

**Vulnerability to shear stress caused by altered peri-endothelial matrix is a key feature  
of Moyamoya disease**

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## Supplementary Table 1

### Primers used for RT-PCR

Gene Name	5'-primer	3'-primer
VE-Cadherin	5'-TGGATGAGAATGACAATGCC-3'	5'-TTGAACTTCACGTTTCGTGG-3'
CD31	5'-AGGTGTTGGTGGAAGGAGTG-3'	5'-CGTGTAGTTGCCACTGTGCT-3'
vWF	5'-AACACCTTTGATGGGAGCAT-3'	5'-TCACTCTCTTGCCATTCTGGA-3'
$\alpha$ -SMA	5'-AGAACATGGCATCATCACCA-3'	5'-TACATGGCTGGGACATGGAA-3'
HAS-2	5'-GTCTCAAATTCATCTGATCTC-3'	5'-ACATTCCTTAAGTAGTCTGG-3'
CD105	5'- CCACTAGCCAGGTCTCGAAG -3'	5'- GATGCAGGAAGACACTGCTG -3'
Oct-4	5'-GACAGGGGGAGGGGAGGAGCTAGG-3'	5'-CTTCCCTCCAACCAGTTGCCCCAAAC-3'
$\beta$ -actin	5'- TCCTGTGGCATCCACGAAACT -3'	5'- GAAGCATTGCGGTGGACGAT -3'

## Supplementary Table 2

### Primers used for real-time PCR

Gene Name	5'-primer	3'-primer
hXylT1	5'-AAGAAGCCACCGAGTAGAC-3'	5'-TGGACACATCCTTCCCGT-3'
hXylT2	5'-CGCTACAAGCTGGCGAT-3'	5'-ACTGTCTGTGTCCTTGGAAC-3'
hGlcAT-I	5'-CCTGCCTACTATCTATGTTGTTAC-3'	5'-ACCACCAGGTGTGTGAA-3'
hChSy1	5'-GGTTTGGCATCACGTGTAT-3'	5'-ACTCCTACTTCCTGGCTC-3'
hChPF	5'-GACCCTCATTTCCGAAGTG-3'	5'-GCCAGATGGCTGGTATT-3'
hChGn-1	5'-AGAAGAAATAAATGAAGTCAAAGGAATAC-3'	5'-GAAGTAGTAGTCCACATCACAG-3'
hUST	5'-ACCATGGACCACCTCCTAGTAA-3'	5'-CACACTTGCCTACCCTGTTGTA-3'
hC4ST-1	5'-AAACGCCAGCGGAAGAA-3'	5'-GGGATGGCAGAGTGAGTAGA-3'
hC4ST-2	5'-GAGGGAAAGTTCTTTGTTTAAGTG-3'	5'-CGGCCTTAACAGCCATAAT-3'
hC6ST-1	5'-CAGGGGAGGAAGAGGAAGAG-3'	5'-CCCTGCTGGTTGAAGAACTC -3'
hD4ST	5'-ACTCAGAGGACAGGGCTA-3'	5'-ACCAAGTGGACTGCATAATC-3'
hGalNAc4S-6ST	5'-TCGTGTGGACAGTAAGCAGAT-3'	5'-TGTAAGAAGCCATTACCAAGGTC-3'
hGAPDH	5'-ATGGGTGTGAACCATGAGAAGTA-3'	5'-GGCAGTGATGGCATGGAC -3'

