Clinical Policy Title:	ivacaftor
Policy Number:	RxA.190
Drug(s) Applied:	Kalydeco®
Original Policy Date:	02/07/2020
Last Review Date:	08/28/2024
Line of Business Policy Applies to:	All lines of Business (except Medicare)

Criteria

I. Initial Approval Criteria

A. Cystic Fibrosis (CF) (must meet all)**:**

1. Diagnosis of CF confirmed by all of the following (a, b, c, and d);
   a. Clinical symptoms consistent with CF in at least one organ system, or positive newborn screen or genetic testing for siblings of patients with CF;
   b. Evidence of CFTR dysfunction confirmed by one of the following (i or ii):
      i. Elevated sweat chloride ≥ 60 mmol/L;
      ii. Genetic testing confirming the presence of two disease-causing mutations in CFTR gene, one from each parental allele;
   c. Presence of one mutation in the CFTR gene responsive to ivacaftor based on clinical and/or in vitro assay data;
   d. Confirmation that a homozygous F508del mutation in the CFTR gene is not present;
2. Kalydeco is not prescribed concurrently with other CFTR modulators (e.g., Orkambi®, Symdeko®, Trikafta™).

Approval Duration

All Lines of Business (except Medicare): 6 months

II. Continued Therapy Approval

A. Cystic Fibrosis (must meet all)**:**

1. Auto-approval based on lookback functionality within the past 120 days as a proxy for member responding positively to therapy.

Approval Duration

All Lines of Business (except Medicare): 12 months

References

1. Mogayzel PJ, Naureckas ET, Robinson KA, et al. Cystic fibrosis pulmonary guidelines: Chronic medications for maintenance of lung health. Am J Respir Crit Care Med. 2013; 187(7): 680-689. Available at: https://pubmed.ncbi.nlm.nih.gov/23540878/. Accessed August 28, 2024.
2. Farrell PM, White TB, Ren CL et al. Diagnosis of cystic fibrosis: Consensus guidelines from the Cystic Fibrosis Foundation. J Pediatr. 2017; 181S: S4-15. Available at: https://pubmed.ncbi.nlm.nih.gov/28129811/. Accessed August 28, 2024.

Review/Revision History	Review/Revised Date	P&T Approval Date
Policy established.	01/2020	02/07/2020
Policy was reviewed: 1. Policy title table was updated. 2. Line of Business Policy Applies to was update to all lines of business. 3. Continued Therapy criteria II.A.1 was rephrased to "Currently receiving medication that has been authorized by RxAdvance..." 4. Initial Approval criteria: Commercial approval duration was updated from length of benefit to 6 months. 5. Continued Approval criteria: Commercial approval duration was updated from length of benefit to 12 months. 6. References were updated.	06/23/2020	09/14/2020
Policy was reviewed: 1. Last Review Date was updated. 2. Initial approval criteria was updated for minimum age of 4 months. 3. Initial and continuation criteria were updated to include maximum dose for patients age 4 months to less than 6 months. 4. References were updated. • Dosage regimen updated to include: Not recommended in patients less than 4 months of age. • Reduce dose in patients 6 months and older with moderate or severe hepatic impairment. • Not recommended in patients 4 months to less than 6 months of age with hepatic impairment.	02/22/2021	06/10/2021
Policy was reviewed: 1. Initial Approval Criteria I.A.3: Updated to add Prescribed by or in consultation with a pulmonologist or a physician who specializes in the treatment of CF. 2. Continued Therapy Approval Criteria II.A.1 was rephrased to "Member is currently receiving medication that has been authorized by RxAdvance...". 3. References were review and updated.	01/18/2022	04/18/2022
Policy was reviewed: 1. Initial Approval Criteria, I.A : Updated to include new diagnostic criteria Diagnosis of CF to	01/18/2023	04/13/2023

Diagnosis of CF confirmed by all of the following (a, b, c, and d):; a. Clinical symptoms consistent with CF in at least one organ system, or positive newborn screen or genetic testing for siblings of patients with CF; b. Evidence of CFTR dysfunction confirmed by one of the following (i or ii): i. Elevated sweat chloride ≥ 60 mmol/L; ii. Genetic testing confirming the presence of two disease-causing mutations in CFTR gene, one from each parental allele; c. Presence of one mutation in the CFTR gene responsive to ivacaftor based on clinical and/or in vitro assay data; d. Confirmation that a homozygous F508del mutation in the CFTR gene is not present. 2. Initial Approval Criteria I.A and Continued Therapy Criteria I.B: Updated to add Kalydeco is not prescribed concurrently with other CFTR modulators (e.g., Orkambi®, Symdeko®, Trikafta™). 3. References were review and updated.		
Policy was reviewed.	10/19/2023	10/19/2023
Policy was reviewed: 1. Removed age restrictions. 2. Removed prescriber restrictions. 3. Removed dose restrictions. 4. Updated Continued therapy approval with auto-approval based on lookback functionality within the past 120 days. 5. Removed other reauthorization requirements including positive response to therapy. 6. Updated approval duration verbiage. 7. References were reviewed and updated.	08/28/2024	09/13/2024