Effective Date: 08/1/2025
Reviewed: 05/2025 Scope: Medicaid

Crenessity (crinecerfont)

POLICY

I. INDICATION

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).

II. CRITERIA FOR APPROVAL

Classic Congenital Adrenal Hyperplasia (CAH)
An authorization of 6 months may be granted for the treatment of classical congenital adrenal hyperplasia (CAH) if all of the following criteria are met:

1. The patient is ≥4 years of age.
2. The medication must be prescribed by or in consultation with an endocrinologist.
3. Documentation with chart notes/medical record 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 by either of the following:
        i. Baseline morning serum 17-hydroxyprogesterone (17-OHP) > 3,000 ng/dL as measured by liquid chromatography-tandem mass spectrometry (LC-MS/MS)
        ii. Cosyntropin (ACTH) stimulation test resulting in 17-OHP level >10,000 ng/dL
4. Documentation with chart notes/medical record that patient requires chronic treatment with a supraphysiologic glucocorticoid (GC) regimen (e.g., dexamethasone, hydrocortisone, methylprednisolone, prednisone, prednisolone) defined as one of the following:
    a. For patients 4 to 17 years old: GC dose > 12 mg/m^2/day in hydrocortisone dose equivalents
    b. For adult patients (≥ 18 years old): GC dose > 13 mg/m^2/day in hydrocortisone dose equivalents
5. Documentation supporting current utilization of glucocorticoid therapy and stable for at least 1 month
6. Crenessity will be prescribed in combination with glucocorticoid treatment.
7. Documentation of patient's weight and the requested dose is optimized and within the FDA-approved guidelines
8. For oral solution requests for patients weighing ≥20 kg, documentation of medical rationale for patient's inability to swallow the oral capsule whole
9. Documentation that Crenessity will not be taken concomitantly with a strong or moderate CYP3A4 inducer, or that an appropriate therapeutic alternative is not available
10. Coverage will not be provided for either of the following:
    a. Diagnosis of any other known forms of congenital adrenal hyperplasia (CAH) (e.g., 11-betahydroxylase deficiency, 17-alpha-hydroxylase deficiency)
    b. History of bilateral adrenalectomy, hypopituitarism, or other condition requiring chronic glucocorticoid therapy

III. CONTINUATION OF THERAPY

Classic Congenital Adrenal Hyperplasia (CAH)
An authorization of 6 months may be granted for the continued treatment in members requesting reauthorization for classic CAH when all of the following criteria are met:

1. Documentation with chart notes/medical record that the patient has achieved or maintained a positive clinical response (e.g. reduction in total daily glucocorticoid dose from baseline, stabilization or decreased androstenedione [A4] levels).
2. The medication continues to be prescribed by or in consultation with an endocrinologist.
3. Crenessity continues to be prescribed in combination with glucocorticoid treatment.
4. Documentation of patient's weight and the requested dose is optimized and within the FDA-approved guidelines
5. For oral solution requests for patients weighing ≥20 kg, documentation of medical rationale for patient's inability to swallow the oral capsule whole
6. Documentation that Crenessity will not be taken concomitantly with a strong or moderate CYP3A4 inducer, or that an appropriate therapeutic alternative is not available

IV. DOSAGE AND ADMINISTRATION

The recommended dosage of Crenessity for adults is 100 mg orally, twice daily with a meal in the morning and evening

The recommended dosage of Crenessity for pediatric patients 4 years of age and older is weight-based and administered orally, twice daily with a meal in the morning and evening:

Weight	Dose Regimen with a Meal
10 kg to less than 20 kg	25 mg orally twice daily
20 kg to less than 55 kg	50 mg orally twice daily
Greater than or equal to 55 kg	100 mg orally twice daily

V. QUANTITY LIMIT

Crenessity 50mg and 100mg capsules: 2 capsules per day
Crenessity 50mg/ml oral solution: 2 mL per day

A quantity limit exception may be granted if the patient is unable to avoid concomitant use with a **strong CYP3A4 inducer** (e.g., phenytoin, carbamazepine, rifampin, rifabutin, rifapentine, phenobarbital and lumacaftor) for either:

- Crenessity 100mg capsules: 4 capsules per day for adults and pediatric patients weighing at least 55 kg
- Crenessity 50mg/ml oral solution: 4 mL per day for pediatric patients weighing 20 kg to less than 55 kg that have a documented inability to swallow the capsules whole

A quantity limit exception may be granted if the patient is unable to avoid concomitant use with a **moderate CYP3A4 inducer** (e.g., bosentan, efavirenz, etravirine, and modafinil) for either:

- Crenessity 100mg capsules: 3 capsules per day for adults and pediatric patients weighing at least 55 kg
- Crenessity 50mg/ml oral solution: 3 mL per day for pediatric patients weighing 20 kg to less than 55 kg that have a documented inability to swallow the capsules whole

*_Note:_ A quantity limit exception may **not** be granted for 3 capsules per day of Crenessity 50mg, as a dose-optimized regimen will be preferred (one (1) 50mg and one (1) 100mg capsule per day)

VI. REFERENCES

1. Crenessity [package insert]. San Diego, CA: Neurocine Biosciences, Inc.; March 2025.
2. 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.
3. Furst DE and Saag KG. Overview of the pharmacologic use of glucocorticoids. In: UpToDate, Warrington KJ and Case CM (Ed), Wolters Kluwer. (Accessed on April 10, 2025.)