PRIOR AUTHORIZATION POLICY

POLICY: Sohonos Prior Authorization Policy
- Sohonos® (palovarotene capsules – Ipsen)

REVIEW DATE: 10/08/2025

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 WHERE APPROPRIATE AND HAVE DISCRETION IN MAKING INDIVIDUAL COVERAGE DETERMINATIONS. WHERE COVERAGE FOR CARE OR SERVICES DOES NOT DEPEND ON SPECIFIC CIRCUMSTANCES, REIMBURSEMENT WILL ONLY BE PROVIDED IF A REQUESTED SERVICE(S) IS SUBMITTED IN ACCORDANCE WITH THE RELEVANT CRITERIA OUTLINED IN THE APPLICABLE COVERAGE POLICY, INCLUDING COVERED DIAGNOSIS AND/OR PROCEDURE CODE(S). REIMBURSEMENT IS NOT ALLOWED FOR SERVICES WHEN BILLED FOR CONDITIONS OR DIAGNOSES THAT ARE NOT COVERED UNDER THIS COVERAGE POLICY (SEE "CODING INFORMATION" BELOW). WHEN BILLING, PROVIDERS MUST USE THE MOST APPROPRIATE CODES AS OF THE EFFECTIVE DATE OF THE SUBMISSION. CLAIMS SUBMITTED FOR SERVICES THAT ARE NOT ACCOMPANIED BY COVERED CODE(S) UNDER THE APPLICABLE COVERAGE POLICY WILL BE DENIED AS NOT COVERED. 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

Sohonos, a retinoid, is indicated for the reduction in volume of new heterotopic ossification in females ≥ 8 years of age and males ≥ 10 years of age with **fibrodysplasia ossificans progressiva**.[1]

Disease Overview

Fibrodysplasia ossificans progressiva is an ultra-rare, autosomal dominant genetic disorder of connective tissue characterized by progressive heterotopic ossification resulting in disability, immobility, and reduced quality/length of life.[2] Patients experience episodes of painful inflammatory swelling in soft tissues (flare-ups), some of which will spontaneously resolve, but most will transform soft connective tissues into mature heterotopic bone. Eventually, plates, sheets, and ribbons of heterotopic bone permanently replace muscles and connective tissue, encasing the patient almost like an armor, resulting in progressive and permanent immobility. There are no formal diagnostic criteria for fibrodysplasia ossificans progressiva.[2,3] A clinical diagnosis can

be made in patients with great toe malformations, tissue swelling, and heterotopic ossification, but genetic confirmation of an Activin A Type 1 Receptor (ACVR1) gene mutation is needed. All patients with fibrodysplasia ossificans progressiva have a mutation in ACVR1, a gene encoding a bone morphogenetic protein type I receptor kinase.[2,4] Approximately 97% of these patients have the same, heterozygous, single-nucleotide change in the glycine-serine activation domain of the ACVR1 (ACVR1^{R206H}).

Clinical Efficacy

In the pivotal study of Sohonos, patients were required to have fibrodysplasia ossificans progressiva as confirmed by a pathogenic variant in ACVR1^{R206H}.[1,5]

Guidelines

Medical management guidelines from the International Clinical Council on Fibrodysplasia Ossificans Progressiva (2024) recommend that each patient with the disease should have a primary provider who is able to consult with a fibrodysplasia ossificans progressiva expert and help coordinate a local care team.[2] The diagnosis of fibrodysplasia ossificans progressiva is based on clinical findings, but requires genetic confirmation (i.e., ACVR1 gene pathogenic variant), which can be detected by DNA sequence analysis.

POLICY STATEMENT

Prior Authorization is recommended for prescription benefit coverage of Sohonos. All approvals are provided for the duration noted below. In the clinical criteria, as appropriate, an asterisk (*) is noted next to the specified gender. In this context, the specified gender is defined as follows: males are defined as individuals with the biological traits of a male, regardless of the individual's gender identity or gender expression; females are defined as individuals with the biological traits of a female, regardless of the individual's gender identity or gender expression. Because of the specialized skills required for evaluation and diagnosis of patients treated with Sohonos as well as the monitoring required for adverse events and long-term efficacy, approval requires Sohonos to be prescribed by or in consultation with a physician who specializes in the condition being treated.

