Medical Policy Bulletin
Title:
Canakinumab (Ilaris®)
Policy #:
MA08.101c

The Company makes decisions on coverage based on the Centers for Medicare and Medicaid Services (CMS) regulations and guidance, benefit plan documents and contracts, and the member's medical history and condition. If CMS does not have a position addressing a service, the Company makes decisions based on Company Policy Bulletins. Benefits may vary based on contract, and individual member benefits must be verified. The Company determines medical necessity only if the benefit exists and no contract exclusions are applicable. Although the Medicare Advantage Policy Bulletin is consistent with Medicare's regulations and guidance, the Company's payment methodology may differ from Medicare.

When services can be administered in various settings, the Company reserves the right to reimburse only those services that are furnished in the most appropriate and cost-effective setting that is appropriate to the member's medical needs and condition. This decision is based on the member's current medical condition and any required monitoring or additional services that may coincide with the delivery of this service.

This Policy Bulletin document describes the status of CMS coverage, medical terminology, and/or benefit plan documents and contracts at the time the document was developed. This Policy Bulletin will be reviewed regularly and be updated as Medicare changes their regulations and guidance, scientific and medical literature becomes available, and/or the benefit plan documents and/or contracts are changed.

Policy

Coverage is subject to the terms, conditions, and limitations of the member's Evidence of Coverage.

The Company reserves the right to reimburse only those services that are furnished in the most appropriate and cost-effective setting that is appropriate to the member's medical needs and condition.

In the absence of coverage criteria from applicable Medicare statutes, regulations, NCDs, LCDs, CMS manuals, or other Medicare coverage documents, this policy uses internal coverage criteria developed by the Company in consideration of peer-reviewed medical literature, clinical practice guidelines, and/or regulatory status.

MEDICALLY NECESSARY

The following indications will be addressed in this policy. Please see the specific criteria below for the Medical Necessity Criteria:

- Cryopyrin-Associated Periodic Syndromes (CAPS) including: Familial Cold Autoinflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS)
- Tumor Necrosis Factor (TNF) receptor--Associated Periodic Syndrome (TRAPS)
- Hyperimmunoglobulin D (Hyper-IgD) Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD)
- Familial Mediterranean Fever (FMF)
- Adult-Onset Still's Disease (AOSD)
- Systemic Juvenile Idiopathic Arthritis (SJIA)

PERIODIC FEVER SYNDROMES
Canakinumab (Ilaris®) is considered medically necessary and, therefore, covered for any of the following indications when **all** of the corresponding criteria, including Dosing and Frequency Requirements, are met:

Cryopyrin-Associated Periodic Syndromes (CAPS), Including Familial Cold Autoinflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS)

- Documented diagnosis of CAPS confirmed by molecular genetic testing that reveals pathogenic variation(s) in the *NLRP3* (also known as CIAS1) gene (e.g., variation of p.Leu353Pro or p.Leu305Pro for FCAS; variation of R260W or T348M for MWS).

- Documented presence of symptoms associated with CAPS (e.g., fever or chills, rash, joint or muscle pain, eye discomfort or redness, fatigue, headache)
- Individual is 4 years of age or older.
- Canakinumab (Ilaris®) will not be used in combination with tumor necrosis factor (TNF) inhibitors (e.g., adalimumab [Humira], etanercept [Enbrel], or infliximab [Remicade]) or other IL-1 blocking agents (e.g., rilonacept [Arcalyst] and anakinra [Kineret]).
- Individual does not have chronic or active infection, including HIV, Hepatitis B, Hepatitis C or tuberculosis.
- Dosing and Frequency: Administration is every 8 weeks.
    - For individuals with body weight greater than 40 kg, dose is 150 mg.
    - For individuals with body weight 15 to 40 kg, dose is 2 mg/kg. If inadequate response, the dose may be increased to 3 mg/kg.

Continuation Therapy (after at least 24 weeks of therapy)
Canakinumab (Ilaris®) is considered medically necessary and, therefore, covered for continuation therapy following at least 24 weeks of therapy when the individual meets all of the following criteria:

- Individual meets the Dosing and Frequency criteria outlined above.
- Documented reduction in total number of fever episodes and reduction in severity of other symptoms, compared to baseline.

