

**Supplementary Table 1.** Characteristics of mutated genes of MODY cohort

Characteristic	GCK (n=13)	HNF1A (n=10)	KCNJ11 (n=2)	ABCC8 (n=2)	HNF1B (n=2)	HNF4A (n=4)	SLC19A2 (n=1)
<b>Clinical data</b>							
Gender, boys	10	3	2	2	0	2	1
Age at diagnosis, yr	5.7±4.1	10.7±4.5	0.5±0.4	6.1±8.5	4.2±5.7	5.4±2.4	13.0
Glycemia at diagnosis, mg/dL	133±56	199±186	635±248	100±34	108±NA	180±210	615
HbA1c at diagnosis, %	6.3±0.6	7.9±2.7	10.8±2.2	6.0±1.6	5.4±NA	6.5±3.0	7.9
IDAA1c score	7.0±1.7	7.6±2.1	8.3±2.8	6.0±1.5	6.7±1.9	6.3±2.3	8.15
GTAA1c score	4.2±1.2	4.2±1.8	5.0±1.6	2.2±NA	2.7±0	2.9±1.2	5.27
TIR <sub>70-180</sub> , %	80.7±8.0	70.5±23.2	69.1±18.5	90.0±NA	88.3±2.3	83.6±16.8	NA
<b>Antidiabetic treatment</b>							
Insulin	1	4	1	0	1	1	1
Oral antidiabetic	1	2	1	1	0	1	0
<b>DIAMODIA criteria</b>							
Strong criteria	13/13	9/9	2/2	2/2	1/1	4/4	1/1
Absence of anti-islet antibodies	12/13	7/10	1/2	2/2	2/2	3/4	1/1
IDAA1c <9	8/9	7/9	1/2	1/1	2/2	0/4	0/1
GTAA1c <4.5	0/13	0/10	1/2	1/2	1/2	4/4	0/1
Age at diagnosis ≤6 mo							
Weak criteria	11/13	9/10	1/2	1/2	2/2	4/4	1/1
First-degree relative with diabetes							
<b>C-peptide</b>							
Positivity	13/13	9/10	0/2	2/2	2/2	4/4	1/1
Extra-pancreatic manifestations	3/12	5/10	0/2	0/1	2/2	1/3	1/1
<b>Absence of</b>							
KA at diagnosis	12/13	9/10	0/2	2/2	2/2	4/4	1/1
Neonatal hypoglycemia	0/12	2/10	1/2	1/2	0/2	2/4	1/1

Values are presented as mean ± standard deviation.

MODY, maturity-onset diabetes of the young; GCK, glucokinase; HNF1A, HNF1 homeobox A; KCNJ11, potassium inwardly rectifying channel subfamily J member 11; ABCC8, ATP binding cassette subfamily C member 8; HNF1B, HNF1 homeobox B; HNF4A, hepatocyte nuclear factor 4 alpha; SLC19A2, solute carrier family 19 member 2; NA, not applicable; HbA1c, glycosylated hemoglobin; IDAA1c, insulin-dose adjusted A1c; GTAA1c, glycemic target-adjusted A1c; TIR<sub>70-180</sub>, time in range (70–180 mg/dL); DIAMODIA, DIAGnose MOnogenic DIAbetes; KA, ketoacidosis.