

DRUG POLICY

Crenessity® (crinecerfont)

BENEFIT APPLICATION

Benefit determinations are based on the applicable contract language in effect at the time the services were rendered. Exclusions, limitations or exceptions may apply. Benefits may vary based on contract, and individual member benefits must be verified. Wellmark determines medical necessity only if the benefit exists and no contract exclusions are applicable. This medical policy may not apply to FEP. Benefits are determined by the Federal Employee Program.

DESCRIPTION

The intent of the policy is to provide coverage consistent with product labeling, FDA guidance, standards of medical practice, evidence-based drug information, and/or published guidelines. The indications below including FDA-approved indications and compendial uses are considered covered benefits provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications

Crenessity is indicated as adjunctive treatment to glucocorticoid replacement to control androgens in adults and pediatric patients 4 years of age and older with classic congenital adrenal hyperplasia (CAH).

Limitations of Use

Coverage will not be provided for members with any of the following exclusions:

- Diagnosis of any other known forms of congenital adrenal hyperplasia (CAH) (e.g., 11-beta-hydroxylase deficiency, 17-alpha-hydroxylase deficiency)
- History of bilateral adrenalectomy, hypopituitarism, or other condition requiring chronic glucocorticoid therapy

POLICY

Required Documentation

Submission of the following information is necessary to initiate the prior authorization review:

1. Chart notes or medical records confirming diagnosis of classic congenital adrenal hyperplasia (CAH) by any of the following:
 - A. Genetic test to confirm presence of pathogenic variants in CYP21A2
 - B. Lab tests confirming 21-hydroxylase deficiency [e.g., baseline morning serum 17-hydroxyprogesterone (17-OHP) measurement by liquid chromatography-tandem mass spectrometry (LC-MS/MS), cosyntropin (ACTH) stimulation test, adrenal steroid profile]

2. Chart notes, medical record documentation, or claims history supporting current utilization of glucocorticoid therapy and stable for at least 1 month
3. Chart notes or medical record documentation confirming the member has achieved or maintained a positive clinical response to treatment (e.g., reduction in glucocorticoid therapy)
4. Chart notes or medical record documentation confirming the member is unable to swallow capsules if oral solution is requested for doses exceeding 100 mg per day

Prescriber Specialties

The requested medication must be prescribed by or in consultation with an endocrinologist.

Criteria for Initial Approval

Crenessity (crinicerfont) may be considered **medically necessary** for the treatment of classic congenital adrenal hyperplasia (CAH) when **ALL** the following criteria are met:

1. Patient is 4 years of age or older
2. The diagnosis is confirmed by any of the following:
 - A. Genetic test to confirm presence of pathogenic variants in CYP21A2
 - B. Lab tests confirming 21-hydroxylase deficiency [e.g., baseline morning serum 17-hydroxyprogesterone (17-OHP) measurement by liquid chromatography-tandem mass spectrometry (LC-MS/MS), cosyntropin (ACTH) stimulation test, adrenal steroid profile]
3. Member has diagnosis of classic congenital adrenal hyperplasia (CAH)
4. Member is currently receiving glucocorticoid therapy and stable for at least 1 month
5. Member is experiencing one of the following:
 - A. Member requires a supraphysiologic glucocorticoid dose to control their disease
 - i. Pediatric: total glucocorticoid dose >12 mg/m²/day in hydrocortisone dose equivalents (Appendix A)
 - ii. Adult: total glucocorticoid dose >13 mg/m²/day in hydrocortisone dose equivalents (Appendix B)
 - B. Patient has uncontrolled androgen levels despite maximally tolerated glucocorticoid dose
6. For doses greater than 100 mg per day requesting oral solution formulation, documentation is provided that patient is unable to swallow capsule whole due to one of the following:
 - A. Age
 - B. Dysphagia
 - C. Oral/motor difficulties
 - D. Medications are administered through a feeding tube

Initial approval will be for 6 months

Continuation of Therapy

The continuation of Crenessity (crinicerfont) may be considered **medically necessary** for the treatment classic congenital adrenal hyperplasia (CAH) when the member has achieved or maintained a positive clinical response (e.g., reduction in glucocorticoid therapy).

Approval will be for 12 months

Other

Crenessity (crinicerfont) is considered **not medically necessary** for members who do not meet the criteria set forth above.

Members currently receiving the requested medication as samples or via the manufacturer's patient assistance program will be required to meet the criteria for initial approval. This ensures that members are treated equally regardless of their provider's ability to access medication samples.

Dosing and Administration

Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines.

Quantity Limits Apply

Medication	Standard Limit	FDA Recommended Standard Dosing	FDA Recommended Dosing with Moderate CYP3A4 Inducer	FDA Recommended Dosing with Strong CYP3A4 Inducer
Crenessity (crinecerfont) 25 mg capsule	60 capsules per 30 days	Adults and pediatric patients ≥ 55 kg: 100 mg orally twice daily Pediatric: 10 kg to <20 kg: 25 mg orally twice daily 20 kg to <55 kg: 50 mg orally twice daily	Adults and pediatric patients ≥ 55 kg: 100 mg in the morning and 200 mg in the evening Pediatric: 10 kg to <20 kg: 25 mg in the morning and 50 mg in the evening 20 kg to <55 kg: 50 mg in the morning and 100 mg in the evening	Adults and pediatric patients ≥ 55 kg: 200 mg orally twice daily Pediatric: 10 kg to <20 kg: 50 mg orally twice daily 20 kg to <55 kg: 100 mg orally twice daily
Crenessity (crinecerfont) 50 mg capsule	60 capsules per 30 days			
Crenessity (crinecerfont) 100 mg capsule	120 capsules per 30 days			
Crenessity (crinecerfont) 50 mg/mL oral solution	120 mL per 30 days			

Appendices

Appendix A: Hydrocortisone 12 mg/m2/day dose equivalency

Medication	Equivalent Dose
Dexamethasone	0.45 mg/m2/day
Prednisone	3 mg/m2/day
Prednisolone	3 mg/m2/day

Appendix B: Hydrocortisone 13 mg/m2/day dose equivalency

Medication	Equivalent Dose
Dexamethasone	0.49 mg/m2/day
Prednisone	3.25 mg/m2/day
Prednisolone	3.25 mg/m2/day

PROCEDURES AND BILLING CODES

To report provider services, use appropriate CPT* codes, Alpha Numeric (HCPCS level 2) codes, Revenue codes, and/or ICD diagnostic codes.

- No applicable codes

REFERENCES

- Crenessity [package insert]. San Diego, CA: Neurocrine Biosciences, Inc.; December 2024
- Speiser PW, Arlt W, Auchus RJ, et al. Congenital Adrenal Hyperplasia Due to Steroid 21-Hydroxylase Deficiency: An Endocrine Society Clinical Practice Guideline. J Clin Endocrinol Metab. 2018;103:4043-4088.
- Sarafoglou K, Kim MS, Lodish M, et al. Phase 3 Trial of Crinicerfont in Pediatric Congenital Adrenal Hyperplasia. N Engl J Med. 2024;391:493-503.
- Auchus RJ, Hamidi O, Pivonllo R, et al. Phase 3 Trial of Crinicerfont in Adult Congenital Adrenal Hyperplasia. N Engl J Med. 2024;391(6):604-514.
- Merke DP, Auchus RJ. Congenital Adrenal Hyperplasia Due to 21-Hydroxylase Deficiency. N Engl J Med. 2020;383:1248-61.

POLICY HISTORY

Policy #: 05.05.78

Original Effective Date: May 21, 2025

Reviewed: January 2026

Revised:

Current Effective Date: May 21, 2025