Louisiana Medicaid
Cystic Fibrosis~~, Oral~~

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

Additional Point-of-Sale edits may apply.

*These agents may have **Black Box Warnings** and/or may be subject to **Risk Evaluation and Mitigation Strategy (REMS)** under FDA safety regulations. Please refer to individual prescribing information for details.*

NOTE: These agents (except for Bronchitol®) are mutation-specific targeted therapy that are indicated to treat only the cystic fibrosis transmembrane conductance regulator (CFTR) mutation(s) listed in the prescribing information. Identification of the indicated genotype is required in order to receive treatment with one of these agents. If the recipient's genotype is unknown, an FDA-cleared CF mutation test should be used to confirm the presence of a CFTR mutation, followed by verification with bi-directional sequencing when recommended by the mutation test instructions.

Elexacaftor/Tezacaftor/Ivacaftor with Ivacaftor (Trikafta®)

Approval Criteria

- The recipient is 6~~12~~ 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 **stated on the request**: The recipient has **AT LEAST ONE** *F508del* mutation in the CFTR gene, or a mutation in the CFTR gene that is responsive to elexacaftor/tezacaftor/ivacaftor with ivacaftor based on *in vitro* data; **AND**
- If the request is for a non-preferred agent - **ONE** of the following is required:
    - The recipient has had a *treatment failure* with at least one preferred drug that is appropriate to use for the condition being treated; **OR**
    - The recipient has had an *intolerable side effect* to at least one preferred drug that is appropriate to use for the condition being treated; **OR**
    - The recipient has *documented contraindication(s)* to the preferred drugs that are appropriate to use for the condition being treated; **OR**
    - There is *no preferred product that is appropriate* to use for the condition being treated; **OR**
    - The prescriber states that the recipient is currently using the requested medication and has had a positive clinical response to treatment; **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 inappropriate concomitant drug therapies or disease states that limit the use of Trikafta® and will not receive Trikafta® in combination with any medication that is contraindicated or not recommended per FDA labeling.

Reauthorization Criteria

- The recipient continues to meet 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

Ivacaftor (Kalydeco®)

Approval Criteria

- The recipient is 4 months old or older on the date of the request with a documented diagnosis of cystic fibrosis; **AND**
- The following is **true** and is **stated on the request**: The recipient has one mutation in the CFTR gene that is responsive to ivacaftor based on clinical and/or *in vitro* assay data; **AND**
- If the request is for a non-preferred agent - **ONE** of the following is required:
  - The recipient has had a *treatment failure* with at least one preferred drug that is appropriate to use for the condition being treated; **OR**
  - The recipient has had an *intolerable side effect* to at least one preferred drug that is appropriate to use for the condition being treated; **OR**
  - The recipient has *documented contraindication(s)* to the preferred drugs that are appropriate to use for the condition being treated; **OR**
  - There is *no preferred product that is appropriate* to use for the condition being treated; **OR**
  - The prescriber states that the recipient is currently using the requested medication and has had a positive clinical response to treatment; **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 ~~i~~Initial and ~~r~~Reauthorization ~~a~~Approval: 12 months

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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 **stated on the request**: The recipient is homozygous for the *F508del* mutation in the CFTR gene; **AND**
- If the request is for a non-preferred agent - **ONE** of the following is required:
    - The recipient has had a *treatment failure* with at least one preferred drug that is appropriate to use for the condition being treated; **OR**
    - The recipient has had an *intolerable side effect* to at least one preferred drug that is appropriate to use for the condition being treated; **OR**
    - The recipient has *documented contraindication(s)* to the preferred drugs that are appropriate to use for the condition being treated; **OR**
    - There is *no preferred product that is appropriate* to use for the condition being treated; **OR**
    - The prescriber states that the recipient is currently using the requested medication and has had a positive clinical response to treatment; **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 ~~i~~Initial and ~~r~~Reauthorization ~~a~~Approval: 12 months

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Mannitol (Bronchitol®)

