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One Island Health System

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PEI Pharmacare Bulletin

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NEW PRODUCT(S) ADDED TO THE PEI PHARMACARE FORMULARY (EFFECTIVE DATE: (NOVEMBER 1, 2021)

Product (Generic name)	Product (Brand name)	Strength	Dosage Form	DIN	MFR
Elexacaftor / Tezacaftor / Ivacaftor and Ivacaftor	Trikafta	100 mg/ 50 mg/ 75 mg tablet and 150 mg tablet	Tablet	02517140	VER
Criteria	For the treatment of cystic fibrosis (CF) in patients 12 years of age and older who meet all of the following criteria: • Confirmed diagnoses of CF with at least one F508del mutation in the cystic fibrosis transmembrane conductance regulator (CFTR gene); AND • Patient has been optimized on best supportive care for their CF prior to starting Trikafta; AND • Has a baseline percent predicted forced expiratory volume in 1 second (ppFEV1) that is less than or equal to 90%[1] before starting Trikafta; AND • Prescribed by a clinical specialist affiliated with a Canadian cystic fibrosis centre. [1]Case-by-case consideration may be provided for patients not meeting lung function criteria. The following measurements must be completed prior to initiating treatment with Trikafta: • Baseline spirometry measurements of FEV1 in liters and percent predicted (within the last 3 months); AND • Number of days treated with oral and/or intravenous (IV) antibiotics for pulmonary exacerbations in the previous 6 months OR number of pulmonary exacerbations requiring oral and/or IV antibiotics in the previous 6 months; AND • Number of CF-related hospitalizations in the previous 6 months; AND • Weight, height, and body mass index (BMI); AND				

	• CFQ -R Respiratory Domain score. **Exclusion criteria:** • Patient has undergone lung transplantation; OR • Patient is using Trikafta as combination therapy with another cystic fibrosis transmembrane conductance regulator (CFTR) modulator. Initial approval duration: 7 months **Initial renewal criteria:** Renewal of funding will be considered in patients demonstrating <u>at least ONE</u> of the following improvements after 6 months of treatment with Trikafta; 1. Improvement of percent predicted FEV1 by 5% or more above the baseline measurement; OR 2. A decrease in the total number of days for which the patient received treatment with oral and/or IV antibiotics for pulmonary exacerbations compared with the 6-month period prior to initiating treatment OR a decrease in the total number of pulmonary exacerbations requiring oral and/or IV antibiotics compared with the 6-month period prior to initiating treatment; OR 3. Decreased number of CF-related hospitalizations in the 6 months after initiation of Trikafta treatment compared with the 6-month period prior to initiating Trikafta; OR 4. No decline in BMI at 6 months compared with the baseline BMI assessment; OR 5. Improvement by 4 points or more in the CFQ-R Respiratory Domain scale compared to baseline scores. **Subsequent renewal criteria:** For patients who have met the initiation criteria and initial renewal criteria. • Ongoing renewal of funding will be provided for those who are continuing to benefit from therapy with Trikafta and who do not meet any of the exclusion criteria. • At the time of renewal application, please include the patient's most recent ppFEV1 and a clinical update to confirm the treatment benefits or response experienced by the patient. Approval Duration of renewals: 1 year Approved doses: 12 years of age and older: 2 tablets (each containing elexacaftor/ tezacaftor/ ivacaftor 100mg/ 50mg/ 75mg) taken in the morning & one tablet (ivacaftor 150mg) taken in the evening approximately 12 hours apart.
Program Eligibility	High Cost Drug Program, Catastrophic Drug Program

CRITERIA UPDATE

Effective November 1, 2021, the Special Authorization criteria for Kalydeco 150 mg Tablet will be updated to the following:

For the treatment of cystic fibrosis in patients meeting the following criteria;

- the patient is at least 6 years old and has one of the following mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene: G551D, G1244E, G1349D, G178R, G551S, S1251N, S1255P, S549N or S549R; OR
- the patient is at least 18 years old with an R117H mutation in the CFTR gene.

Renewal criteria[1]:

Renewal requests will be considered in patients with documented response to treatment as evidenced by the following:

(a) In cases where the patient's sweat chloride levels prior to commencing therapy were above 60mmol/L:
- the patient's sweat chloride level fell below 60mmol/L; or
- the patient's sweat chloride level is 30% lower than the level reported in a previous test;

(b) In cases where the patient's sweat chloride levels prior to commencing therapy were below 60mmol/L:
- the patient's sweat chloride level is 30% lower than the level reported in a previous test; or
- the patient demonstrates a sustained absolute improvement in FEV_1 of at least 5% when compared to the FEV_1 test conducted prior to the commencement of therapy. FEV_1 will be compared with the baseline pre-treatment level one month and three months after starting treatment.

Clinical Notes:

- The patient's sweat chloride level and FEV_1 must be provided with each request
- A sweat chloride test must be performed within a few months of starting ivacaftor therapy to determine if sweat chloride levels are reducing.
    - If the expected reduction occurs, a sweat chloride test must be performed again 6 months after starting therapy to determine if the full reduction has been achieved. Thereafter, sweat chloride levels must be checked annually.
    - If the expected reduction does not occur, a sweat chloride test should be performed again one week later. If the criteria are not met, coverage will be discontinued.

Claim Notes:

- Approved dose: 150mg every 12 hours
- Approval period: 1 year

[1]It should be noted that, while baseline sweat chloride levels and FEV1 are not required to meet initial approval criteria for Kalydeco, these parameters are used to evaluate the effect of Kalydeco at the time of renewal. To avoid delays, the prescriber should submit a copy of the mutation report, recent baseline sweat chloride levels before starting Kalydeco, and recent baseline FEV1 with the initial request for funding of Kalydeco. These baseline values will be used to evaluate the patient's response to therapy at the time of renewal and would be logistically difficult to obtain once treatment is initiated.

INFORMATION

Coverage for Orkambi (ivacaftor/lumacaftor) 100mg/125mg & 150mg/188 granule packets and 100mg/125mg & 200mg/125mg tablets may be available through the High Cost Drug Plan and Catastrophic Drug Plan for the treatment of cystic fibrosis patients who meet certain medical criteria. Please contact the PEI Pharmacare Program office at 1-877-577-3737 for more information regarding coverage availability and the Special Authorization application process for this product.