Clinical Policy: Omaveloxolone (Skyclarys)

Reference Number: PA.CP.PHAR.590
Effective Date: 06/2023
Last Review Date: 05/2023

Description
Omaveloxolone (Skyclarys™) is a nuclear factor erythroid 2–related factor 2 (Nrf2) pathway activator.

FDA Approved Indication(s)
Skyclarys is indicated for the treatment of Friedreich's ataxia (FA) in adults and adolescents aged 16 years and older.

Policy/Criteria
Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.

It is the policy of PA Health & Wellness® that Skyclarys is **medically necessary** when the following criteria are met:

I. Initial Approval Criteria

A. Friedreich's Ataxia (must meet all)**:**

1. Diagnosis of FA;
2. Documentation of genetic testing confirming FXN gene mutation;
3. Prescribed by or in consultation with a neurologist;
4. Age ≥ 16 years;
   (see Appendix D);
5. Member has ability to swallow capsules;
6. Dose does not exceed both of the following (a and b):
   a. 150 mg per day;
   b. 3 capsules per day.

   Approval duration: 6 months

B. Other diagnoses/indications

1. Refer to the off-label use policy if diagnosis is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized): PA.CP.PMN.53

II. Continued Therapy

A. Friedreich's Ataxia (must meet all)**:**

1. Currently receiving medication via PA Health & Wellness benefit and documentation supports positive response to therapy or the Continuity of Care policy (PA.LTSS.PHAR.01) applies;
2. Member is responding positively to therapy as evidenced by, including but not limited to, improvement in any of the following parameters: FA symptoms (see Appendix D), maximal exercise testing, or mFARS score;
3. Dose does not exceed both of the following (a and b):

a. 150 mg per day;
b. 3 capsules per day.

Approval duration: 6 months

B. Other diagnoses/indications (must meet 1 or 2)**:**

1. Currently receiving medication via PA Health & Wellness benefit and documentation supports positive response to therapy or the Continuity of Care policy (PA.LTSS.PHAR.01) applies.
   Approval duration: Duration of request or 12 months (whichever is less); or
2. Refer to the off-label use policy for the relevant line of business if diagnosis is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized): PA.CP.PMN.53

III. Diagnoses/Indications for which coverage is NOT authorized:

A. Non-FDA approved indications, which are not addressed in this policy, unless there is sufficient documentation of efficacy and safety according to the off label use policies – PA.CP.PMN.53

IV. Appendices/General Information

Appendix A: Abbreviation/Acronym Key
FA: Friedreich's ataxia
FDA: Food and Drug Administration
mFARS: modified functional assessment rating scale
Nrf2: nuclear factor erythroid 2–related factor 2

Appendix B: Therapeutic Alternatives
Not applicable

Appendix C: Contraindications/Boxed Warnings
None reported

Appendix D: General Information

- FA is a progressive, life-shortening ataxia which has cardinal symptoms of progressive gait and limb ataxia, lower limb areflexia, extensor plantar responses and dysarthria. In addition to ataxia, FA may cause fatigue, cardiomyopathy, and metabolic disturbances.
- Examples of clinically significant cardiac disease include the following: clinically significant congenital or acquired valvular disease, pericardial constriction, restrictive or congestive cardiomyopathy, symptomatic coronary disease, history of hospitalization for heart failure in last five years, cardiac insufficiency defined as New York Heart Association Class > 2, history of atrial fibrillation, history of unstable arrhythmias.
- The mFARS is a disease specific, exam-based neurological rating scale which includes assessment of bulbar function, upper limb coordination, lower limb coordination, and upright stability. The mFARS has a maximum cumulative value of

93 points, where higher cumulative scores signify greater degree of disability. The rating scale is provided below:

Neurologic assessment type (maximum points)	Description (points)
Bulbar (5)	Cough (2) Speech (3)
Upper limb coordination (36)	Finger-finger (3+3) Nose-finger (4+4) Dysmetria (4+4) Rapid movement (3+3) Finger taps (4+4)
Lower limb coordination (16)	Heel-shin slide (4+4) Heel-shin tap (4+4)
Upright stability (36)	Sitting position (4) Stance feet apart (4) Stance feet apart with eyes closed (4) Stance feet together (4) Stance feet together with eyes closed (4) Tandem stance (4) Stance dominant foot (4) Tandem walk (3) Gait (5)

- Maximal exercising testing is defined as being able to ride an exercise ergometer at approximately 60 rpm against no added resistance for 3 minutes.
- Pes cavus is an orthopedic condition that is defined by the elevation of the longitudinal arch of the foot. In part 1 of the MOXIe study, patients with pes cavus did not show a statistically significant improvement with Skyclarys treatment, while patients with pes cavus were excluded from the full analysis set of the part 2 of the study.

V. Dosage and Administration

Indication	Dosing Regimen	Maximum Dose
FA	150 mg PO QD	150 mg/day

VI. Product Availability

Capsule: 50 mg

VII. References

1. Skyclarys Prescribing Information. Plano, TX: Reata Pharmaceuticals, Inc.; February 2023. Available at: https://www.accessdata.fda.gov/drugsatfda_docs/label/2023/216718Orig1s000lbl.pdf?utm_medium=email&utm_source=govdelivery. Accessed February 28, 2023.
2. Lynch DR, Chin MP, Delatycki MB, et al. Safety and Efficacy of Omaveloxolone in Friedreich Ataxia (MOXIe Study). Ann Neurol. 2021;89(2):212-225. https://doi.org/10.1002/ana.25934

3. Lynch DR, Farmer J, Hauser L, et al. Safety, pharmacodynamics, and potential benefit of omaveloxolone in Friedreich ataxia. Ann Clin Transl Neurol. 2018;6(1):15-26. Published 2018 Nov 10. https://doi.org/10.1002/acn3.660
4. Corben LA, Lynch D, Pandolfo M, et al. Consensus clinical management guidelines for Friedreich ataxia. Orphanet J Rare Dis 9, 184 (2014). https://doi.org/10.1186/s13023-014-0184-7
5. Rummey C, Corben LA, Delatycki MB, et al. Psychometric properties of the Friedreich Ataxia Rating Scale. Neurol Genet 2019;5e371. https://doi.org/10.1212/NXG.0000000000000371

Reviews, Revisions, and Approvals	Date	P&T Approval Date
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