CNS feature	Pre SU transfer (n)	Post SU transfer (n)	Post transfer only (n)	Improvement on SU (n)
Any	13	13	0	7
DD	10	10	1	3
LD	7	11	2	1
ADHD	6	8	2	1
Epilepsy	4	3	2	2**§
Muscle weakness	2	2	0	0
Anxiety	2	2	0	0
Sleep problems	2	2	1	0
Spasticity	2	2	0	0
Autism	0	0	0	N/A
Other*	5	4	0	1***

Supplementary Table S1. Neurological features present before and after transfer to sulfonylureas, and features that improved on SU transfer

*'Other' CNS features prior to sulfonylurea transfer in addition to the specific features listed consisted of obsessive-compulsive disorder (OCD) with mild Tourette's, encopresis, hypertonia, hypotonia and an abnormal electroencephalogram (EEG) (in the absence of a diagnosis of epilepsy). These features were also present after sulfonylurea transfer with the exception of hypotonia (but not known if this was tested).

**both individuals had seizures at time of diagnosis only which may have been attributable to cerebral oedema (Table S2)

§individuals treated with anti-epileptic medication not included as 'improved' (Table S2)

***improved background on EEG

CNS = central nervous system, SU = sulfonylurea, DD = developmental delay, LD = learning difficulties, ADHD = attention deficit hyperactivity disorder

Mutation in *ABCC8* gene	**Age at diagnosis of diabetes (weeks)**	**Age at transfer to SU (years)**	**Clinical history**	**Other neurological features present in addition to seizures / epilepsy**	**Neurological features improved on transfer to SU**
Individuals in whom metabolic disturbance at diagnosis may have contributed to seizures					
P45L/G1401R	6	8	Diabetic ketoacidosis at 6 weeks of age with severe dehydration, reduced consciousness, opisthotonus and partial seizures - diagnosed with cerebral edema (23)	Muscle weakness, hypertonia, spasticity, DD, LD, and sleep problems	Improvements in sleep, speech, concentration and schoolwork noted by parents and teachers. No epilepsy at most recent follow-up.
V215A/V215A	9	0.5	Focal seizures around time of diagnosis: in left arm 2 days before admission and in left arm and leg 2 days after admission	DD (mild), LD (mild)	No seizures since 2 months of age but on antiepileptic drugs. Other features (DD / LD) identified only after SU transfer.
V86G/N	5	3	Seizures only at time of diagnosis: none since.	DD, LD, ADHD, anxiety	Slight improvement. No anxiety post SU transfer and no further seizures. Main problem currently is speech delay / difficulties at school.
Individuals in whom seizures are attributable to another (non-metabolic) cause					
L1295F/N	12	6	One seizure due to starting treatment with dexmethylphenidate for ADHD (no further seizures on stopping drug)	DD, LD, ADHD, muscle weakness	No change.
L135P/N	6	10	Viral meningoencephalitis at 6 weeks of age, treated with Depakine 200-300mg at clinic follow-up prior to SU transfer	Spastic paraplegia, DD, LD, sleep problems	No change. Epilepsy not reported at recent follow-up but has had treatment with antiepileptic drugs

Supplementary Table S2. Clinical details of patients in whom seizures / epilepsy may have been attributable to factors other than the genetic mutation. SU = sulfonylurea, DD = developmental delay, LD = learning difficulties, ADHD = attention deficit hyperactivity disorder

Outcome on SU treatment	***ABCC8*-PNDM (n=21)**	***KCNJ11*-PNDM (n=81)**
Patients independent of insulin at 10 years (%)	86	93
Median HbA1c at 10 years - paired (%)	6.5 (n=16)	6.4 (n=64)
Median SU dose required at 10 years - paired (mg/kg/day glyburide)	0.25 (n=16)	0.23 (n=64)
Median BMI at 10 years SDS (kg/m^2)	-0.75 (n=13)	-0.22 (n=72)
Frequency of neurological features at 10 years (%)	62 (n=21)	64 (n=81)
Improvement in neurological features after SU transfer (%)	54 (n=13)	47 (n=38)
Number of episodes of severe hypoglycemia on SU only over 10 years	0 (n=18)	0 (n=75)
Frequency of side effects (%)	11 (n=18)	14 (n=81)
Frequency of diabetes complications (%)	11 (n=18)	9 (n=81)

Supplementary Table S3. Comparison of long-term outcomes in individuals with mutations in the *KCNJ11* and *ABCC8* genes.

Comparative data on *KCNJ11*-PNDM taken from Bowman et al Lancet D&E 2018 (11) SU = sulfonylurea, PNDM = permanent neonatal diabetes mellitus, BMI = body mass index, SDS = standard deviation score