

Supporting Information for:

Peters plus syndrome mutations affect the function and stability of human β 1,3-glucosyltransferase

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Table S1. Site-directed mutagenesis primers for human B3GLCT construct.

Clone	Primer name	Sequence (5' → 3')
D349N	AI73 f	AGTCATTGTGA <u>AT</u> GATGATACATTAATAAGTATCTCCAGGCTCC
	AI74 r	GTATCATCAT <u>T</u> CACAATGACTAACCATGCTGTTTTGTCCTG
G393E	KZ21 f	CACGGGAGA <u>A</u> GGAGGAATGGTCTTCAGCAGAG
	KZ22 r	ATTCCTCCT <u>T</u> CTCCCGTGATGTAGCTGTAGCC
G394E	KZ23 f	GGGAGGAGA <u>A</u> AGGAATGGTCTTCAGCAGAGAAG
	KZ24 r	ACCATTCCCT <u>T</u> CTCCTCCCGTGATGTAGCTGT
Q457R	KZ07 f	TTCTCATCGG <u>G</u> TCCCATATCGTTCCACAAAC
	KZ08 r	ATGGGAAC <u>CC</u> GATGAGAAAGGTAGTCCTTAGG
T179S	KZ41 f	CGAGAATCCT <u>T</u> CAGTTTTTAAGTATCCAGACTTTG
	KZ42 r	CTTAAAACTGA <u>AG</u> GATTCTCGGAAAAGGCATAATG
V245M	KZ43 f	GTACCAATGAC <u>A</u> TGGACTTCTACTGTGCTACCACATTC
	KZ44 r	GTAGAAGTCC <u>A</u> TGTCATTGGTACAAAACTCAGGCACTG
R337H	KZ45 f	TTTCTGAATC <u>A</u> TAGCCAGGACAAAACAGCATGG
	KZ46 r	GTCCTGGCTAT <u>G</u> ATTCAGAAATCTTTCCAAAATGG
Y366*	KZ17 f	AGCTGTTA <u>A</u> AGACTCCGGCGAGCCTGTGTTT
	KZ18 r	GCCGGAGT <u>C</u> TAAACAGCTAAGCAAGTGCTG
R412*	KZ19 f	AGTAAATGT <u>T</u> GATGCTACAGCAATGATGCTC
	KZ20 r	CTGTAGCAT <u>C</u> AACATTTACTGGCGAGAA
E132A	KZ49 f	CTTCTGTG <u>CC</u> GAAGAGACAAGAATACAGATTCCAAAACCTTGG
	KZ50 r	CTTGTCTCT <u>T</u> CGGCACAGAAGAAAATCCAAGATGAATTTCTGCTA
D421A	KZ55 f	TCCCGATG <u>CC</u> ATGGTCCTGGGAATGTGCTTTAG
	KZ56 r	AGGACCATG <u>G</u> CATCGGGAGCATCATTGCTGTAGC
CT1	AI210 f	GAGTTTTGTACCAATGACGTGGACTTCTAC
	AI214 r (pcDNA3.1) AI213 r (pSecTag2C)	AGCCAAACCAAAGCCAGGGAG GGCCGCGTCACCAGTGG
CT2	AI211 f	ACATTCCATTCTTTTCTACCGCTTTGTAG
	AI214 r (pcDNA3.1) AI213 r (pSecTag2C)	AGCCAAACCAAAGCCAGGGAG GGCCGCGTCACCAGTGG

CT3	AI212 f	AAGCCAGTGAAGAAGAAGGATATTTTTGTTG
	AI214 r (pcDNA3.1) AI213 r (pSecTag2C)	AGCCAAACCAAAGCCAGGGAG GGCCGCGTCACCAGTGG
NT1	AI337 f	CTGAGTTTTG <u>A</u> ACCAATGACGTGGACTTCTACTGTG
	AI338 r	CGTCATTGGT <u>T</u> CAAACTCAGGCACTGGGGTC
NT2	AI339 f	TGTGCTACCTG <u>A</u> TTCATTCTTTTCTACCGCTTTGTAGAAAGCC
	AI340 r	GAATGGAAT <u>C</u> AGGTAGCACAGTAGAAGTCCACGTCATTGG
NT3	AI341 f	GCTTTGTAGAT <u>A</u> GCCAGTGAAGAAGAAGGATATTTTTGTTGC
	AI342 r	TTCACTGGCTAT <u>T</u> CTACAAAGCGGTAGAAAAGAATGGAATG

Supporting Information Figure Legends:

Supporting information

Figure S1. Alignment of B3GLCT sequences from multiple species. Multiple sequence alignment of B3GLCT from human (UNIPROT Q6Y288), mouse (UNIRPOT Q8BHT6), zebrafish (UNIRPOTA0A068F9P7), fruitfly (UNIPROT X2JDC2), and starfish (NCBI REFSEQ XP_038056415.1) were aligned with the MAFFT software/algorithm. Red background highlights regions of 100% sequence identity within the shown sequences; red letters indicate regions of 100% sequence similarity; blue rectangles enclose sets of conserved residues. Annotated domains are highlighted under amino acid sequences.

Figure S2. Time-dependent *in vitro* enzyme assay to determine the linear phase of the transfer reaction catalyzed by B3GLCT. 0.5 μ g purified B3GLCT, 50 μ M UDP-Glc and three different concentrations of pNP-Fuc were analyzed. Reactions were stopped by freezing on dry ice. All reactions were incubated at 37 °C.

