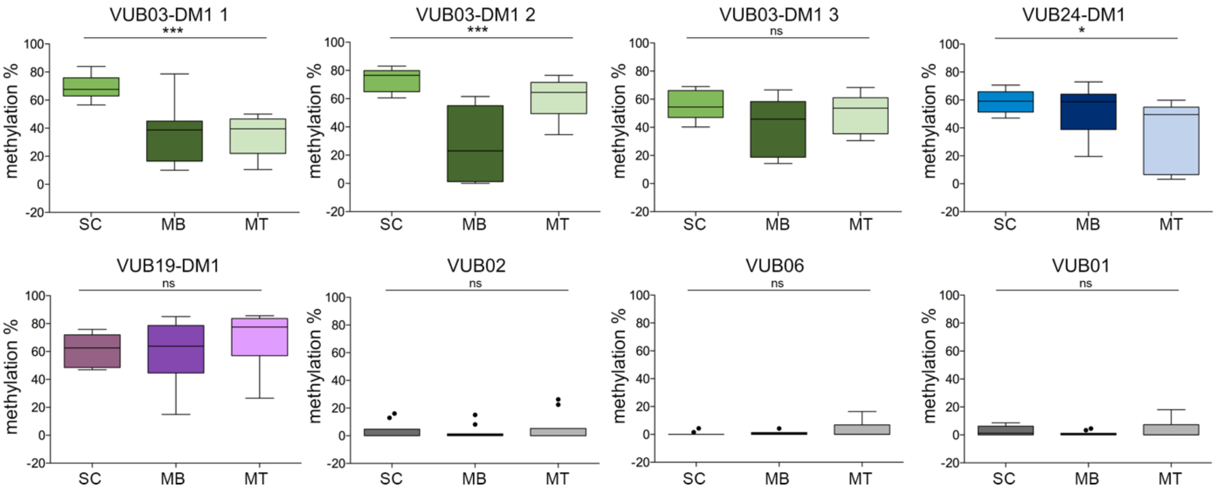

**Fig. S4.** The CpG methylation downstream of the CTG repeat in DM1 samples and non-DM1 samples over the course of differentiation.

The downstream methylation is shown for 11 CpG sites after massive parallel sequencing for 3 DM1 and 3 non-DM1 cell lines. The methylation status of the downstream region decreases for some lines (One-way Anova VUB03-DM1 1 and 2 $p<0.0001$, VUB24-DM1 $p= 0.0214$) other lines remain unchanged (One-way Anova VUB03-DM1 3 $p=0.0913$, VUB19-DM1 $p=0.4399$, VUB02 $p=0.5037$, VUB06 $p=0.0753$, VUB01 $p=0.2927$). Abbreviations: SC: hESC; MB: myoblasts; MT: myotubes

