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## ADPKD Treatment and New European Guidelines on Tolvaptan

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Tolvaptan (V2 receptor antagonist) represents the first treatment of ADPKD targeting disease-specific mechanisms. Position statement of ERA on the use of tolvaptan in ADPKD from 2016 was recently (in 2022) updated based on new data from clinical studies in patients with later stage of ADPKD and the expanding practical experience with tolvaptan. Consensus statement is aimed at nephrologists in clinical practice. Treatment with tolvaptan should be started in all adult patients with ADPKD  $\leq$  55 years of age with an eGFR  $\geq$  25 ml/min/1.73 m<sup>2</sup> and rapidly progressive disease based on the rate of loss of eGFR ( $\geq$  3 ml/min/1.73 m<sup>2</sup>) or Mayo classification, PROPKD score and clinical presentation using hierarchical decision. Treatment should be discontinued when eGFR approaches kidney failure with the need of renal replacement therapy. On the other hand, treatment with tolvaptan should not be started in patients with well preserved eGFR indexed for age. Initial dose of tolvaptan should be titrated to 90/30 mg/day if tolerated. Adverse effect of treatment with tolvaptan, especially polyuria and its practical consequences, including impacts on lifestyle and the risk of dehydration should be discussed with the patient before the start of treatment and reduced sodium intake should be recommended. Liver function must be monitored monthly during the first 18 months of treatment, patients with serious liver toxicity related to tolvaptan should not be re-exposed to tolvaptan. Increased fluid intake should not be recommended as an alternative to tolvaptan, but despite lacking evidence patients with ADPKD not treated with tolvaptan should adhere to low-salt diet (3-5 g/day) and high water intake (3-4 L/day). Accumulating evidence demonstrates longterm efficacy and safety of treatment with tolvaptan. It is important to enhance the access to this treatment to all patients who can benefit from the amelioration of the rapid rate of loss of eGFR.

**Keywords:** polycystic kidney disease, progression, tolvaptan, treatment