Figure S1. DNA analyses for RNF213 RNF213 SNP : G>A ss179362673

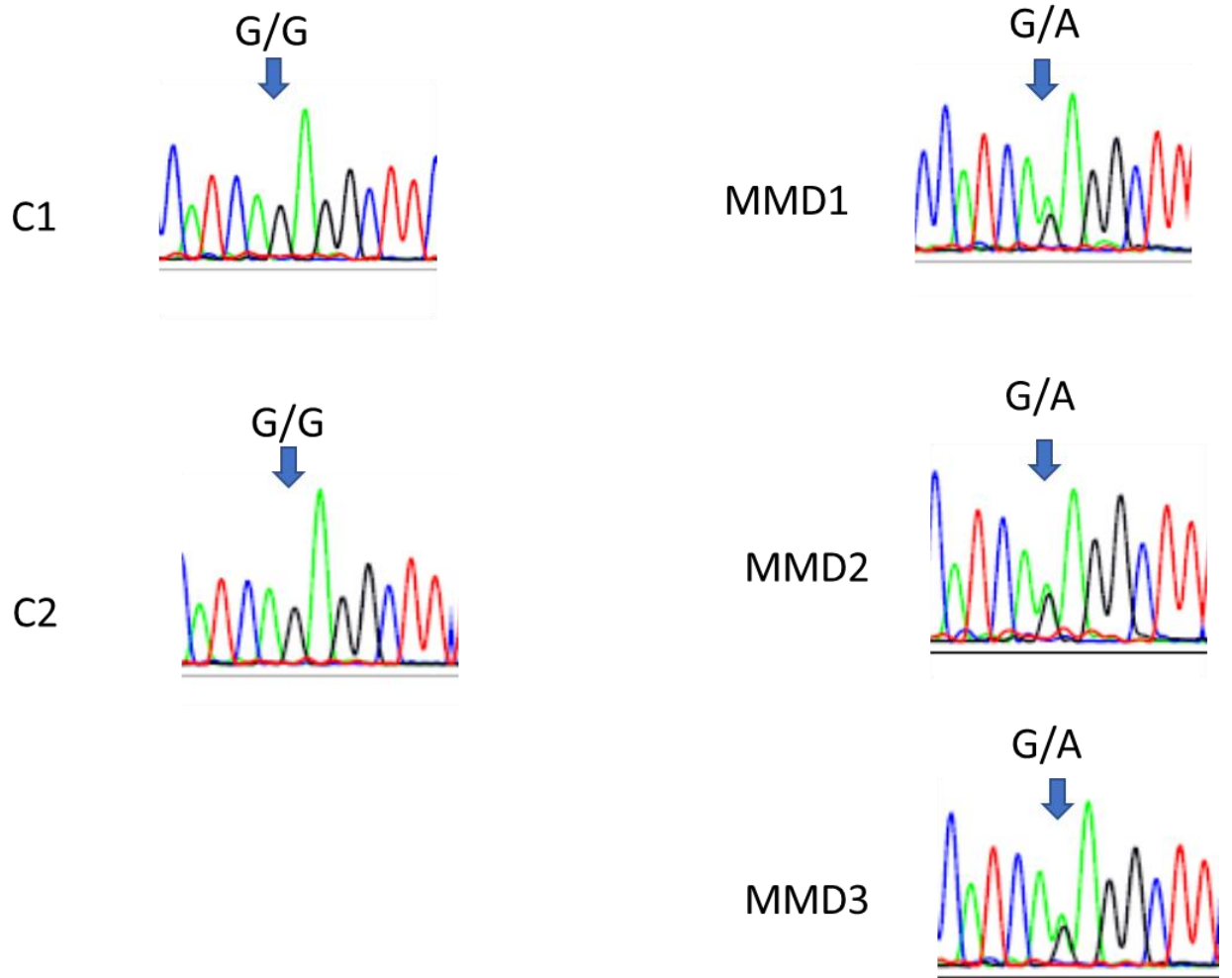


Figure S2. Three-dimensional models of arterial branches.

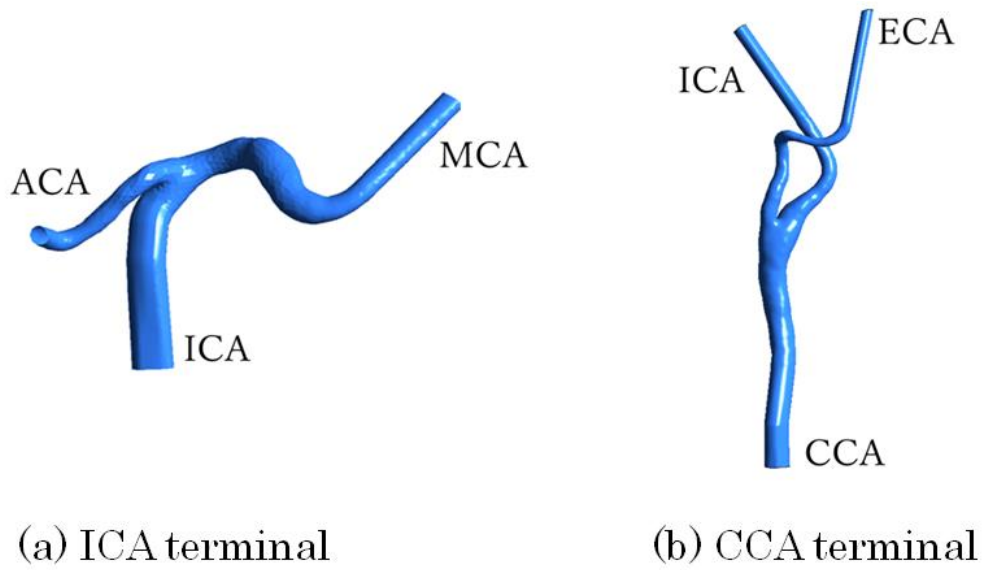


Figure S3. Biosynthesis of chondroitin sulfate.

