**Table S3.** Three inherited *UPK3A* mutations identified in the duplex collecting system subgroup. Results of in silico analysis, online database queries, and renal ultrasound in parents.

|  |  |  |  |  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| Duplex collecting system and VUR case ID | location | position | amino acid | mutation (cDNA) | mutation (protein) | Sanger sequencing result in 96 control chromosomes | HGMD | ENTREZ SNP | domain | Grantham difference | align GVGD result (Grantham variation, Grantham Deviation) | previously published | renal ultrasound in parents |
| 81079 | exon 3 | 211 | 71 | c.211A>G | p.Ile71Val | absent | not reported | not reported | luminal | 29 | ClassC0\*,179,47 | no urinary tract abnormalities detected |
| 27658 | exon 6 | 811 | 271 | c.811C>T | p.Arg271Trp | absent | not reported | not reported | cytoplasmic | 101 | Class C0\*,243,26 | Parent that carries *UPK3A* mutation: central complex interrupted by a parenchymal ridge, suspect for a duplex collecting system. Other parent: no relevant abnormalities. |
| 81152 | exon 6 | 818 | 273 | c.818C>T | p.Pro273Leu | absent | reported: accession# CM056713 | not reported | cytoplasmic | 98 | Class C0\*, 208,63 | Jenkins et al. 2006: "probably behaves like wildtype" (Jenkins (2005) J Am Soc Nephrol 16, 2141) | no consent for renal ultrasound |

\* align GVGD class C0 means "probably neutral variant”