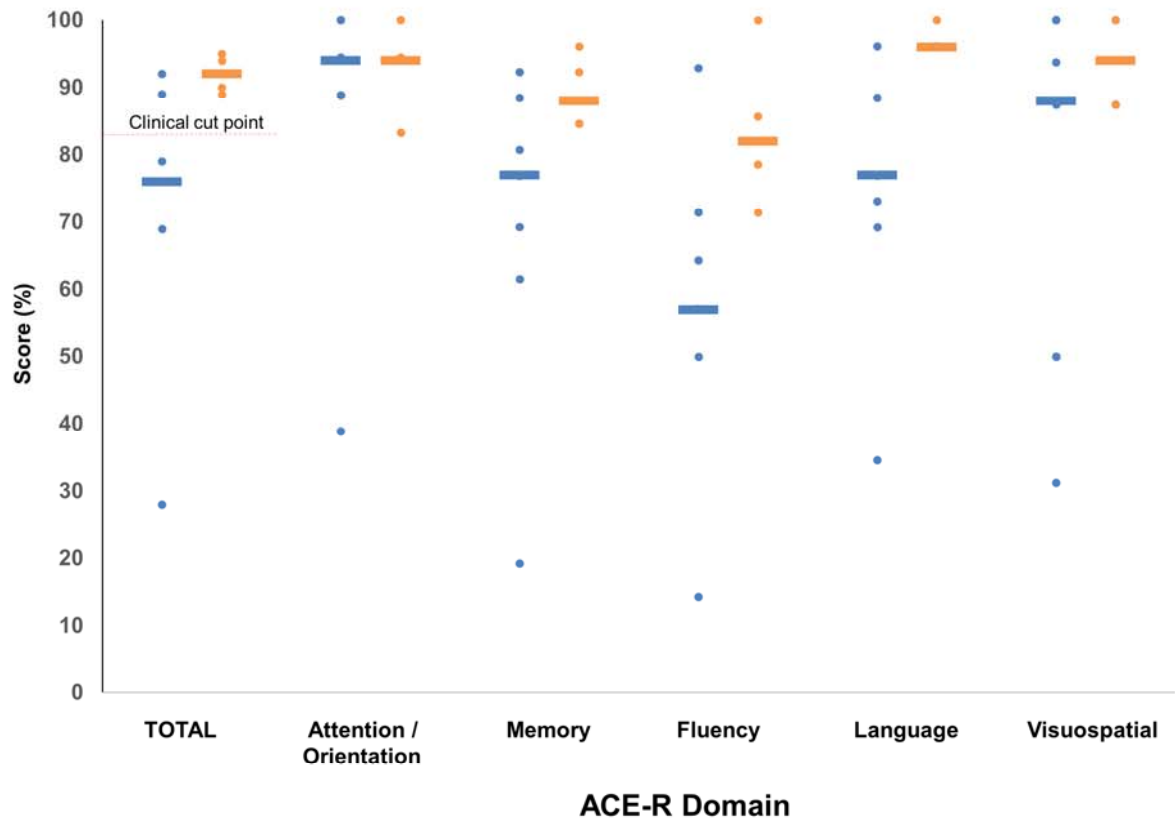


SUPPLEMENTARY DATA

Supplementary Figure S1. ACE-R in *KCNJ11* individuals (blue), n=7 and *INS* individuals (orange), n=4. Scores are presented as percentages of maximum possible score for each subsection. A total score of ≤ 82 is suggests clinically relevant cognitive impairment.



SUPPLEMENTARY DATA

Supplementary Table S1. Clinical characteristics of participants with *KCNJ11* mutations excluded from analysis

NK = not known, D=delayed, MS = Mainstream school, SS = Special school, LS = learning support, E = employed, UE = unemployed, MR = medically retired

Case ID (mutation)	Sex	Age (years)	Age at diabetes diagnosis (weeks)	Age at genetic diagnosis (years)	Age at SU transfer (years)	Treatment (total daily dose)	Development (type of delay)	Seizures (cause, if known)	Learning difficulties / support / education	Employment status (job)	Psychiatric history	Neurological features
<i>KCNJ11</i> 9 (V59M)	F	44	8	35	36	Glyburide (60mg)	D (global)	Yes – hypoglycaemia on insulin	Unable to speak / read / write, SS	UE	Nil	Spastic tetraparesis, able to follow 1-step commands only, unable to speak but able to vocalise noises.
<i>KCNJ11</i> 10 (L164P)	M	44	16	39	40	Glyburide (dose NK)	D (motor)	No	LS (through school), MS	MR (previously E - Post Office)	Bipolar disorder – two hospital admissions. On ziprasidone, fluoxetine, metoclopramide.	Cogging pursuit eye movement, hypomimia, reduced arm swing, cogwheel rigidity of arms, mild rigidity of legs. Difficulty with motor sequences.

