STATE OF WEST VIRGINIA
DEPARTMENT OF HEALTH AND HUMAN RESOURC
BUREAU FOR MEDICAL SERVICES

Office of Pharmacy Service
Prior Authorization Criteria

EMFLAZA™ (deflazacort)
Effective 1/01/2018

Prior Authorization Request Form

EMFLAZA is a corticosteroid indicated for the treatment of Duchenne muscular dystrophy (DMD) in patients 5 years of age and older.

Prior authorization requests for may be approved if the following criteria have been satisified:

1. Diagnosis of Duchenne muscular dystrophy (DMD); **AND**

2. Patient ≥ 5 years old; **AND**

3. Patient must have a documented history of at least 12-months continuous therapy with prednisone; **AND**

4. Documentation must be submitted indicating that the patient has experienced significant adverse effects associated with prednisone therapy. Documentation must include a detailed description of the adverse effect; as the side effect profiles are similar between deflazacort and prednisone, prior authorization shall only be granted for those patients experiencing side effects where deflazacort shows an improved profile.

5. Request must be accompanied with a baseline 6-minute walk distance (6MWD); **AND**

6. Initial authorizations shall be for 90 days. Continuation requests may be granted a 12-month approval if significant improvement is demonstrated in either the patient's adverse effect profile or 6MWD.

References

1.) Griggs RC, Miller JP, Greenberg CR, et al. Efficacy and safety of deflazacort vs prednisone and placebo for Duchenne muscular dystrophy. *Neurology*. 2016;87(20):2123-2131
2.) Lexi-Comp drug monograph for deflazacort (Reviewed 8/22/2017)
3.) Efficacy and safety of deflazacort vs prednisone and placebo for Duchenne muscular dystrophy. Neurology. 2016 Nov 15; 87(20): 2123–2131.
4.) UpToDate article: Treatment of Duchenne and Becker muscular dystrophy. Updated July 18, 2017.

v2018.2a – BMT updated 05/10/2018
DUR Board Approval: 11/15/2018

STATE OF WEST VIRGINIA
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Office of Pharmacy Service
Prior Authorization Criteria

EXONDYS 51® (eteplirsen)
Effective 06/01/2018

Prior Authorization Request Form

EXONDYS 51 is an antisense oligonucleotide indicated for the treatment of Duchenne muscular dystrophy (DMD) in patients who have a confirmed mutation of the DMD gene that is amenable to exon 51 skipping. This indication is approved under accelerated approval based on an increase in dystrophin in skeletal muscle observed in some patients treated with EXONDYS 51. **A clinical benefit of EXONDYS 51 has not been established.** Continued approval for this indication may be contingent upon verification of a clinical benefit in confirmatory trials.

Prior authorization requests for Exondys 51 must be submitted as a medical claim and require review by the Medical Director and are will only be considered if the following criteria are met:

7. Patient must have a confirmed mutation of a DMD gene that is amenable to exon 51 skipping (chart notes required); **AND**

8. Patient must be currently taking a corticosteroid OR have a contraindication to corticosteroids; **AND**

9. Prior authorization requests must be accompanied with peer-reviewed literature **confirming** clinical benefit of this medication in patients diagnosed with DMD. Preliminary trial data may be submitted for consideration but does not assure approval.; **AND**

10. Appropriate and validated baseline function test results must be submitted with the initial request for therapy. These tests may include any of the following:

    a. Ambulatory patients: Six-minute walk test (6MWDT) of > 180 meters.
    b. Non-ambulatory patients: Brooke Upper Extremity Function Scale (of 5 or less) AND a Forced Vital Capacity of ≥ 30% of predicted value.

Prior authorization approvals will be for 6 months. Continuation requests must provide clinical documentation of efficacy as evidenced by improvement or stabilization of functions compared to baseline measures.

