Supplementary Table 1: Clinical characteristics of the six NDM subjects sequenced for the *INS* promoter, negative for any missense mutations in *KCNJ11*, *ABCC8* and *INS*, or chromosome 6q24 abnormalities.

Proband-	**1**	**2**	**3**	**4**	**5**	**6**
Sex	F	F	F	M	M	M
Ethnic group	Turkish	Turkish	Morocco	Algeria	Iraq	Iraq
Birth weight (g/percentile)	1540/<3	1760/<3	1890/<3	2890/<2	NA	2240/<3
Gestation (wk)	37	40	37	39	42	41
Status at presentation						
NDM onset (d [days] m[months] or Y[years])	1d	2d	11d	3Y4m	1d	31d
Presentation	SGM	SGM	SGM	SPUPD and weight loss	NA	SGM and weight loss
Blood glucose (mmol/l)	14.8	10	28.8	72	NA	10.7
Antibodies	0	0	0	NA	0	NA
Pancreatic ultrasonography	N	N	N	NA	Aplasia	Agenesis
Neuropsychological assessment	N	N	N	Mental retardation	N	N
Other manifestations	No	No	Asthma	Macrocrania, megalencephalia, renal malformation, dysmorphy, visual defect	NA	Squeletal dysplasia
Age at remission (m[months])	5m / 24m	1.5m	NA	NA	NA	NA
Age at recurrence (m[months])	22m / 28m	NA	NA	NA	NA	NA
Current status						
Age (m[months] or Y[years])	28m	8Y3m	7Y7m	16Y	16Y6m	6Y3m
Weight (kg/percentile)	11.8/50	NA	21.5/25	50.6/10	48.3/3	NA
Type of insulin therapy	Injection	NA	Pump	Injection	Pump	Pump
Dose (units/day)	1.2	NA	20	50	NA	NA
Dose (units/kg/day)	0.08	NA	0.9	1	NA	NA
HbA1c in % (nl<5,9%)	6.2	NA	8.5	8	7.3	6.7

NA: not available / **N**: normal
SGM: systematic glucose monitoring / **SPUPD**: polyuria-polydipsia

Supplementary Table 2: Clinical and metabolic characteristics of the Family-1.

Family-1	**Proband**		**Sister**	**Father**	**Mother**	**Paternal grandfather**	**Paternal grandmother**
Sex	F		F	M	F	M	F
Ethnic group	Turkish		Turkish	Turkish	Turkish	Turkish	Turkish
Current status							
Age (m[months] or y[years])	28m		4y	26y	28y	53Y	51Y
BMI (kg.m^{-2}/percentile)	15.4/10		15.2/50	27.8	20.6	30	37
History of diabetes	Yes		No	No	No	Yes	Yes
Insulin treatment	Yes		No	No	No	No	No
Oral anti-diabetic drugs	No		No	No	No	Biguanide	Biguanide + Sulfonylureas
HbA1c (%)	5.4-5.6 / 6.6-6.2 *		5.1	5.2	4.7	6.9	7.3
Neuropsychological assessment	N		N	N	N	N	N
Oral glucose tolerance test	**NB1**	**NB2**					
T0: Glucose (mmol/l)/Insulin (mUI/l)/ Peptide C (ng/ml)	2.8 / 2 / 0.7	2.8 / 1 / 0.3	3.6 / NA / 1.1	4.7 / 6 / 1.6	4.2 / 5 / 1.6	NA	NA
T30: Glucose/Insulin/Peptide C	7 / 9 / 1.8	6.4 / 6 / 0.8	NA	8 / 70 / 7.4	6.2 / 34 / 3.8	NA	NA
T60: Glucose/Insulin/Peptide C	7.4 / 11 / 2.1	7.2 / 6 / 0.7	NA	7.7 / 57 / 7.5	6 / 32 / 5	NA	NA
T90: Glucose/Insulin/Peptide C	6.7 / 13 / 2.2	5.7 / 4 / 0.9	NA	6.4 / 37 / 6.9	7.8 / 51 / 7.8	NA	NA
T120: Glucose/Insulin/Peptide C	2.8 / 3 / 1	6.5 / 4 / 0.9	NA	6 / 38 / 5.7	7.5 / 50 / 7.4	NA	NA
T180: Glucose/Insulin/Peptide C	3.2 / 2 / 0.7	5.1 / 4 / 0.8	NA	6 / 41 / 7.2	7.2 / 51 / 8.7	NA	NA
Proinsulin (pmol/l) / Proinsulin-Insulin ratio (%)	**NB1**	**NB2**					
T0 [2.5 – 14 / 5 – 50][†]	1.7 / 5.6	NA	2.1 / 9.5	5.1 / 20	2.5 / 2.6	NA	NA
T30 [8.5 – 56 / 4 – 24][†]	3.9 / 18	NA	6.7 / 12	18.8 / 7	5.9 / 5	NA	NA
T120 [12 – 70 / 12 – 76][†]	3.4 / 28	NA	9.9 / 41	25.6 / 18	18.4 / 10	NA	NA

