

Supplementary Table 1. Questionnaire on Fabry nephropathy

Category	Questions
1) Respondent details	Age Sex Specialist
2) Experience with Fabry patients	Have you ever treated Fabry patients in the past or present? Are you currently treating patients with Fabry disease? If you treated a patient with Fabry disease, was it a classic phenotype? How many Fabry disease patients are you currently treating?
3) Timing of kidney biopsy	Do you think kidney biopsy is necessary in patients with Fabry disease?
4) Need for a Fabry registry	Are you planning to register in the Fabry registry of the Korean Nephrology Society?
5) Lyso-Gb3 measurements	Do you measure the concentration of lyso-Gb3 (globotriaosphingosine) or lyso-Gb3 as a biomarker for Fabry disease? When should the concentration of lyso-Gb3 be measured as a biomarker for Fabry disease? – For screening When should the concentration of lyso-Gb3 be measured as a biomarker for Fabry disease? – For diagnosis When should the concentration of lyso-Gb3 be measured as a biomarker for Fabry disease? – Before treatment When should the concentration of lyso-Gb3 be measured as a biomarker for Fabry disease? – After treatment When should the concentration of lyso-Gb3 be measured as a biomarker for Fabry disease? – All When should lyso-Gb3 be measured if insurance is provided? – Screening When should lyso-Gb3 be measured if insurance is provided? – Diagnosis When should lyso-Gb3 be measured if insurance is provided? – Before treatment When should lyso-Gb3 be measured if insurance is provided? – After treatment When should lyso-Gb3 be measured if insurance is provided? – 6 mo When should lyso-Gb3 be measured if insurance is provided? – 12 mo
6) Interval and preferred tests for kidney function	At what frequency should tests to monitor renal kidney function in Fabry disease patients be performed? What tests are used to monitor renal kidney function in Fabry disease patients? – Proteinuria What tests are used to monitor kidney function in Fabry disease patients? – eGFR by serum creatinine What tests are used to monitor kidney function in Fabry disease patients? – α -Gal A activity What tests are used to monitor kidney function in Fabry disease patients? – cystatin C What tests are used to monitor kidney function in Fabry disease patients? – lyso-Gb3 What tests are used to monitor kidney function in Fabry disease patients? – Kidney biopsy What tests should be performed if Fabry nephropathy is suspected? – Proteinuria and eGFR If Fabry nephropathy is suspected, what tests should be performed? – α -Gal A activity If Fabry nephropathy is suspected, what tests should be performed? – lyso-Gb3 If Fabry nephropathy is suspected, what tests should be performed? – Kidney biopsy If Fabry nephropathy is suspected, what tests should be performed? – Gene test
7) Definition	What is your definition of Fabry nephropathy?
8) Other treatments of Fabry nephropathy beyond ERT	What additional therapies are available to treat Fabry nephropathy besides enzyme therapy? – ARB/ACEi What additional therapies are available to treat Fabry nephropathy besides enzyme therapy? – Low-protein diet What additional therapies are available to treat Fabry nephropathy besides enzyme therapy? – Low-salt diet What additional therapies are available to treat Fabry nephropathy besides enzyme therapy? – Low-salt diet When do you believe that ERT is required for a patient with variant Fabry nephropathy?

ACEi, angiotensin-converting enzyme inhibitor; ARB, angiotensin receptor blocker; eGFR, estimated glomerular filtration rate; ERT, enzyme replacement therapy; lyso-Gb3, globotriaosphingosine; KSN, Korean Society of Nephrology.