

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 (%)
<i>Onset at 0-6 m</i>														
1	P	2.90	39	AGA	<i>KCNJ11</i>	p.Arg201His	38.9	0.07	9.5	/	Yes	8.6	Still on SU	6.2
2	P	1.20	35	SGA	<i>KCNJ11</i>	p.Arg201His	35.5	0.70	/	Moderate anemia	Yes	3.7	Still on SU	5.8
3	P	1.40	35	SGA	<i>KCNJ11</i>	p.Arg201His	33.5	0.28	/	Moderate anemia	Yes	3.7	Still on SU	5.8
4	T	2.30	36	AGA	<i>KCNJ11</i>	p.Arg201His	20.4	0.05	5.8	/	Yes	6.9	Remission at 4 m	5.1
5	P	2.60	38	AGA	<i>KCNJ11</i>	p.Arg201His	27.8	0.04	10.5	DKA	Yes	2.6	Still on SU	5.6
6	P	2.60	39	SGA	<i>KCNJ11</i>	p.Arg201His	40.2	0.11	10.7	DKA	Yes	1.7	Still on SU	5.8
7	P	2.30	36	AGA	<i>KCNJ11</i>	p.Arg201Cys	33.0	0.05	8.2	/	/	/	INS for 2 y	Defaulted follow-up
8	P	2.60	38	AGA	<i>KCNJ11</i>	p.Arg201Cys	33.8	0.24	5.7	DKA	Yes	1.5	Still on SU	6.1
9	T	3.40	38	AGA	<i>KCNJ11</i>	p.Arg201Cys	39.7	0.05	8.1	DKA	Yes	3.7	Remission at 8 m	5.1
10	P	2.50	38	SGA	<i>KCNJ11</i>	p.Val59Met	21.3	0.05	9.8	/	/	2.0	INS for 1.5 y, developmental delay	Defaulted follow-up
11	P	2.70	40	SGA	<i>KCNJ11</i>	p.Val59Met	28.5	0.90	14.0	/	Yes	5.5	Still on SU, mild developmental delay	5.9
12	P	2.30	37	SGA	<i>KCNJ11</i>	p.Val59Met	29.0	0.08	14.0	DKA Developmental delay	Yes	3.2	Still on SU, developmental delay	6.1
13	P	2.80	41	SGA	<i>KCNJ11</i>	p.Val59Met	39.8	0.06	13.1	Congenital laryngeal wheezing, developmental delay	Yes	2.4	Still on SU, developmental delay	5.8
14	P	2.70	35	AGA	<i>KCNJ11</i>	p.Val59Met	31.2	0.10	8.9	DKA, Congenital laryngeal wheezing,	Yes	3.1	Still on SU, developmental	6.0

