PHARMACY POLICY STATEMENT
Marketplace

DRUG NAME	Jynarque (tolvaptan)
BENEFIT TYPE	Pharmacy
STATUS	Prior Authorization Required

Jynarque, initially approved by the FDA in 2018, is a selective vasopressin V2-receptor antagonist indicated to slow kidney function decline in adults at risk of rapidly progressing autosomal dominant polycystic kidney disease (ADPKD). Decreased binding of vasopressin to the V2-receptor in the kidney lowers adenylate cyclase activity resulting in a decrease in intracellular cAMP concentrations. Decreased cAMP concentrations prevent aquaporin 2 containing vesicles from fusing with the plasma membrane, which in turn causes an increase in urine water excretion, an increase in free water clearance (aquaresis) and a decrease in urine osmolality.

ADPKD is a genetic disorder that leads to the formation of cysts in the kidneys. It is sometimes referred to as "adult PKD" as it is usually diagnosed between 30 and 50 years of age. Symptoms include high blood pressure, flank pain, blood in the urine and poor function of the kidneys that gets worse over time. Organs other than the kidneys can become involved, but ultimately ADPKD leads to end stage renal disease.

Jynarque is only available through a Risk Evaluation and Mitigation Strategy (REMS) program due to the potential for fatal liver injury.

Jynarque (tolvaptan) will be considered for coverage when the following criteria are met:

Autosomal Dominant Polycystic Kidney Disease (ADPKD)

For **initial** authorization:

1. Member is at least 18 years of age; AND
2. Medication must be prescribed by or in consultation with a nephrologist; AND
3. Member has autosomal dominant polycystic kidney disease (ADPKD) confirmed by imaging (e.g. ultrasound, CT scan, or MRI scan) or genetic testing documented in chart notes; AND
4. Member is at risk of rapidly progressing disease, defined as having **ONE** of the following:
   a) Mayo classification 1D or 1E;
   b) Mayo classification 1C with **ONE** of the following:
      i)