Approval Criteria

- The recipient is 18 years of age or older on the date of the request with a documented diagnosis of cystic fibrosis; **AND**
- The medication is prescribed by, or the request states that this medication is being prescribed in consultation with, a pulmonologist; **AND**
- The recipient has a documented failure of, or intolerance to, or contraindication to treatment with hypertonic saline; **AND**
- If the request is for the 7-day or 4-week treatment pack, the prescriber **states on the request** that the recipient has successfully completed the Bronchitol Tolerance Test (BTT); **AND**
- The prescriber **states on the request** that Bronchitol® is prescribed concurrently with a short-acting bronchodilator; **AND**
- If the request is for a non-preferred agent - **ONE** of the following is required:
    - The recipient has had a *treatment failure* with at least one preferred drug that is appropriate to use for the condition being treated; **OR**
    - The recipient has had an *intolerable side effect* to at least one preferred drug that is appropriate to use for the condition being treated; **OR**
    - The recipient has *documented contraindication(s)* to the preferred drugs that are appropriate to use for the condition being treated; **OR**
    - There is *no preferred product that is appropriate* to use for the condition being treated; **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 the requested medication and will not receive the requested medication 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**
- If the request is for Bronchitol Tolerance Test (BTT), reauthorization is not permitted; **AND**
- The prescriber **states on the request** that Bronchitol® is prescribed concurrently with a short-acting bronchodilator; **AND**
- The prescriber **states on the request** that there is evidence of a positive response to treatment.

Duration of approval:
Bronchitol Tolerance Test (BTT): 1 week

Treatment Pack (7-day or 4-week): 6 months

Tezacaftor/Ivacaftor (Symdeko®)

Approval Criteria

- The recipient is 6 years of age or older on the date of the request; **AND**
- The recipient has a diagnosis of cystic fibrosis (CF) and **ONE** of the following is **stated on the request**:
  - The recipient is homozygous for the *F508del* mutation; **OR**
  - The recipient has at least one mutation in the CFTR gene that is responsive to tezacaftor/ivacaftor based on *in vitro* data and/or clinical evidence; **AND**
- If the request is for a non-preferred agent - **ONE** of the following is required:
  - The recipient has had a *treatment failure* with at least one preferred drug that is appropriate to use for the condition being treated; **OR**
  - The recipient has had an *intolerable side effect* to at least one preferred drug that is appropriate to use for the condition being treated; **OR**
  - The recipient has *documented contraindication(s)* to the preferred drugs that are appropriate to use for the condition being treated; **OR**
  - There is *no preferred product that is appropriate* to use for the condition being treated; **OR**
  - The prescriber states that the recipient is currently using the requested medication and has had a positive clinical response to treatment; **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 Symdeko® and will not be receiving Symdeko® 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 ~~i~~Initial and ~~r~~Reauthorization ~~a~~Approval: 12 months

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References

Bronchitol (mannitol) [package insert]. Cary, NC: Chiesi USA, Inc; October 2020. https://www.accessdata.fda.gov/drugsatfda_docs/label/2020/202049s000lbl.pdf

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

Mogayzel PJ Jr, 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.

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 2020. https://pi.vrtx.com/files/uspi_tezacaftor_ivacaftor.pdf

Trikafta (elexacaftor/tezacaftor/ivacaftor) [package insert]. Boston, MA: Vertex Pharmaceuticals Incorporated; June 2021~~December 2020~~.
https://pi.vrtx.com/files/uspi_elexacaftor_tezacaftor_ivacaftor.pdf

Revision / Date	**~~Date~~Implementation Date**
Single PDL Implementation	May 2019
Modify age to 6 years or older – Symdeko® / November 2019	November 2019
Removed Fee-for-Service, modified formatting, added revision table, removed footer, combined all cystic fibrosis agent criteria into one document / January 2020	January 2020
Formatting changes, updated references / July 2020	July 2020
Modified age to 4 months for Kalydeco®, updated reference, formatting changes / October 2020	April 2021~~October 2020~~
Updated indications for Kalydeco® and Symdeko®, formatting changes, updated references / February 2021	~~February 2021~~July 2021
Added Trikafta® criteria to this document, updated references / April 2021	~~April 2021~~July 2021
Added Bronchitol® criteria, updated references, formatting changes / May 2021	July 2021
Modified age to 6 years or older – Trikafta® / June 2021	July 2021