**Supplementary Table 1: Clinical features of the ADPKD patients in two Chinese families**

|  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| Patient | | Gender | Age | Onset age | Blood pressure | Renal size | Renal function | Kidney cysts | Extrarenal cysts |
| **Pedigree 1** | II 2 | Female | 60 | 23 | High | Renal transplantation due to renal failure | | | Hepatic cysts |
|  | II 3 | Male | 57 | 25 | High | Massive enlargement | Renal insufficiency | Bilateral renal cysts | — |
|  | II 5 | Male | 55 | 25 | High | Massive enlargement | Renal insufficiency | Bilateral renal cysts | — |
|  | II 8 | Female | 51 | 22 | High | Massive enlargement | Uremia | Bilateral renal cysts | Hepatic cysts |
|  | III 2 | Male | 32 | 24 | High | Mild enlargement | Normal | Bilateral renal cysts | — |
|  | III 3 | Female | 30 | 22 | Normal | Mild enlargement | Normal | Bilateral renal cysts | — |
|  | III 4 | Female | 29 | 26 | Normal | Mild enlargement | Normal | Bilateral renal cysts | Hepatic cysts |
|  | III 6 | Female | 27 | 25 | Normal | Mild enlargement | Normal | Bilateral renal cysts | — |
|  | III 8 | Female | 25 | 23 | Normal | Mild enlargement | Normal | Bilateral renal cysts | — |
| **Pedigree 2** | II 2 | Female | 64 | 26 | High | Massive enlargement | Uremia | Bilateral renal cysts | — |
|  | II 5 | Female | 58 | 27 | High | Massive enlargement | Renal insufficiency | Bilateral renal cysts | — |
|  | III 2 | Female | 40 | 25 | Normal | Mild enlargement | Normal | Bilateral renal cysts | — |
|  | III 3 | Male | 38 | 25 | Normal | Mild enlargement | Normal | Bilateral renal cysts | — |
|  | III 6 | Female | 32 | 26 | Normal | Mild enlargement | Normal | Bilateral renal cysts | — |

Note: I 1 and II 9 in pedigree 1 as well as I 1 in pedigree 2 died of renal failure.