KALYDECO® (ivacaftor)

LENGTH OF AUTHORIZATION: Up to 6 months

REVIEW CRITERIA:

- Patient must be $\geq 6$ months old.
- Must have a diagnosis of Cystic Fibrosis confirmed via “health conditions” or medical records.
- Must have genetic testing confirming the presence of one cystic fibrosis transmembrane conductance regulator (CFTR) mutation.
  - (It is not effective in CF patients with two copies (homozygous) of the F508del mutation ($F508del/F508del$) in the CF gene and should not be approved for these patients)
- Baseline liver function tests prior to initiating therapy that are less than 3 times the upper limit of normal, then every 3 months the first year, then annually
- Baseline ophthalmic examination to monitor lens opacities/cataracts
- 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 6 months to $< 6$ years of age and weighing 5kg to $< 7$kg: One 25 mg packet mixed with 1 teaspoon (5 mL) of soft food or liquid and administered orally every 12 hours with fat-containing food.
- Pediatric patients 6 months to less than 6 years of age and weighing 7 kg to $< 14$ kg: One 50 mg packet mixed with 1 teaspoon (5 mL) of soft food or liquid and administered orally every 12 hours with fat-containing food.
- Pediatric patients 6 months to $< 6$ years of age and 14 kg or greater: One 75 mg packet mixed with 1 teaspoon (5 mL) of soft food or liquid and administered orally every 12 hours with fat-containing food.
- Adults and pediatric patients age 6 years and older: 150 mg tablet taken orally every 12 hours with fat-containing food.
- Dosage form: 25mg, 50mg, 75mg granule packets; 150 mg tablets