Figure S3. Neither the B3GLCT C-GT nor N-GT-like domains expressed alone. (A) Domain map of human B3GLCT with amino acids cutoff for N-GT-like domain and C-GT domain constructs. Each N-GT-like domain (NT1, NT2, NT3) or C-GT domain construct (CT1, CT2, CT3) contained amino acids ending or beginning as indicated. SP, signal peptide; N-GT-like, N-terminal GT-like domain; C-GT, C-terminal GT domain; REEL, C-terminus containing KDEL-like motif. **(B)** Plasmids encoding ADAMTS20 TSR2-8-Myc-His₆ and GFP were co-transfected into WT or *B3GLCT*^{-/-} HEK293T cells. Rescue experiments were performed by co-transfection

with plasmids encoding full-length WT B3GLCT (wtB3), or the CT1, CT2 or CT3 constructs, all with the REEL ER-retention motif. Media and cell lysates were analyzed by Western blot probed with anti-Myc (red, 50 kDa) to detect ADAMTS20 TSR2-8 (TSR2-8), and anti-B3GLCT for B3GLCT WT or mutants (green, 50 kDa, mutants expected <50kDa). Anti-GFP for transfection and loading control (red, 25 kDa). **(C)** Plasmids encoding WT, NT1, NT2, and NT3, all generated with the N-His6-B3GLCT construct, were co-transfected into WT HEK293 cells with GFP plasmid. Media and cell lysates were analyzed by Western blot probed with anti-His₆-tag (red, WT 50 kDa, mutants <50 kDa) and anti-GFP for transfection (green, 25 kDa).

Figure S4. Coomassie blue staining of purified WT and PTRPLS/PTRPLS-like mutant enzymes. Bovine serum albumin (BSA) standards and purified B3GLCT WT, PTRPLS (black) and PTRPLS-like (blue) mutants were separated by 10% SDS-PAGE and stained with Coomassie blue for imaging. For purified enzymes, 5 μ L of eluate was loaded in each lane.

Figure S5. B3GLCT with PTRPLS mutations did not rescue secretion of ADAMTS20 TSR2-8 while PTRPLS-like mutations did. Plasmids encoding ADAMTS20 TSR2-8-Myc-His₆ (TSR2-8) and GFP were co-transfected into wild type (WT) and *B3GLCT* knockout (*B3GLCT*^{-/-}) HEK293T cells. PTRPLS mutations are colored in black and PTRPLS-like mutations in blue. Rescue experiments were performed by co-transfection with a plasmid encoding full-length B3GLCT WT, G393E, and G394E **(A)**, Q457R and T179S **(B)**, and V245M and R337H **(C)** (B3GLCT) with serial dilutions. Serial dilutions of B3GLCT plasmids were performed starting with 0.24 μ g of plasmid diluted 5-, 10-, and 20-fold. Media and cell lysates were analyzed by Western blot probed with anti-Myc (red) to detect ADAMTS20 TSR2-8, anti-

B3GLCT to detect endogenous or transfected B3GLCT (green, 50 kDa), and anti-GFP for transfection and loading control (green, 25 kDa).

Figure S6. B3GLCT mutants G393E and Q457R destabilized the enzyme. Melting curve (A) and first derivative showing the melting peak (B) of WT and mutants were analyzed using the thermo shift protein stability assays described in Experimental Procedures. 10% glycerol in TBS was used as negative control to monitor the background. Arrows in (B) point to reduced melting temperature of G393E (blue) and Q457R (orange).

Figure S7. PTRPLS nonsense mutations at Y366 and R412 result in deletion of important catalytic residues. Ribbon presentation of structures for B3GLCT C-GT Y366* (A) and R412* (B). Light blue ribbon represented the backbone of B3GLCT C-GT. Light blue, residues without mutations; salmon, residues at which had PTRPLS mutations; yellow, residues at which had PTRPLS-like mutations (blue letters).

Figure S8. Internal core structures of B3GLCT C-GT aligned with the cores of MFNG and B3GNT2. Protein backbone alignment between B3GLCT C-GT model (tan), MFNG (PDB 2J0B, blue) and B3GNT2 (PDB 7JHN, purple). Structure alignment was visualized with Chimera.

Figure S9. Sequence alignment revealed that B3GLCT C-GT has conserved motifs of GT31 family enzymes. Amino acid sequence of human B3GLCT (UNIPROT Q6Y288) was aligned to

the sequences of human B3GNT2 (UNIPROT Q9NY97) and mouse MFNG (UNIPROT O09008). DxD motif, dark blue; Glycine rich loop, magenta; xED motif, purple; C-His, orange.

Figure S10. B3GLCT is active in the absence of divalent cations. *In vitro* enzyme assays of B3GLCT with UDP-Glc and Fuc-*O*-TSR3 were supplied with various divalent metal ions at three different concentrations. 10x stock solutions of MnCl₂, MgCl₂, CaCl₂ and EDTA were supplemented to each reaction to make final in-reaction concentrations as described. Statistical analysis was performed with One-way ANOVA in Prism 7. Error bars, standard deviation, n=3; *p<0.05; **p<0.01.