													developmental delay	delay
15	P	2.90	40	AGA	<i>KCNJ11</i>	p.Val59Met	38.8	0.05	12.2	DKA	Yes	2.3	Still on SU, mild developmental delay	5.8
16	T-R	3.10	38	AGA	<i>KCNJ11</i>	p.Glu229Lys	22.9	0.05	6.9	/	/	7.5	Remission at 8m; Relapse 6 y	7.3
17	T	2.60	38	AGA	<i>KCNJ11</i>	p.Glu229Lys	22.5	0.05	5.0	/	Yes	4.8	Remission at 4 m	4.9
18	P	2.50	39	SGA	<i>KCNJ11</i>	p.Cys42Arg	33.5	0.08	14.0	DKA	No	2.7	Still on INS	8.6
19	P	2.30	40	SGA	<i>KCNJ11</i>	p.Ala18Gly	24	0.10	4.5	/	Yes	1.4	Still on SU	6.1
20	P	3.10	38	AGA	<i>KCNJ11</i>	p.Tyr330Cys	29.5	0.12	11.7	DKA	Yes	1.9	Still on SU	5.8
21	P	2.40	36	AGA	<i>KCNJ11</i>	p.Cys42Arg	19.9	0.21	9.1	/	Yes	1.5	Still on SU	5.9
22	T	2.20	37	SGA	/	/	24.3	0.05	7.2	/	/	7.2	Remission at 4.5 m	4.9
23	T	2.50	38	AGA	<i>ABCC8</i>	p.Ile395Phe	32.6	0.33	4.0	/	No	8.3	Remission at 5.5 m	5.3
24	T-R	3.00	38	AGA	<i>ABCC8</i>	p.Gly1255Ser	44.0	0.42	7.9	/	/	12.2	Remission 5.5 m; Relapse 8y and treated with SU, at age of 11y, treated with insulin.	8.9
25	T	1.38	30	SGA	/	/	24.0	0.71	4.7	Cutaneous hemangioma	/	2.1	Remission at 3.5 m	5.7
26	T	2.50	38	AGA	<i>ABCC8</i>	p.Gly251Glu p.Ile467Phe	27.8	0.26	9.0	DKA	Yes	1.3	Remission at 9 m	5.8
27	P	2.00	39	SGA	Chromosome 6q24	pUPD	15.0	0.36	7.8	Convex forehead, upturned nose	No	7.8	Still on INS	6.6
28	T	1.80	38	SGA	Chromosome 6q24	pUPD	32.2	0.40	4.6	Left testicular hydrocele, macroglossia	Yes	4.0	Remission at 4 m	5.3
29	T	2.00	41	SGA	Chromosome 6q24	pUPD	18.3	0.05	6.2	Moderate anemia	Yes	8.2	Remission at 3 m	5.1
30	T	1.80	41	SGA	Chromosome	154-Kb	31.0	0.05	6.1	Mild anemia	No	4.6	Remission at 7.5 m	4.8

					6q24.2	duplication								
31	T-R	1.90	39	SGA	Chromosome 1p36.23p36.12	11.76-Mb deletion	15.8	0.45	No data	VSD, epicanthal folds, anteverted nares, broad eyebrows, distichiasis and long eyelashes	Yes	5.9	Remission at 10 m, seizures 1y2m, developmental delay; Relapse 4.5 y and treated with INS	6.3
32	P	2.80	38	AGA	Chromosome 17p13.3	183-Kb duplication	27.8	0.06	12.8	DKA	No	3.1	Still on INS	7.9
33	p	2.70	38	AGA	<i>GCK</i>	IVS4+2T>A	8.0	1.36	5.2	/	/	6.4	Diet	6.7
34	p	2.70	38	AGA	<i>GCK</i>	p.Val182Met	8.5	1.58	5.8	/	/	7.6	Diet	6.5
35	P	2.95	38	AGA	<i>GCK</i>	p.Thr228Met	7.6	1.07	4.8	/	/	1.8	Diet	6.5
36	P	2.50	41	SGA	<i>INS</i>	p.Gly32Ser	48.4	0.66	9.8	/	/	6.2	Still on INS	8.5
37	P	2.50	37	AGA	<i>BSCL2</i>	p.Glu189* p.Ile262Hisfs*12	21.0	14.2	7.4	Hypertriglyceridemia, hyperinsulinemia, hepatomegaly and widespread acanthosis nigricans	/	5.5	Diet + INS	7.7
38	P	2.70	40	SGA	/	/	35.0	0.05	6.1	Mild anemia	/	3.5	Still on INS	7.3
39	P	2.80	38	AGA	/	/	23.0	0.39	5.9	DKA	/	4.5	Still on INS	8.1
40	T-R	2.80	38	AGA	/	/	22.0	0.26	7.8	Moderate anemia	/	11.0	Remission at 4 m; Relapse 8 y and treated with SU	7.9
41	T	2.40	38	SGA	/	/	27.8	0.10	6.1	Macroglossia	/	5.1	Remission at 4 m	5.2
42	T	2.60	38	AGA	/	/	28.2	1.00	6.5	/	/	4.5	Remission at 4 m	4.7
43	T	2.70	38	AGA	/	/	15.5	0.92	4.5	/	/	4.9	Remission at 7.5m	5.1
Onset at 6-12 m														
44	P	2.50	40	SGA	<i>KCNJ11</i>	p.His46Arg	30.4	0.05	14.0	DKA, Cutaneous hemangioma	Yes	2.1	Still on SU	6.0
45	P	2.70	40	SGA	<i>INS</i>	p.Gly32Ser	27.0	0.08	13.9	DKA	/	2.2	Still on INS	7.9

