CYSTIC FIBROSIS TRANSMEMBRANE CONDUCTANCE REGULATORS

Included Products: Kalydeco (Ivacaftor), Orkambi (Ivacaftor/Lumacaftor), Symdeko (Ivacaftor/Tezacaftor), Trikafta (Elexacaftor/Ivacaftor/Tezacaftor)

Created: 05/21/2012 Revised: 09/12/2024 *Reviewed*: 09/12/2024 *Updated*: 10/01/2024

Cystic Fibrosis			
Initial Criteria		**If yes**	**If no**
1.	Does the member have a diagnosis of cystic fibrosis?	Continue to #2.	Do not approve.
2.	Is the request from a pulmonologist?	Continue to #3.	Do not approve.
3.	Is the requested product appropriate in the patient's age?	Continue to #4.	Do not approve.
4.	Does the member have an FDA indicated mutation confirmed with an FDA-cleared genetic test? a. Trikafta: at least one F508del mutation in the CFTR gene or with Trikafta responsive mutation in the CFTR gene (must review FDA label directly) b. Symdeko: Homozygous for F508del mutation or with Symdeko responsive mutation in CFTR gene (must review FDA label directly) c. Orkambi: Homozygous for F508del mutation d. Kalydeco: Ivacaftor responsive mutation in CFTR gene (must review FDA label directly)	Continue to #5.	Do not approve.
5.	Approve for 6 months.		
Renewal Criteria		**If yes**	**If no**
1.	Is this the first renewal following the original 6 month approval?	Continue to #2.	Continue to #3.
2.	Did the member demonstrate a documented objective response by one of the following? a. A lack of decline in FEV1 verified with documentation b. A reduction in the incidence of pulmonary exacerbations c. A significant improvement in BMI by 10% from baseline	Continue to #3.	Do not approve. Not medically necessary.

3.	Is the request for Symdeko?	Continue to #4.	Continue to #5.
4.	Has there been monitoring of liver function testing completed?	Continue to #5.	Review case with medical director.
5.	Has the member shown compliance with fill history and documentation of ongoing oversight and cystic fibrosis management by the prescriber?	Continue to #6.	Review case with medical director.
6.	Approve for 12 months.		