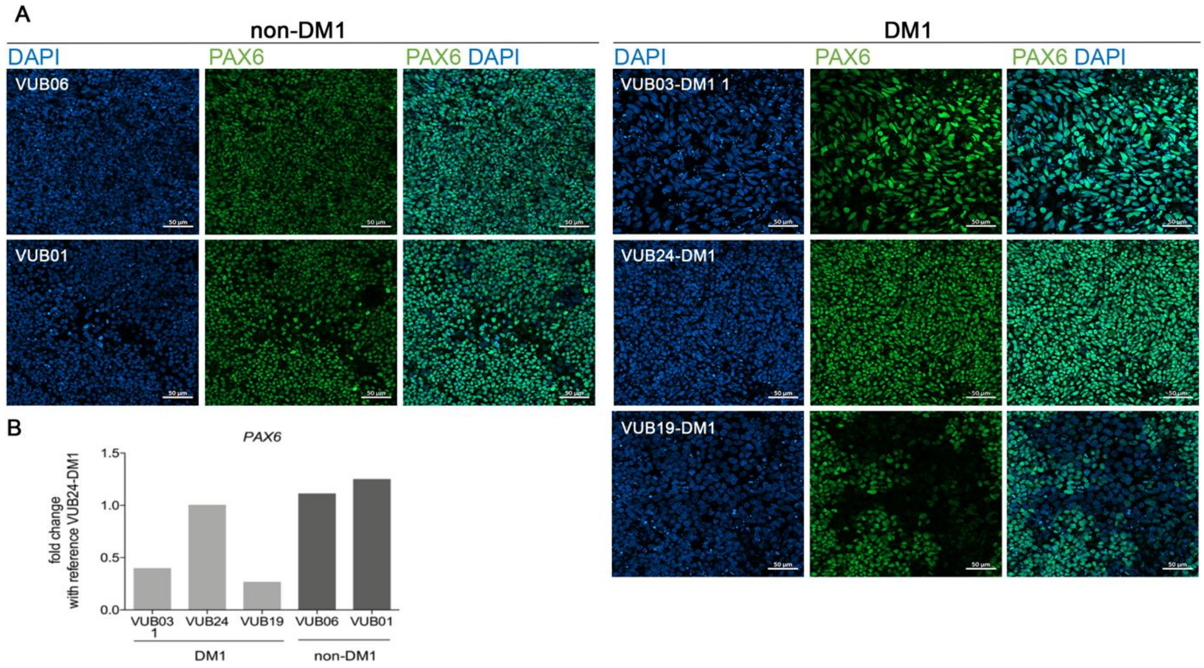

**Fig. S5.** Characterization of the cells after neuroectoderm differentiation.

**(A)** Immunostaining for the neuroectoderm marker PAX6 for 2 control non-DM1 cell lines and 3 DM1 cell lines after 12 days of neuroectoderm differentiation. **(C)** *PAX6* mRNA expression neuroectoderm cells relative to the DM1 reference sample VUB24. *UBC* and *GUSB* gene expression was used for normalization. The experiment was only performed once.

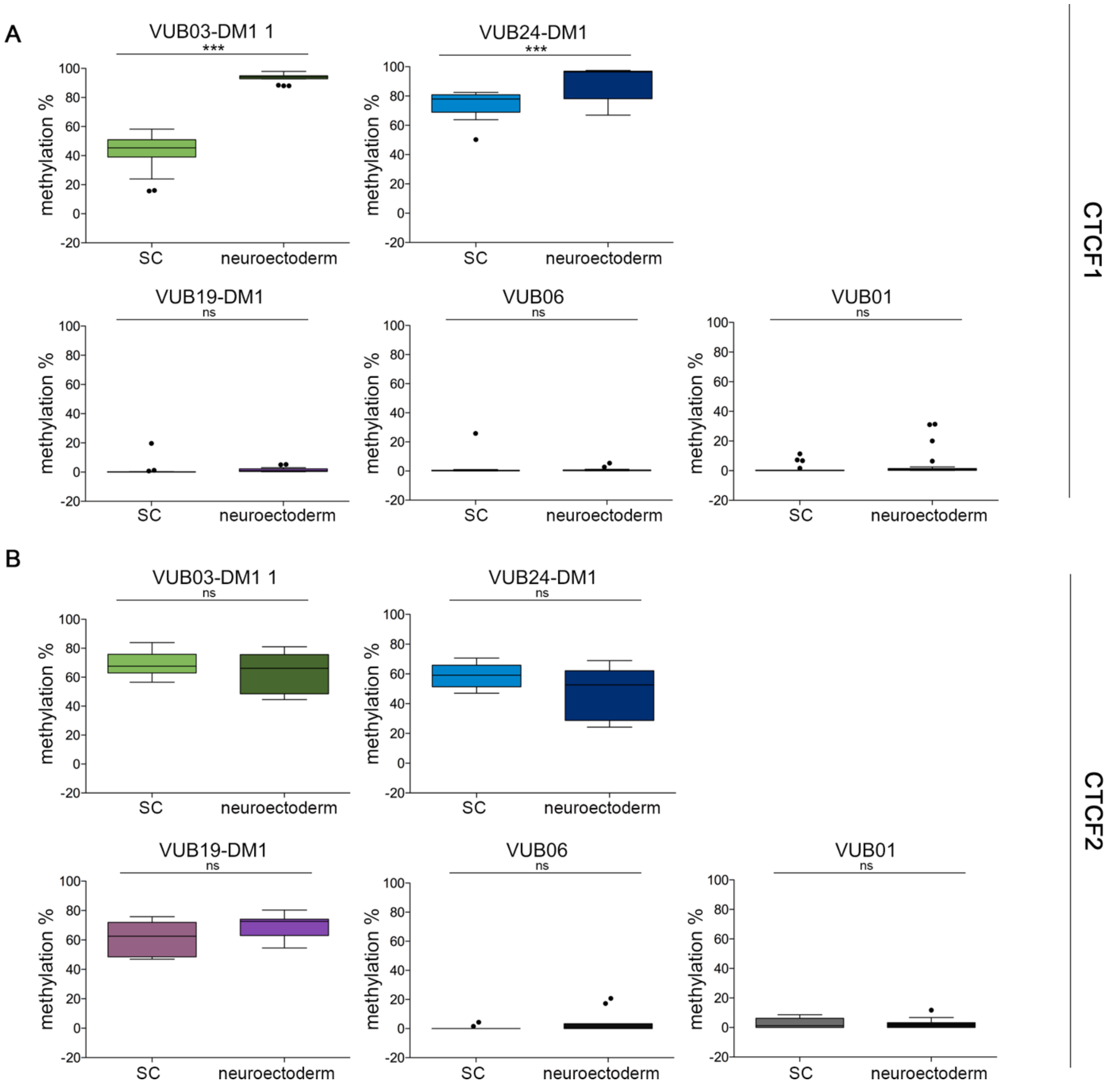

**Fig. S6.** The upstream CpG methylation tends to increase over neuroectoderm differentiation in DM1 samples, the downstream CpG region remains unchanged.

