

Table S1. Clinical and genetic data of the 71 subjects

Case	Subtype	Birth weight (kg)	Gestational age (w)	Birth size [#]	Causative gene	Variant	At diagnosis				Treatment and evolution			
							FPG (mmol/L)	C peptide (µg/L)	HbA1c (%)	Combined with other problems	SU-R	Years of follow-up	Evolution	HbA1c at the last follow-up (%)
Onset at 0-6 m														
	P	4.2	36	AGA	KCNJ11	p Arg ₁₁₀ His	5.2	0.2	6.5		Yes	12	Still on SU	6.5
1	P	3.8	35	SGA	KCNJ11	p Arg ₁₁₀ His	5.5	0.2		Moderate anemia	Yes	12	Still on SU	6.5
2	P	3.5	35	SGA	KCNJ11	p Arg ₁₁₀ His	5.8	0.1		Moderate anemia	Yes	12	Still on SU	6.5
3	T	4.5	36	AGA	KCNJ11	p Arg ₁₁₀ His	5.0	0.3	6.0		Yes	12	Remission at 1 m	6.5
4	P	4.0	36	AGA	KCNJ11	p Arg ₁₁₀ His	5.2	0.2	6.2	DKA	Yes	12	Still on SU	6.5
5	P	4.2	36	SGA	KCNJ11	p Arg ₁₁₀ His	5.5	0.2		DKA	Yes	12	Still on SU	6.5
6	P	4.0	36	AGA	KCNJ11	p Arg ₁₁₀ Cys	5.2	0.2	6.0			12	INS for 1 y	Defaulted follow-up
7	P	4.0	36	AGA	KCNJ11	p Arg ₁₁₀ Cys		0.1	6.5	DKA	Yes	12	Still on SU	
8	T	4.5	36	AGA	KCNJ11	p Arg ₁₁₀ Cys		0.3		DKA	Yes	12	Remission at 1 m	6.5
9	P	3.8	36	SGA	KCNJ11	p Val ₁₁₀ Met ₁₁₀	5.5	0.2				12	INS for 1 y, developmental delay	Defaulted follow-up
10	P	4.0	36	SGA	KCNJ11	p Val ₁₁₀ Met ₁₁₀	5.2	0.2			Yes	12	Still on SU, mild developmental delay	6.5
11	P	4.0	36	SGA	KCNJ11	p Val ₁₁₀ Met ₁₁₀	5.5	0.2		DKA Developmental delay	Yes	12	Still on SU, developmental delay	
12	P	4.0	36	SGA	KCNJ11	p Val ₁₁₀ Met ₁₁₀		0.2		Congenital laryngeal wheezing, developmental delay	Yes	12	Still on SU, developmental delay	6.5
13	P	4.0	36	AGA	KCNJ11	p Val ₁₁₀ Met ₁₁₀	5.5	0.2		DKA, Congenital laryngeal wheezing,	Yes	12	Still on SU, developmental	6.5

developmental delay										delay
	P		AGA	KCNJ11	p Val Met	 	DKA	Yes		Still on SU, mild developmental delay
	T-R		AGA	KCNJ11	p Glu Lys	 				Remission at m Relapse y
	T		AGA	KCNJ11	p Glu Lys	 		Yes		Remission at m
	P		SGA	KCNJ11	p Cys Arg	 	DKA	No		Still on INS
	P		SGA	KCNJ11	p Ala Gly	 		Yes		Still on SU
	P		AGA	KCNJ11	p Tyr Cys	 	DKA	Yes		Still on SU
	P		AGA	KCNJ11	p Cys Arg	 		Yes		Still on SU
	T	 	SGA			 				Remission at m
	T	 	AGA	ABCC8	p Ile Phe	 		No		Remission at m
										Remission Relapse y and treated with SU, at age of y, treated with insulin
	T		SGA			 	Cutaneous hemangioma			Remission at m
	T	 	AGA	ABCC8	p Gly Glu p Ile Phe	   	DKA	Yes		Remission at m
	P	 	SGA	Chromosome q	pUPD	 	Convex forehead, upturned nose	No		Still on INS
	T		SGA	Chromosome q	pUPD	 	Left testicular hydrocele, macroglossia	Yes		Remission at m
	T	 	SGA	Chromosome q	pUPD	 	Moderate anemia	Yes		Remission at m
	T		SGA	Chromosome	Kb	 	Mild anemia	No		Remission at m

q11 duplication										
T-R		SGA	Chromosome p15p13	1.5-Mb deletion		No data	VSD, epicanthal folds, anteverted nares, broad eyebrows, distichiasis and long eyelashes	Yes		Remission at 18 m, seizures 24 m, developmental delay Relapse 2 y and treated with INS
P		AGA	Chromosome p16	1-Kb duplication			DKA	No		Still on INS
p		AGA	GCK	IVS 2, Thr 241 A						Diet
p		AGA	GCK	p Val 105 Met						Diet
P		AGA	GCK	p Thr 105 Met						Diet
P		SGA	INS	p Gly 31 Ser						Still on INS
P		AGA	BSCL2	p Glu 112 Hisfs p Ile 113			Hypertriglyceridemia, hyperinsulinemia, hepatomegaly and widespread acanthosis nigricans			Diet INS
P		SGA					Mild anemia			Still on INS
P		AGA					DKA			Still on INS
T-R		AGA					Moderate anemia			Remission at 1 m Relapse 2 y and treated with SU
T		SGA					Macroglossia			Remission at 1 m
T		AGA								Remission at 1 m
T		AGA								Remission at 1 m
Onset at 6-12 m										
P		SGA	KCNJ11	p His 191 Arg			DKA, Cutaneous hemangioma	Yes		Still on SU
P		SGA	INS	p Gly 31 Ser			DKA			Still on INS

	P		AGA	Chromosome 12p11.23 1.5 Mb duplication		DKA, Auricular malformation	Still on INS
	P		AGA				Still on INS
	P		AGA				Still on INS
	P		AGA			DKA	Still on NS
	P		AGA			DKA	Still on INS
	P		AGA				Still on INS
	P		AGA				Still on INS
Onset at 1-2 y							
	P		AGA			DKA	Still on INS
	P		AGA			DKA	Still on INS
	P		AGA			DKA	Still on INS
	P		AGA				Still on INS
	P		AGA			DKA	Still on INS
	p		AGA				Still on INS
	P		AGA			DKA	Still on INS
	P		AGA				Still on INS
	P		AGA				Still on INS
	P		AGA			DKA	Still on INS
Onset at 2-3 y							
	P		AGA	INS p Arg Cys		DKA	Still on INS
	P		AGA				Still on INS
	P		AGA				Still on INS
	P		AGA			DKA	Still on INS
	P		AGA			DKA	Still on INS
	P		AGA			DKA	Still on INS
	P		AGA			Mild DKA	Still on INS
	P		AGA			Mild DKA	Still on INS
	P		AGA			DKA	Still on INS

P, permanent T, transient T-R, transient-relapse FPG, fasting plasma glucose w, week d, day m, month y, year SU, sulphonylurea SU-R, sulphonylurea response AGA