Figure S11. B3GLCT N-GT-like domain adopts a GT-A fold. (A) Protein backbone alignment between B3GLCT N-GT-like domain (tan), B3GLCT C-GT model (green), MFNG (PDB 2J0B, blue) and B3GNT2 (PDB 7JHN, purple). Structure alignment was visualized with Chimera. (B) The DxD motif, G-loop, catalytic aspartic acid, and C-His for B3GLCT N-GT-like domain model (tan), C-GT domain model (green) are aligned with MFNG (PDB 2J0B, blue), B3GNT2 (PDB 7JHN, purple). DxD motifs, G-loops, and catalytic aspartic acid of all four domains also overlaid, whereas C-His of the N-GT domain is missing.

Figure S1

1 10 20 30 40
 HUMAN MRPPAC.....WVLLAPPA**LLALL**LCSLAFGLASEDTKKEVK.QSQDLEKSGI
 MOUSE MR.....PPA**LLALL**FCSSAAFALMSEIEKEKVT.PSQDLRQSSL
 ZEBRAFISH **ML**SIKQNGYSVTVYFFLW.....S**LVAFIT**AK.....GDEVENPAKVHGTDKPSSG.
 FRUITFLY **MP**TRIC.....VLFGLL.....C**LA**SL**CG**CS.....NKPG.
 STARFISH **MA**ASIV.....A**EVGLIT**GVFAATF.....DPYFPRDLTVAEPNL

50 60 70 80 90
 HUMAN SRKNDIDLK**GIVFVI**Q**SQSN**S**FHAKRAEQ**...**LKKSITKQ**AAD**LTQE**..L**PS**..V**LLLH**
 MOUSE PGRHNDIDLK**EIVFVI**Q**SQSN**S**FHAKRAEQ**...**LKKNILKQ**AAN**LQD**..L**PR**..V**LLLH**
 ZEBRAFISH ...SQDLDR**EIVFVI**Q**SQRN**S**FHVRQA**EK...**LRKDLLLQ**QT**LQES**..P**V**..I**LLLH**
 FRUITFLY**EVLVVI**AC...P**HPQA**AR**SDCLA**L**SHNVLE**Q**QRA**L**ELAGIF**P**EDF**V**LKLVH**
 STARFISH EGHHTATGE**DLVFIR**S**QQA**S**PHDQWAKQ**...**VKADILQ**Q**YRI**L**ELG**..N**PQ**..V**LLLH**

100 110 120 130 140 150
 HUMAN **QLAKQE****GAWTILPLPHF**SVT...Y**SRNS**S**SWIFF****FCEEE****TRT**..Q**IPKLE**T**LRRYD**P**SKEW**
 MOUSE **QLAKQE****GAWTILPLPHF**SVT...Y**SKNS**A**WIFF****FCEEE****TRL**..Q**IPRLD**T**LRRYD**P**SKEW**
 ZEBRAFISH **TL**SDNE**G**W**STILPLPR**L**SSQ**...F**GKNS**S**SWIM****FLEED****TRV**..K**LQKLE**H**V**L**KKF**D**RK**E**W**
 FRUITFLY **VM**HEL**FNS**W**ITMDALPH**L**R**AQ**ARV**L**GAR**T**EWITWC**Q**HNT**R**V**S**SLRGL**E**LQ**L**RRQN**P**RELA**
 STARFISH **QQ**Y**EF**S**G**V**WITLPVLP**K**L**N**ED**...E**GQ**..A**KWIF****FCEEQ****TRV**..S**VH**E**GLQ**V**L**T**KYN**P**Q**E**W**

N-GT-like Domain

160 170 180 190 200 210
 HUMAN **FLGKALHDE**E**ATI**I**H**H**YAF**S**ENPTV**V**KYP**D**F**A**AGWALS**I**FLVN**K**LTKRL**...K**SE**S**LK**S
 MOUSE **FLGKALYDE**E**STI**I**H**H**YAF**S**ENPTV**V**KYP**D**F**A**AGWALS**I**PLVN**K**LAKRL**...K**SE**A**LK**S
 ZEBRAFISH **FLGKPLHDE**E**STI**I**H**H**YAF**S**ENPTA**E**Y**P**D**F**S**A**GWALS**I**FLIN**R**L**A**S**K**I**...E**E**E**P**L**K**S
 FRUITFLY **FYGHALYDA**E**ATI**I**H**H**YAF**S**NYKDP**Q**RE**F**Y**P**M**L**S**A**GVVF**T**G**A**LLRR**L**ADLV**A**P**S**G**Q**N**I**V**H**S**
 STARFISH **FLGRALQ**D**R**A**ASVITH**H**YR**E**H**D**DP**S**K**S**Y**P**D**F**E**A**GW**L**I**S**TGL**L**R**G**LAD**R**W**...E**TE**E**H**R**M**

220 230 240
 HUMAN **DFTIDLKHE**E**ALYI**W**D**K...G**GG**P**ELT**P**V**P**EE****CT**...N**D**V**D**F**Y**...
 MOUSE **DFTIDLKHE**E**ALYI**W**D**K...G**GG**P**AL**T**P**V**PE****CT**...E**D**V**D**P**R**...
 ZEBRAFISH **DFTIDLKHE**V**ALYI**W**D**K...G**GK**P**RL**T**G**V**P**E**L****CT**..A**E**H**N**K**R**A**Q**S...
 FRUITFLY **D**F**S**I**D**A**S**H**E**L**A**R**E**F**I**W**D**N**V**S**P**D**P**H**I**S**T**P**I**S**G**G**I**L**L**K**S**A**S**Y**I**C**S**T**P**T**S**V**P**N**R**K**L**P**C**...
 STARFISH **D**F**S**I**D**V**K**H**E**L**A**Y**I**L**N**D...G**N**G**T**R**L**T**A**L**P**A**L**C**A**G...N**V**E**E**N**P**A**P**R**S**D**S**E**I**R

