

ORKAMBI (ivacaftor/lumacaftor) PA Criteria

1. What is the patient's diagnosis? Cystic Fibrosis (CF), ICD 10 code: _____

Complete below information for applicable situation, Initiation or Continuation of therapy.

This is **INITIATION** of therapy. *(Please answer the following questions):*

a. Yes No The patient is 6 years of age or older.

b. Yes No Submission of laboratory results or other supportive documentation confirming that patient is homozygous for the F508del mutation in the cystic fibrosis transmembrane regulator (CFTR) gene?

c. Yes No Will the baseline ALT/AST and bilirubin levels be drawn and monitored every 3 months for the first year with adjustment in dosing dependent on severity of liver function?

d. Yes No Ivacaftor and lumacaftor is being prescribed by or in consultation with a prescriber who specializes in treating CF patients. Name of CF treating specialist/consult _____

Initial authorization will be issued for 6 months.

This is **CONTINUATION** of therapy. (Please answer the following questions):

•If yes, when was the requested medication started? _____

1. Ivacaftor and lumacaftor will be approved based on both of the following criteria:

•Provider attests that in comparison to baseline, the patient has achieved a clinically meaningful response while on ivacaftor and lumacaftor therapy to one of the following:

a. Yes No Lung function as demonstrated by improvement in percent predicted expiratory volume in 1 second (ppFEV1)?

b. Yes No Body mass index (BMI)

c. Yes No Pulmonary exacerbations

d. Yes No Quality of life as demonstrated by Cystic Fibrosis Questionnaire-Revised (CFQ-R) respiratory domain score

-AND

2. Yes No Will there be annual testing of ALT/AST and bilirubin levels after the first year of therapy?

3. Yes No Ivacaftor/lumacaftor is being prescribed by or in consultation with a prescriber who specializes in treating CF patients. Name of CF treating specialist/consult _____

Reauthorization will be issued for 12 months.