Louisiana Medicaid
Cystic Fibrosis, Oral

The *Louisiana Uniform Prescription Drug Prior Authorization Form* should be utilized to request clinical authorization for ivacaftor (Kalydeco®), lumacaftor/ivacaftor (Orkambi®) and tezacaftor/ivacaftor (Symdeko®).

Additional Point-of-Sale edits may apply.

Ivacaftor (Kalydeco®)

Approval Criteria

- The recipient is ~~4~~6 months old or older on the date of the request with a documented diagnosis of cystic fibrosis; **AND**
- The following is ***true*** and is ***noted on the request***:
    - The recipient has one of the following mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene and the ***mutation is listed on the request***; **AND**

List of CFTR Gene Mutations that Produce CFTR Protein and are Responsive to KALYDECO®					
E56K	G178R	S549R	S977F	F1074L	2789+5G→A
P67L	E193K	G551D	F1052V	D1152H	3272-26A→G
R74W	L206W	G551S	K1060T	G1244E	3849+10kbC→T
D110E	R347H	D579G	A1067T	S1251N	
D110H	R352Q	711+3A→G	G1069R	S1255P	
R117C	A455E	E831X	R1070Q	D1270N	
R117H	S549N	S945L	R1070W	G1349D	

    - The recipient is not homozygous for the F508del mutation in the CFTR gene; **AND**
- By submitting the authorization request, the prescriber attests to the following:
    - The prescribing information for the requested medication has been thoroughly reviewed, including any Black Box Warning, Risk Evaluation and Mitigation Strategy (REMS), contraindications, minimum age requirements, recommended dosing, and prior treatment requirements; **AND**
    - All laboratory testing and clinical monitoring recommended in the prescribing information have been completed as of the date of the request and will be repeated as recommended; **AND**
    - The recipient has no concomitant drug therapies or disease states that limit the use of Kalydeco® and will not be receiving Kalydeco® in combination with any medication that is contraindicated or not recommended per FDA labeling.

Reauthorization Criteria

- The recipient continues to meet all initial approval criteria; **AND**
- The prescriber ***states on the request*** that there is evidence of a positive response to treatment.

Duration of Initial and Reauthorization Approval: 12 months

Lumacaftor/Ivacaftor (Orkambi®)

Approval Criteria

- Recipient is 2 years of age or older on the date of the request with a documented diagnosis of cystic fibrosis; **AND**
- The following is ***true*** and is ***noted on the request***: The recipient is homozygous for the F508del mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene; **AND**
- By submitting the authorization request, the prescriber attests to the following:
    - The prescribing information for the requested medication has been thoroughly reviewed, including any Black Box Warning, Risk Evaluation and Mitigation Strategy (REMS), contraindications, minimum age requirements, recommended dosing, and prior treatment requirements; **AND**
    - All laboratory testing and clinical monitoring recommended in the prescribing information have been completed as of the date of the request and will be repeated as recommended; **AND**
    - The recipient has no concomitant drug therapies or disease states that limit the use of Orkambi® and will not be receiving Orkambi® in combination with any medication that is contraindicated or not recommended per FDA labeling.

Reauthorization Criteria

- The recipient continues to meet all initial approval criteria; **AND**
- The prescriber ***states on the request*** that there is evidence of a positive response to treatment.

Duration of Initial and Reauthorization Approval: 12 months

Tezacaftor/Ivacaftor (Symdeko®)

Approval Criteria

- Recipient is 6 years of age or older; **AND**
- Recipient has a diagnosis of cystic fibrosis (CF) and **ONE** of the following is ***documented on the request***:
    - The recipient is homozygous for the *F508del* mutation; **OR**
    - The recipient 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 or clinical evidence.

List of CFTR Gene Mutations Responsive to Tezacaftor/Ivacaftor							
A1067T	A455E	D110E	D110H	D1152H	D1270N	D579G	E193K
E56K	E831X	F1052V	F1074L	K1060T	L206W	P67L	R1070W
R117C	R347H	R352Q	R74W	S945L	S977F	F508del (2 copies)	
2789+5G→A		3272-26A→G		3849+10kbC→T		711+3A→G	

Reauthorization Criteria

- The recipient continues to meet all initial approval criteria; **AND**

- The prescriber ***states on the request*** that there is evidence of a positive response to treatment.

Duration of Initial and Reauthorization Approval: 12 months

References

Kalydeco (ivacaftor) [package insert]. Boston, MA: Vertex Pharmaceuticals Incorporated; ~~April 2019~~September 2020. https://pi.vrtx.com/files/uspi_ivacaftor.pdf

Orkambi (lumacaftor/ivacaftor) [package insert]. Boston, MA: Vertex Pharmaceuticals Incorporated; July 2019. https://pi.vrtx.com/files/uspi_lumacaftor_ivacaftor.pdf

Symdeko (tezacaftor/ivacaftor) [package insert]. Boston, MA: Vertex Pharmaceuticals Inc; December 2019. https://pi.vrtx.com/files/uspi_tezacaftor_ivacaftor.pdf

Revision	Date
Single PDL Implementation	May 2019
Modify age to 6 years or older – Symdeko®	November 2019
Removed Fee-for-Service, modified formatting, added revision table, removed footer, combined all cystic fibrosis agent criteria into one document	January 2020
Formatting changes, updated references	July 2020
Modified age to 4 months for Kalydeco®, updated reference, formatting changes	October 2020