

Flanagan SE, Patch A-M, Mackay DJG, Edghill EL, Gloyn AL, Robinson D, Shield JPH, Temple K, Ellard S, Hattersley AT: Mutations in ATP-sensitive K<sup>+</sup> channel genes cause transient neonatal diabetes and permanent diabetes in childhood or adulthood. *Diabetes* 56:1930–1937, 2007

One proband within this cohort has subsequently been shown to have a 6q24 duplication inherited from her paternal grandmother. Thus, the prevalence of 6q24 TNDM within the cohort is 72% (70/97) and not 71% (69/97). This patient and her unaffected father are also heterozygous for the novel *KCNJ11* gene variant, R365H. This was assumed to be pathogenic, since the arginine residue at codon 365 is conserved through evolution, was not present in 200 control chromosomes, and had not been previously reported in patients with hyperinsulinism. The significance of this variant is now uncertain, and it may be a rare polymorphism of no clinical significance.