Linker

250 260 270 280
 HUMANC**ATT**F**H**S**F**L**P**L**C**R**K**P**V**K**K**K**D**I**F**V**AV**K**T**C**K**K**F**H**G**D**R**I**P**I**V**K
 MOUSEC**VTT**F**H**S**F**L**P**L**C**G**V**P**V**K**K**E**E**I**F**V**AV**K**T**C**K**K**F**H**A**D**R**I**P**I**V**K
 ZEBRAFISHC**ATT**V**S**H**S**P**L**C**G**E**P**V**K**I**E**N**I**F**V**AV**K**T**C**K**K**F**H**S**D**R**I**P**V**V**K**
 FRUITFLY ..L**H**A**Q**P**E**E**P**L**T**L**G**Q**R**R**N**G**C**E**HT**T**G**S**H**...I**Y**F**A**I**K**T**C**K**K**F**H**K**E**R**I**P**I**E
 STARFISH K**A**L**N**K**A**I**R**D**G**P...Q**E**E**D**G**C**V**S**A**H**P**K**G**L**P**T**C**A**E**A**V**L**K**E**D**IL**F**AV**K**T**C**K**K**F**H**K**D**R**V**P**I**V**Q

290 300 310 320 330 340
 HUMAN **Q**T**W**E**S**Q**AS**L**I**E**Y**S**D**Y**T**E**N**S**I**P**T**V**D**I**G**I**P**N**T**D**R**G**H**C**G**K**T**F**A**I**L**E**R**F**L**N**R**..S**Q**D**K**T**A**W**L**V
 MOUSE **K**T**W**A**A**Q**AS**L**I**E**Y**S**D**Y**A**E**T**A**I**P**T**V**D**I**G**I**P**N**T**D**R**G**H**C**G**K**T**F**A**I**L**E**K**F**L**N**H**..S**H**N**K**I**S**W**L**V
 ZEBRAFISH **K**T**W**G**K**Q**AS**L**I**E**Y**S**D**Y**A**D**E**P**I**P**T**I**N**L**G**I**P**N**T**E**R**G**H**C**G**K**T**F**A**I**L**R**R**F**L**S**S**..H**V**P**R**T**D**W**L**L
 FRUITFLY **R**T**W**A**A**D**AR**N**R**R**Y**S**D**V**A**D**V**G**I**P**A**I**G**T**I**G**I**P**N**V**Q**T**G**H**C**A**K**M**A**I**L**Q**L**S**L**K**D**I**G**K**Q**L**D**I**R**W**L**
 STARFISH **Q**T**W**G**K**H**T**S**H**I**V**F**V**S**D**V**Q**D**D**T**I**P**T**I**A**S**G**V**P**N**T**E**R**G**H**C**G**K**L**F**A**I**F**E**M**F**N**S**K**..P**E**Y**L**K**Y**S**W**L**V**

C-GT Domain

350 360 370 380 390
 HUMAN **I**V**D**D**D**T**L**I.....S**I**S**R**L**Q**H**LL**S**C**Y**D**S**G**E**P**V**F**L**G**E**R**Y**G**Y**G**..L**G**T**G**..G**Y**S**Y**I**T**G**G**
 MOUSE **I**V**D**D**D**T**L**I.....S**I**S**R**L**R**H**LL**S**C**Y**D**S**S**D**P**V**F**L**G**E**R**Y**G**Y**G**..I**G**T**G**..G**Y**S**Y**V**T**G**G**
 ZEBRAFISH **I**V**D**D**D**T**L**I.....S**L**P**R**V**Q**A**LL**S**C**Y**E**S**S**E**P**L**C**L**G**E**R**Y**G**Y**G**..I**G**Q**G**..G**Y**S**Y**I**T**G**G**
 FRUITFLY **I**V**D**D**D**T**L**L**S**L**H**L**I**H**T**H**L**P**T**..S**V**P**R**V**S**A**LL**C**R**H**N**A**T**E**L**V**L**L**G**Q**R**Y**G**Y**R**..L**H**A**P**D**G**F**N**Y**H**T**G**G
 STARFISH **V**A**D**D**D**T**L**L.....S**V**A**R**L**R**A**LL**S**C**Y**N**A**K**K**L**V**F**L**G**E**R**Y**G**Y**G**H**L**K**L**G**W**G**Y**D**V**L**T**G**G**

