

PHARMACY POLICY STATEMENT

Marketplace

DRUG NAME	Joenja (leniolisib)
BENEFIT TYPE	Pharmacy
STATUS	Prior Authorization Required

Joenja, approved by the FDA in 2023, is a small molecule kinase inhibitor indicated for the treatment of activated phosphoinositide 3- of age and older. It selectively targets signaling by inhibiting its hyperactive subunit. APDS is also known as p110 -activating mutation causing senescent T cells, lymphadenopathy, and immunodeficiency (PASLI). It is an ultra-rare disease caused by variants in either of the genes encoding the (*PIK3CD* in APDS1 or *PIK3R1* in APDS2) which leads to . This results in disrupted immune cell development and function. Clinical manifestations include infections, nonmalignant lymphoproliferation, autoimmunity (e.g., cytopenias), enteropathy, bronchiectasis, and increased risk of lymphoma. In a Phase 3 study, Joenja met the coprimary endpoints of reducing lymphadenopathy and normalizing immune cell subsets. It is the first FDA-approved drug for APDS.

Joenja (leniolisib) will be considered for coverage when the following criteria are met:

Activated Phosphoinositide 3-Kinase D

For **initial** authorization:

1. Member is at least 12 years of age; AND
2. Medication must be prescribed by or in consultation with an immunologist or hematologist; AND
3. Member has a diagnosis of APDS confirmed by an APDS- documented variant in either *PIK3CD* or *PIK3R1*; AND
4. Chart notes must show clinical findings/manifestations of APDS (e.g., history of repeated oto-sino-pulmonary infections); AND
5. Chart notes must document at least 1 measurable nodal lesion by CT or MRI; AND
6. Joenja is NOT being prescribed concurrently with an immunosuppressive medication.
7. **Dosage allowed/Quantity limit:** Weight 45 kg or greater: 70 mg orally twice daily. (QL: 60 tablets per 30 days)

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:

1. Chart notes must show positive clinical response such as reduced lymph node size, increased naïve B cell percentage (out of total B cells), reduced spleen volume, or improved cytopenias.

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Joenja (leniolisib) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
04/24/2023	New policy for Joenja created.

References:

1. Joenja [prescribing information]. Pharming Technologies B.V.; 2023.
2. A, et al. A randomized, placebo-controlled phase 3 trial of *Blood*. 2023;141(9):971-983. doi:10.1182/blood.2022018546
3. Rao VK, Webster S, Dalm