References

5.) Exondys 51 Package Insert (Sarepta Therapeutics) – Revised 2/2018
6.) Lexicomp monograph for Exondys 51 – reviewed 5/10/2018
7.) Birnrant et al. Lancet Neurol. 2018 March; 17(3): 251-267. Diagnosis and management of Duchenne muscular dystrophy, part 1: diagnosis, and neuromuscular, rehabilitation, endocrine, and gastrointestinal and nutritional management
8.) Mendell, JR et al. Ann Neurol 2016;79:257–271. Longitudinal Effect of Eteplirsen versus Historical Control on Ambulation in Duchenne Muscular Dystrophy
9.) Kinane, TB et al. Journal of Neuromuscular Diseases 5 (2018) 47–58 Long-Term Pulmonary Functionin Duchenne Muscular Dystrophy: Comparison of Eteplirsen-Treated Patients to Natural History
10.) Clinical Trials:
a. https://clinicaltrials.gov/ct2/show/NCT01396239?term=eteplirsen&rank=6
b. https://clinicaltrials.gov/ct2/show/NCT01540409

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Office of Pharmacy Service
Prior Authorization Criteria

KALYDECO®
(ivacaftor)
Prior Authorization Request Form
Effective 6/01/2018

KALYDECO is a cystic fibrosis transmembrane conductance regulator (CFTR) potentiator indicated for the treatment of cystic fibrosis (CF) in patients age 2 years and older who have one mutation in the CFTR gene that is responsive to ivacaftor based on clinical and/or in vitro assay data.

Criteria for Approval

1) Individual must be two (2) years or older; **AND**
2) Patient must have a confirmed diagnosis of Cystic Fibrosis; **AND**
3) Patient must be determined have at least one mutation in the CFTR gene which is responsive to ivacaftor as confirmed by an FDA-approved CF mutation test; **AND**
4) Patient must have a documented baseline AST, ALT and FEV_1 (forced expiratory volume in one second) presented with the prior authorization request; **AND**
5) Patient must NOT be homozygous for the F508del mutation in the CFTR gene; **AND**
6) Dosage does not exceed 150 mg twice daily for ages 6 and up; **OR**
7) For patients ages 2 to less than 6 years, dosage should be weight-based and may not exceed 75 mg twice daily.
8) Patients under the age of 18 must have undergone a baseline ophthalmic examination to monitor for lens opacities/cataracts.

Prior authorizations will be for every 6 months in the first year, followed thereafter by an annual prior authorization.

Criteria for Continuation of Therapy

1) Patients under the age of 18 must have follow up ophthalmic examinations at least annually (documentation required); **AND**
2) Patient must have LFTs/bilirubin monitored every 6 months for the first year of treatment and annually thereafter (documentation required); **AND**
3) Serum ALT or AST < 5 times the upper limit of normal (ULN); **OR**
4) Serum ALT or AST < 3 times the ULN with bilirubin < 2 times the ULN.

References

1) Kalydeco package insert revised 7/2017
2) Lexi-Comp Clinical Application 05/09/2018

v2018.2a – BMT updated 05/10/2018
DUR Board Approval: 11/15/2018

STATE OF WEST VIRGINIA
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Office of Pharmacy Service
Prior Authorization Criteria

ORKAMBI®
(lumacaftor/ivacaftor)
Prior Authorization Request Form
Effective 6/01/2018

Orkambi is a combination drug containing lumacaftor and ivacaftor that is indicated for the treatment of cystic fibrosis in patients age 6 years and older who are **homozygous** for the ***F508del*** mutation in the CFTR gene.

Criteria for Approval

9) Individual is 6 years or older; **AND**
10) Patient must have a confirmed diagnosis of Cystic Fibrosis; **AND**
11) Patient must be determined to be **homozygous** for the ***F508del*** mutation in the CFTR gene as confirmed by an FDA-approved CF mutation test; **AND**
12) Patient must have a documented baseline AST, ALT and FEV_1 (forced expiratory volume in one second) presented with the prior authorization request; **AND**
13) Patients under the age of 18 must have undergone a baseline ophthalmic examination to monitor for lens opacities/cataracts.

Prior authorizations will be for every 6 months in the first year, followed thereafter by an annual prior authorization.