**(A)** Methylation levels of the CpG sites upstream of the CTG repeat. The upstream methylation is shown for 23 CpG sites and all epi-alleles were analyzed after massive parallel sequencing for 3 DM1 cell lines and 2 non-DM1 cell lines (one-way Anova). **(B)** Methylation levels of the CpG sites downstream of the CTG repeat. The upstream methylation is shown for 23 CpG sites and all epi-alleles were analyzed after massive parallel sequencing for 3 DM1 cell lines and 2 non-DM1 cell lines (one-way Anova).

**Table S7.** Splicing defects comparing DM1 myoblasts versus myotubes
https://figshare.com/s/491658bfa49266b27e9a

**Table S8.** Splicing defects comparing DM1 stem cells versus myotubes
https://figshare.com/s/491658bfa49266b27e9a

**Table S9.**: Splicing defects comparing non-DM1 lines versus VUB03_DM1
https://figshare.com/s/491658bfa49266b27e9a

**Table S10.**: Splicing defects comparing non-DM1 myotubes versus DM1 myotubes
https://figshare.com/s/491658bfa49266b27e9a

**Table S11.** Splicing comparing non-DM1 myotubes versus non-DM1 myotubes
https://figshare.com/s/491658bfa49266b27e9a

**Table S12.** Splicing defects comparing non-DM1 myotubes versus DM1 myotubes
https://figshare.com/s/491658bfa49266b27e9a

**Table S13.**: Splicing defects comparing non-DM1 stem cells versus DM1 stem cells
https://figshare.com/s/491658bfa49266b27e9a

**Table S14.** Splicing comparing non-DM1 stem cells versus non-DM1 myotubes
https://figshare.com/s/491658bfa49266b27e9a

**Table S15.** Probes, assays and primers for qRT-PCR.

| **Gene** | **TaqMan Assay/ Sequence** |
|---|---|
| *MYOG* | Hs01072232_m1 |
| *MYOD1* | Hs00159528_m1 |
| *IL6* | Hs00174131_m1 |
| *TRL3* | Hs01551079_g1 |
| *IRF7* | Hs01014809_g1 |
| *PAX6* | Hs00240871_m1 |
| *GUSB* | Hs99999908_m1 |
| *UBC* | Forward 5'-CGCAGCCGGGATTTG-3' |
|  | Reverse 5'-TCAAGTGACGATCACAGCGA-3' |
|  | Probe 6-FAM-TCGCAGTTCTTGTTTGTG-MGB |
| *GAPDH* | Forward 5’-ATGGAAATCCCATCACCATCTT-3’ |
|  | Reverse 5’-CGCCCCACTTGATTTTGG-3’ |
|  | Probe 6-FAM-CAGCAGCGAGATCC-MGB |

**Table S16.** List of used antibodies and manufacturers.

| **Primary antibodies** | **Species** | **Application** | **Company** | **Cat#** |
|---|---|---|---|---|
| HNK-1-FITC (B3GAT1) | Mouse | FACS | AVIVA SYSTEMS BIOLOGY | OASA02271 |
| C-MET-APC (anti-HGF R/C-met) | Mouse | FACS | R&D systems | FAB3582A |
| MF20 | Mouse | ICC | DSHB | NS |
| PAX6 | Mouse | ICC | Abcam | ab78545 |
| **Secondary antibodies** | **Species** | **Application** | **Company** | **Cat#** |
| Alexa Fluor 488 | Goat-anti-Mouse | ICC | Invitrogen | A11001 |

**Table S17.** List of primers.

Primer sequences for the amplification and massive parallel sequencing of CTCF1 and CTCF2. ‘F’ indicates the forward primers, ‘R’ the reverse primers. Number ‘1’ following ‘F’ or ‘R’ indicate the first round of the nested PCR. For the second PCR round, primers with Miseq at the end of the target name were used.

| **Target name** | **Primer sequence** |
|---|---|
| CTCF1 F1 | 5'-TGTYGTYGTTTTGGGTTGTATTG-3' |
| CTCF1 R1 | 5'-CAACATTCCYGACTACAAAAACCCTT-3' |
| CTCF2 F1 | 5'-TTYGGTTAGGTTGAGGTTT-3' |
| CTCF2 R1 | 5'-TTAACAAAAACAAATTTCCC-3' |
| CTCF1 F Miseq | 5’-TCGTCGGCAGCGTCAGATGTGTATAAGAGACAGGTTGTATTGGGTTGGTGGTTTA-3’ |
| CTCF1 R Miseq | 5’- GTCTCGTGGGCTCGGAGATGTGTATAAGAGACAG CTACAAAAACCCTTYGAACCC-3’ |
| CTCF2 F Miseq | 5’-TCGTCGGCAGCGTCAGATGTGTATAAGAGACAGTAAATTGTAGGTTTGGGAAG-3’ |
| CTCF2 R Miseq | 5’-GTCTCGTGGGCTCGGAGATGTGTATAAGAGACAGTTAACAAAAACAAATTTCCC-3’ |
| DM101 | 5'-CTTCCCAGGCCTGCAGTTTGCCCATCCA-3' |
| DM102 | 5'-GAACGGGGCTCGAAGGGTCCTTGT-3' |