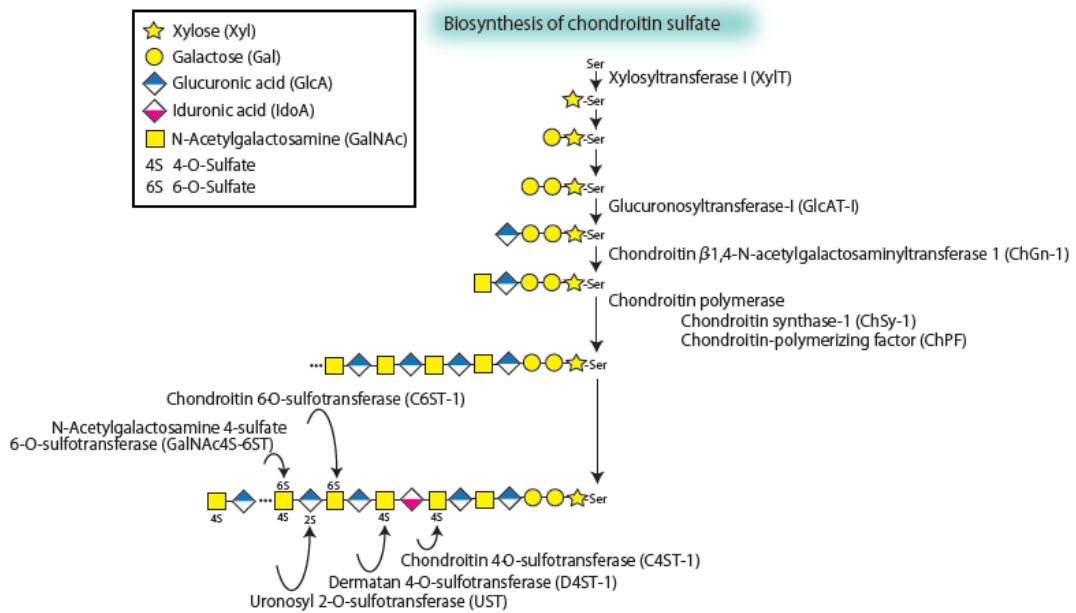
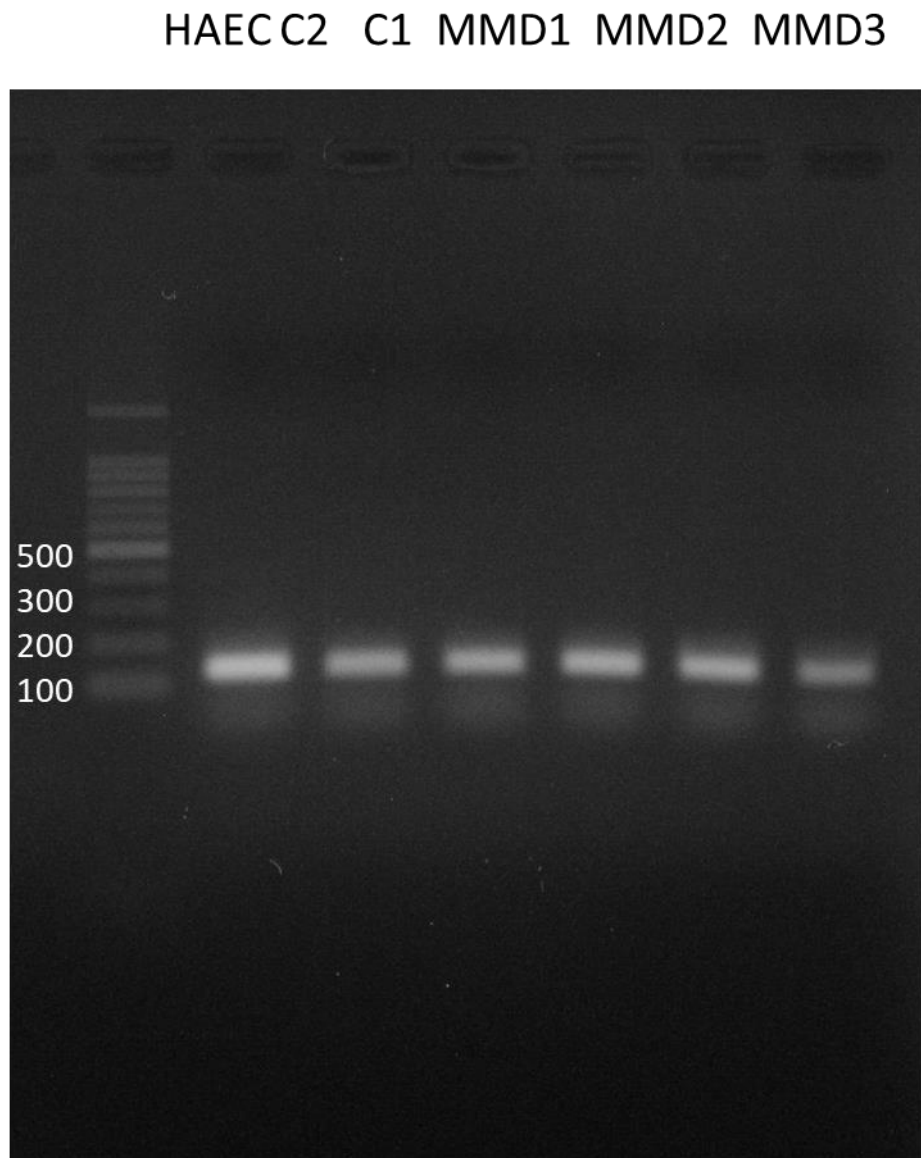