***** HbA1C at TNDM remissions / HbA1c at diabetes recurrences

[†] [normal values] (personal communication from Dr. Didier Chevenne [Robert-Debré Hospital, Paris, France])

NA: not available / **N**: normal

NB1: measurement performed during the first remission phase (with no insulin treatment)

NB2: measurement performed during the second remission phase (with no insulin treatment)

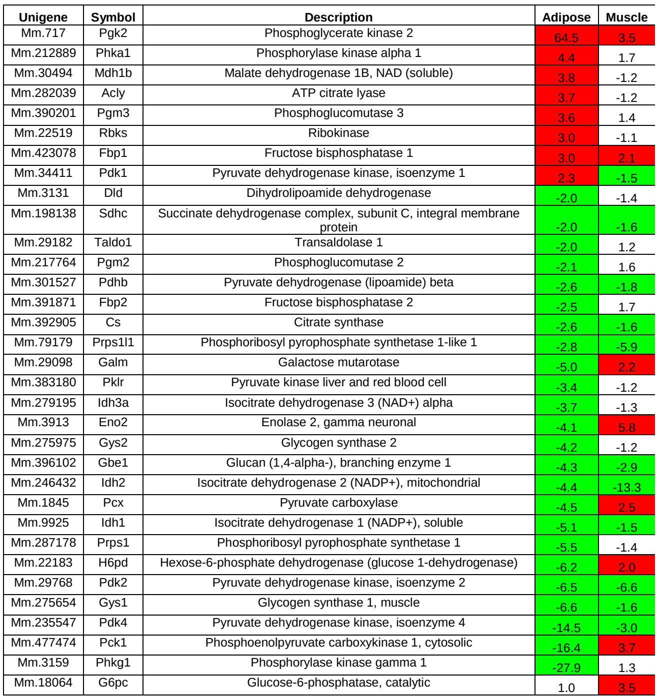
Supplemental Table 3: Expression of all the *KLF* genes in human islets (n = 3) and sorted beta cells (n = 3), through microarray

	Human beta cells		**Human islets**	
Gene	Median level of expression (MLEβ)	% genes which level is below (Eβ)	Median level of expression (MLEi)	% genes which level is below (Ei)
KLF1	NE	NE	NE	NE
KLF2	**659.8**	**74%**	**2581.7**	**93%**
KLF3	48.5	30%	48.9	33%
KLF4	**245.2**	**46%**	**960.4**	**80%**
KLF5	**380.2**	**59%**	**439.9**	**61%**
KLF6	**1839.6**	**90%**	**3532.0**	**95%**
KLF7	12.9	5%	NE	NE
KLF8	NE	NE	NE	NE
KLF9	**386.8**	**59%**	**690.3**	**73%**
KLF10	53.1	5%	NE	NE
KLF11	**339.5**	**55%**	**358.3**	**55%**
KLF12	NE	NE	NE	NE
KLF13	**507.8**	**67%**	**551.1**	**67%**
KLF14	NE	NE	NE	NE
KLF15	14.5	6%	20.2	16%
KLF16	NE	NE	NE	NE

NE: not expressed

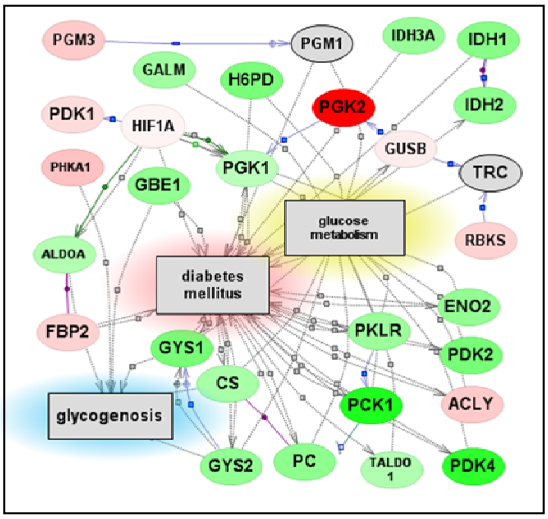
Supplementary Figure 1:

A.