400 410 420 430 440 450
 HUMAN **G**C**M**V**S**S**R**E**A**V**R**R**L**L**A**S**K**R**C**R**C**Y**S**N**D**A**P**D**D**M**V**L**G**M**C**F**S**G**L**G**T**F**V**T**H**S**P**L**F**H**Q**A**R**P**V**D**Y**P**K**D**Y**
 MOUSE **G**G**M**V**S**S**R**E**A**I**R**R**L**L**V**S**S**C**R**C**Y**S**N**D**A**P**D**D**M**V**L**G**M**C**F**S**G**L**G**V**P**V**T**H**S**P**L**F**H**Q**A**R**P**V**D**Y**P**K**D**Y
 ZEBRAFISH **G**G**M**L**S**S**R**E**A**V**V**Q**L**L**S**S**G**N**C**R**C**Y**S**N**D**A**P**D**D**M**V**L**G**M**C**L**N**S**L**R**V**P**V**T**H**S**P**L**F**H**Q**A**R**P**D**Y**A**R**D**F
 FRUITFLY **A**G**I**V**L**S**L**P**L**V**R**L**I**V**Q**R**C**S**C**P**S**A**S**A**P**D**D**M**I**L**G**M**C**L**Q**A**L**L**V**P**A**I**H**V**A**G**M**H**Q**A**R**P**Q**D**Y**A**G**E**L**
 STARFISH **G**G**M**I**S**S**R**A**G**I**Q**L**L**A**T**G**C**K**N**K**D**E**D**P**D**D**M**I**F**G**M**C**T**K**R**H**M**I**P**I**T**H**S**P**L**F**H**Q**A**R**P**S**D**Y**A**V**G**Y

460 470 480 490
 HUMAN **L**S**H**Q**V**P**I**S**F**H**K**H**W**N**I**D**P**V**K**V**Y**F**T**W**L**A.....P**S**D**E**D**K**A**R**Q**E**T**Q**K**G**F.....R**E**E**L**...
 MOUSE **L**A**H**Q**I**P**V**S**F**H**K**H**W**H**I**D**P**V**K**V**Y**L**T**W**L**A.....P**S**E**E**D**Q**A**T**Q**E**T**Q**K**D**P.....R**E**E**L**...
 ZEBRAFISH **L**S**H**Q**T**P**I**S**F**H**K**H**W**N**I**D**P**A**V**F**N**K**W**L.....K**N**E**K**H**T**E**G**L**Q**K**S**T.....R**E**E**L**...
 FRUITFLY **L**Q**L**H**A**P**L**T**F**H**K**H**W**N**I**D**P**E**H**T**Y**R**R**W**L**G**S**M**V**N**R**S**A**P**L**A**A**H**K**E**R**Q**P**A**G**P**L**H**M**T**M**G**R**H**S**L**A**A**G**
 STARFISH **L**Q**H**Q**T**P**V**S**F**H**K**H**L**N**A**D**P**L**A**V**Y**R**D**W**E**R.....H**A**D**E**V**D**S**R**T**H**Q**A**G**E**.....R**E**E**L**...

Figure S2

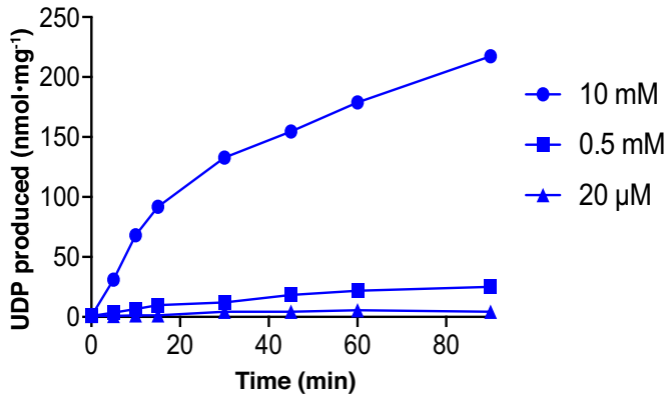
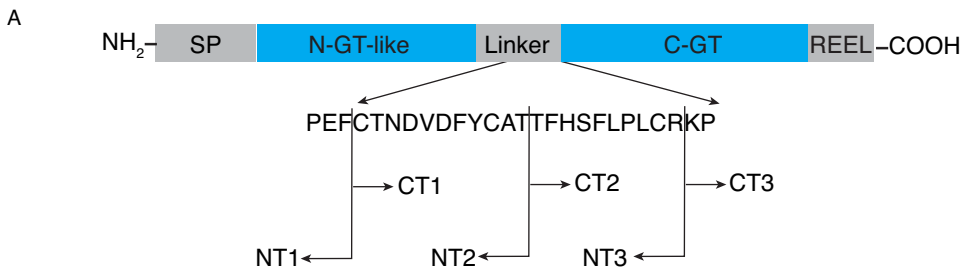
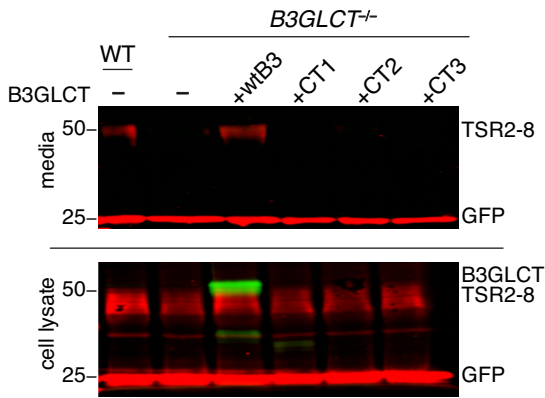