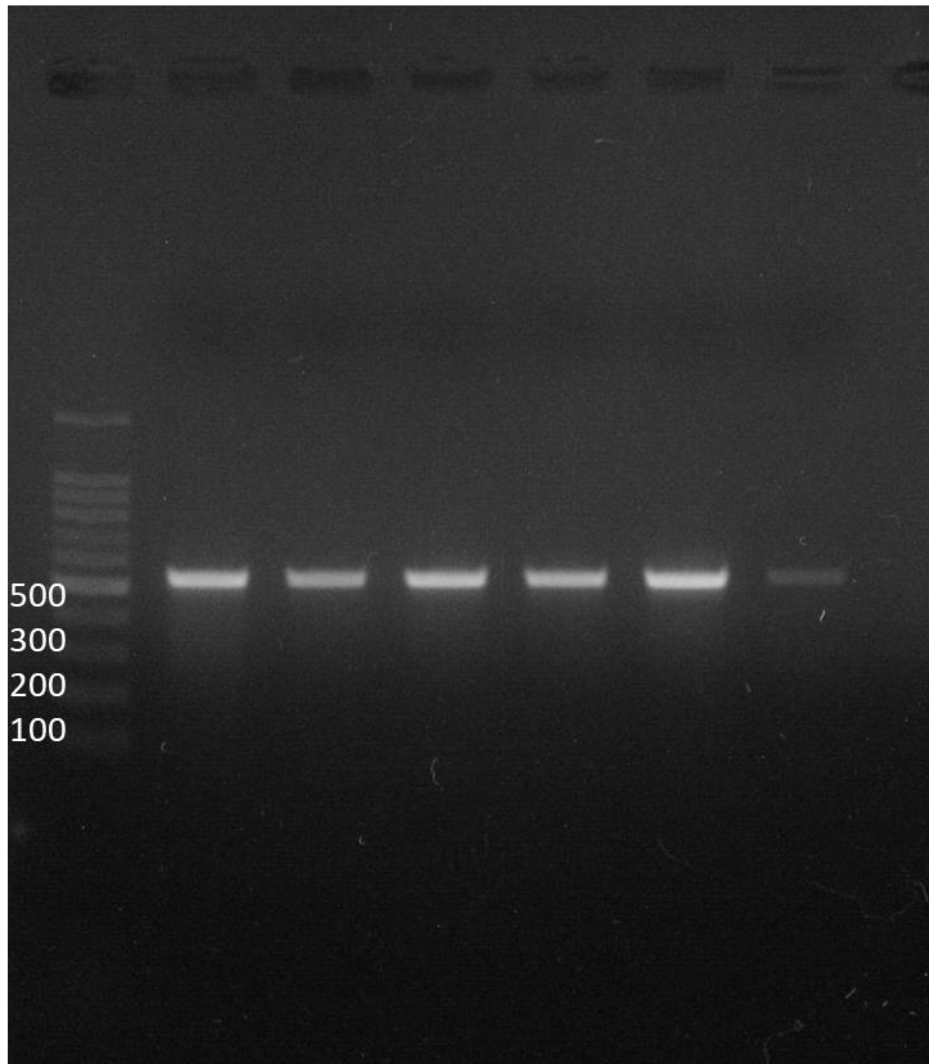
Figure S4. RT-PCR results of endothelial-differentiated iPSCs