Supplementary Table S2. Individual scores on subtests of the Wechsler Memory Scale 4th edition (WMS-IV), Wechsler Abbreviated Scale of Intelligence (WASI), Wechsler Adult Intelligence Scale 4th edition (WAIS-IV), and Colour Trails Test (CTT) I and II. ND = Not done.

Patient	Mutation	WMS-IV (Z-score)				WASI (Z-score)				WAIS-IV (Z-score)		Colour Trails Test	
		Verbal paired associates (immediate)	Verbal paired associates (delayed)	Visual reproduction (immediate)	Visual reproduction (delayed)	Vocabulary	Matrix reasoning	IQ (raw score)	IQ (Z-score)	Digit span	Cancellation	CTT 1	CTT 2
<i>KCNJ11</i> 1	G53S	-1.67	-2	-3	-1.67	-2.3	-1.8	65	-2.33	-2.33	-1	-2.5	-1.7
<i>KCNJ11</i> 2	R201H	0	0.33	0	0.33	0.5	-2.3	101	0.067	0.33	0	-0.9	0.8
<i>KCNJ11</i> 3	R201H	0.33	1	0.67	1.33	1.6	-3.2	99	-0.067	-1.33	0	-0.1	0.6
<i>KCNJ11</i> 4	R201C	0	0.33	-1.33	-1.33	-0.9	-0.9	86	-0.933	-2	-1	-1.3	0.1
<i>KCNJ11</i> 5	R201C	-1.67	-1.67	-1.33	-0.33	0.1	-4	76	-1.6	-1.67	-1	-2.1	-3
<i>KCNJ11</i> 6	V252G	-2	-2.67	-3	-3	-3.2	-4.8	55	-3	-3	-3	ND	ND
<i>KCNJ11</i> 7	V59M	0	0.66	-3	-2.33	-1.1	-4.4	65	-2.33	-3	-2	-3	-1.9
<i>KCNJ11</i> 8	V59M	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND
<i>INS</i> 1	C43F	0.67	1.33	0.33	1	2.1	0.6	124	1.6	0.33	2.67	0.13	0.6
<i>INS</i> 2	F48C	1.67	1.33	0.67	0	0.9	0.6	113	0.867	0	3	0.6	0.8
<i>INS</i> 3	G75C	-1	-0.67	1	0	0.2	0.8	109	0.6	-1	3	1.2	1.2
<i>INS</i> 4	H29D	0	0	-1.33	-0.67	-0.4	-0.7	90	-0.67	0	0.67	-1.1	-1

SUPPLEMENTARY DATA

Supplementary Table S3. Individual scores on Autism Spectrum Quotient (AQ), Hospital Anxiety and Depression Scale (HADS), Controlled Oral Word Association Test (COWAT), and subtests of the Visual Object and Space Perception battery (VOSP) and Addenbrooke's Cognitive Examination-Revised (ACE-R). ND = Not done.

Patient	Mutation	AQ raw score	HADS raw score		COWAT Z-score	VOSP Score (Pass/Fail)				ACE-R raw score					
			HADS A	HADS D		Incomplete Letters	Object Decision	Dot Counting	Cube Analysis	Total (/100)	Attention / orientation (/18)	Memory (/26)	Fluency (/14)	Language (/26)	Visuospatial (/16)
<i>KCNJ11 1</i>	G53S	33	9	1	-2.21	F	P	P	F	69	16	20	7	18	8
<i>KCNJ11 2</i>	R201H	12	6	6	-1.31	P	P	P	P	89	17	23	10	23	16
<i>KCNJ11 3</i>	R201H	17	9	2	0.79	P	P	P	P	92	18	21	13	25	15
<i>KCNJ11 4</i>	R201C	32	9	7	-1.91	P	F	P	P	76	18	16	8	20	14
<i>KCNJ11 5</i>	R201C	24	10	5	-1.72	P	F	P	F	79	16	18	9	20	16
<i>KCNJ11 6</i>	V252G	ND	14	8	ND	P	F	F	F	28	7	5	2	9	5
<i>KCNJ11 7</i>	V59M	25	9	7	-1.92	P	P	F	F	76	17	24	8	19	8
<i>KCNJ11 8</i>	V59M	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND
<i>INS 1</i>	C43F	5	1	0	-0.77	P	P	P	P	95	18	24	11	26	16
<i>INS 2</i>	F48C	15	12	4	-0.86	P	P	P	P	89	15	25	10	25	14
<i>INS 3</i>	G75C	26	6	1	-0.42	P	P	P	P	94	17	22	14	25	16
<i>INS 4</i>	H29D	30	17	7	-0.79	P	F	P	P	90	17	22	12	25	14