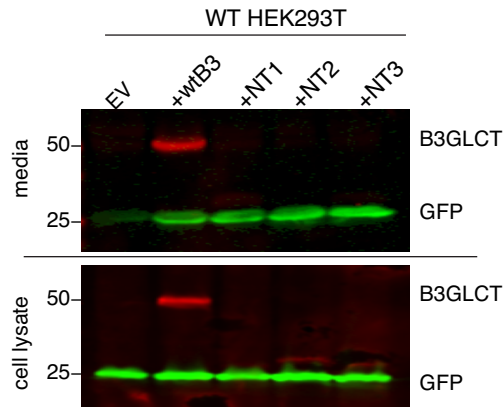
Figure S3

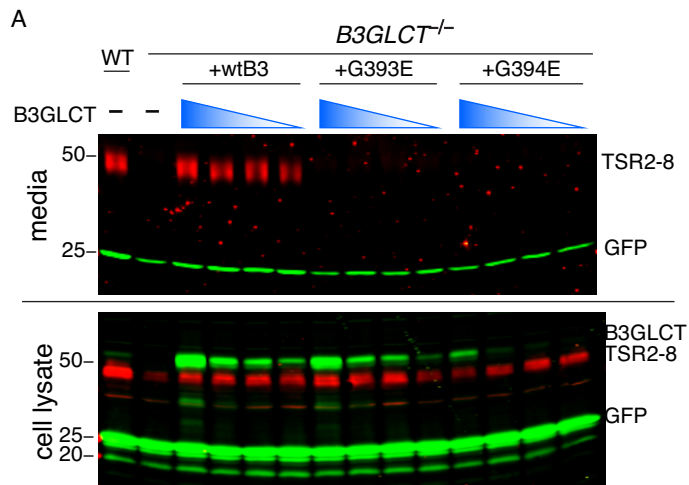
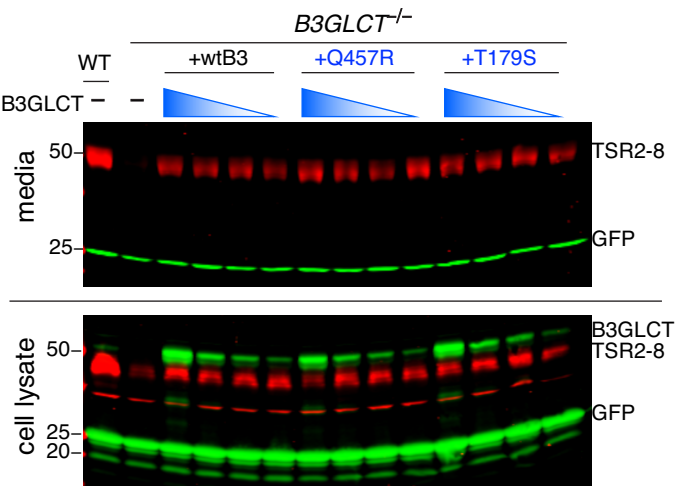
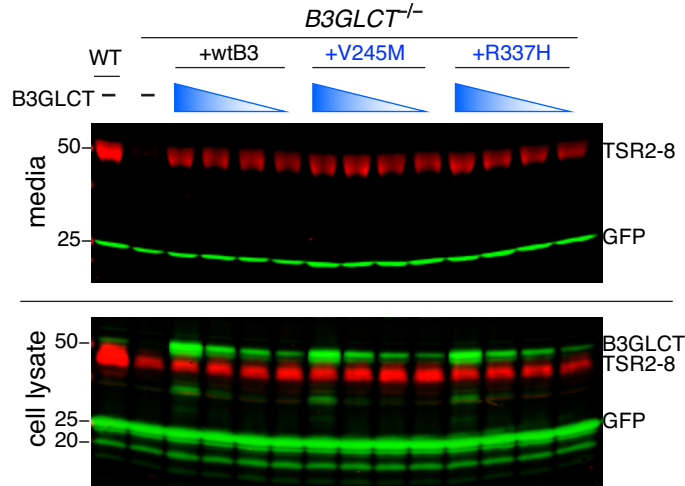


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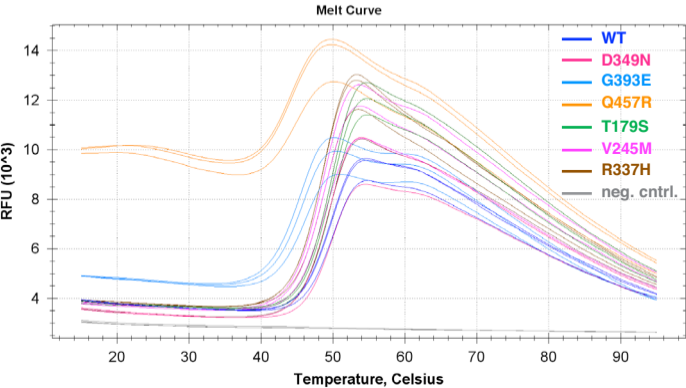
C