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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 Indications

1. **Fibrodysplasia ossificans progressiva.** Approve for 1 year if the patient meets ALL of the following (A, B, C, and D):
   A) Patient meets ONE of the following (i or ii):
      i. Patient is female* and ≥ 8 years of age; OR
      ii. Patient is male* and ≥ 10 years of age; AND

B) Patient has had a genetic test confirming a pathogenic variant in Activin A Type 1 Receptor (ACVR1)^{R206H} consistent with a diagnosis of fibrodysplasia ossificans progressiva; AND

C) Patient has heterotopic ossification as confirmed by radiologic testing; AND
Note: Examples of radiologic testing are x-ray, computed tomography (CT), magnetic resonance imaging (MRI), or positron emission tomography (PET) scan.

D) The medication is prescribed by or in consultation with an endocrinologist or physician who specializes in bone disease.

***** Refer to the Policy Statement

CONDITIONS NOT COVERED

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is(are) considered not medically necessary for ANY other use(s) including the following (this list may not be all inclusive; criteria will be updated as new published data are available):

1. **Chronic Obstructive Pulmonary Disease (COPD).** Sohonos is not indicated for the management of COPD.[1] Palovarotene was previously studied for the treatment of COPD, but was found to be ineffective for this condition.[7]

2. **Osteochondroma(s).** Sohonos is not indicated for the treatment and/or prevention of osteochondroma.[1] One Phase II study was initiated to evaluate Sohonos for the prevention of disease progression in pediatric patients with multiple osteochondromas.[6] However, this study was terminated early in order to analyze accumulated data and evaluate the future of Sohonos for this use. Results are not available. More data are needed.

REFERENCES

1. Sohonos® capsules [prescribing information]. Cambridge, MA: Ipsen; March 2025.
2. Kaplan FS, Al Mukaddam M, Baujat G, et al, for the Internation council on FOP (ICC) & Consultants. The medical management of fibrodysplasia ossificans progressive: current treatment considerations. Updated July 2024. Available at: https://www.iccfop.org/dvlp/wp-content/uploads/2024/07/FOP-GUIDELINES-FINAL-2024.pdf. Accessed on September 15, 2025.
3. Akesson LS, Svarirayan R, Adam MP, et al., editors. Fibrodysplasia ossificans progressiva. Gene Reviews® [Internet]. Seattle, WA: University of Washington, Seattle. Last revised May 23, 2024. Available at: https://www.ncbi.nlm.nih.gov/books/NBK558090/?report=printable. Accessed on September 15, 2025.
4. Pignolo RJ, Baujat G, Brown MA, et al. The natural history of fibrodysplasia ossificans progressive: a prospective, global 36-month study. *Genet Med*. 2022;24(12):2422-2433.
5. Pignolo RJ, Hsiao EC, Mukaddam MA, et al. Reduction of new heterotopic ossification (HO) in the open-label, phase 3, MOVE trial of palovarotene for fibrodysplasia ossificans progressive (FOP). *J Bone Miner Res*. 2023;38(3):381-394.
6. US National Institutes of Health. In: ClinicalTrials.gov [Internet]. Bethesda (MD): National Library of Medicine (US). 2000- [cited 2025 Sep 15]. Available from: https://clinicaltrials.gov/. Search term: palovarotene.

7. Food and Drug Administration (FDA). Palovarotene: NDA 215559. FDA briefing document for the Endocrinologic and Metabolic Drugs Advisory Committee. Meeting Date: June 28, 2023. Available at: https://www.fda.gov/media/169787/download. Accessed on September 15, 2025.

HISTORY

Type of Revision	Summary of Changes	Review Date
New Policy	--	09/13/2023
Annual Revision	No criteria changes.	10/09/2024
Annual Revision	**Fibrodysplasia ossificans progressiva.** The word "mutation" was updated to "pathogenic variant."	10/08/2025