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Policy Number: C16583-A

Ofev (nintedanib)

PRODUCTS AFFECTED

Ofev (nintedanib)

COVERAGE POLICY

Coverage for services, procedures, medical devices, and drugs are dependent upon benefit eligibility as outlined in the member's specific benefit plan. This Coverage Guideline must be read in its entirety to determine coverage eligibility, if any. This Coverage Guideline provides information related to coverage determinations only and does not imply that a service or treatment is clinically appropriate or inappropriate. The provider and the member are responsible for all decisions regarding the appropriateness of care. Providers should provide Molina Healthcare complete medical rationale when requesting any exceptions to these guidelines.

Documentation Requirements:
Molina Healthcare reserves the right to require that additional documentation be made available as part of its coverage determination; quality improvement; and fraud; waste and abuse prevention processes. Documentation required may include, but is not limited to, patient records, test results and credentials of the provider ordering or performing a drug or service. Molina Healthcare may deny reimbursement or take additional appropriate action if the documentation provided does not support the initial determination that the drugs or services were medically necessary, not investigational, or experimental, and otherwise within the scope of benefits afforded to the member, and/or the documentation demonstrates a pattern of billing or other practice that is inappropriate or excessive.

DIAGNOSIS:
Idiopathic pulmonary fibrosis (IPF), Chronic fibrosing interstitial lung diseases (ILDs) with a progressive phenotype, Systemic sclerosis-associated interstitial lung disease (SSc-ILD)

REQUIRED MEDICAL INFORMATION:
This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved. If a drug within this policy receives an updated FDA label within the last 180 days, medical necessity for the member will be reviewed using the updated FDA label information along with state and federal requirements, benefit being administered and formulary preferencing. Coverage will be determined on a case-by case basis until the criteria can be updated through Molina Healthcare, Inc. clinical governance. Additional information may be required on a case-by-case basis to allow for adequate review. When the requested drug product for coverage is dosed by weight, body surface area or other member specific measurement, this data element is required as part of the medical necessity review. The Pharmacy and Therapeutics Committee has determined that the drug benefit shall be a mandatory generic and that generic drugs will be dispensed whenever available.

A. ALL INDICATIONS:
   1. Prescriber attests that member is a non-smoker or is actively working to quit smoking in order to not alter the efficacy profile of Ofev (nintedanib) per the FDA label
      AND

2. Prescriber to (or the clinical reviewer has found that) the member not having any FDA labeled contraindications that haven't been addressed by the prescriber within the documentation submitted for review [Contraindications to Ofev (nintedanib) include: concomitant use of P-gp and CYP3A4 inducers (e.g., rifampin, carbamazepine, phenytoin, and St. John's wort) with Ofev should be avoided as these drugs may decrease exposure to nintedanib, avoid becoming pregnant, avoid smoking.]

B. IDIOPATHIC PULMONARY FIBROSIS:
1. Documented diagnosis of idiopathic pulmonary fibrosis (IPF)
AND
2. Documentation diagnosis was confirmed by the presence of usual interstitial pneumonia (UIP) via high-resolution computed tomography (HRCT) AND/OR Surgical lung biopsy OR transbronchial lung cryobiopsy (TBLC). [DOCUMENTATION REQUIRED: *Submit chest HRCT study, pathology report if a surgical lung biopsy was performed, or TBLC result*]
AND
3. Prescriber attests that member does not have other known causes of interstitial lung disease.
   a) No significant environmental exposure known to cause pulmonary fibrosis (e.g., drugs, asbestos, beryllium, radiation, raising birds/livestock, and metal)
   AND
   b) No known explanation for interstitial lung disease (e.g., radiation, sarcoidosis, hypersensitivity pneumonitis, bronchiolitis obliterans organizing pneumonia, human immunodeficiency virus (HIV), viral hepatitis, and cancer),
   AND
   c) No diagnosis of any connective tissue disease known to cause interstitial lung disease (e.g., scleroderma, polymyositis/dermatomyositis, systemic lupus erythematosus, and rheumatoid arthritis)
   AND
4. Documented baseline Forced Vital Capacity (%FVC) ≥ 50% of expected value for the member [DOCUMENTATION REQUIRED]
AND
5. Documented baseline diffusing capacity of the lungs for carbon monoxide (%DLCO) ≥ 30% of expected value for the member [DOCUMENTATION REQUIRED]