A. Cadherin



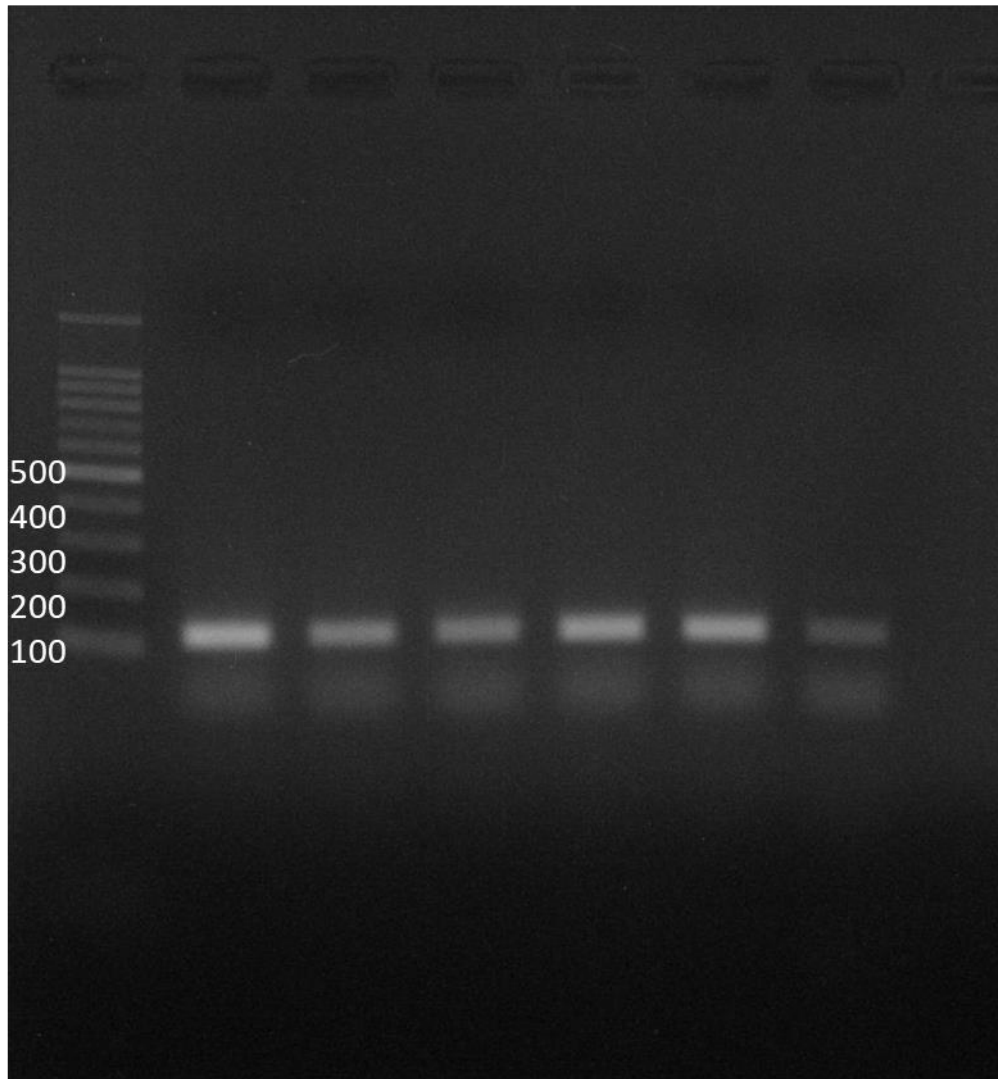
B. CD31

HAEC C2 C1 MMD1 MMD2 MMD3



C. vWF

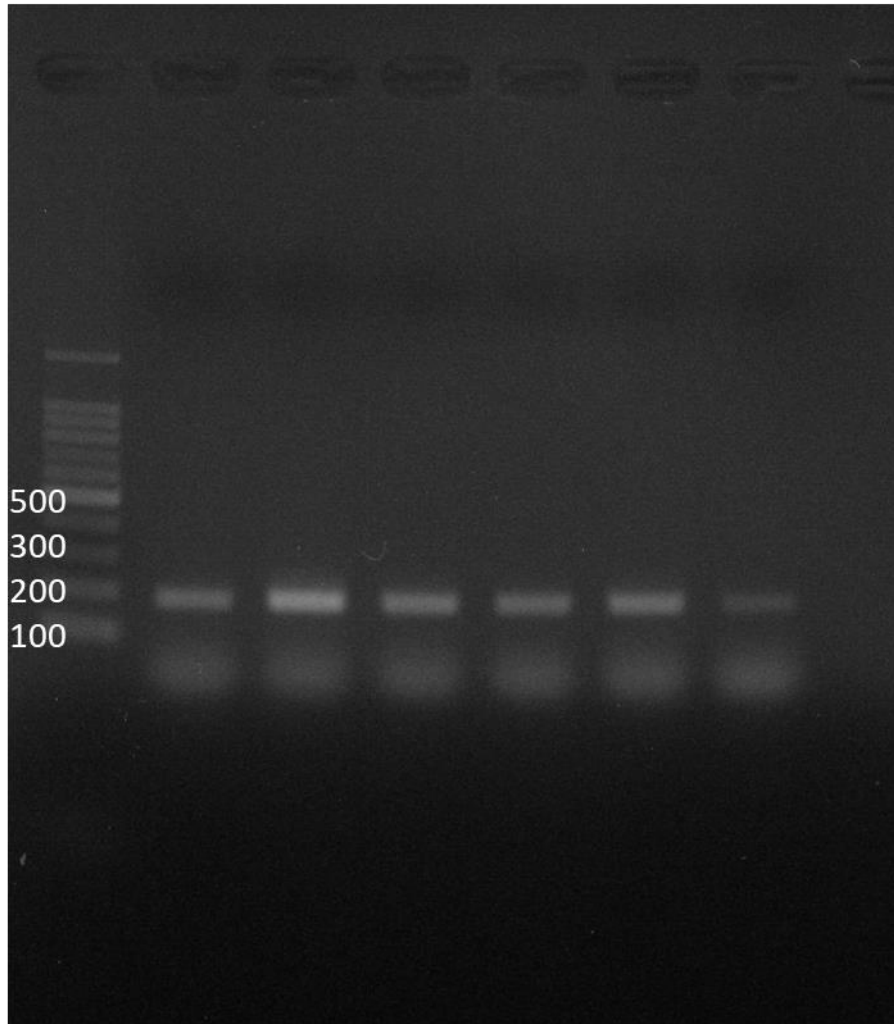
HAEC C2 C1 MMD1 MMD2 MMD3