Tumor Necrosis Factor (TNF) Receptor--Associated Periodic Syndrome (TRAPS)

- Documented diagnosis of TRAPS confirmed by molecular genetic testing that reveals pathogenic variation(s) in the *TNFRSF1A* gene (e.g., R92Q, T50M, C29F, C30R/S, C33G/Y, Y38C, C52F, C55S, C70R/Y, C88R/Y).
- Documented presence of all of the following characteristics of TRAPS:
    - Chronic or recurrent disease (i.e., more than 6 fever episodes per year)
    - C-reactive protein (CRP) level of more than 10 mg/L
    - Physician's global assessment (PGA) score of 2 or higher for fever, skin rash, musculoskeletal pain, abdominal pain, and eye manifestations
- Individual is 2 years of age or older.
- Canakinumab (Ilaris®) will not be used in combination with tumor necrosis factor (TNF) inhibitors (e.g., adalimumab [Humira], etanercept [Enbrel], or infliximab [Remicade]) or other IL-1 blocking agents (e.g., rilonacept [Arcalyst] and anakinra [Kineret]).
- Individual does not have chronic or active infection, including HIV, Hepatitis B, Hepatitis C or tuberculosis.
- Dosing and Frequency:
    - For individuals with body weight less than or equal to 40 kg, dose is 2 mg/kg administered every 4 weeks. If inadequate response, the dose can be increased to 4 mg/kg every 4 weeks.
    - For individuals with body weight greater than 40 kg, the dose is 150 mg administered every 4 weeks. If inadequate response, the dose can be increased to 300 mg every 4 weeks.

Continuation Therapy (after at least 16 weeks of therapy)
Canakinumab (Ilaris®) is considered medically necessary and, therefore, covered for continuation therapy following at least 16 weeks of therapy when the individual meets all of the following criteria:

- Individual meets the Dosing and Frequency criteria outlined above.
- Physician's global assessment (PGA) score less than 2 for fever, skin rash, musculoskeletal pain, abdominal pain, and eye manifestations.
- Documented C-reactive protein (CRP) level of 10 mg/L or less, or a reduction by 70% or more from baseline.

Hyperimmunoglobulin D (Hyper-IgD) Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD)*

- Documented presence of all of the following characteristics of HIDS/MKD:
    - Documented history of at least three fever episodes in a six-month period when not receiving prophylactic treatment
    - C-reactive protein (CRP) level of more than 10 mg/L
    - Physician's global assessment (PGA) score of 2 or higher for fever, abdominal pain, lymphadenopathy, aphthous ulcers
- Individual is 2 years of age or older.

- Canakinumab (Ilaris®) will not be used in combination with tumor necrosis factor (TNF) inhibitors (e.g., adalimumab [Humira], etanercept [Enbrel], or infliximab [Remicade]) or other IL-1 blocking agents (e.g., rilonacept [Arcalyst] and anakinra [Kineret]).
- Individual does not have chronic or active infection, including HIV, Hepatitis B, Hepatitis C or tuberculosis.
- Dosing and Frequency:
    - For individuals with body weight less than or equal to 40 kg, dose is 2 mg/kg administered every 4 weeks. If inadequate response, the dose can be increased to 4 mg/kg every 4 weeks.
    - For individuals with body weight greater than 40 kg, the dose is 150 mg administered every 4 weeks. If inadequate response, the dose can be increased to 300 mg every 4 weeks.

*If genetic variation(s) in the *MVK* gene is noted in documentation, the above clinical criteria must also be met.

Continuation Therapy (after at least 16 weeks of therapy)
Canakinumab (Ilaris®) is considered medically necessary and, therefore, covered for continuation therapy following at least 16 weeks of therapy when the individual meets all of the following criteria:

- Individual meets the Dosing and Frequency criteria outlined above.
- Physician's global assessment (PGA) score less than 2 for fever, skin rash, musculoskeletal pain, abdominal pain, and eye manifestations.
- Documented C-reactive protein (CRP) level of 10 mg/L or less, or a reduction by 70% or more from baseline.