C. SYSTEMIC SCLEROSIS-ASSOCIATED INTERSTITIAL LUNG DISEASE (SSc-ILD):
1. Documented diagnosis of systemic sclerosis- associated interstitial lung disease
AND
2. Documentation of chest high resolution computed tomography (HRCT) scan confirming diagnosis of interstitial lung disease [DOCUMENTATION REQUIRED]
AND
3. Documentation of FVC greater than or equal to 40% of predicted AND a DLCO 30-89% of predicted. [DOCUMENTATION REQUIRED]
AND
4. Documentation of treatment failure, serious side effects, clinical contraindication, or will be concurrently taking mycophenolate mofetil

D. PROGRESSIVE FIBROSING INTERSTITIAL LUNG DISEASE:
1. Documented diagnosis of an interstitial lung disease associated with a progressive fibrosing phenotype includes connective tissue disease-related ILDs (CTD-ILDs) such as those related to rheumatoid arthritis (RA-ILD), and polymyositis/dermatomyositis; ILD related to chronic sarcoidosis; chronic hypersensitivity pneumonitis (HP); idiopathic non-specific interstitial pneumonia (iNSIP); and unclassifiable ILD [DOCUMENTATION REQUIRED]
AND
2. Documentation member has fibrosing lung disease affecting more than 10% of lung volume on high-resolution computed tomography (CT) [DOCUMENTATION REQUIRED]
AND

3. Documentation of FVC greater than or equal to 45% of predicted AND a DLCO 30% to less than 80% of predicted [DOCUMENTATION REQUIRED]

CONTINUATION OF THERAPY:

A. ALL INDICATIONS:

1. Adherence to therapy at least 85% of the time as verified by the prescriber or member medication fill history OR adherence less than 85% of the time due to the need for surgery or treatment of an infection, causing temporary discontinuation

   AND
2. Documentation of positive clinical response or disease stabilization as demonstrated by one of the following [DOCUMENTATION REQUIRED]:
   a. < 10% decline in percent predicted FVC [NOTE: A >10% decline in FVC over a 12- month period indicates disease progression and continuation of treatment will not be authorized]
   OR
   b. < 15% decline in predicted DLCO during a 6-month period

   AND
3. Prescriber attests that member continues to be a non-smoker or is actively working to quit smoking in order to not alter the efficacy profile of Ofev (nintedanib) per the FDA label

   AND
4. Prescriber attests to or clinical reviewer has found no evidence of intolerable adverse effects or drug toxicity

DURATION OF APPROVAL:

Initial authorization: 6 months, Continuation of therapy: 12 months

PRESCRIBER REQUIREMENTS:

Prescribed by, or in consultation with, a board-certified pulmonologist or rheumatologist. [If prescribed in consultation, consultation notes must be submitted with initial request and reauthorization requests]

AGE RESTRICTIONS:

18 years of age and older

QUANTITY:

150 mg twice daily

PLACE OF ADMINISTRATION:

The recommendation is that oral medications in this policy will be for pharmacy benefit coverage and patient self-administered.

DRUG INFORMATION

ROUTE OF ADMINISTRATION:

Oral

DRUG CLASS:

Pulmonary Fibrosis Agents - Kinase Inhibitors

FDA-APPROVED USES:

Indicated in adults for treatment of idiopathic pulmonary fibrosis (IPF), treatment of chronic fibrosing interstitial lung diseases (ILDs) with a progressive phenotype, slowing the rate of decline in pulmonary function in patients with systemic sclerosis-associated interstitial lung disease (SSc-ILD)

COMPENDIAL APPROVED OFF-LABELED USES:

None

APPENDIX

APPENDIX:

Diffusing Capacity for Carbon Monoxide (DLCO)

Carbon monoxide diffusing capacity (DLCO); DLCO measures the ability of the lungs to transfer gas from inhaled air to the red blood cells in pulmonary capillaries.