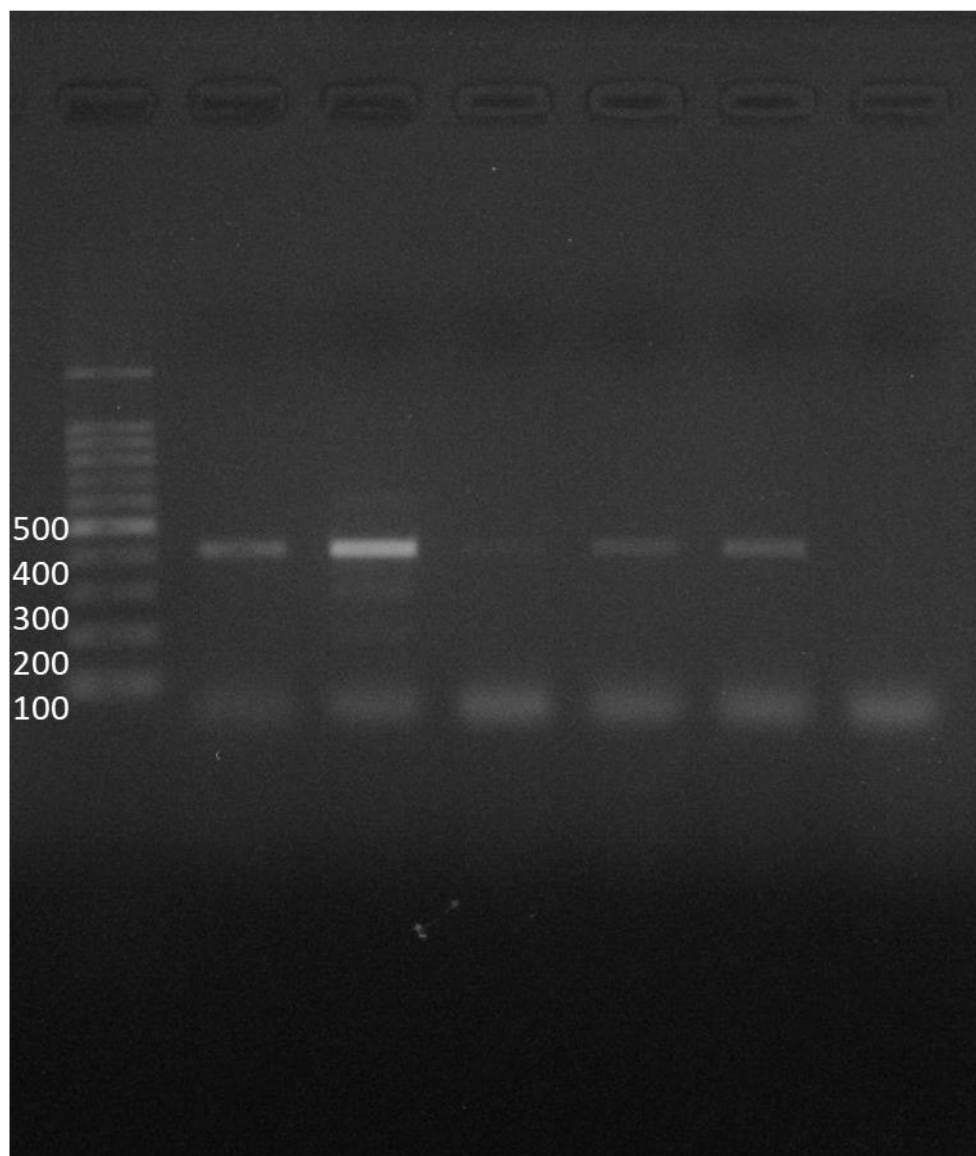
D.  $\alpha$ -SMA

HAEC C2 C1 MMD1 MMD2 MMD3

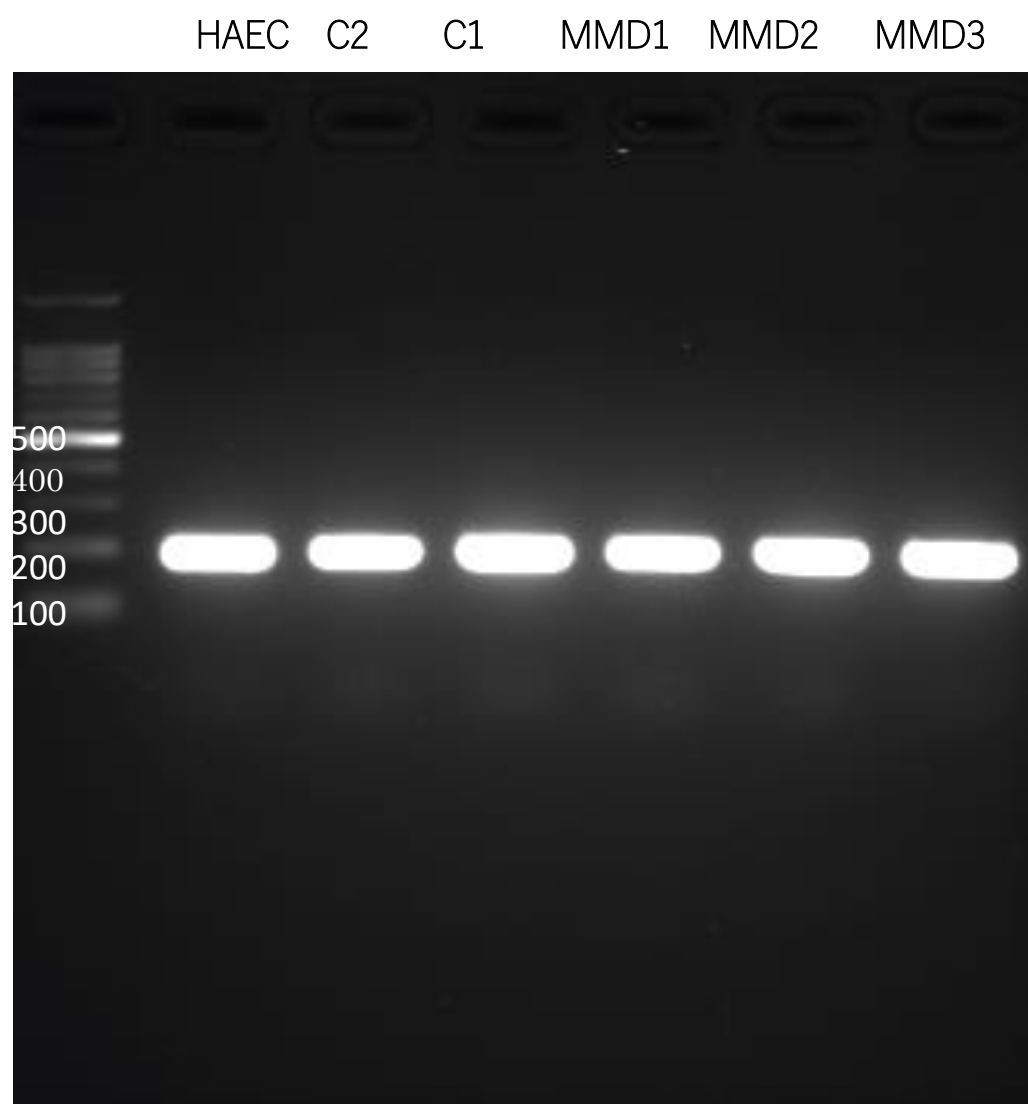


E. HAS2