**B****C**

FigureS6

A



B

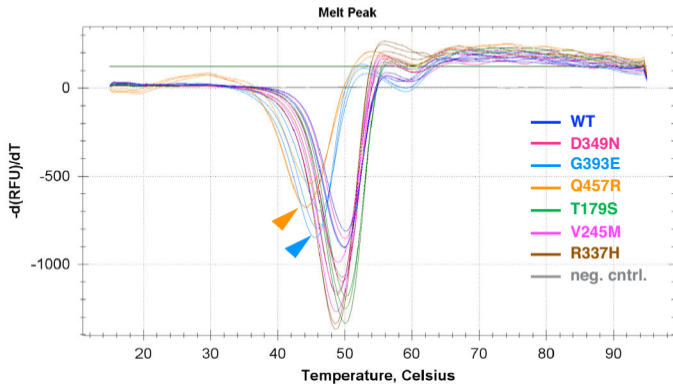
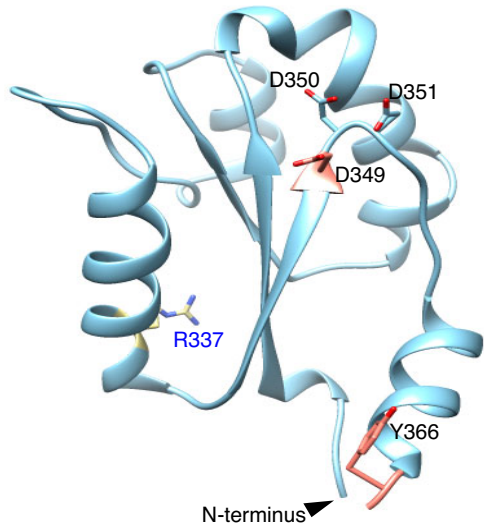


