

Supplemental Table 1. Patient profiles at the time of renal biopsy

Older brother

Height 162.5 cm, weight 57.7 kg, blood pressure 102/50 mmHg (right), 108/48 mmHg (left)
Urinalysis: pH 6.0, urinary protein (2+), hematuria (-), urinary sugar (-), urinary sediment:
RBC 0-2/HPF, WBC 2-4/HPF, no specific cast, urinary NAG 3.6 U/L, urinary beta-2-
microglobulin 61 µg/L

Peripheral blood: WBC 5100/µl (seg 50%, lym 43%, mono 5%, eosino 2%), RBCs 475*10⁴/µl,
Hb 14.3 g/dl, Plts 24.9*10⁴/µl, erythrocyte sedimentation rate 11 mm/h

Biochemical tests: total protein 6.9 g/dl, albumin 4.1 g/dl (alpha2-globulin 9.5%, gamma-
globulin 14.9%), total bilirubin 0.5 mg/dl, total cholesterol 181 mg/dl, BUN16 mg/dl, Cre
0.5 mg/dl, Ccr96 ml/min, Na 142 mEq/L, K 4.0 mEq/L Cl 105 mEq/l Ca 9.1 mg/dl, P 3.9
mg/dl, CPK 6750 U/L, myoglobin 707 ng/ml,

Immunological test: CRP 0.3 mg/dl, ANA(-), IgG 1122 mg/dl, IgA 149 mg/dl, IgM 68 mg/dl,
C3 86 mg/dl, C4 19 mg/dl, CH50 40.7 U/ml, serum beta-2-microglobulin 1.1 mg/dl

Infection test: HBs Antigen(-), HBc antibody (-), HIV antibody (-)

Younger brother

Height 154.5 cm, weight 66.1 kg, blood pressure 108/54 mmHg (right), 110/66 mmHg (left)
Urinalysis: pH 6.0, urinary protein (2+), hematuria (-), urinary sugar (-), urinary sediment:
RBC 2-4/HPF, WBC 0-2/HPF, hyaline cast 3/HPF, granular cast 1/HPF, albuminuria 106
mg/day (at the age of 33) Urinary NAG 6.9 U/L, Urinary beta-2-microglobulin 106 µg/L

Peripheral blood: WBC 7200/ul (seg 54%, lym 40%, mono 5%, eosino 1%) RBCs 532*10⁴/µl,
Hb 15.6 g/dl, Plts 28.9*10⁴/µl, erythrocyte sedimentation rate 6 mm/h

Biochemical tests: total protein 8.0 g/dl, albumin 4.8 g/dl (alpha2-globulin 9.6%, gamma-
globulin 17.8%), total bilirubin 0.4 mg/dl, total cholesterol 218 mg/dl, BUN10 mg/dl, Cre
0.6 mg/dl, Ccr139 ml/min, Na 143 mEq/L, K 4.3 mEq/L Cl 103 mEq/l Ca 9.7 mg/dl, P 4.1
mg/dl, CPK 541 U/L (at the age of 33), myoglobin 124 ng/ml

Immunological test: CRP 0.3 mg/dl, ANA<20, IgG 1622 mg/dl, IgA 161 mg/dl, IgM 125
mg/dl, C3 109 mg/dl, C4 22 mg/dl, CH50 41.8 U/ml

Infection test: HBs Antigen(-), HBc antibody (-), HIV antibody (-)