Pharmacy Medical Necessity Guidelines: Cushing's Disease Agents

Effective: January 1, 2025

Prior Authorization Required	√	Type of Review – Care Management	
Not Covered		Type of Review – Clinical Review	√
Pharmacy (RX) or Medical (MED) Benefit	RX	Department to Review	RXUM
These pharmacy medical necessity guidelines apply to the following: ☒ Tufts Health RITogether – A Rhode Island Medicaid Plan		**Fax Numbers:** RXUM: 617.673.0988	

Note: This guideline does not apply to Medicare Members (includes dual eligible Members).

OVERVIEW

FOOD AND DRUG ADMINISTRATION-APPROVED INDICATIONS

Korlym (mifepristone) is a cortisol receptor blocker indicated to control hyperglycemia secondary to hypercortisolism in adult patients with endogenous Cushing's syndrome who have type 2 diabetes mellitus or glucose intolerance and have failed surgery or are not candidates for surgery. Korlym (mifepristone) should not be used in the treatment of patients with type 2 diabetes unless it is secondary to Cushing's syndrome.

Recorlev (levoketoconazole) is a cortisol synthesis inhibitor indicated for the treatment of endogenous hypercortisolism in adult patients with Cushing's syndrome for whom surgery is not an option or has not been curative.

Signifor (pasireotide) is a somatostatin analog indicated for the treatment of adult patients with Cushing's disease for whom pituitary surgery is not an option or has not been curative.

Cushing's disease is a rare, aggressive endocrine condition that is caused by an ACTH-secreting pituitary adenoma. Cushing's disease can occur spontaneously in children and adults, and it typically occurs after puberty with similar frequency in males and females. Most adults are diagnosed between 25 years of age and 45 years of age.

Generally, the treatment of choice for Cushing's syndrome is curative surgery with selective pituitary or ectopic corticotrophin tumor resection. Second-line treatments include more radical surgery, radiation therapy (for Cushing's disease), medical therapy, and bilateral adrenalectomy.

Medical therapy is often required when surgery is delayed, contraindicated, or unsuccessful.

COVERAGE GUIDELINES

Korlym (mifepristone)

The plan may authorize coverage of **Korlym (mifepristone)** for Members when all the following criteria are met, and limitations do not apply:

Initial Authorization Criteria

1. Documented diagnosis of hyperglycemia secondary to endogenous Cushing's syndrome

AND

2. Documented diagnosis of type 2 diabetes or glucose intolerance

AND

3. Documentation that surgery (e.g., pituitary surgery, adrenal surgery) is not an option or has not been curative for the Member

AND

4. The prescribing physician is an endocrinologist

AND

5. Member is at least 18 years of age

Reauthorization Criteria

1. Documented diagnosis of hyperglycemia secondary to endogenous Cushing's disease

AND

2. Documented diagnosis of type 2 diabetes or glucose intolerance

AND

3. The prescribing physician is an endocrinologist

AND

4. Member is at least 18 years of age

AND

5. Documentation of improved or stable glucose tolerance

Recorlev (levoketoconazole)

The plan may authorize coverage of **Recorlev (levoketoconazole)** for Members when all the following criteria are met, and limitations do not apply:

Initial Authorization Criteria

1. Documented diagnosis of hypercortisolemia secondary to Cushing's disease

AND

2. Documentation that surgery (e.g., pituitary surgery, adrenal surgery) is not an option or has not been curative for the Member

AND

3. The prescribing physician is an endocrinologist

AND

4. Member is at least 18 years of age

Reauthorization Criteria

1. Documented diagnosis of hypercortisolemia secondary to Cushing's disease

AND

2. The prescribing physician is an endocrinologist

AND

3. Member is at least 18 years of age

AND

4. Documentation of a reduction in baseline 24-hour urinary free cortisol levels

Signifor (pasireotide)

The plan may authorize coverage of **Signifor (pasireotide)** for Members when all the following criteria are met, and limitations do not apply:

Initial Authorization Criteria

1. Documented diagnosis of Cushing's disease

AND

2. Documentation that pituitary surgery is not an option or has not been curative for the Member

AND

3. The prescribing physician is an endocrinologist

AND

4. Member is at least 18 years of age

Reauthorization Criteria

1. Documented diagnosis of Cushing's disease

AND

2. The prescribing physician is an endocrinologist

AND

3. Member is at least 18 years of age

AND

4. Documentation of a reduction in baseline 24-hour urinary free cortisol levels

LIMITATIONS

- Initial coverage of a Cushing's Disease Agent will be authorized for three (3) months. Reauthorization of a Cushing's Disease Agent will be provided in 12-month intervals.
- Members new to the plan stable on a Cushing's Disease agent should be reviewed against Reauthorization Criteria.

CODES

Medical billing codes may not be used for these medications. These medications must be obtained via the Member's pharmacy benefit.

REFERENCES

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APPROVAL HISTORY

September 13, 2022: Reviewed by Pharmacy & Therapeutics Committee.

- August 8, 2023: Removed the Limitations "The plan does not cover Korlym (mifepristone) for the treatment of patients with type 2 diabetes that is not secondary to Cushing's syndrome", "The plan does not cover Signifor (pasireotide) for any conditions not listed in the Pharmacy Coverage Guidelines above, unless there is sufficient documentation of efficacy and safety in the published literature", "Coverage for Signifor (pasireotide) will be limited to 60 ampules per 30 days", and "Coverage of Korlym (mifepristone) will be limited to 120 units per 30 days for Commercial Products and Tufts Health Direct."
- October 8, 2024: Removed Isturisa coverage is moved to non-formulary status effective 1/1/25 (eff 12/31/2024)

BACKGROUND, PRODUCT AND DISCLAIMER INFORMATION

Pharmacy Medical Necessity Guidelines have been developed for determining coverage for plan benefits and are published to provide a better understanding of the basis upon which coverage decisions are made. The plan makes coverage decisions on a case-by-case basis considering the individual member's health care needs. Pharmacy Medical Necessity Guidelines are developed for selected therapeutic classes or drugs found to be safe, but proven to be effective in a limited, defined population of patients or clinical circumstances. They include concise clinical coverage criteria based on current literature review, consultation with practicing physicians in the service area who are medical experts in the particular field, FDA and other government agency policies, and standards adopted by national accreditation organizations. The plan revises and updates Pharmacy Medical Necessity Guidelines annually, or more frequently if new evidence becomes available that suggests needed revisions.

For self-insured plans, coverage may vary depending on the terms of the benefit document. If a discrepancy exists between a Pharmacy Medical Necessity Guideline and a self-insured Member's benefit document, the provisions of the benefit document will govern.

Treating providers are solely responsible for the medical advice and treatment of members. The use of this policy is not a guarantee of payment or a final prediction of how specific claim(s) will be adjudicated. Claims payment is subject to member eligibility and benefits on the date of service, coordination of benefits, referral/authorization and utilization management guidelines when applicable, and adherence to plan policies and procedures and claims editing logic.

Provider Services