Familial Mediterranean Fever (FMF)**

- Documented presence of all of the following characteristics of FMF:
    - Documented history of at least one fever episode per month
    - C-reactive protein (CRP) level of more than 10 mg/L
    - Physician's global assessment (PGA) score of 2 or higher for fever, abdominal pain, skin rash, chest pain, arthralgia, arthritis
    - Fulfillment of Tel-Hashomer Diagnostic Criteria for FMF as follows: two or more major symptoms, or one major plus two minor symptoms, as listed below:
        - Major Symptoms
            - Recurrent febrile episodes with serositis (peritonitis, synovitis or pleuritis)
            - Amyloidosis of AA type without a predisposing disease
            - Favorable response to regular colchicine treatment
        - Minor Symptoms
            - Recurrent febrile episodes
            - Erysipelas-like erythema
            - FMF in a first-degree relative
- Documented contraindication, failure, or intolerance to colchicine.
- Individual is 2 years of age or older.
- Canakinumab (Ilaris®) will not be used in combination with tumor necrosis factor (TNF) inhibitors (e.g., adalimumab [Humira], etanercept [Enbrel], or infliximab [Remicade]) or other IL-1 blocking agents (e.g., rilonacept [Arcalyst] and anakinra [Kineret]).
- Individual does not have chronic or active infection, including HIV, Hepatitis B, Hepatitis C or tuberculosis.
- Dosing and Frequency:
    - For individuals with body weight less than or equal to 40 kg, dose is 2 mg/kg administered every 4 weeks. If inadequate response, the dose can be increased to 4 mg/kg every 4 weeks.
    - For individuals with body weight greater than 40 kg, the dose is 150 mg administered every 4 weeks. If inadequate response, the dose can be increased to 300 mg every 4 weeks.

**If genetic variation(s) in the *MEFV* gene is noted in documentation, the above clinical criteria must also be met.

Continuation Therapy (after at least 16 weeks of therapy)
Canakinumab (Ilaris®) is considered medically necessary and, therefore, covered for continuation therapy following at least 16 weeks of therapy when the individual meets all of the following criteria:

- Individual meets the Dosing and Frequency criteria outlined above.
- Physician's global assessment (PGA) score less than 2 for fever, skin rash, musculoskeletal pain, abdominal pain, and eye manifestations.
- Documented C-reactive protein (CRP) level of 10 mg/L or less, or a reduction by 70% or more from baseline.

STILL'S DISEASE (ADULT-ONSET STILL'S DISEASE [AOSD], SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS [SJIA])

Canakinumab (Ilaris®) is considered medically necessary and, therefore, covered for the treatment of active Still's disease, including AOSD or SJIA when the following criteria are met:

- Documented diagnosis of moderate to severely active AOSD or SJIA characterized by all of the following symptoms:
    - chronic arthritis with two or more active joints involved (for SJIA) or four or more active joints involved (for AOSD)
    - daily, but intermittently, high spiking fever
    - additionally, accompanied by one or more of the following: rash, hepatosplenomegaly, lymphadenopathy, serositis
- Individual is 2 years of age or older.
- There is documentation of inadequate response, intolerance, or contraindication to at least one of the following: nonsteroidal anti-inflammatory drugs (NSAIDS), corticosteroids, other disease-modifying anti-rheumatic drugs (DMARDs) (e.g. anakinra, methotrexate, tocilizumab, tumor necrosis factor [TNF] inhibitors).
- Canakinumab (Ilaris®) will not be used in combination with tumor necrosis factor (TNF) inhibitors (e.g., adalimumab [Humira], etanercept [Enbrel], or infliximab [Remicade]) or other IL-1 blocking agents (e.g., rilonacept [Arcalyst] and anakinra [Kineret]).
- Individual does not have chronic or active infection, including HIV, Hepatitis B, Hepatitis C or tuberculosis.
- Dosing and Frequency: Individuals with body weight 7.5 kg or above, dose is 4 mg/kg (maximum of 300 mg) administered every 4 weeks.

Continuation Therapy (after at least 32 weeks of therapy)

Canakinumab (Ilaris®) is considered medically necessary and, therefore, covered for continuation therapy following at least 32 weeks of therapy when the individual meets all of the following criteria:

- Individual meets the Dosing and Frequency criteria outlined above.
- Documented improvement or stability of AOSD or SJIA (e.g., reduction in the signs and symptoms [including number of active joints involved], and reduction in corticosteroid dose).

NOT MEDICALLY NECESSARY

When molecular genetic testing of the relevant genes listed above reveals established "benign variation(s)" or "wild-type genotype", canakinumab (Ilaris®) is considered not medically necessary and, therefore, not covered because the available published peer-reviewed literature does not support its use in the treatment of this disease.