HAEC C2 C1 MMD1 MMD2 MMD3



F. CD105



G.  $\beta$ -actin

HAEC C2 C1 MMD1 MMD2 MMD3

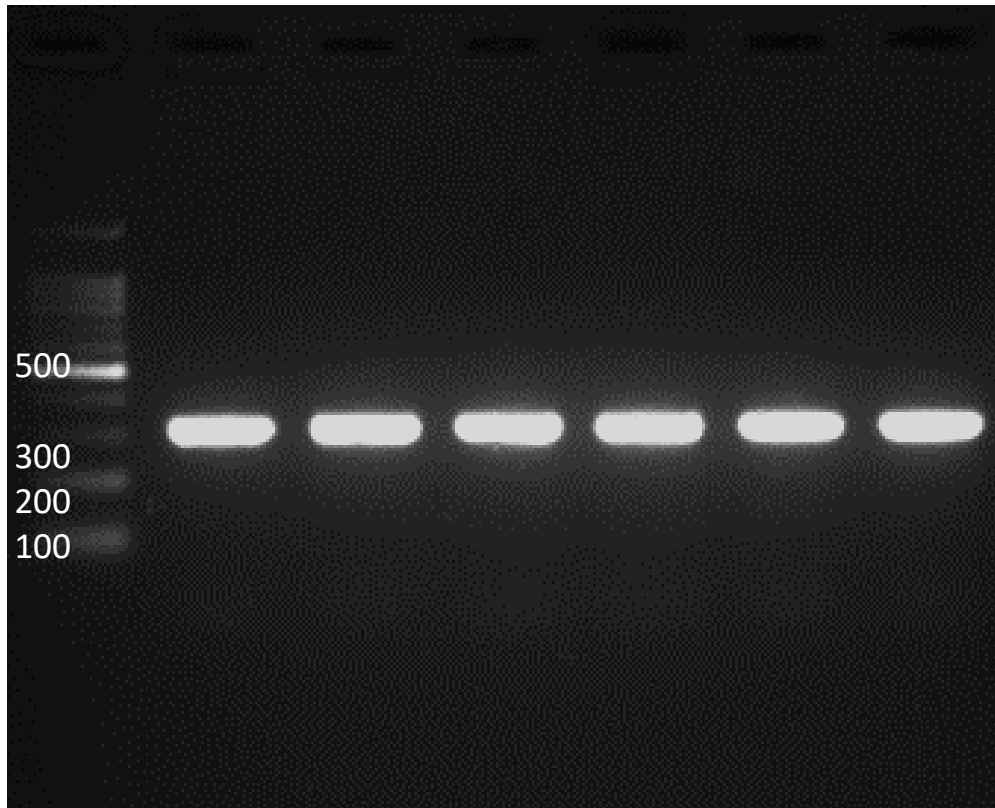
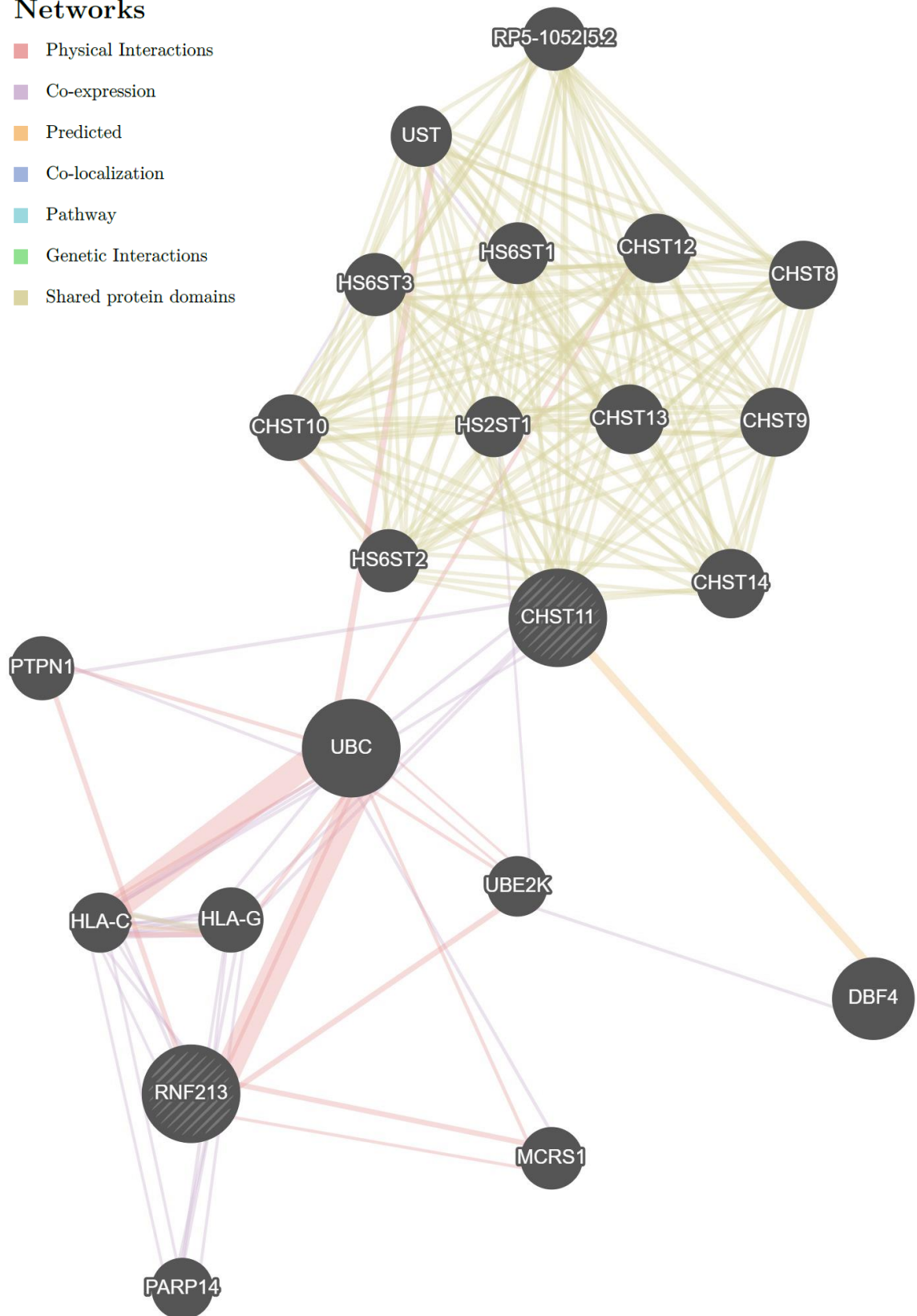


