Division: Pharmacy Policy	Subject: Prior Authorization Criteria
Original Development Date: Original Effective Date: Revision Date:	March 30, 2018 June 18, 2018

SYMDEKO® (tezacaftor/ivacaftor)

LENGTH OF AUTHORIZATION: Up to 6 months

REVIEW CRITERIA:

- Patient must be ≥ 12 years old.
- Must have a diagnosis of Cystic Fibrosis confirmed via "health conditions" or medical records.
- Patient is homozygous for the *F508del* mutation or patient has at least one mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene that is responsive to tezacaftor/ivacaftor based on *in vitro* data and/or clinical evidence. The CFTR mutations that are responsive to tezacaftor/ivacaftor include *E56K, P67L, R74W, D110E, D110H, R117C, E193K, L206W, R347H, R352Q, A455E, D579G, 711+3A→G, E831X, S945L, S977F, F1052V, K1060T, A1067T, R1070W, F1074L, D1152H, D1270N, 2789+5G→A, 3272-26A→G, and 3849+10kbC→T.*
- If a patient's genotype is unknown, an FDA-cleared cystic fibrosis mutation test should be used to detect the presence of CFTR mutation followed by verification with bi-directional sequencing when recommended by the mutation test instructions for use.
- Baseline liver function tests prior to initiating therapy, then every 3 months the first year, then annually.
- Baseline ophthalmic examination to monitor lens opacities/cataracts in pediatric patients 12 through 17 years of age, not required in adults 18 years and older.
- Baseline documented percent predicted FEV1 within the previous 30 days

CONTINUATION OF THERAPY:

- Disease response as indicated by one or more of the following:
  - Decreased pulmonary exacerbations compared to pretreatment baseline
  - Improvement or stabilization of lung function (as measured by percent predicted FEV1) compared to baseline or decrease in the rate of decline of lung function.
  - Weight gain
  - Improvement in quality of life
- Patient has not received a lung transplant.
- Patient has not experienced unacceptable toxicity from the drug.

DOSING and ADMINISTRATION:

- One tablet (tezacaftor 100mg/ivacaftor 150mg) in the morning and one tablet of 150 mg ivacaftor in the evening, approximately 12 hours apart.