PRIOR AUTHORIZATION POLICY

POLICY: Pulmonary Arterial Hypertension – Winrevair Prior Authorization Policy
- Winrevair™ (sotaterept-csrk subcutaneous injection – Merck)

REVIEW DATE: 04/10/2024

INSTRUCTIONS FOR USE

THE FOLLOWING COVERAGE POLICY APPLIES TO HEALTH BENEFIT PLANS ADMINISTERED BY CIGNA COMPANIES. CERTAIN CIGNA COMPANIES AND/OR LINES OF BUSINESS ONLY PROVIDE UTILIZATION REVIEW SERVICES TO CLIENTS AND DO NOT MAKE COVERAGE DETERMINATIONS. REFERENCES TO STANDARD BENEFIT PLAN LANGUAGE AND COVERAGE DETERMINATIONS DO NOT APPLY TO THOSE CLIENTS. COVERAGE POLICIES ARE INTENDED TO PROVIDE GUIDANCE IN INTERPRETING CERTAIN STANDARD BENEFIT PLANS ADMINISTERED BY CIGNA COMPANIES. PLEASE NOTE, THE TERMS OF A CUSTOMER'S PARTICULAR BENEFIT PLAN DOCUMENT [GROUP SERVICE AGREEMENT, EVIDENCE OF COVERAGE, CERTIFICATE OF COVERAGE, SUMMARY PLAN DESCRIPTION (SPD) OR SIMILAR PLAN DOCUMENT] MAY DIFFER SIGNIFICANTLY FROM THE STANDARD BENEFIT PLANS UPON WHICH THESE COVERAGE POLICIES ARE BASED. FOR EXAMPLE, A CUSTOMER'S BENEFIT PLAN DOCUMENT MAY CONTAIN A SPECIFIC EXCLUSION RELATED TO A TOPIC ADDRESSED IN A COVERAGE POLICY. IN THE EVENT OF A CONFLICT, A CUSTOMER'S BENEFIT PLAN DOCUMENT ALWAYS SUPERSEDES THE INFORMATION IN THE COVERAGE POLICIES. IN THE ABSENCE OF A CONTROLLING FEDERAL OR STATE COVERAGE MANDATE, BENEFITS ARE ULTIMATELY DETERMINED BY THE TERMS OF THE APPLICABLE BENEFIT PLAN DOCUMENT. COVERAGE DETERMINATIONS IN EACH SPECIFIC INSTANCE REQUIRE CONSIDERATION OF 1) THE TERMS OF THE APPLICABLE BENEFIT PLAN DOCUMENT IN EFFECT ON THE DATE OF SERVICE; 2) ANY APPLICABLE LAWS/REGULATIONS; 3) ANY RELEVANT COLLATERAL SOURCE MATERIALS INCLUDING COVERAGE POLICIES AND; 4) THE SPECIFIC FACTS OF THE PARTICULAR SITUATION. EACH COVERAGE REQUEST SHOULD BE REVIEWED ON ITS OWN MERITS. MEDICAL DIRECTORS ARE EXPECTED TO EXERCISE CLINICAL JUDGMENT AND HAVE DISCRETION IN MAKING INDIVIDUAL COVERAGE DETERMINATIONS. COVERAGE POLICIES RELATE EXCLUSIVELY TO THE ADMINISTRATION OF HEALTH BENEFIT PLANS. COVERAGE POLICIES ARE NOT RECOMMENDATIONS FOR TREATMENT AND SHOULD NEVER BE USED AS TREATMENT GUIDELINES. IN CERTAIN MARKETS, DELEGATED VENDOR GUIDELINES MAY BE USED TO SUPPORT MEDICAL NECESSITY AND OTHER COVERAGE DETERMINATIONS.

CIGNA NATIONAL FORMULARY COVERAGE:

OVERVIEW

Winrevair, an activin signaling inhibitor, is indicated for the treatment of **pulmonary arterial hypertension (PAH)** [World Health Organization {WHO} Group 1] in adults to increase exercise capacity, improve WHO functional class (FC), and reduce the risk of clinical worsening events.

Disease Overview

PAH is a serious but rare condition with an estimated prevalence of 10.6 cases per 1 million adults in the US.[2] It is classified within WHO Group 1 pulmonary hypertension among the five different groups that are recognized. In this progressive disorder, the small arteries in the lungs become narrowed, restricted, or blocked. This causes the heart to work harder to pump blood, leading to activity impairment. Although the mean age of diagnosis is between 36 and 50 years, patients of any age may be affected. PAH is defined as a mean pulmonary artery pressure (mPAP) > 20 mmHg (at rest) with a pulmonary arterial wedge pressure (PAWP) ≤ 15 mmHg and a pulmonary vascular resistance > 2 Wood units measured by cardiac catheterization.[3,4] Despite the introduction of many PAH-specific therapies, mortality associated with PAH remains high. When stratified into intermediate and high risk PAH, the 3-year mortality rate was reported as 18% to 20% for intermediate risk and 28% to 55% for high risk PAH.[5]

Guidelines

Various guidelines for PAH are available; however, none address Winrevair yet.

