



Shaping Tomorrow's Nephrology: Insight-Driven Kidney Care

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Epidemiology and Longitudinal Change in Renal Involvement in Pediatric Patients with Tuberous Sclerosis Complex: A Single-Center Retrospective Study

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Objectives : Tuberous sclerosis complex (TSC) is an autosomal dominant disorder characterized by multiorgan tumor development, with renal manifestations including angiomyolipoma (AML, 70%), multiple cysts (35%). These renal lesions may lead to hypertension and progressive renal dysfunction, necessitating regular monitoring. This study aimed to evaluate longitudinal changes in renal function and renal tumors in pediatric TSC patients at a single center.

Methods : This retrospective study included patients diagnosed with TSC at Pusan National University Yangsan Hospital from 2008 to December 2025. Data collected included genetic mutation type, renal function, urinalysis, renal imaging, and history of everolimus or sirolimus use. Primary outcomes included mortality, renal function decline, initiation of dialysis, and kidney transplantation. Statistical analyses were performed using SPSS 30.

Results : A total of 78 patients were enrolled (male 48.7%, female 51.3%); mean age at diagnosis was 61.8 ± 68.4 months. Genetic mutations were confirmed in 46.2% (TSC1: 22.2%, TSC2: 72.2%, TSC2/PKD1: 5.6%). Everolimus was prescribed in 32.1%. Initial renal imaging was performed in 83.8%, of whom 52.3% had AML or renal cysts at diagnosis (AML maximal size: 1.3 ± 1.5 cm; cyst size: 0.8 ± 0.8 cm). At last follow-up (mean 93.2 months, $n=50$), AML size increased to 1.5 ± 1.6 cm and cysts to 1.3 ± 1.6 cm. Initial eGFR was 93.4 ± 36.5 mL/min/1.73m²; proteinuria ($\geq 1+$) was present in 7.5% with no hematuria. Final eGFR was 113.6 ± 27.8 mL/min/1.73m²; proteinuria in 18.2% and hematuria in 6.8%. Among 17 patients treated with oral everolimus (mean duration 50.3 ± 4.23 months), 69.2% showed AML size reduction or stabilization after 33.4 ± 36.1 months. Two patients died from neurological complications (status epilepticus, brain hemorrhage); one patient developed ESRD requiring kidney transplantation.

Conclusions : TSC requires continuous monitoring of renal tumors and renal function from the time of diagnosis, given the long-term risk of progression to hypertension and CKD. A multidisciplinary approach is essential for optimal management.