The normal values for CO diffusing capacity vary widely between laboratories, and both absolute values and their reproducibility are largely influenced by the measurement technique.

Therefore, this measurement is most useful if the patient's lung function changes are followed consistently by the same laboratory.

Forced vital capacity (FVC) is a widely used measure of disease status and a common endpoint in clinical trials in patients with idiopathic pulmonary fibrosis. FVC is measured via spirometry.

High-resolution computed tomography (HRCT; also called thin-section CT scanning) provides more detail than either chest radiography or conventional CT scanning, with an overall sensitivity of 95 percent and a specificity approaching 100 percent. Compared to chest radiography, HRCT can more accurately assess the pattern and distribution of diffuse lung disease, which may be beneficial when trying to narrow the differential diagnosis or define a target for lung biopsy.

Table 1. The American College of Rheumatology/European League Against Rheumatism criteria for the classification of systemic sclerosis (SSc)*

Item	Sub-item(s)	Weight/score†
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints *(sufficient criterion)*	–	9
Skin thickening of the fingers *(only count the higher score)*	Puffy fingers Sclerodactyly of the fingers (distal to the metacarpophalangeal joints but proximal to the proximal interphalangeal joints)	2 4
Fingertip lesions *(only count the higher score)*	Digital tip ulcers Fingertip pitting scars	2 3
Telangiectasia	–	2
Abnormal nailfold capillaries	–	2
Pulmonary arterial hypertension and/or interstitial lung disease *(maximum score is 2)*	Pulmonary arterial hypertension Interstitial lung disease	2 2
Raynaud's phenomenon	–	3
SSc-related autoantibodies (anticentromere, anti–topoisomerase I [anti–Scl-70], anti–RNA polymerase III) *(maximum score is 3)*	Anticentromere Anti–topoisomerase I Anti–RNA polymerase III	3

* These criteria are applicable to any patient considered for inclusion in an SSc study. The criteria are not applicable to patients with skin thickening sparing the fingers or to patients who have a scleroderma-like disorder that better explains their manifestations (e.g., nephrogenic sclerosing fibrosis, generalized morphea, eosinophilic fasciitis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft-versus-host disease, diabetic cheiroarthropathy).

† The total score is determined by adding the maximum weight (score) in each category. Patients with a total score of ≥9 are classified as having definite SSc.

BACKGROUND AND OTHER CONSIDERATIONS

BACKGROUND:

Idiopathic pulmonary fibrosis (IPF) is defined as a specific form of chronic, progressively fibrosing Idiopathic Interstitial Pneumonias, occurring primarily in older adults, limited to the lungs, and associated with the histopathological and/or radiologic pattern of unspecified interstitial pneumonia (UIP). IPF is a form of chronic interstitial fibrosis that is limited to the lung and is the most common idiopathic interstitial pneumonia, one type of interstitial lung disease, and is characterized histopathologic ally by usual interstitial pneumonia. IPF is characterized by the loss of alveolar structure through apoptosis of epithelial and endothelial cells, infiltration of inflammatory cells into interstitial and alveolar spaces, proliferation of fibroblasts, and excessive deposition of interstitial collagens. The cause is unknown; however, fibrosis appears to be preceded and provoked by a chronic inflammatory process.

Demographically, males predominate, and diagnosis occurs between the fifth and seventh decade of life. Progression of the disease is variable among individuals, ranging from a rapid decline to a steady decrement in lung function that can last several years. Progressive fibrosis ultimately leads to death with a median survival of 3 to 5 years after diagnosis. Clinical manifestations include dyspnea, reduced lung volume, and impaired gas exchange. It is progressive and irreversible, with a mean survival of approximately 2 to 4 years and patients with acute exacerbations have ≥ 60% in-hospital mortality.