EXPERIMENTAL/INVESTIGATIONAL

When molecular genetic testing of the relevant genes listed above reveals "likely pathogenic" or "variations of unknown significance (VUS)", the use of canakinumab (Ilaris®) is considered experimental/investigational and, therefore, not covered because the safety and/or effectiveness of this service cannot be established by review of the available published peer-reviewed literature.

All other uses for canakinumab (Ilaris®) are considered experimental/investigational and, therefore, not covered unless the indication is supported as an accepted off-label use, as defined in the Company medical policy on off-label coverage for prescription drugs and biologics.

DOSING AND FREQUENCY REQUIREMENTS

The Company reserves the right to modify the Dosing and Frequency Requirements listed in this policy to ensure consistency with the most recently published recommendations for the use of canakinumab (Ilaris®). Changes to these guidelines are based on a consensus of information obtained from resources such as, but not limited to: the US Food and Drug Administration (FDA); Company-recognized authoritative pharmacology compendia; or published peer-reviewed clinical research. The professional provider must supply supporting documentation (i.e., published peer-reviewed literature) in order to request coverage for an amount of canakinumab (Ilaris®) outside of the Dosing and Frequency Requirements listed in this policy. For a list of Company-recognized pharmacology compendia, view our policy on off-label coverage for prescription drugs and biologics.

Accurate member information is necessary for the Company to approve the requested dose and frequency of this

drug. If the member's dose, frequency, or regimen changes (based on factors such as changes in member weight or incomplete therapeutic response), the provider must submit those changes to the Company for a new approval based on those changes as part of the utilization management activities. The Company reserves the right to conduct post-payment review and audit procedures for any claims submitted for canakinumab (Ilaris®).

REQUIRED DOCUMENTATION

The individual's medical record must reflect the medical necessity for the care provided. These medical records may include, but are not limited to: records from the professional provider's office, hospital, nursing home, home health agencies, therapies, and test reports.

The Company may conduct reviews and audits of services to our members, regardless of the participation status of the provider. All documentation is to be available to the Company upon request. Failure to produce the requested information may result in a denial for the drug.

When coverage of canakinumab (Ilaris®) is requested outside of the Dosing and Frequency Requirements listed in this policy, the prescribing professional provider must supply documentation (i.e., published peer-reviewed literature) to the Company that supports this request.

Guidelines

There is no Medicare coverage determination addressing ramucirumab (Cyramza®); therefore, the Company policy is applicable.

PRESCRIBING INFORMATION

Canakinumab (Ilaris®) is administered via subcutaneous injection every 4 weeks for:

- Adult-Onset Still's Disease (AOSD)
- Familial Mediterranean Fever (FMF)
- Hyperimmunoglobulin D (Hyper-IgD) Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD)
- Systemic Juvenile Idiopathic Arthritis (SJIA)
- Tumor Necrosis Factor (TNF) receptor--Associated Periodic Syndrome (TRAPS)

Canakinumab (Ilaris®) is administered via subcutaneous injection every 8 weeks for Cryopyrin-Associated Periodic Syndromes (CAPS) including: Familial Cold Autoinflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS).

Live vaccines should not be given concurrently with canakinumab (Ilaris®).

DIAGNOSTIC TOOL

The physician's global assessment (PGA) classifies clinical signs and symptoms associated with each disease with the use of a 5-point scale with scores of 0 (none), 1 (minimal), 2 (mild), 3 (moderate), and 4 (severe).

BENEFIT APPLICATION

Subject to the terms and conditions of the applicable Evidence of Coverage, canakinumab (Ilaris®) is covered under the medical benefits of the Company's Medicare Advantage products when the medical necessity criteria listed in this medical policy are met.

US FOOD AND DRUG ADMINISTRATION (FDA) STATUS

Canakinumab (Ilaris®) was approved by the FDA on June 17, 2009 for the treatment of Cryopyrin-Associated Periodic Syndromes (CAPS), in adults and children 4 years of age and older including Familial Cold Autoinflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS). Supplemental approvals for canakinumab (Ilaris®) have since been issued by the FDA.

Description

The cytokine interleukin-1β (IL-1β) is a key mediator of the inflammatory response; it is hypothesized that excessive release of activated IL-1β that drives inflammation. Canakinumab (Ilaris®) is a recombinant monoclonal antibody that binds to human IL-1β and neutralizes its activity by blocking its interaction with IL-1 receptors, thus reducing the inflammatory response pathway.