Figure S5. Network analysis by GeneMANIA

### Networks

- Physical Interactions
- Co-expression
- Predicted
- Co-localization
- Pathway
- Genetic Interactions
- Shared protein domains



<b>Gene</b>	<b>Description</b>	<b>Rank</b>
RNF213	ring finger protein 213 [Source:HGNC Symbol;Acc:HGNC:14539]	N/A
CHST11	carbohydrate sulfotransferase 11 [Source:HGNC Symbol;Acc:HGNC:17422]	N/A
UBC	ubiquitin C [Source:HGNC Symbol;Acc:HGNC:12468]	1
DBF4	DBF4 zinc finger [Source:HGNC Symbol;Acc:HGNC:17364]	2
CHST13	carbohydrate sulfotransferase 13 [Source:HGNC Symbol;Acc:HGNC:21755]	3
CHST12	carbohydrate sulfotransferase 12 [Source:HGNC Symbol;Acc:HGNC:17423]	4
CHST14	carbohydrate sulfotransferase 14 [Source:HGNC Symbol;Acc:HGNC:24464]	5
CHST9	carbohydrate sulfotransferase 9 [Source:HGNC Symbol;Acc:HGNC:19898]	6
CHST8	carbohydrate sulfotransferase 8 [Source:HGNC Symbol;Acc:HGNC:15993]	7
CHST10	carbohydrate sulfotransferase 10 [Source:HGNC Symbol;Acc:HGNC:19650]	8
HLA-G	major histocompatibility complex, class I, G [Source:HGNC Symbol;Acc:HGNC:4964]	9
PTPN1	protein tyrosine phosphatase, non-receptor type 1 [Source:HGNC Symbol;Acc:HGNC:9642]	10
RP5-1052I5.2		11
HS6ST2	heparan sulfate 6-O-sulfotransferase 2 [Source:HGNC Symbol;Acc:HGNC:19133]	12
HS6ST3	heparan sulfate 6-O-sulfotransferase 3 [Source:HGNC Symbol;Acc:HGNC:19134]	13
MCRS1	microspherule protein 1 [Source:HGNC Symbol;Acc:HGNC:6960]	14
HS6ST1	heparan sulfate 6-O-sulfotransferase 1 [Source:HGNC Symbol;Acc:HGNC:5201]	15
PARP14	poly(ADP-ribose) polymerase family member 14 [Source:HGNC Symbol;Acc:HGNC:29232]	16
UST	uronyl 2-sulfotransferase [Source:HGNC Symbol;Acc:HGNC:17223]	17
HS2ST1	heparan sulfate 2-O-sulfotransferase 1 [Source:HGNC Symbol;Acc:HGNC:5193]	18
UBE2K	ubiquitin conjugating enzyme E2 K [Source:HGNC Symbol;Acc:HGNC:4914]	19
HLA-C	major histocompatibility complex, class I, C [Source:HGNC Symbol;Acc:HGNC:4933]	20