Complications of IPF include heart failure, pulmonary embolism, pulmonary arterial hypertension, and lung cancer. Risk factors for IPF include cigarette smoking, genetic variants of a number of genes, exposure to metal and wood dust, and possibly gastroesophageal reflux and exposure to certain viruses.

Pharmacologic treatments have been limited and traditional approaches have included various anti-inflammatory and immunosuppressive agents; however, these approaches do not seem to be effective and are no longer considered part of routine maintenance care. Treatment has predominantly been limited to supportive care, including oxygen therapy and pulmonary rehabilitation. Lung transplantation is also an option for selected patients. Five-year survival is approximately 20-30%. Prior to the FDA approval of pirfenidone (Esbriet) and nintedanib (Ofev) in October 2014, no medications were approved for the treatment of IPF. These two agents work by different mechanisms of action, and neither agent is curative, their simultaneous FDA approval in 2014 was based on the potential for nintedanib and pirfenidone to reduce the rate of the inexorable decline in lung function in IPF.

Nintedanib is a tyrosine kinase inhibitor shown to inhibit the vascular endothelial growth factor receptors (VEGFR- 1, VEGFR-2, and VEGFR-3), fibroblast growth factor receptor (FGFR1, FGFR2, and FGFR3), and platelet-derived growth factor receptor (PDGFR) alpha and beta. Nintedanib blocked intracellular signaling and prevented proliferation, migration, and transformation of fibroblasts implicated in idiopathic pulmonary fibrosis pathogenesis.

The efficacy and safety of nintedanib was evaluated in two replicate phase 3 trials: INPULSIS-1 and INPULSIS-2. INPULSIS-1 and INPULSIS-2 were identical in design. Both trials were randomized, double-blind, placebo-controlled studies comparing treatment with nintedanib 150 mg twice daily to placebo for 52 weeks in patients with IPF. Patients were randomized in a 3:2 ratio to either nintedanib 150 mg or placebo twice daily for 52 weeks. The primary endpoint was the annual rate of decline in Forced Vital Capacity (FVC). Nintedanib was found to significantly reduce absolute FVC decline in both trials, however, a <10% decline, which is thought to be evidence of reducing clinical disease progression was significant in only INPULSIS-1 and not in INPULSIS-2.

Nintedanib was frequently associated with diarrhea, with a rate of 61.5% treatment group vs 18.6% placebo group in INPULSIS-1, and 63.2% treatment group vs 18.3% placebo group in INPULSIS-2. Effectiveness is defined as improvement or maintenance (<10% decline in percent predicted FVC or < 200 ml decrease in FVC) of disease. The primary outcome evaluated was the annual rate of decline in FVC in milliliters (mL). Nintedanib significantly reduced the annual rate of FVC decline compared with placebo in the INPULSIS-1 (-114.7 vs -239.9 mL) and the INPULSIS-2 (-113.6 vs -207.3 mL) randomized trials (N=1066) of patients 40 years or older diagnosed with idiopathic pulmonary fibrosis within the previous 5 years.

FVC is considered reliable, valid, and responsive measures of disease status as well as independent predictors of survival in patients with IPF. A decline in FVC is consistent with disease progression and is

predictive of reduced survival time.
FVC can be used as an objective measure of IPF disease progression, and a ≥10% FVC decline over 12 months has traditionally been used as a predictor of mortality risk. One study showed that a ≥10% FVC decline at 12 months was linked to a ≥2.4x greater risk of mortality. ATS/ERS/JRS/ALAT Consensus Guidelines (2011) indicate that a change in absolute forced vital capacity (FVC) of 10% [with or without a concomitant change in carbon monoxide diffusing capacity (DLCO)] or a change in absolute DLCO of 15% (with or without a concomitant change in FVC) is a surrogate marker of mortality and is evidence of disease progression.

