



Effective Date 11/1/2023
Next Review Date... 11/1/2024
Coverage Policy Number IP0287

Tolvaptan (Jynarque®)

Table of Contents

Overview1
Medical Necessity Criteria1
Reauthorization Criteria2
Authorization Duration2
Conditions Not Covered.....2
Background.....2
References3

Related Coverage Resources

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.

Overview

This policy supports medical necessity review for tolvaptan tablets (Jynarque®).

Receipt of sample product does not satisfy any criteria requirements for coverage.

Medical Necessity Criteria

Tolvaptan (Jynarque) is considered medically necessary when the following are met:

- 1. Autosomal Dominant Polycystic Kidney Disease. Individual meets ALL of the following criteria (A, B, C and D):
A. 18 years of age or older
B. At risk of rapidly-progressing autosomal dominant polycystic kidney disease (for example, reduced or declining renal function, high or increasing total kidney volume [height adjusted], Mayo classes 1C, 1D, or 1E)
C. Does not have Stage 5 or end stage chronic kidney disease

D. Medication is prescribed by, or in consultation with, a nephrologist

When coverage is available and medically necessary, the dosage, frequency, duration of therapy, and site of care should be reasonable, clinically appropriate, and supported by evidence-based literature and adjusted based upon severity, alternative available treatments, and previous response to therapy.

Reauthorization Criteria

Tolvaptan (Jynarque) is considered medically necessary for continued use when initial criteria are met AND there is documentation of beneficial response.

Authorization Duration

Initial approval duration: up to 12 months.

Reauthorization approval duration: up to 12 months.

Conditions Not Covered

Any other use is considered experimental, investigational or unproven, including the following (this list may not be all inclusive):

- 1. Individual 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 individuals 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 individuals with heart failure and syndrome of inappropriate antidiuretic hormone (SIADH). Samsca should be used for this condition.

Background

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 \geq 3.0 mL/min/1.73 m² over a period of \geq 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}

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.

"Cigna Companies" refers to operating subsidiaries of Cigna Corporation. All products and services are provided exclusively by or through such operating subsidiaries, including Cigna Health and Life Insurance Company, Connecticut General Life Insurance Company, Evernorth Behavioral Health, Inc., Cigna Health Management, Inc., and HMO or service company subsidiaries of Cigna Health Corporation. © 2023 Cigna.