Unigene	Symbol	Description	Adipose	Muscle
Mm.717	Pgk2	Phosphoglycerate kinase 2	64.5	3.5
Mm.212889	Phka1	Phosphorylase kinase alpha 1	4.4	1.7
Mm.30494	Mdh1b	Malate dehydrogenase 1B, NAD (soluble)	3.8	-1.2
Mm.282039	Acly	ATP citrate lyase	3.7	-1.2
Mm.390201	Pgm3	Phosphoglucomutase 3	3.6	1.4
Mm.22519	Rbks	Ribokinase	3.0	-1.1
Mm.423078	Fbp1	Fructose bisphosphatase 1	3.0	2.1
Mm.34411	Pdk1	Pyruvate dehydrogenase kinase, isoenzyme 1	2.3	-1.5
Mm.3131	Dld	Dihydrolipoamide dehydrogenase	-2.0	-1.4
Mm.198138	Sdhc	Succinate dehydrogenase complex, subunit C, integral membrane protein	-2.0	-1.6
Mm.29182	Taldo1	Transaldolase 1	-2.0	1.2
Mm.217764	Pgm2	Phosphoglucomutase 2	-2.1	1.6
Mm.301527	Pdhb	Pyruvate dehydrogenase (lipoamide) beta	-2.6	-1.8
Mm.391871	Fbp2	Fructose bisphosphatase 2	-2.5	1.7
Mm.392905	Cs	Citrate synthase	-2.6	-1.6
Mm.79179	Prps1l1	Phosphoribosyl pyrophosphate synthetase 1-like 1	-2.8	-5.9
Mm.29098	Galm	Galactose mutarotase	-5.0	2.2
Mm.383180	Pklr	Pyruvate kinase liver and red blood cell	-3.4	-1.2
Mm.279195	Idh3a	Isocitrate dehydrogenase 3 (NAD+) alpha	-3.7	-1.3
Mm.3913	Eno2	Enolase 2, gamma neuronal	-4.1	5.8
Mm.275975	Gys2	Glycogen synthase 2	-4.2	-1.2
Mm.396102	Gbe1	Glucan (1,4-alpha-), branching enzyme 1	-4.3	-2.9
Mm.246432	Idh2	Isocitrate dehydrogenase 2 (NADP+), mitochondrial	-4.4	-13.3
Mm.1845	Pcx	Pyruvate carboxylase	-4.5	2.5
Mm.9925	Idh1	Isocitrate dehydrogenase 1 (NADP+), soluble	-5.1	-1.5
Mm.287178	Prps1	Phosphoribosyl pyrophosphate synthetase 1	-5.5	-1.4
Mm.22183	H6pd	Hexose-6-phosphate dehydrogenase (glucose 1-dehydrogenase)	-6.2	2.0
Mm.29768	Pdk2	Pyruvate dehydrogenase kinase, isoenzyme 2	-6.5	-6.6
Mm.275654	Gys1	Glycogen synthase 1, muscle	-6.6	-1.6
Mm.235547	Pdk4	Pyruvate dehydrogenase kinase, isoenzyme 4	-14.5	-3.0
Mm.477474	Pck1	Phosphoenolpyruvate carboxykinase 1, cytosolic	-16.4	3.7
Mm.3159	Phkg1	Phosphorylase kinase gamma 1	-27.9	1.3
Mm.18064	G6pc	Glucose-6-phosphatase, catalytic	1.0	3.5

B.



Supplementary Figure 1: *Klf11*$^{-/-}$ mice display alterations in metabolic enzymes in adipose tissue. Data from global expression profiling (GeneChip, Affymetrix®, Santa Clara, CA) obtained from both wild-type and *Klf11*$^{-/-}$ adipose tissue was filtered using the Pathway Studio® software (Ariadne Genomics, Inc., Rockville, MD) to detect alterations in diabetes-associated genes or those involved in glucose metabolism. **A.** Metabolic genes, differentially expressed in *Klf11*$^{-/-}$ adipose and muscle tissue are shown as fold-change of expression in *Klf11* knock-out (KO)/wild-type (WT) mice. Gene symbols and Unigene numbers are also provided. **B.** Processing of expression data using the Pathway Studio® pathway reconstruction algorithm illustrates how these genes are linked in single pathway that converge on two relevant processes, glucose metabolism/glycogenosis and diabetes (grey boxes). Shades of green indicate genes that were down-regulated in the *Klf11*$^{-/-}$ animals compared to wild-type, whereas shades of red illustrate genes that were up-regulated in *Klf11*$^{-/-}$ versus wild-type. Grey circles represent 2 nodes (PGM1 and TRC) automatically added by Pathway Studio® to link the pathway. These results demonstrate that, in addition to defective insulin transcription in the pancreas, *Klf11*$^{-/-}$ mice also display alterations in gene expression within peripheral tissue.