

Supplementary Material

Supplementary References

Supplementary information is available at *KI Report's* website.

Supplementary Methods

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Supplementary Methods

Use of patient urine samples was in accordance with Addenbrooke's Hospital Research and Development Committee rules. Urine samples from patients with Dent1 disease were stored in liquid nitrogen for up to 14 years before analysis and thawed only once. Measurements of UfRBP4 and creatinine were performed as described previously⁹. Results from some of the patients reported here were included in previously described grouped results for inherited RFS⁹.

Calculation of the GSC for fRBP4 was based on the previous model⁸. The value of the median UfRBP4 expressed as mg/mmol creatinine was multiplied by 14.7 mmol to approximate 24 hr excretion^{S8} and then divided by 179 L (being 124 mL/min x60x24/1,000)^{S9} to approximate 24 hr GFR. The resulting value, representing the concentration of fRBP4 in mg/L in the glomerular ultrafiltrate, was then divided by the established value for normal mean plasma fRBP4, 5.8 mg/L, to estimate the GSC of fRBP4.

To determine the molar concentrations of fRBP4 and albumin in the glomerular ultrafiltrate, the respective normal plasma concentrations in $\mu\text{mol/L}$ (mg/L) of 0.276, (5.8) and 687, (45,000) are multiplied by the estimated GSCs of 0.097 and 7.7×10^{-5} , respectively⁸. This yields the glomerular ultrafiltrate molar concentrations of fRBP4 and albumin as 26.8 and 52.9

nmol/L, respectively. The total molar concentration of protein in the glomerular ultrafiltrate, calculated from the GSC of fRBP4 estimated here and the other values previously reported, is 314 nmol/l so the mole% of fRBP4 and albumin are 8.5% and 16.8%, respectively.

Supplementary References

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