

<b>Clinical Policy Title:</b>	lumacaftor/ivacaftor
<b>Policy Number:</b>	RxA.435
<b>Drug(s) Applied:</b>	Orkambi®
<b>Original Policy Date:</b>	03/06/2020
<b>Last Review Date:</b>	10/19/2023
<b>Line of Business Policy Applies to:</b>	All lines of business (except Medicare)

## Criteria

### I. Initial Approval Criteria

#### A. Cystic Fibrosis (must meet all):

1. Diagnosis of CF;
2. Age  $\geq$  1 years;
3. Prescribed by or in consultation with a pulmonologist or a physician who specializes in the treatment of cystic fibrosis;
4. Member is homozygous for the F508del mutation in the CFTR gene;
5. Dose does not exceed one of the following (a, b, c, or d):
  - a. Age 1 to 2 years and one of the following (i, ii, or iii):
    - i. Weight 7 kg to < 9 kg: lumacaftor 150 mg/ivacaftor 188 mg per day;
    - ii. Weight 9 kg to < 14 kg: lumacaftor 200 mg/ivacaftor 250 mg per day;
    - iii. Weight  $\geq$  14 kg: lumacaftor 300 mg/ivacaftor 376 mg per day;
  - b. Age 2 to 5 years and one of the following (i or ii):
    - i. Weight < 14 kg: lumacaftor 200 mg/ivacaftor 250 mg per day;
    - ii. Weight  $\geq$  14 kg: lumacaftor 300 mg/ivacaftor 376 mg per day;
  - c. Age 6 to 11 years: lumacaftor 400 mg/ivacaftor 500 mg per day;
  - d. Age  $\geq$  12 years: lumacaftor 800 mg/ivacaftor 500 mg per day.

#### Approval Duration

**Commercial:** 12 months

**Medicaid:** 12 months

### II. Continued Therapy Approval

#### A. Cystic Fibrosis (must meet all):

1. Member is currently receiving medication that has been authorized by RxAdvance or the member has met initial approval criteria listed in this policy;
2. If request is for a dose increase, new dose does not exceed one of the following (a, b, c, or d):
  - a. Age 1 to 2 years and one of the following (i, ii, or iii):
    - i. Weight 7 kg to < 9 kg: lumacaftor 150 mg/ivacaftor 188 mg per day;
    - ii. Weight 9 kg to < 14 kg: lumacaftor 200 mg/ivacaftor 250 mg per day;
    - iii. Weight  $\geq$  14 kg: lumacaftor 300 mg/ivacaftor 376 mg per day;
  - b. Age 2 to 5 years and one of the following (i or ii):
    - i. Weight < 14 kg: lumacaftor 200 mg/ivacaftor 250 mg per day;
    - ii. Weight  $\geq$  14 kg: lumacaftor 300 mg/ivacaftor 376 mg per day;

This clinical policy has been developed to authorize, modify, or determine coverage for individuals with similar conditions. Specific care and treatment may vary depending on individual need and benefits covered by the plan. This policy is not intended to dictate to providers how to practice medicine, nor does it constitute a contract or guarantee regarding payment or results. This document may contain prescription brand name drugs that are trademarks of pharmaceutical manufacturers that are not affiliated with RxAdvance.

- c. Age 6 to 11 years: lumacaftor 400 mg/ivacaftor 500 mg per day;
- d. Age ≥ 12 years: lumacaftor 800 mg/ivacaftor 500 mg per day.

**Approval Duration**

**Commercial:** 12 months

**Medicaid:** 12 months

**References**

1. Mogayzel PJ, Naureckas ET, Robinson KA, et al. Cystic fibrosis pulmonary guidelines: Chronic medications for maintenance of lung health. Am J Respir Crit Care Med. 2013; 187(7): 680-689. Available at: <https://pubmed.ncbi.nlm.nih.gov/23540878/>. Accessed April 24, 2023.
2. Ren CL, Morgan RL, Oermann C, et al. Cystic Fibrosis Foundation pulmonary guidelines: Use of cystic fibrosis transmembrane conductance regulator modulator therapy in patients with cystic fibrosis. Ann Am Thorac Soc. 2018; 15(3): 271-280. Available at: <https://pubmed.ncbi.nlm.nih.gov/29342367/>. Accessed April 24, 2023.
3. Farrell PM, White TB, Ren CL et al. Diagnosis of cystic fibrosis: Consensus guidelines from the Cystic Fibrosis Foundation. J Pediatr. 2017; 181S: S4-15. Available at: <https://pubmed.ncbi.nlm.nih.gov/28129811/> Accessed April 24, 2023.
4. Alexander S, Alshafi K, Al-Yaghchi C, et al. Clinical Guidelines: Care of Children with Cystic Fibrosis. Royal Brompton and Harefield NHS. 2020;(8):22-23. Available at: <https://www.rbht.nhs.uk/sites/nhs/files/Corporate%20governance/CF%20guideline%202020%20FINAL.pdf>. Accessed April 25, 2023.
5. Cystic Fibrosis Foundation: Clinical Care Guidelines. Available at: <https://www.cff.org/medical-professionals/clinical-care-guidelines>. Accessed April 25, 2023.

Review/Revision History	Review/Revised Date	P&T Approval Date
Policy established.	01/2020	03/06/2020
Policy was reviewed: 1) Policy title was updated. 2) Continued Therapy Approval criteria II.A.1 was rephrased. 3) References were updated.	03/06/2020	09/14/2020
Policy was reviewed: 1. Initial Approval Criteria and Continued Therapy Approval criteria were updated to remove HIM approval duration. 2. Continued Therapy Approval Criteria II.A.1 was rephrased to " Member is currently receiving medication that has been authorized by RxAdvance...". 3. References were reviewed and updated.	7/1/2021	9/14/2021
Policy was reviewed: 1. Initial Approval Criteria I.A.3: Updated to add prescriber criteria. 2. References were reviewed and updated.	04/04/2022	07/18/2022
Policy was reviewed:	04/25/2023	07/13/2023

<ol style="list-style-type: none"> <li>1. Initial Approval Criteria, I.A.7.a: Updated to include new age specific dosing criteria "Age 1 to 2 years".</li> <li>2. Continued Therapy Approval Criteria, II.A.4.a: Updated to include new age specific dosing criteria "Age 1 to 2 years".</li> <li>3. References were reviewed and updated.</li> </ol>		
<p>Policy was reviewed.</p>	<p>10/19/2023</p>	<p>10/19/2023</p>