CRYOPYRIN-ASSOCIATED PERIODIC SYNDROMES

Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Autoinflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS), are rare genetic syndromes cause by variation(s) in the *NLRP3* gene (also known as CIAS1 gene) which results in hyperactive cryopyrin proteins and an inappropriate inflammatory response. FCAS is characterized by fever episodes typically lasting 1 day resulting in skin rash, fever, chills, joint pain, conjunctivitis, nausea, sweating, drowsiness, headache, thirst, or headaches after exposure to cold temperatures or other environmental triggers. FCAS typically begins in infancy and occurs throughout the rest of the individual's life. MWS is characterized by fever episodes typically lasting 1-3 days resulting in skin rash, fever, chills, joint pain, conjunctivitis, or severe headaches with vomiting. MWS flare-ups may begin in infancy or early childhood. The triggers of MWS are unknown but are thought to either arise spontaneously or be triggered by cold, heat, fatigue, or other stresses. Individuals with MWS may develop hearing loss and kidney damage due to amyloidosis.

Lachmann 2009 evaluated the safety and effectiveness of canakinumab (Ilaris®) in the treatment of CAPS with a three-part, 48-week, double-blind, placebo-controlled, randomized withdrawal study. Part 1 was an 8-week open label trial where 35 individuals with MWS received a single dose of 150 mg of canakinumab (Ilaris®) subcutaneously. A complete response was defined as a global assessment of no or minimal disease activity by a physician, an assessment of no or minimal rash, and a value for both serum C-reactive protein (CRP) and serum amyloid A protein (SAA) that was within the normal range (<10 mg per liter for both measures). At the end of 8 weeks, 34 of the 35 patients (97%) achieved a complete response to canakinumab (Ilaris®). Thirty-one of those who achieved a complete response entered Part 2, a randomized, double-blind, placebo controlled withdrawal period where participants received either 150 mg of canakinumab (Ilaris®) or placebo every 8 weeks for up to 24 weeks. The primary outcome measure was the proportion of individuals with a relapse of CAPS during canakinumab (Ilaris®) treatment, as compared with placebo, in part 2. Relapse was defined as a value for either CRP or SAA of more than 30 mg per liter, accompanied by a physician's assessment of global disease activity that was greater than minimal or that was minimal and accompanied by a rash that was assessed as more than minimal. None of those who received canakinumab (Ilaris®) had disease flares, compared to 81% of those who received placebo (P<0.001). At the end of part 2, median C-reactive protein (CRP) and serum amyloid A protein (SAA) values were normal (<10 mg per liter for both measures) in patients receiving canakinumab but were elevated in those receiving placebo (P<0.001 and P = 0.002, respectively). Part 3 was a 16-week open-label active treatment period. Clinical and biochemical remission of CAPS was sustained in 28 of the 29 patients (97%) who completed part 3.

An open-label trial was performed in individuals ages 4 to 74 years old with either MWS or FCAS phenotypes of CAPS. Results showed a clinically significant improvemTUMent of signs and symptoms and in normalization of high CRP and SAA in a majority of the individuals within one week of treatment with canakinumab (Ilaris®).

TUMOR NECROSIS FACTOR RECEPTOR-ASSOCIATED PERIODIC SYNDROME

Tumor necrosis factor (TNF) receptor--associated periodic syndrome (TRAPS) is a rare genetic disorder characterized by chronic or recurrent disease (i.e., more than 6 fever episodes per year), fever, chills, abdominal pain, skin rash, periorbital edema, conjunctivitis, musculoskeletal pain, high C-reactive protein (CRP), arthralgia, and myalgia**.** TRAPS may cause inflammation in various areas of the body including the heart muscle, certain joints, throat, or mucous membranes, amyloidosis, and kidney failure. TRAPS is caused by pathogenic variations in the *TNFRSF1A* gene that causes the tumor necrosis factor receptor 1 (TNFR1) protein to misfold which prevents its binding and subsequent signaling to the tumor necrosis factor (TNF) protein. The misfolded proteins forms clumps and cause the production of cytokines through the inflammation pathway.