The two Phase III (INPULSIS-1 and INPULSIS-2) trials which the FDA approval of nintedanib was based upon included only patients 40 years of age or older.
Forced vital capacity (FVC): Nintedanib has only been studied in subjects with mild-to-moderate IPF (percent predicted FVC greater than 50% and a percent predicted DLCO between 30-79%). The safety and efficacy in patients with more severe IPF disease is unknown. There is little data to indicate to what extent nintedanib is effective in patients with severe IPF (FVC < 50%).
Carbon monoxide diffusing capacity (DLCO): All patients included in the studies had a clinically and radiologically confirmed diagnosis of IPF and mild-to-moderate disease, with a baseline percent predicted FVC ≥ 50% and percent predicted DLCO 30% to 79%. The safety and efficacy in patients with more severe disease is unknown.
Liver function tests (ALT, AST, and bilirubin) should be conducted prior to initiation of treatment and monthly for 3 months, and every 3 months thereafter and as clinically indicated. In clinical trials, Ofev was associated with elevations of liver enzymes that were reversible with dose modification or interruption and not associated with clinical signs or symptoms of liver injury. Ofev also associated with increases in bilirubin. The safety and efficacy of Ofev in patients with moderate (Child-Pugh class B) and severe (Child-Pugh class C) hepatic impairment have not been studied and is therefore not recommended for these patients:
Cigarette smokers: Smoking may decrease exposure to nintedanib; members should stop smoking prior to treatment and avoid smoking during therapy as smoking can lower blood levels of nintedanib.
Pregnancy & Lactation: Based on the mechanism of action and adverse events observed in animal reproduction studies, nintedanib may be expected to cause fetal harm if used during pregnancy.
Women of reproductive potential should use adequate contraception during and at least 3 months after the last dose of therapy; Pregnancy status should be obtained before treatment and pregnancy should be avoided while receiving treatment with nintedanib . Based on animal studies, nintedanib may reduce female fertility.

Clinical Practice Guidelines
The American Thoracic Society (ATS), through a cooperative assessment with several other international organizations, provides guidance on the diagnosis and treatment of IPF. The official clinical practice guideline of the ATS was approved by the ATS, May 2015, The European Respiratory Society (ERS), April 2015, The Japanese Respiratory Society (JRS), April 2015, AND The Latin American Thoracic Association (ALAT), April 2015
A multidisciplinary, dynamic approach, with the input of clinicians, radiologists, and pathologists has been shown to improve diagnostic accuracy and is strongly recommended. (ATS 2015/ERS/JRS/ALAT)
Requirements for diagnosis of IPF include exclusion of other known causes of interstitial lung disease, presence of a pattern of usual interstitial pneumonia on high resolution CT (HRCT), and/or a specific combination of HRCT findings and lung biopsy findings.
Endpoints for assessing the disease and highlights vital capacity (VC) are one of the factors that can be used to assess response to therapy. A favorable response can be defined as a >10% increase in VC over 3-6 months. A stable response is within a 10% change, positively or negatively from baseline, in VC over 3-6 months. A >10% decrease in VC over that same time period is identified as a failure to respond. (ATS).

Many trials will assess change in forced vital capacity (FVC), usually centering around 10%, and change in carbon monoxide diffusing capacity (DLCO). Clinical practice will similarly monitor these two parameters over time to determine progression of the disease.
The ATS statement does not specifically recommend any one treatment as superior. While there is no guidance on sequential therapies, both nintedanib and pirfenidone are conditionally recommended. The ATS statement identifies conditional recommendations as recognizing that different choices will be appropriate for individual patients and providers should help each member arrive at a management decision consistent with his or her values and preferences. Providers should explicitly interpret that although drug therapy may slow rate of loss of lung function, this may also not necessarily make an individual member feel better (ATS 2015).

