



PRIOR AUTHORIZATION POLICY

POLICY: Tolvaptan Products – Jynarque Prior Authorization Policy

- Jynarque® (tolvaptan tablets – Otsuka)

REVIEW DATE: 06/28/2023

INSTRUCTIONS FOR USE

THE FOLLOWING COVERAGE POLICY APPLIES TO HEALTH BENEFIT PLANS ADMINISTERED BY CIGNA COMPANIES. CERTAIN CIGNA COMPANIES AND/OR LINES OF BUSINESS ONLY PROVIDE UTILIZATION REVIEW SERVICES TO CLIENTS AND DO NOT MAKE COVERAGE DETERMINATIONS. REFERENCES TO STANDARD BENEFIT PLAN LANGUAGE AND COVERAGE DETERMINATIONS DO NOT APPLY TO THOSE CLIENTS. COVERAGE POLICIES ARE INTENDED TO PROVIDE GUIDANCE IN INTERPRETING CERTAIN STANDARD BENEFIT PLANS ADMINISTERED BY CIGNA COMPANIES. PLEASE NOTE, THE TERMS OF A CUSTOMER'S PARTICULAR BENEFIT PLAN DOCUMENT [GROUP SERVICE AGREEMENT, EVIDENCE OF COVERAGE, CERTIFICATE OF COVERAGE, SUMMARY PLAN DESCRIPTION (SPD) OR SIMILAR PLAN DOCUMENT] MAY DIFFER SIGNIFICANTLY FROM THE STANDARD BENEFIT PLANS UPON WHICH THESE COVERAGE POLICIES ARE BASED. FOR EXAMPLE, A CUSTOMER'S BENEFIT PLAN DOCUMENT MAY CONTAIN A SPECIFIC EXCLUSION RELATED TO A TOPIC ADDRESSED IN A COVERAGE POLICY. IN THE EVENT OF A CONFLICT, A CUSTOMER'S BENEFIT PLAN DOCUMENT ALWAYS SUPERSEDES THE INFORMATION IN THE COVERAGE POLICIES. IN THE ABSENCE OF A CONTROLLING FEDERAL OR STATE COVERAGE MANDATE, BENEFITS ARE ULTIMATELY DETERMINED BY THE TERMS OF THE APPLICABLE BENEFIT PLAN DOCUMENT. COVERAGE DETERMINATIONS IN EACH SPECIFIC INSTANCE REQUIRE CONSIDERATION OF 1) THE TERMS OF THE APPLICABLE BENEFIT PLAN DOCUMENT IN EFFECT ON THE DATE OF SERVICE; 2) ANY APPLICABLE LAWS/REGULATIONS; 3) ANY RELEVANT COLLATERAL SOURCE MATERIALS INCLUDING COVERAGE POLICIES AND; 4) THE SPECIFIC FACTS OF THE PARTICULAR SITUATION. COVERAGE POLICIES RELATE EXCLUSIVELY TO THE ADMINISTRATION OF HEALTH BENEFIT PLANS. COVERAGE POLICIES ARE NOT RECOMMENDATIONS FOR TREATMENT AND SHOULD NEVER BE USED AS TREATMENT GUIDELINES. IN CERTAIN MARKETS, DELEGATED VENDOR GUIDELINES MAY BE USED TO SUPPORT MEDICAL NECESSITY AND OTHER COVERAGE DETERMINATIONS.

CIGNA NATIONAL FORMULARY COVERAGE:

OVERVIEW

Jynarque, a selective vasopressin V₂-receptor antagonist, is indicated to slow kidney function decline in adults at risk of rapidly-progressing **autosomal dominant polycystic kidney disease** (ADPKD).¹

Disease Overview

ADPKD is a heterogeneous, inherited kidney disorder associated with the development of kidney cysts, which result in kidney pain, hypertension, renal failure, and other clinical sequelae.²⁻⁵ The condition is a common cause of end-stage renal disease; however, other organs are also impacted (e.g., hepatic and vascular systems). Progressive kidney enlargement occurs; however, manifestations generally do not occur until later in life (fourth decade) due to compensatory renal mechanisms. If a parent has the condition, a child has a 50% chance of inheritance. Approximately 600,000 people in the US have this condition.