DeBenedetti 2018 evaluated the safety and effectiveness of canakinumab (Ilaris®) in the treatment of genetically confirmed TRAPS, HIDS/MKD, or colchicine-resistant FMF in a randomized, Phase 3 cohort study called CLUSTER. Participants received either 150 mg of canakinumab (Ilaris®) or placebo every 4 weeks. The primary outcome was complete response, defined as resolution of baseline flare at day 15 (Physician's global assessment [PGA] score less than 2, plus CRP level of ≤10 mg/L or a reduction by ≥70% from baseline) and no new flare (PGA score of ≥2 and CRP level of ≥30 mg per liter) until week 16. At week 16, a statistically significant percentage of participants receiving canakinumab (Ilaris®) had a complete response than those receiving placebo: 61% vs. 6% of those with FMF (P<0.001), 35% versus 6% of those with HIDS/MKD (P = 0.003), and 45% versus 8% of those with TRAPS (P =

0.006).

HYPERIMMUNOGLOBULIN D SYNDROME/MEVALONATE KINASE DEFICIENCY

Hyperimmunoglobulin D (Hyper-IgD) syndrome (HIDS) is the less severe form of a metabolic disorder known as mevalonate kinase deficiency (MKD). It is characterized by periodic episodes of fever, abdominal pain, lymphadenopathy, and aphthous ulcers. HIDS/MKD usually begins during infancy. Most episodes last several days and occur periodically throughout life. The frequency of episodes and their severity vary greatly from person to person. Some individuals have no symptoms between attacks, while others have persistent symptoms from frequent attacks. Episodes can occur spontaneously or be triggered by vaccinations, infections, and/or emotional or physical stress. Amyloidosis, abdominal adhesions, and very rarely joint contractures may occur. HIDS/MKD is caused by variations in the *MVK* gene which provides instructions for making the mevalonate kinase enzyme, thus the variations result in a partial enzyme deficiency.

See DeBenedetti 2018 Study (above) for study summary.

FAMILIAL MEDITERRANEAN FEVER
Familial Mediterranean fever (FMF) is an inherited condition characterized by recurrent episodes of painful inflammation in the abdomen, lungs, or joints. FMF is characterized by periodic episodes of fever, abdominal pain, skin rash, chest pain, arthralgia, and arthritis that last one to three days and vary in severity. FMF usually begins in childhood or the teenage years, but in some cases, the initial attack occurs much later in life. The length of time between attacks is also variable and can range from days to years. During these periods, affected individuals usually have no signs or symptoms related to the condition. Colchicine is usually the first-line agent for treatment. Without treatment, amyloidosis and kidney failure may occur. FMF is caused by variations in the *MEFV* gene which provides instructions for making a protein called pyrin (also known as marenostrin), which is found in white blood cells, thus the variations result in reduced pyrin activity, which disrupts control of the inflammation process.

See DeBenedetti 2018 Study (above) for study summary.

STILL'S DISEASE

ADULT-ONSET STILL'S DISEASE
Adult-Onset Still's Disease (AOSD) is a rare inflammatory disorder characterized by episodes of high, spiking daily fevers, rash, joint or muscle pain, sore throat, hepatosplenomegaly, lymphadenopathy, and serositis. It is considered a continuum of systemic juvenile idiopathic arthritis (SJIA). AOSD primarily affects individuals 16-35 years of age and is considered a rare disease, unlike SJIA. The severity and frequency of episodes will vary between individuals. The treatment of AOSD may include nonsteroidal anti-inflammatory drugs (NSAIDS), glucocorticoids, or disease-modifying anti-rheumatic drugs (DMARDs) (e.g. anakinra, canakinumab, methotrexate, tocilizumab, tumor necrosis factor [TNF] inhibitors).

