



Supplement Figure 1: (A) BON4 cells transduced with various NEUROG3-expressing constructs with coexpressing GFP and stained with anti-FLAG antibody and DAPI. (Line - 25 μ m) **(B)** BON4 cells transfected with SLC5A1 promoter reporter and increasing concentration of NEUROG3 expression vector. **(C)** Western blot with anti-FLAG of BON4 cells expressing various NEUROG3 variants. **(D)** Gel shift assay was performed using nuclear extracts of BON4 cells expressing NEUROG3^{WT}, NEUROG3^{R93L}, NEUROG3^{R107S} and NEUROG3^{NULL}, and a biotin labeled duplex oligonucleotide, NEUROD1 E-box1. **(E)** BON4 NEUROG3^{DN} cells cotransfected with increasing concentrations of NEUROG3^{WT} and the NEUROD-1 E-box1 reporter.

Supplement Table 1:

PMID#	Mutation	Sex	Outcome	Age reported	Diarrhea - age of onset	IDDM - age of onset	C-peptide detected	In vitro test	NEUROD1 promoter; EMSA	Animal
16855267	p.R107S	M	Died; (MVTx)	3 yr	< 1 week	No	No	Yes	HeLa; Yes; Yes	Xenopus
16855267	p.R93L	M	Alive	8 yr	< 1 week	8 yr	No	Yes	HeLa; Yes; Yes	Xenopus
16855267	p.R93L	M	Alive	9 yr	< 1 week	8 yr	No	Yes	HeLa; Yes; Yes	Xenopus
21378176	p.E28X p.L135L	F	alive	7 yr	< 1 week	20 days	Yes	Yes	P19; Yes	Chick
27533310	p.L135P	F	Alive	24 yr	congenital	6 yr*	Yes	No	/	/
27533310	p.L135P	M	Alive	17 yr	congenital	13 yr	Yes	No	/	/
27533310	p.R107S	F	Alive	18 yr	congenital	14 yr	Yes	No	/	/
27533310	p.R107S	M	Alive	23 yr	congenital	No	Yes	No	/	/
24134759	p.171FsX68	F	Alive	20 mo	< 10 days	No	ND	No	/	/
21490072	p.E123X	F	Died; sepsis	10 mo	< 1 week	5.5 mo	Yes	Yes	293T; Yes	No
28940958	p.Q4X	M	Alive	5 yr	< 1 week	2 mo; insulin d/c 13 days; intermittent	Yes	No	/	/
28940958	p.Q4X	F	Alive	7 yr	< 2 week		Yes	No	/	/
26541772	p.R93L	M	Alive	2.5 yr	< 2 week	No	Yes	No		
28724572	p.R107S	M		16 yr	< 2 week	11 yr	ND	No	/	/
Current	p.P39PfsX38 p.H145Pfs*9	M	Alive	16 yr	2 week	3 yr	Yes	Yes	BON4; Yes; Yes	No
Current	4	F	Alive	14 yr	2 week	7 yr	Yes	Yes	BON4; Yes; Yes	No

Supplement Table 2: Oligonucleotides (vectors)

Cloning/EMSA Oligo names	Oligo sequences
NEUROG3 BamH1 5'	ACGGATCCATGACGCCTCAACCCTCGGGTG
NEUROG3-FLAG EcoR1 3'	ATGAATTCTCACTTGTTCATCGTCGTCCTTGTAGTCCAGAAAATCTGAGAAAGCCA
NEUROG3-H144PfsX94-FLAG EcoR1 3'	ATGAATTCTCACTTGTTCATCGTCGATCCTTGTAGTCCGGCTCCCGGCTC
p21 promoter (-592/14) Kpn1 5'	CGGGTACCTCAAGATGCTTTGTTGGGGTG
p21 promoter (-592/14) BglII 3'	CGAGATCTGCCAGCTCCGGCTCCACAAGGA

Supplement Table 3: Oligonucleotides (EMSA, qPCR)

Gene	5' Oligo	3' Oligo	qPCR Method
NEUROG3	CCTCGGAAGACGAAGTGACC	AGTGCCAACTCGCTCTTAGG	Sybr Green
NEUROG3-H144PfsX94	CGCAATCGAATGCACAACCT	CTATGCGCAGCGTTTGAGTC	Sybr Green
CHGA	CGGATCCTTTCCATTCTGAG	TCATCTTCAAACCGCTGTG	Sybr Green
PCSK1	TGGTTTGGAGTGGAATCACA	GTCCCGTGTTTGTCTCGTT	Sybr Green
Tubulin	GCTGCGGACCGAGAAACT	CTTGGCCCCGATCTGGTTG	Sybr Green

Supplement Table 4: Antibodies

Antibody	Species	Vendor	Isotype	Dilution	Mass	Cat. No.
FLAG	Human	Sigma Aldrich	Mouse IgG1	1/1000		F3165
NEUROG3	Human	DSHB	Mouse IgG	1/1000	23 kDa	F25A1B3
CHGA	Human	Santa Cruz Biotechnology	Goat IgG	1/1000	68-80 kDa	SC-1488
PCSK1	Human	ABNOVA	Mouse IgG	1/1000	70 kDa	H00005122-M02
TUBULIN	Human	DSHB	Mouse IgG	1/1000	55 kDa	AA4.3
Anti-rabbit IgG-HRP	Rabbit	GE Healthcare	IgG-HRP	1/2000		NA9340V
Anti-mouse IgG-HRP	Mouse	GE Healthcare	IgG-HRP	1/2000		NA9310V
Anti-goat IgG-HRP	Goat	Santa Cruz Biotechnology	IgG-HRP	1/2000		SC-2020
Ki67	Human	Leica Biosystems	Mouse IgG1	1/50	345 kDa	PA0230
Alexa Fluor 647	Mouse	Life Technologies	IgG	1/1000		A31571