Supplemental Table 1. Clinical and long-term outcome characteristics and genetic data of non-pancreatectomised CHI patients excluded from the study.

#Pat	Gender/ Gestational age (w)/ Z-score birth wight	Age of onset (d)	Start - Stop DZX	Age removal NGT (y)	DZX responsive	Resolved (y)	Current Medica-tion	Out-come	Cu-rrent age (y)	Cu-rrent BMI	PET-TC scan	*ABCC8*/SUR1 variant	Zygosity (segre-gation)	Pathogenicity (ACMG criteria[1])	First report (HGMD[2])
10	F/38/1	1	39d-0.6y	4	No	4	Insulin	D	20	31	NA	c.4078G>T/ p.(Val1360Leu)	Het (NA)	LP	This study
15	F/41/-0.1	72	72d-2y	2	Yes	2	Off	NG	16		NA	c.-49G>C	Het (Mo)	B	Bellanné-Chantelot *et al.* 2010(1)
25	F/31.6/-0.2	1	40d-0.5y	0.2	Yes	0.5	Off	NG	4	16.7	NA	c.1332+4C>T	Het (Fa)	VUS	This study
26	F/40/2.3	300	NA-NA	NA	NA	1	Off	NG	2	16	NA	c.2797C>T/ p.(Arg933Ter)	Het (Fa)	P	Fernández-Marmiesse *et al.* 2006(2)

1: American College of Medical Genetics (ACMG) criteria revised in Varsome, last revision May 2022. 2: first time reported, revised in Human Gene Mutation Database (HGMD), last revision December 2021; #Pat: patient number; w: weeks; d: days; y: years old; F: female; NA: non-available data; D: diabetes; NG: Normoglycemia; Het: heterozygous; Comp het: compound heterozygous; Fa: father; Mo: mother; P: pathogenic; LP: likely pathogenic; VUS: variant of uncertain significance; B: benign.

Supplemental Table 2. Description and *in silico* prediction studies of the variants of uncertain significance in the *ABCC8* gene described in this study.

***ABCC8* cDNA change (NM_000352.6)**	**SUR1 Amino acid change (NP_000343.2)**	**Type of variants / consequence**	**Pathogenicity (ACMG criteria[1])**	**SUR1 domain**	**Amino acid conservation (Clustal Omega)**	**Change of amino acid properties**	**Protein stability (I-Mutant[2], sequence)**	**Protein stability (I-Mutant[2], structure 6C3P)**	**Patient**	**First report (HGMD[3])**
c.5C>T	p.(Pro2Leu)	SNV/missense	VUS	N-terminal	++	Yes	0.19	-0.53	21 (Het/Comp het?)	This study
c.4720G>A	p.(Ala1574Thr)	SNV/missense	VUS	NBD2	+++	Yes	-0.85	-1.80	7 (Comp het)	This study

1: American College of Medical Genetics (ACMG) criteria revised in Varsome, last revision May 2022. 2: I-Mutant website tool to predict protein stability change due to missense variants, result in free energy change value (DDG), DDG<0: decreased stability, DDG>0: increased stability; 3: first time reported, revised in Human Gene Mutation Database (HGMD), last revision December 2021; VUS: variant of uncertain significance; NBD: nucleotide binding domain; +++: highly conserved, 0 changes in analysed species; ++: conserved, 1-2 changes in analysed species.

References:

1. **Bellanné-Chantelot C, Saint-Martin C, Ribeiro M-J, Vaury C, Verkarre V, Arnoux J-B, Valayannopoulos V, Gobrecht S, Sempoux C, Rahier J, Fournet J-C, Jaubert F, Aigrain Y, Nihoul-Fékété C, Lonlay P de.** ABCC8 and KCNJ11 molecular spectrum of 109 patients with diazoxide-unresponsive congenital hyperinsulinism. *J. Med. Genet.* 2010;47(11):752–759.

2. **Fernández-Marmiesse A, Salas A, Vega A, Fernández-Lorenzo JR, Barreiro J, Carracedo A.** Mutation spectra of ABCC8 gene in Spanish patients with Hyperinsulinism of Infancy (HI). *Hum. Mutat.* 2006;27(2):214.