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Current Treatment Status of Fabry Nephropathy in Korea

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Fabry disease is a rare genetic disease that affects multiple organs, including the heart, brain, skin, and kidneys. It is a lysosomal storage disease with a deficiency of α-galactosidase A, an enzyme encoded by the GLA gene that located on the X chromosome. After the introduction of enzyme replacement therapy (ERT) in Korea, some case or case series studies were reported to show the use of ERT in Korean patients. However, detailed studies on the current status and outcomes are not yet available. Especially, after the introduction of migalastat, an oral medication for the treatment Fabry disease, there might be changes in the status of treatment. To investigate the current treatment status and outcomes of Korean patients with Fabry disease, we analyzed data from the National Health Insurance Service from 2002 to 2021. As there is no specific code for Fabry disease, we collected data from patients with ICD code E752, a code for other sphingolipidosis. Among them, we selected samples who treated with ERT (agalsidase beta, Fabryzyme; agalsidase alfa, Replagal) or migalastat (Galafold). A total of 3,791 patients had ICD code E752, and 228 patients were remained because they received treatment with ERT or migalastat. Of them, 120 patients were male, and 108 were female. The mean age at diagnosis was 36.8 years in men and 49.9 years in women. The median time from diagnosis to treatment was 75 days in men and 103.5 days in women. I will discuss further findings on the current treatment status of Korean patients with Fabry disease.

Keywords: Fabry disease, Korean, Treatment