**Suppl Table1** Characteristics of 28 children with Wilson disease

| **No.** | **Gender** | **Ferenci score** | ***ATP7B* Genotype** | **Age at diagnosis (y)** | **24-hr UCE pre-PCT****(µmol/day)** | **24-hr UCE post-PCT****(µmol/day)** | **UCCR****(µmol/ mmol)** | **AST (IU/L)** | **ALT** **(IU/L)** | **Duration of follow-up****(y)** | **First treatment** | **Last treatment (mg/kg/day)** |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| 1 | Female | 8 | Compound heterozygous for the E1064A:H1069Q | 17.26 | 1.8 | 19.5 | NA | NA | 32 | 14.39 | D-pen | D-pen 13.9 |
| 2 | Male | 8 | Compound heterozygous for the p.M769fs, c.2304\_2305 insC mutation in exon 8 and the p.R1319x, c.3955c>T mutation in exon 19  | 12.17 | 8.7 | 32.7 | NA | 34 | 104 | 6.90 | D-pen | Trientine 20.0 |
| 3 | Male | 8 | Homozygous for the p.A1003V, c.3008C>T mutation in exon 13  | 15.59 | 30.9 | 67.7 | NA | NA | 182 | 10.94 | D-pen | Trientine 25.0 |
| 4 | Female | 4 | Homozygous for the p.Pro840Leu, c.2519C>T mutation in exon 10  | 11.79 | NA | NA | NA | NA | 33 | 13.79 | D-pen | D-pen 12.7 |
| 5 | Male | 10 | Compound heterozygous for the p.F491fs: C.1708-1G>C mutation in exon3 and c.1708-1G>C in intron4 | 8.54 | 3.3 | 18.1 | NA | NA | 71 | 12.70 | D-pen | D-pen 16.0 |
| 6 | Male | 6 | No mutation found | 11.34 | 5.1 | 25.2 | NA | NA | 320 | 12.25 | D-pen | LT (day59) |
| 7 | Male | 10 | Compound heterozygous for the p.Gln289X, c. 865 C>T and the Ala 1003Thr, c.3007G>A mutation in exon 2 and 13  | 10.39 | 4 | 13.2 | NA | NA | 63 | 11.13 | D-pen | D-pen 12.5 |
| 8 | Female | 9 | Homozygous for the p.Leu1368fs, c4102\_4103de1lCTinsAGTCCC mutation in exon 20  | 3.84 | 0.4 | 5.1 | NA | 450 | 385 | 9.84 | D-pen | D-pen 17.2 |
| 9 | Female | 6 | Homozygous for the p.Pro840Leu, c.2519C>T mutation in exon 10  | 16.92 | NA | 13.8 | NA | 40 | 26 | 10.59 | D-pen | D-pen 4.2 |
| 10 | Male | 10 | Homozygous for the p.Gly1000fs, c.2997dupC mutation in exon 13  | 11.27 | NA | 12.1 | NA | 67 | 90 | 2.65 | D-pen | D-pen 23.9 |
| 11 | Female | 10 | Compound heterozygous for the c.2731-2A>G mutation in intron 11 and the p.Thr977Met, C2930C>T mutation in exon 13  | 7.96 | 28.3 | 48.6 | NA | 92 | 164 | 9.92 | D-pen | Trientine 23.9 |
| 12 | Female | 7 | Compound heterozygous for the p.Gly1061Glu, c.3182G>A mutation and the p.Phe491fs,c.1470delC mutation in exon 3  | 11.51 | NA | NA | NA | NA | 93 | 10.05 | D-pen | Trientine 34.7 |
| 13 | Female |  10 | Homozygous for the p.Gly1341Asp, c.4022G>A mutation in exon 20  | 4.22 | 0.92 | 11.3 | NA | 511 | 221 | 8.60 | D-pen | D-pen 19.0 |
| 14 | Female |   9 | Homozygous for the pHis1069Gln, c.3207C>A mutation in exon  | 9.34 | 2.3 | 28.6 | NA | 362 | 187 | 6.70 | D-pen | Trientine 10.3 |
| 15 | Female |  8 | Homozygous for a mutations in ATP7B gene compatible with Wilson disease | 16.46 | 128.7 | 132.2 | NA | 21 | 74 | 7.76 | D-pen | LT (day7) |
| 16 | Male | 9  | Homozygous for the p.(His1069)Gln, c.3207C>A mutation in exon 14  | 6.72 | 1.4 | 15.5 | NA | 436 | 202 | 6.98 | D-pen | D-pen 17.4 |
| 17 | Male | 6  | Compound heterozygous for p.(Arg778Leu), c.233G>T mutation in exon 8 and the p.(Lys838fs), c.2513del mutation in exon 10  | 15.22 | NA | NA | 1.7 | 14 | 20 | 6.29 | D-pen | D-pen 19.5 |
| 18 | Male | 9  | Homozygous for the[(Ile1102Thr), c.3305T>C mutation in exon 15 | 12.52 | 3.7 | 32.4 | NA | 43 | 60 | 5.85 | D-pen | Trientine 35.3 |
| 19 | Male | 11  | Homozygous for the p.(Cys271\*), c.813C>A mutation in exon 2  | 9.79 | NA | NA | 10.8 | 379 | 127 | 5.30 | D-pen | Trientine 22.9 |
| 20 | Male | 6 | No mutation found | 13.18 | 9.1 | 36.9 | NA | 54 | 116 | 5.22 | D-pen | Trientine 9.2 |
| 21 | Male | 10 | Compound heterozygous for 3 significant variants [p.(Pro992Leu), p.(Ile1148Thr), p.(Gln1142His)]  | 9.72 | 2.13 | 21.36 | NA | 104 | 116 | 4.85 | D-pen | D-pen 16.4 |
| 22 | Female | 10  | Homozygous for damaging variant c.2807T>A | 6.98 | 3.22 | 6.08 | NA | 222 | 129 | 4.19 | D-pen | D-pen 35.9 |
| 23 | Female | 9 | Compound heterozygous for 2 damaging variants [p.(Asn1270Ser) and p.(ALa1003Val)] | 14.12 | 2.51 | 30.42 | NA | hemolysed | hemolysed | 3.61 | D-pen | D-pen 22.1 |
| 24 | Male | 10  | Homozygous mutation for p.(Cys271Ter) | 13.42 | 16.73 | 61.66 | NA | 64 | 100 | 3.55 | D-pen | D-pen 14.0 |
| 25 | Male | 9  | Homozygous for p.(Ile582Arg) | 14.33 | 3.54 | 43.59 | NA | NA | 144 | 2.51 | D-pen | D-pen 20.8 |
| 26 | Male |  10 | Homozygous for the c.-441\_-427del (a 15 nucleotide deletion within the ATP7B promotor region) | 9.46 | 6.78 | 12.1 | NA | 680 | 206 | 1.39 | D-pen | D-pen 19.3 |
| 27 | Male | 8 | Compound heterozygous for two damaging variants p.(Met769Val), p.(Gln289Ter) | 0.00 | 3.27 | 27.27 | NA | 814 | 1162 | 2.49 | D-pen | D-pen 18.7 |
| 28 | Male | 8  | Homozygous for the c.1745T>G variant | 12.17 | 3.09 | 30.5 | NA | 172 | 76 | 2.39 | D-pen | D-pen 17.2 |