

PHARMACY POLICY STATEMENT

BILLING CODE	J3590
BENEFIT TYPE	Medical
SITE OF SERVICE ALLOWED	Home/Office/Outpatient
STATUS	Prior Authorization Required

Ryplazim (plasminogen, human-tvmh) is plasma-derived human plasminogen indicated for the treatment of patients with plasminogen deficiency type 1 (hypoplasminogenemia). It was approved by the FDA on June 4, 2021 and is the first approved treatment for plasminogen deficiency type 1.

Individuals with this disease lack a protein called plasminogen, which is responsible for the ability of the body to break down fibrin clots. Plasminogen deficiency leads to an accumulation of fibrin, causing the development of growths (lesions) that can impair normal tissue and organ function and may lead to blindness when these lesions affect the eyes. Ligneous conjunctivitis (LC) appears to be the most common clinical manifestation and is characterized by inflamed, woody growths on the conjunctival membranes that, if left untreated, can result in visual impairment or blindness.

Treatment with Ryplazim temporarily increases plasminogen levels in blood. The effectiveness and safety of Ryplazim (plasminogen) is primarily based on one single-arm, open-label (unblinded) clinical trial enrolling 15 adult and pediatric patients with plasminogen deficiency type 1. All patients received Ryplazim administered every two to four days for 48 weeks. The effectiveness of Ryplazim was demonstrated by at least 50% improvement of their lesions in all 11 patients who had lesions at baseline, and absence of recurrent or new lesions in any of the 15 patients through the 48 weeks of treatment.

Ryplazim (plasminogen, human-tvmh) will be considered for coverage when the following criteria are met:

For authorization:

1. Member must be at least 11 months old; **Medication** must be prescribed by a hematologist or medical geneticist;
2. Member has a documented history of disease-related lesions and symptoms consistent with the clinical diagnosis of hypoplasminogenemia; AND
3. Documentation of baseline plasminogen activity level < 45%.
4. **Medication** must be administered at a dose of 6.6 mg/kg body weight given intravenously every 2-4 days.

If all the above requirements are met, the medication will be approved for 12 weeks.

For :

Ryplazim will be reauthorized when chart notes show at least one of the following:

- a) Absence of recurrent or new lesions
- b) Decrease in the lesion number and/or size
- c) Increase in trough plasminogen activity level by at least 10% from baseline

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

