curative

Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) Prior Authorization Drug List A

Drug(s) Applied:	Kalydeco, Trikafta, Symdeko, Orkambi

Criteria:
Drug(s) Applied will be approved when the requested medication is being used for an FDA approved indication and all of the following criteria are met:

I. Initial Approval Criteria

A. Cystic Fibrosis, homozygous F508del mutation or mutation responsive (Kalydeco, Symdeko, Orkambi) as indicated by chart notes within past 6 months:

1. Diagnosis of Cystic Fibrosis as confirmed by genetic testing with ONE of the following:
   (1) Homozygous for F508del mutation in the CFTR gene (Symdeko or Orkambi) according, **or**
   (2) A mutation responsive to the requested drug based on clinical and/or in vitro assay data (Symdeko or Kalydeco)
2. Patient's age is within FDA labeling for the requested indication for the requested agent, **and**
3. Patient will NOT be using the requested agent in combination with another CFTR modulator agent for the requested indication, **and**
4. Prescriber is a specialist, or has consulted with a specialist in the area of the patient's diagnosis (e.g. cystic fibrosis, pulmonologist)

Approval Duration: 6 months

B. Cystic Fibrosis, at least 1 F508del mutation or mutation responsive (Trikafta) as indicated by chart notes within past 6 months:

1. Diagnosis of Cystic Fibrosis as confirmed by genetic testing with ONE of the following:
   a) Presence of ≥1 F508del mutation in the CFTR gene, **or**
   b) A mutation responsive to the requested drug based on clinical and/or in vitro assay data, **and**
2. Patient's age is within FDA labeling for the requested indication for the requested agent, **and**
3. Patient will NOT be using the requested agent in combination with another CFTR modulator agent for the requested indication, **and**

4. Prescriber is a specialist, or has consulted with a specialist, in the area of the patient's diagnosis (e.g. cystic fibrosis, pulmonologist), **and**
5. Documentation of liver function test (LFT) results, including alanine aminotransferase (ALT), aspartate aminotransferase (AST), alkaline phosphatase, and bilirubin within 1 month prior to starting the requested drug, **and**
6. Patient does not have severe hepatic impairment (Child-Pugh Class C)

Approval Duration: 6 months

II. Continued Therapy Approval

A. Cystic Fibrosis, homozygous F508del mutation or mutation responsive (Kalydeco, Symdeko, Orkambi), as indicated by chart notes within past 6 months:

1. Patient has been previously approved for the requested agent through the plan's Prior Authorization process, or meets Initial Approval criteria **and**
2. Confirmed clinical improvement or stabilization with the requested agent from baseline (prior to treatment with the requested agent) as evidenced through at least one of the following:
    a) Improvement in FEV1or lung function tests, such as Cystic Fibrosis Questionnaire-Revised (CFQ-R) Respiratory Domain score, **or**
    b) Increase in weight/BMI, **or**
    c) Improvement in sweat chloride, **or**
    d) Reduced number of pulmonary exacerbations and/or pulmonary infections, **and**
3. The patient will NOT be using the requested agent in combination with another CFTR modulator agent for the requested indication, **and**
4. Prescriber is a specialist, or has consulted with a specialist, in the area of the patient's diagnosis (e.g. cystic fibrosis, pulmonologist)

Approval Duration: 12 months

B. Cystic Fibrosis, at least 1 F508del mutation or mutation responsive (Trikafta), as indicated by chart notes within past 6 months:

1. Patient has been previously approved for the requested agent through the plan's Prior Authorization process, or meets Initial Approval criteria **and**
2. Confirmed clinical improvement or stabilization with the requested agent from baseline (prior to treatment with the requested agent) as evidenced through at least one of the following:
    a) Improvement in FEV1or lung function tests, such as Cystic Fibrosis Questionnaire-Revised (CFQ-R) Respiratory Domain score, **or**
    b) Increase in weight/BMI, **or**

c) Improvement in sweat chloride, **or**
d) reduced number of pulmonary exacerbations and/or pulmonary infections, **and**

3. Patient will NOT be using the requested agent in combination with another CFTR modulator agent for the requested indication, **and**
4. Prescriber is a specialist, or has consulted with a specialist, in the area of the patient's diagnosis (e.g. cystic fibrosis, pulmonologist), **and**
5. Documentation within the 365 days of liver function test (LFT) results, including alanine aminotransferase (ALT), aspartate aminotransferase (AST), alkaline phosphatase, and bilirubin, **and**
6. ONE of the following:
   a) Patient does not have evidence of elevated LFT results, **or**
   b) Patient is being followed by a hepatologist with notes indicating benefit outweighs risk, **and**
7. Patient does not have severe hepatic impairment (Child-Pugh Class C)

Approval Duration: 12 months

Policy Owned by: Curative PBM team