Reference number(s)
2572-A

Specialty Guideline Management tolvaptan-Jynarque

Products Referenced by this Document

Drugs that are listed in the following table include both brand and generic and all dosage forms and strengths unless otherwise stated. Over-the-counter (OTC) products are not included unless otherwise stated.

Brand Name	Generic Name
Jynarque	tolvaptan

Indications

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-approved Indications[1,2]

Jynarque is indicated to slow kidney function decline in adults at risk of rapidly progressing autosomal dominant polycystic kidney disease (ADPKD).

All other indications are considered experimental/investigational and not medically necessary.

Documentation

Submission of the following information is necessary to initiate the prior authorization review:

- Imaging used for diagnosis and confirmation of rapidly progressing disease (e.g., ultrasonography, magnetic resonance imaging [MRI], computed tomography [CT]).
- Genetic testing results, if applicable.

Prescriber Specialties

This medication must be prescribed by or in consultation with a nephrologist or a specialist in the treatment of ADPKD.

Coverage Criteria[1-8]

Authorization of 12 months may be granted for treatment of autosomal dominant polycystic kidney disease (ADPKD) when all of the following criteria are met:

- The member is 18 years of age or older with a diagnosis of ADPKD as confirmed by any of the following:
  - In members aged 18 to less than 40 years with a first degree relative with ADPKD: greater than or equal to 3 cysts (unilateral or bilateral) using any radiologic method.[7]
  - In members aged 40 to less than 60 years with a first degree relative with ADPKD: greater than or equal to 2 cysts per kidney using any radiologic method.[7]
  - In members aged 60 years or older with a first degree relative with ADPKD: greater than or equal to 4 cysts per kidney using any radiologic method.[7]
  - In members with no family history (no first degree relative with disease): positive genetic test for ADPKD (i.e., mutation in *PKD1* or *PKD2* gene).[4]
- The member's estimated glomerular filtration rate (eGFR) is greater than or equal to 25 milliliters per minute per 1.73 square meters and one of the following criteria is met:
  - Member has or is at risk for rapidly progressing disease as confirmed by height-adjusted total kidney volume compatible with Mayo class 1C, 1D, or 1E disease[1]
  - Member has a historical rate of eGFR decline of $\geq$ 3 milliliters per minute per 1.73 square meters. [8]

Continuation of Therapy

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization for an indication listed in the coverage criteria section when the member has demonstrated a beneficial response to Jynarque therapy (e.g., slowed kidney function decline, decreased kidney pain) and the member's estimated glomerular filtration rate (eGFR) is greater than or equal to 25 mL/min/1.73m^2.

References

1. Jynarque [package insert]. Rockville, MD: Otsuka America Pharmaceutical, Inc.; March 2025.
2. Tolvaptan [package insert]. Naples, FL: Lupin Pharmaceuticals, Inc.; May 2025.

3. Torres VE, Devuyst O, Chapman AB, et al; for the REPRISE Trial Investigators. Rationale and design of a clinical trial investigating tolvaptan safety and efficacy in autosomal dominant polycystic kidney disease. Am J Nephrol. 2017;45(3):257-266.
4. Chapman AB, Devuyst O, Eckardt KU, et al. Autosomal-dominant polycystic kidney disease (ADPKD): executive summary from a Kidney Disease: Improving Global Outcomes (KDIGO) Controversies Conference. Kidney Int. 2015;88(1):17-27.
5. Srivastava A, Patel N. Autosomal dominant polycystic kidney disease. Am Fam Physician. 2014 Sep 1;90(5):303-307.
6. Müller RU, Messchendorp AL, Birn H, et al. An update on the use of tolvaptan for autosomal dominant polycystic kidney disease: consensus statement on behalf of the ERA Working Group on Inherited Kidney Disorders, the European Rare Kidney Disease Reference Network and Polycystic Kidney Disease International. Nephrol Dial Transplant. 2022;37(5):825-839.
7. Pei Y, Obaji J, Dupuis A, et. Al. Unified criteria for ultrasonographic diagnosis of ADPKD. J Am Soc Nephrol. 2009;20:205-212.
8. Kidney Disease: Improving Global Outcomes (KDIGO). KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and treatment of Autosomal Dominant Polycystic Kidney Disease (ADPKD). Kidney Int. 2025 Feb; 107 (2S): S1-S239.