The approval of canakinumab (Ilaris) in adults with AOSD is based on the pharmacokinetic exposure and extrapolation of the established efficacy of canakinumab (Ilaris) in those with SJIA. The efficacy of canakinumab (Ilaris) was also assessed in a randomized, double-blind, placebo-controlled, Phase 2 study that enrolled 36 individuals (22 to 70 years old) diagnosed with AOSD. Individuals with active joint involvement were eligible for enrolment if they fulfilled the AOSD Yamaguchi classification criteria, had a disease activity based on DAS28(ESR) of ≥3.2 at screening, and had ≥4 tender and swollen (28-joint count) joint counts at screening and baseline; half of the participants experienced fever, with lesser occurrences of rash and lymphadenopathy. Prior or concurrent therapies included, nonsteroidal anti-inflammatory drugs (NSAIDS), corticosteroids, other disease-modifying anti-rheumatic drugs (DMARDs) (e.g. anakinra, tocilizumab, tumor necrosis factor [TNF] inhibitors). Participants received either 4 mg/kg canakinumab (Ilaris®) or placebo every 4 weeks for at least 12 weeks. Placebo non-responders at week 12 received canakinumab (Ilaris®) from weeks 12 to 24. Participants who responded to treatment at week 24 were able to enter the open-label, long-term extension phase being treated with canakinumab (Ilaris®). The primary outcome was defined as the proportion of individuals with a clinically relevant reduction of the articular manifestation measured by change in disease activity score (ΔDAS28(ESR)>1.2) at week 12. Twelve individuals in the canakinumab (Ilaris®) and seven individuals in the placebo group showed a reduction of the DAS28(ESR) of more than 1.2 at week 12 (66.7% vs 41.2%, respectively). The difference in the DAS28(ESR) response rate of 25.5% (95% CI −10.3% to 55.9%) was not statistically significant (p=0.18, Fisher's test) and the primary outcome was not met. Canakinumab (Ilaris) led to an improvement of several other outcome measures in AOSD. The authors determined that the efficacy data (reduction in disease activity) were generally consistent with the results of a pooled efficacy analysis of individuals with SJIA.

SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS

Systemic juvenile idiopathic arthritis (SJIA), also known as Still's disease, is a rare subtype of juvenile idiopathic arthritis where the individual exhibits systemic effects, such as fever spikes, rash, hepatosplenomegaly, lymphadenopathy, serositis. SJIA is thought to be driven by proinflammatory cytokines such as IL-1β. Treatment of SJIA may include nonsteroidal anti-inflammatory drugs (NSAIDS), glucocorticoids, or disease-modifying anti-rheumatic drugs (DMARDs) (e.g. anakinra, methotrexate).

Ruperto (2012) evaluated the safety and effectiveness of canakinumab (Ilaris®) in the treatment of individuals 2 to 19 years of age with SJIA and active systemic features of fever, two or more active joints involved, C-reactive protein greater than 30 mg/L, and glucocorticoid dose of less than 1 mg/kg/day.

Trial 1 was a randomized, double-blind, single-dose, 29-day study, where 84 individuals received 4 mg/kg canakinumab (Ilaris®) or placebo. The primary outcome, JIA ACR 30 response (improvement of 30% or more in at least three of the six core criteria for JIA, worsening of more than 30% in no more than one of the criteria, and resolution of fever.) At day 15 in trial 1, 36 individuals (84%) in the canakinumab (Ilaris®) group had an adapted JIA ACR 30 response, as compared to 4 individuals (10%) in the placebo group; P<0.001). Those who had a response in Trial 1 were enrolled in Trial 2 on day 29.

Trial 2 was a two-part withdrawal design. During Part 1, 177 individuals received 4 mg/kg canakinumab (Ilaris®) every 4 weeks for 12 to 32 weeks. Those receiving glucocorticoids at enrollment, were permitted to undergo glucocorticoid tapering from week 9 through week 28, if there was at least an adapted JIA ACR 50 response (indicating the absence of fever and an improvement of ≥50% in at least three of the six core criteria for JIA, with a worsening of >30% in no more than one of the criteria). Part 2 was the withdrawal phase where individuals who had at least an adapted JIA ACR 30 response that was sustained and who were not receiving glucocorticoids or who had undergone successful glucocorticoid tapering and were receiving a stable dose of glucocorticoids were randomly assigned in a 1:1 ratio, in a double-blind fashion, to continued treatment with canakinumab (Ilaris®) or to placebo. In this withdrawal phase, individuals who had a disease flare were treated again with canakinumab (Ilaris®) in an open-label fashion. All patients who did not undergo glucocorticoid tapering, who had no response during the open-label phase, or who had a flare during the withdrawal phase could enter the ongoing long-term, open-label extension phase of the trial. The primary outcome was time to flare of systemic JIA. Among the 100 patients (of 177 in the open-label phase) who underwent randomization in the withdrawal phase, the risk of flare was lower among individuals who continued to receive canakinumab (Ilaris®) than among those who were switched to placebo (74% of individuals in the canakinumab group had no flare, vs. 25% in the placebo group, P = 0.003). The average glucocorticoid dose was reduced from 0.34 to 0.05 mg/kg/day, and glucocorticoids were discontinued in 42 of 128 individuals (33%).

