| Effective date: 4/1/2020 |
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| Reviewed: 1/2020, 3/2021, 3/2022, 3/2023 |
| Scope: Medicaid |

# SPECIALTY GUIDELINE MANAGEMENT

# JYNARQUE (tolvaptan)

## POLICY

### I. 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 Indication
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.

### II. DOCUMENTATION

Submission of the following information is necessary to initiate the prior authorization review:
A. Imaging used for diagnosis and confirmation of rapidly progressing disease (ultrasonography, magnetic resonance imaging [MRI], computed tomography [CT])
B. Genetic testing results if applicable

### III. CRITERIA FOR INITIAL APPROVAL

Authorization of 6 months may be granted for treatment of autosomal dominant polycystic kidney disease (ADPKD) when all of the following criteria are met:
A. The member is 18 years of age or older and with a diagnosis of ADPKD as confirmed by any of the following:
    1. 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
    2. 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
    3. In members aged 60 or older with a first degree relative with ADPKD: greater than or equal to 4 cysts per kidney using any radiologic method
    4. In members with no family history (no first degree relative with disease): positive genetic test for ADPKD (mutation in PKD1 or PKD2 gene)
B. Prescribed by, or in consultation with a nephrologist or physicians specializing in the management of ADPKD.
C. The 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.

D. The member's estimated glomerular filtration rate (eGFR) is greater than or equal to 25 mL/min/1.73m$^2$.
E. The member does not have any of the following contraindications: liver impairment or injury, member is concurrently taking a strong CYP3A4 inhibitor, member does not have the ability to sense or respond to thirst, abnormal serum sodium (particularly hypernatremia), hypovolemia, concomitant use of diuretics, or uncorrected urinary outflow obstruction.

## IV. CONTINUATION OF THERAPY

Authorization of 6 months may be granted for continued treatment in members requesting reauthorization for an indication listed in Section III 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$^2$.

## V. QUANTITY LIMIT

Jynarque has a quantity limit of 2 tablets per day.

## VI. REFERENCES

1. Jynarque [package insert]. Rockville, MD: Otsuka America Pharmaceutical, Inc.; November 2022.
2. 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.
3. 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.
4. Srivastava A, Patel N. Autosomal dominant polycystic kidney disease. *Am Fam Physician.* 2014 Sep 1;90(5):303-307.
5. Gansevoort RT, Arici M, Benzing T, et al. Recommendations for the use of tolvaptan in autosomal dominant polycystic kidney disease: a position statement on behalf of the ERA-EDTA Working Groups on Inherited Kidney Disorders and European Renal Best Practice. *Nephrol Dial Transplant.* 2016;31(3):337-348.
6. Pei Y, Obaji J, Dupuis A, et. Al. Unified criteria for ultrasonographic diagnosis of ADPKD. *J Am Soc Nephrol.* 2009;20:205-212.