



STATE OF WEST VIRGINIA  
DEPARTMENT OF HEALTH AND HUMAN RESOURCES  
BUREAU FOR MEDICAL SERVICES



Office of Pharmacy Services  
Prior Authorization Criteria

ORKAMBI®  
(lumacaftor/ivacaftor)  
Prior Authorization Request Form  
*Effective 10/28/2022*

ORKAMBI is a combination of ivacaftor, a cystic fibrosis transmembrane conductance regulator (CFTR) potentiator, and lumacaftor, indicated for the treatment of cystic fibrosis (CF) in patients aged 1 year and older who are **homozygous** for the F508del mutation in the CFTR gene. If the patient's genotype is unknown, an FDA-cleared CF mutation test should be used to detect the presence of the F508del mutation on both alleles of the CFTR gene.

**Criteria for Approval:**

- 1) The patient is within the age range as recommended by the FDA label; **AND**
- 2) Patient must have a confirmed diagnosis of Cystic Fibrosis; **AND**
- 3) Patient must be determined to be **homozygous** for the **F508del** mutation in the CFTR gene as confirmed by an FDA-approved CF mutation test; **AND**
- 4) Patient must have a documented baseline AST, ALT and FEV<sub>1</sub> (forced expiratory volume in one second) presented with the prior authorization request; **AND**
- 5) Patients under the age of 18 must have undergone a baseline ophthalmic examination to monitor for lens opacities/cataracts.

***Prior authorizations will be for every 6 months in the first year, followed thereafter by an annual prior authorization.***

**Criteria for Continuation of Therapy**

- 1) Patients under the age of 18 must have follow up ophthalmic examinations at least annually (documentation required); **AND**
- 2) Patient must have LFTs/bilirubin monitored every 6 months for the first year of treatment and annually thereafter (documentation required); **AND**
- 3) Serum ALT or AST < 5 times the upper limit of normal (ULN); **OR**
- 4) Serum ALT or AST < 3 times the ULN with bilirubin < 2 times the ULN.

**References**

- 1) Orkambi package insert revised 7/2019, 9/2022
- 2) Lexi-Comp Clinical Application 11/15/2019



**STATE OF WEST VIRGINIA  
DEPARTMENT OF HEALTH AND HUMAN RESOURCES  
BUREAU FOR MEDICAL SERVICES**