- In 2022, the European Society of Cardiology and the European Respiratory Society (ESC/ERS) updated guidelines on the diagnosis and management of PAH in adults.[3] The treatment algorithm for PAH has been simplified, with a clear focus on individual risk assessment, cardiopulmonary comorbidities, and treatment goals. Generally, combination therapy with a phosphodiesterase type 5 inhibitor (PDE5i) and endothelin receptor antagonist (ERA) are recommended as initial therapy in patients considered low risk. If low-risk status is not achieved within 3 to 6 months, the addition of a prostacyclin analogue is recommended. Combination therapy including an intravenous (IV) prostacyclin analogue is recommended as initial therapy in patients considered high-risk.
- The CHEST guidelines (2019) use the patient's functional class (FC) to determine disease severity instead of the comprehensive risk assessment.[6] The guidelines recommend initiating combination oral therapy with an ERA and a PDE5i for patients with mild or moderate disease (FC II or III) or IV prostacyclin therapy for patients with severe disease (FC III with rapid disease progression or FC IV). Patients who remain symptomatic despite initial treatment should be treated with a second or third agent. Those who fail to improve despite maximal medical therapy should be considered for lung transplant evaluation.

POLICY STATEMENT

Prior Authorization is recommended for prescription benefit coverage of Winrevair. All approvals are provided for the duration noted below. Because of the specialized skills required for evaluation and diagnosis of patients treated with these products as well as the monitoring required for adverse events and long-term efficacy, approval requires these agents to be prescribed by or in consultation with a physician who specializes in the condition being treated.

Documentation: Documentation is required for initiation of therapy as noted in the criteria as **[documentation required]**. Documentation may include, but is not limited to, chart notes and catheterization laboratory reports. For a patient case in which the documentation requirement of the right heart catheterization upon Prior Authorization coverage review for a different medication indicated for WHO Group 1 PAH has been previously provided, the documentation requirement in this *Pulmonary Arterial Hypertension – Winrevair Prior Authorization Policy* is considered to be met.

- **Winrevair™ (sotaterept-csrk subcutaneous injection – Merck)**

is(are) covered as medically necessary when the following criteria is(are) met for FDA-approved indication(s) or other uses with supportive evidence (if applicable):

FDA-Approved Indication

1. **Pulmonary Arterial Hypertension (PAH) [World Health Organization {WHO} Group 1].** Approve for the duration noted if the patient meets ONE of the following (A or B):
   - **A)** Initial Therapy. Approve for 6 months if the patient meets ALL of the following (i, ii, iii, iv and v):
     - **i.** Patients is ≥ 18 years of age; AND
     - **ii.** Patient meets the following (a and b):
       - **a)** The patient has had a right heart catheterization **[documentation required]** (see documentation section above); AND
       - **b)** The results of the right heart catheterization confirmed the diagnosis of WHO Group 1 PAH; AND
     - **iii.** Patient is in Functional Class II or III; AND
     - **iv.** Patient meets ONE of the following (a or b):
       - **a)** Patient is currently receiving at least two other PAH therapies from the following different pharmacologic categories each for ≥ 60 days: phosphodiesterase type 5 inhibitors (PDE5i), endothelin receptor antagonists (ERAs), soluble guanylate cyclase stimulator (sGCs), and prostacyclins; OR
       - **b)** Patient is currently receiving at least one other PAH therapy for ≥ 60 days and is intolerant to combination therapy with a phosphodiesterase type 5 inhibitors (PDE5i), endothelin receptor antagonists (ERAs), soluble guanylate cyclase stimulator (sGCs), or prostacyclin; AND
       Note: Examples of PDE5i include sildenafil and tadalafil. Examples of ERAs include bosentan, ambrisentan, and Opsumit (macitentan tablets). Example of sGCs includes Adempas (riociguat tablets). Examples of prostacyclins include Tyvaso (treprostinil inhalation solution), Tyvaso DPI (treprostinil inhalation solution), Ventavis (iloprost inhalation solution), Orenitram (treprostinil tablets), Uptravi (selexipag tablets), treprostinil injection and epoprostenol injection.
     - **v.** The medication is prescribed by or in consultation with a cardiologist or a pulmonologist.
   - **B)** Patient is Currently Receiving Winrevair. Approve for 1 year if the patient meets BOTH of the following (i and ii):
     - **i.** Patient meets BOTH of the following (a and b):
       - **a)** Patient has had a right heart catheterization; AND
       Note: This refers to prior to starting therapy with a medication for WHO Group 1 PAH.
       - **b)** Results of the right heart catheterization confirmed the diagnosis of WHO Group 1 PAH; AND
     - **ii.** The medication is prescribed by or in consultation with a cardiologist or a pulmonologist.

CONDITIONS NOT COVERED

- **Winrevair™ (sotaterept-csrk subcutaneous injection – Merck)**

is(are) considered experimental, investigational or unproven for ANY other use (criteria will be updated as new published data are available).

REFERENCES

1. Winrevair® subcutaneous injection [prescribing information]. Rahway, NJ: Merck; March 2024.
2. Ruopp NF, Cockrill BA. Diagnosis and treatment of pulmonary arterial hypertension. A review. *JAMA*. 2022;327(14):1379-1391.
3. Humbert M, Kovacs G, Hoeper MM, et al, for the ESC/ERS Scientific Document Group. 2022 ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension. *Eur Heart J*. 2022;43(38):3618-3731.
4. Maron B. Revised definition of pulmonary hypertension and approach to management: a clinical primer. *J Am Heart Assoc*. 2023 April 7. [epub ahead of print].
5. Chang KY, Duval S, Badesch DB, et al. PHAR Investigators Mortality in Pulmonary Arterial Hypertension in the modern era: early insights from the Pulmonary Hypertension Association Registry. *J Am Heart Assoc*. 2022 May 3;11(9):e024969. doi: 10.1161/JAHA.121.024969.
6. Klinger JR, Elliott CG, Levine DJ, et al. Therapy for pulmonary arterial hypertension in adults. Update of the CHEST guideline and Expert Panel Report. *CHEST*. 2019;155(3):565-586.

HISTORY

Type of Revision	Summary of Changes	Review Date
New Policy	-	04/10/2024