The Idiopathic Pulmonary Fibrosis (IPF) and Progressive Pulmonary Fibrosis (PPF) guidelines by the ATS/ERS/JRS/ALAT were updated in 2022. For IPF, antifibrotic therapies like pirfenidone and nintedanib remain central to slowing disease progression. In patients with progressive pulmonary fibrosis not classified as IPF, the guidelines recommend nintedanib as a therapeutic option to reduce decline in lung function. The use of immunosuppressive therapy (except in connective tissue-related ILDs) is generally discouraged due to associated risks. The guidelines also emphasize the importance of supportive care, including pulmonary rehabilitation, oxygen therapy, and early referral for lung transplantation in advanced cases.

The 2023 American Thoracic Society guidelines on the treatment of Systemic Sclerosis–associated Interstitial Lung Disease (SSc-ILD) strongly recommends mycophenolate mofetil as first-line treatment for SSc-ILD due to its effectiveness in stabilizing or improving lung function. Cyclophosphamide is an alternative for patients unable to tolerate MMF, though its use is limited by side effects. The guidelines also suggest considering nintedanib, an antifibrotic agent, to slow lung function decline in progressive disease. The guidelines conditionally recommend tocilizumab. The recommendations emphasize tailoring therapy to individual patient characteristics and closely monitoring for adverse effects.
Supportive care, including pulmonary rehabilitation and managing other systemic manifestations, remains crucial.

CONTRAINDICATIONS/EXCLUSIONS/DISCONTINUATION:
All other uses of Ofev (nintedanib) are considered experimental/investigational and therefore, will follow Molina's Off-Label policy. Contraindications to Ofev (nintedanib) include: concomitant use of P-gp and CYP3A4 inducers (e.g., rifampin, carbamazepine, phenytoin, and St. John's wort) with Ofev should be avoided as these drugs may decrease exposure to nintedanib, avoid becoming pregnant, avoid smoking.

Exclusions/Discontinuation:
Cases of drug-induced liver injury have been observed with Ofev treatment. Conduct liver function tests (ALT, AST, and bilirubin) prior to initiation of treatment with Ofev, at regular intervals during the first three months of treatment, and periodically thereafter or as clinically indicated. Measure liver tests promptly in patients who report symptoms that may indicate liver injury, including fatigue, anorexia, right upper abdominal discomfort, dark urine or jaundice. Dosage modifications or interruption may be necessary for liver enzyme elevations.
Do not use concurrently with Esbriet (pirfenidone).
Do not use concurrently with Actemra (tocilizumab).

OTHER SPECIAL CONSIDERATIONS:
None

CODING/BILLING INFORMATION

CODING DISCLAIMER. *Codes listed in this policy are for reference purposes only and may not be all-inclusive or applicable for every state or line of business. Deleted codes and codes which are not*

effective at the time the service is rendered may not be eligible for reimbursement. Listing of a service or device code in this policy does not guarantee coverage. Coverage is determined by the benefit document. Molina adheres to Current Procedural Terminology (CPT®), a registered trademark of the American Medical Association (AMA). All CPT codes and descriptions are copyrighted by the AMA; this information is included for informational purposes only. Providers and facilities are expected to utilize industry-standard coding practices for all submissions. Molina has the right to reject/deny the claim and recover claim payment(s) if it is determined it is not billed appropriately or not a covered benefit. Molina reserves the right to revise this policy as needed.

HCPCS CODE	DESCRIPTION
NA	

AVAILABLE DOSAGE FORMS:
Ofev CAPS 100MG
Ofev CAPS 150MG

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SUMMARY OF REVIEW/REVISIONS	DATE
REVISION- Notable revisions: Required Medical Information Continuation of Therapy Prescriber Requirements Contraindications/Exclusions/Discontinuation References	Q4 2025
REVISION- Notable revisions: Coding/Billing Information Template Update Required Medical Information Quantity Background References	Q4 2024
REVISION- Notable revisions: Required Medical Information Continuation of Therapy Age Restrictions Quantity FDA-Approved Uses Background Contraindications/Exclusions/Discontinuation Other Special Considerations References	Q4 2023

REVISION- Notable revisions: Required Medical Information Continuation of Therapy Duration of Approval Prescriber Requirements Age Restrictions References	Q4 2022
Q2 2022 Established tracking in new format	Historical changes on file