46	P	3.00	38	AGA	Chromosome 4p15.1	4.78-Mb duplication	29.1	0.25	9.9	DKA, Auricular malformation	/	4.1	Still on INS	8
47	P	3.30	37	AGA	/	/	21.9	0.12	8.7	/	/	2.3	Still on INS	5.8
48	P	3.00	38	AGA	/	/	25.6	0.63	8.6	/	/	5.5	Still on INS	8.1
49	P	3.50	39	AGA	/	/	31.5	0.13	8.6	DKA	/	1.9	Still on NS	7.2
50	P	3.10	38	AGA	/	/	33.0	0.26	10.5	DKA	/	5.0	Still on INS	7.0
51	P	3.30	38	AGA	/	/	39.0	0.06	8.5	/	/	2.3	Still on INS	6.8
52	P	2.80	37	AGA	/	/	29.0	0.18	9.9	/	/	2.4	Still on INS	7.5
Onset at 1-2 y														
53	P	3.90	39	AGA	/	/	37.0	0.12	11.9	DKA	/	6.0	Still on INS	7.6
54	P	3.30	38	AGA	/	/	18.1	0.08	11.2	DKA	/	1.8	Still on INS	7.9
55	P	3.30	39	AGA	/	/	27.8	0.06	14.0	DKA	/	4.5	Still on INS	8.5
56	P	2.70	36	AGA	/	/	21.3	0.05	12.5	/	/	3.7	Still on INS	8.2
57	P	2.50	38	AGA	/	/	33.2	0.17	12.0	DKA	/	2.3	Still on INS	8.9
58	p	3.10	38	AGA	/	/	27.5	0.11	9.8	/	/	2.1	Still on INS	6.8
59	P	3.30	39	AGA	/	/	27.8	0.06	14.0	DKA	/	4.6	Still on INS	8.5
60	P	3.70	39	AGA	/	/	27.5	0.035	10.6	/	/	6.6	Still on INS	8.2
61	P	3.40	40	AGA	/	/	22.9	0.11	12.3	/	/	4.7	Still on INS	7.9
62	P	3.20	38	AGA	/	/	32.2	0.15	11.7	DKA	/	1.7	Still on INS	7.0
Onset at 2-3 y														
63	P	3.20	39	AGA	INS	p.Arg89Cys	28.0	0.28	14.0	DKA	/	2.4	Still on INS	7.6
64	P	3.75	39	AGA	/	/	28.2	0.20	11.3	/	/	7.7	Still on INS	8.1
65	P	3.40	39	AGA	/	/	26.5	0.14	12.8	/	/	4.5	Still on INS	7.9
66	P	3.90	39	AGA	/	/	29.0	0.05	14.0	DKA	/	3.6	Still on INS	7.5
67	P	2.80	38	AGA	/	/	28.9	0.30	9.3	DKA	/	6.3	Still on INS	6.2
68	P	2.60	37	AGA	/	/	29.8	0.05	12.5	DKA	/	4.0	Still on INS	7.8
69	P	2.40	36	AGA	/	/	26.7	0.38	14.0	Mild DKA	/	2.5	Still on INS	7.2
70	P	2.60	38	AGA	/	/	25.2	0.15	8.6	Mild DKA	/	2.7	Still on INS	8.5
71	P	2.90	37	AGA	/	/	25.9	0.08	14.0	DKA	/	2.8	Still on INS	7.8

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, appropriate for gestational age; SGA, small for gestational age; pUPD, paternal uniparental disomy; DKA, diabetic ketoacidosis; INS, insulin; VSD, ventricular septal defect.

The birth size was defined according to Fenton Preterm Growth Charts.