SYMDEKO® (tezacaftor/ivacaftor)

LENGTH OF AUTHORIZATION: Up to 6 months

REVIEW CRITERIA:

- Patient must be ≥ 6 years old.
- Must have a diagnosis of Cystic Fibrosis confirmed via “health conditions” or medical records.
- 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:

- Pediatric patients age 6 to less than 12 years weighing less than 30 kg: one tablet (containing tezacaftor 50 mg/ivacaftor 75 mg) in the morning and one tablet (containing ivacaftor 75 mg) in the evening, approximately 12 hours apart. SYMDEKO should be taken with fat-containing food.

- Adults and pediatric patients age 12 years and older or pediatric patients age 6 to less than 12 years weighing 30 kg or more: one tablet (containing tezacaftor 100 mg/ivacaftor 150 mg) in the morning and one tablet (containing ivacaftor 150 mg) in the evening, approximately 12 hours apart. SYMDEKO should be taken with fat-containing food.