Guidelines

The European Renal Association Working Groups on Inherited Kidney Disorders, the European Rare Kidney Disease Reference Network, and the Polycystic Kidney Disease International published a consensus statement regarding use of tolvaptan in ADPKD (2022).⁷ A confirmed annual estimated glomerular filtration rate decline ≥ 3.0 mL/min/1.73 m² over a period of ≥ 4 years defines rapid progression. Also, a Mayo

Classification of 1D or 1E indicates rapid disease progression. Patients with Mayo Classification of 1C should be further evaluated for additional evidence of rapid disease progression. Total kidney volume changes should not be used as a marker of progression in individual patients. Finally, Jynarque should be discontinued when the patient approaches kidney failure (i.e., the need for renal replacement therapy).

The National Kidney Foundation and the Polycystic Kidney Disease Foundation list tolvaptan as an FDA-approved treatment option for patients with ADPKD.^{5,8}

POLICY STATEMENT

Prior Authorization is recommended for prescription benefit coverage of Jynarque. All approvals are provided for the duration noted below. Due to the specialized skills required for evaluation and diagnosis of patients treated with Jynarque as well as the monitoring required for adverse events and long-term efficacy, approval requires Jynarque to be prescribed by or in consultation with a physician who specializes in the condition being treated.

• **Jynarque® (tolvaptan tablets (Otsuka)) is(are) covered as medically necessary when the following criteria is(are) met for FDA-approved indication(s) or other uses with supportive evidence (if applicable):**

FDA-Approved Indication

1. Autosomal Dominant Polycystic Kidney Disease. Approve for 1 year if the patient meets the following (A, B, C, and D):

A) Patient is ≥ 18 years of age; AND

B) According to the prescriber, the patient has rapidly-progressing autosomal dominant polycystic kidney disease; AND

Note: Examples of rapidly declining renal function include estimated glomerular filtration rate decline of ≥ 3.0 mL/min/1.73 m², and Mayo Classification of 1D or 1E.

C) Patient does not have Stage 5 chronic kidney disease; AND

Note: Stage 5 chronic kidney disease is defined as glomerular filtration rate < 15 mL/min/1.73 m² or receiving dialysis.

D) The medication is prescribed by or in consultation with a nephrologist.

CONDITIONS NOT COVERED

• **Jynarque® (tolvaptan tablets (Otsuka)) is(are) considered experimental, investigational or unproven for ANY other use(s) including the following (this list may not be all inclusive; criteria will be updated as new published data are available):**

- 1. Patient is Currently Receiving Samsca (tolvaptan tablets).** Samsca is a tolvaptan product that is indicated for the treatment of clinically-significant hypervolemic and euvolemic hyponatremia, including patients with heart failure and syndrome of inappropriate antidiuretic hormone (SIADH).⁶ Concomitant use is not recommended.
- 2. Hyponatremia.** Samsca is another tolvaptan product indicated for the treatment of clinically-significant hypervolemic and euvolemic hyponatremia (serum sodium < 125 mEq/L or less marked hyponatremia that is symptomatic and has resisted correction and fluid restriction), including patients with heart failure and SIADH. Samsca should be used for this condition.

REFERENCES

1. Jynarque® tablets [prescribing information]. Rockville, MD: Otsuka; October 2020.
2. 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;88:17-27.
3. Ong ACM, Devuyst O, Knebelmann B, et al, on behalf of the ERA-EDTA Working Group for Inherited Kidney Diseases. Autosomal dominant polycystic kidney disease: the changing face of clinical management. *Lancet.* 2015;385:1993-2002.
4. Harris PC, Torres VE. Polycystic Kidney Disease, Autosomal Dominant. In Adam MP, Ardinger HH, Pagon RA, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2018. Last Updated: July 19, 2018. Available at: <https://www.ncbi.nlm.nih.gov/books/NBK1246/> Accessed on June 9, 2022.
5. National Kidney Foundation. Polycystic kidney disease. Available at: <https://www.kidney.org/atoz/content/polycystic>. Accessed on June 9, 2022.
6. Samsca® tablets [prescribing information]. Rockville, MD: Otsuka; June 2018.
7. Muller 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;37:825-839.
8. Polycystic Kidney Disease Foundation. Tolvaptan. Available at: <https://pkdcure.org/tolvaptan/>. Accessed on June 8, 2022.

HISTORY

Type of Revision	Summary of Changes	Review Date
Annual Revision	Autosomal Dominant Polycystic Kidney Disease: Removed examples of rapidly-progressing autosomal dominant polycystic kidney disease (ADPKD) from criterion. Added note with examples of rapidly-progressing ADPKD. Moved definition of Stage 5 kidney disease from criterion to a note.	06/15/2022
Annual Revision	No criteria changes.	06/28/2023

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