Figure S7

A



B

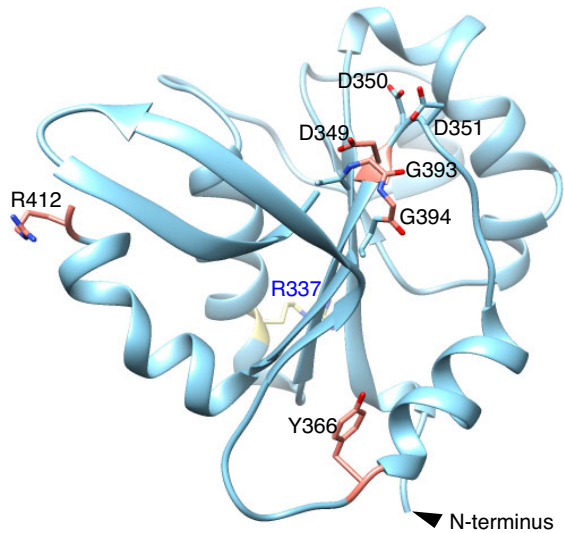


Figure S8

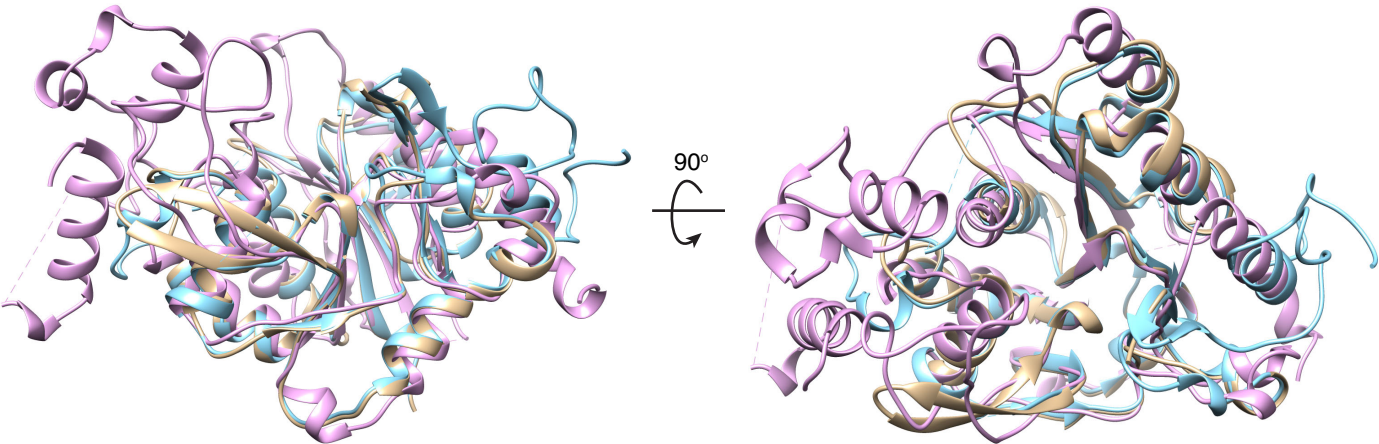


Figure S9

B3GNT2_HUMAN	-----MS-VGRRRIKLLGILM	15
B3GLCT_HUMAN	MRPPACWLLAPPALLALLTCSLAFGLASEDTKKEVKQSQDLEKSGISRKNIDLKGIVF	60
MFNG_MOUSE	-----	0
B3GNT2_HUMAN	MANV----FIYFIMEVSKSS-----S-----QEKNKGGEV IIPKEKFWKIS	52
B3GLCT_HUMAN	VIQSQSNSFHAKRAEQLKKSILKQAADLTQELPSVLLHQLAKQEGAWTILPLLPHFSVT	120
MFNG_MOUSE	-----	0
B3GNT2_HUMAN	TPPEAYWNREQE-----KLNRYNPILSMLTNQTGEAGRLSNISHLNYCE----	97
B3GLCT_HUMAN	YSRNSSWIFFCEEETRIQIPKLLLETLRRYDPSKEWFLGKALHDEEATI IHHYAFSENPTV	180
MFNG_MOUSE	-----	0
B3GNT2_HUMAN	-----PDLRVTSVVTGFN-----NLPDRFKD-	118
B3GLCT_HUMAN	FKYPDFAAGWALS IPLVNKLT KRLKSESLKSDFTIDLKHEIALYIWDKGGGPP LTPVPEF	240
MFNG_MOUSE	-----MHCRLFRGMAGAL---FTLL	17
B3GNT2_HUMAN	--FLLYLRCRN-----YSL LIDQPKCAK---KPFLL LAIKSL-TPHFARRQAIRES	164
B3GLCT_HUMAN	CTNDVDFYCATT-----FHSFLPL-----CRKPVKKKDI FVAVKTCCKK FHGDRIPIVKQT	290
MFNG_MOUSE	CVGLLSLRYHSSLSQRM IQGALRLNQRNPGPLELQLGDI FIAVKTTWAFHRSRLDLLLDT	77
	: : . * : : : * * : : :	
B3GNT2_HUMAN	WGQESNAGNQTVVRVFLGQTPPEDNHPDLS DMLKFE---SEKHQDILMWN YRDTFFNLS	221
B3GLCT_HUMAN	WESQAS-----LIEYSDY TENS IPTVDLG----IPN--TDRGHCGKTFA-----	329
MFNG_MOUSE	WVSRIR-----QQT FIFTD SPDERLQERLGP HLVVTNCSAEHSH PAL SCK-----	122
	* .. : : . *. . : : :	
B3GNT2_HUMAN	LKEVLF LRWVSTSCPDTEFVFKG DDD VFVNTHHILNYLNSLSKTKAKDLFIGDVIHNAGP	281
B3GLCT_HUMAN	----ILERFLNRSQDKTAWLVIV DDD TLISISRLQHLLSCYD--SGEPVFLGER YGY---	380
MFNG_MOUSE	----MAAEFD AFLVSGLRWFCHV DDD NYVNP KALLQLLKTFP--QDRDVYVGKPSLNRPI	176
	: .: :. *** :. : : *. . . : : *	
B3GNT2_HUMAN	HRDKKLKYYIPEVVYSGLYPPYAG GGG FLYSGHLALRLYHITDQVHL-----YP IDDV	334
B3GLCT_HUMAN	-----GLGTGGYSYITG GGG MVFSREAVRLLASKCRC-Y-----SNDAP DDM	422
MFNG_MOUSE	HASEL---QSKNR TKLVRFWFATG GAG FCINRQLALKMVPWASGSHFVDTSALIRL PDDC	233
	: : **.*: . . . : :	**
B3GNT2_HUMAN	YTGMC LQ-KLGLVPEKHKGFR----TFDIEEK N---KNNICSYVDL-----ML----	374
B3GLCT_HUMAN	VLGMCF-SGLGIPVTHSPLFHQARPVDY--PKDYLSHQVPISF H KHWNIDPVKVYFT-WL	478
MFNG_MOUSE	TVGYIIIECKLGGRLQPSPLFHS H LETQLLGA AQLPEQVTL SYGVF----EGKLNVIKLP	289
	* : ** * : . . : : *	
B3GNT2_HUMAN	-----V H SRKPQEMIDIWSQLQSAHLKC-----	397
B3GLCT_HUMAN	APSDEDKARQETQKGFREEL-----	498
MFNG_MOUSE	GPF----SHEEDPSRFRSLHCLLYPDT PWCPLLAAP	321
	:.. . : .	

Figure S10

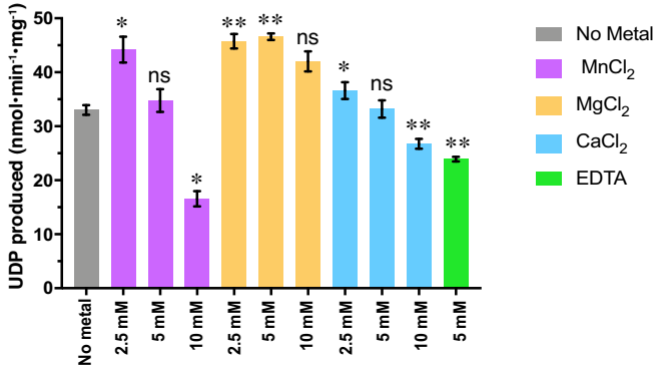
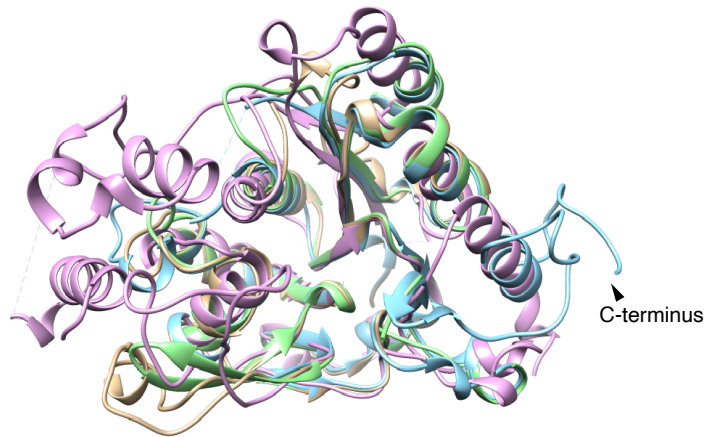


Figure S11

A



B