Criteria for Continuation of Therapy

5) Patients under the age of 18 must have follow up ophthalmic examinations at least annually (documentation required); **AND**
6) Patient must have LFTs/bilirubin monitored every 6 months for the first year of treatment and annually thereafter (documentation required); **AND**
7) Serum ALT or AST < 5 times the upper limit of normal (ULN); **OR**
8) Serum ALT or AST < 3 times the ULN with bilirubin < 2 times the ULN.

References

3) Orkambi package insert revised 9/2016
4) Lexi-Comp Clinical Application 09/30/2016

STATE OF WEST VIRGINIA
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Office of Pharmacy Service
Prior Authorization Criteria

SUBLOCADE® (buprenorphine extended-release injection)
Effective 6/01/2018

Prior Authorization Request Form

SUBLOCADE contains buprenorphine, a partial opioid agonist, and is indicated for the treatment of moderate to severe opioid use disorder in patients who have initiated treatment with a transmucosal buprenorphine -containing product, followed by dose adjustment for a minimum of 7 days.

Prior authorization requests for may be approved if the following criteria are met:

11. Patient must be at least 18 years of age; **AND**

12. Must be prescribed and administered by an addiction specialist solely for the treatment of opioid addiction; **AND**

13. Patient must be stable on buprenorphine therapy (for opioid use disorder) for at least 28 days immediately prior to the request to start Sublocade; **AND**

14. Prior authorization will only be granted for doses that follow the manufacturer's guidelines:

    SubQ: Initial: 300 mg monthly for the first 2 months, after treatment has been inducted and adjusted with 8 to 24 mg of a transmucosal buprenorphine-containing product for a minimum of 7 days. Maintenance: 100 mg monthly, increasing to 300 mg monthly for patients who tolerate the 100 mg dose but do not demonstrate a satisfactory clinical response (as evidenced by self-reported illicit opioid use or urine drug screens positive for illicit opioid use). **Note**: Administer doses at least 26 days apart.

Initial approval of Sublocade will be for 3 months. Additional therapy shall be approved up to 12 months at a time with documentation of satisfactory patient response.

References

11.) Sublocade package insert (Indivior Inc.) Updated 3/2018
12.) Lexicomp monograph for Sublocade (reviewed 5/10/2018)

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Prior Authorization Criteria

SYMDEKO®
(tezacaftor/ivacaftor + ivacaftor)
Prior Authorization Request Form
Effective 6/01/2018

SYMDEKO is a combination of tezacaftor and ivacaftor, indicated for the treatment of patients with cystic fibrosis (CF) aged 12 years and older who are homozygous for the F508del mutation or who have at least one mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene that is responsive to tezacaftor/ivacaftor based on in vitro data and/or clinical evidence.

Criteria for Approval

14) Individual is 12 years or older; **AND**
15) Patient must have a confirmed diagnosis of Cystic Fibrosis; **AND**
16) Patient must be determined to be **homozygous** for the ***F508del*** mutation in the CFTR gene **or** at least one other mutation in the CFTR gene which is responsive to tezacaftor/ivacaftor as confirmed by an FDA-approved CF mutation test; **AND**
17) Patient must have a documented baseline AST, ALT and FEV_1 (forced expiratory volume in one second) presented with the prior authorization request; **AND**
18) Patients under the age of 18 years must have undergone a baseline ophthalmic examination to monitor for lens opacities/cataracts.

Prior authorizations will be for every 6 months in the first year, followed thereafter by an annual prior authorization.

Criteria for Continuation of Therapy

9) Patients under the age of 18 years must have follow up ophthalmic examinations at least annually (documentation required); **AND**
10) Patient must have LFTs/bilirubin monitored every 6 months for the first year of treatment and annually thereafter (documentation required); **AND**
11) Serum ALT or AST < 5 times the upper limit of normal (ULN); **OR**
12) Serum ALT or AST < 3 times the ULN with bilirubin < 2 times the ULN.

References

5) Symdeko package insert revised 2/2018
6) Lexi-Comp Clinical Application 05/09/2018