OFF-LABEL INDICATION
There may be additional indications contained in the Policy section of this document due to evaluation of criteria highlighted in the Company's off-label policy, and/or review of clinical guidelines issued by leading professional organizations and government entities.

References

American College of Rheumatology. Patient/Caregiver: Cryopyrin-Associated Autoinflammatory Syndromes. 03/2019. Available at: https://www.rheumatology.org/I-Am-A/Patient-Caregiver/Diseases-Conditions/Cryopyrin-Associated-Autoinflammatory-Syndrome-CAPS-Juvenile. Accessed June 24, 2020.

American Hospital Formulary Service (AHFS). Drug Information 2020. canakinumab (Ilaris®). [Lexicomp Online Web site]. 12/09/11. Available at: http://online.lexi.com/lco/action/home [via subscription only]. Accessed June 24, 2020.

Beukelman T, Patkar N, Saag K. 2011 American College of Rheumatology Recommendations for the treatment of juvenile idiopathic arthritis: initiation and safety monitoring of therapeutic agents for the treatment of arthritis and systemic features. *Arthritis Care Res.* 2011;63(4):465-482.

Canakinumab (Ilaris®). [prescribing information]. East Hanover, NJ; Novartis Pharmaceuticals Corp; 09/2020. Available at: http://www.ilaris.com/index.jsp. Accessed December 17, 2020.

De Benedetti F, Gattorno M, Anton J, et al. Canakinumab for the Treatment of Autoinflammatory Recurrent Fever Syndromes. [Study and Supplementary Appendix]. *N Engl J Med*. 2018;378(20):1908-1919.

Elsevier's Clinical Pharmacology Compendium. Canakinumab (Ilaris®). 06/17/2020. [Clinical Key Web site]. Available

at: https://www.clinicalkey.com/pharmacology/ [via subscription only]. Accessed June 24, 2020.

Feist E, Quartier P, Fautrel B, et al. Efficacy and safety of canakinumab in patients with Still's disease: exposure-response analysis of pooled systemic juvenile idiopathic arthritis data by age groups. *Clin Exp Rheumatol.* 2018 Jul-Aug;36(4):668-675.

Frosch M, Roth J. New insights in systemic juvenile idiopathic arthritis--from pathophysiology to treatment. *Rheumatology (Oxford).* 2008 Feb;47(2):121-5.

Greco E, Aita A, Galozzi P, et al. The novel S59P mutation in the TNFRSF1A gene identified in an adult onset TNF receptor associated periodic syndrome (TRAPS) constitutively activates NF-êB pathway. *Arthritis Res Ther.* 2015;17:93.

Hedrich CM, Günther C, Aringer M. Morbus Still im Kindes- und Erwachsenenalter [Still's disease in children and adults]. *Hautarzt*. 2017 Jun;68(6):497-511. German.

Hoffman HM, Gregory SG, Mueller JL, Tresierras M, Broide DH, Wanderer AA, et al. Fine structure mapping of CIAS1: identification of an ancestral haplotype and a common FCAS mutation, L353P. *Hum Genet.* 2003;112: 209-216.

Jamilloux Y, Gerfaud-Valentin M, Martinon F, et al. Pathogenesis of adult-onset Still's disease: new insights from the juvenile counterpart*. Immunol Res.* 2015 Feb;61(1-2):53-62.

Kadavath S, Efthimiou P. Adult-onset Still's disease-pathogenesis, clinical manifestations, and new treatment options. *Ann Med.* 2015 Feb;47(1):6-14.

Kedor C, Listing J, Zernicke, et al. Canakinumab for Treatment of Adult-Onset Still's Disease to Achieve Reduction of Arthritic Manifestation (CONSIDER): Phase II, Randomised, Double-Blind, Placebo-Controlled, Multicentre, Investigator-Initiated Trial. *Ann Rheum Dis*. 2020 May 13;annrheumdis-2020-217155.

Kimura Y. Systemic juvenile idiopathic arthritis: Clinical manifestations and diagnosis. 07/12/2018. Available at: https://www.uptodate.com/contents/systemic-juvenile-idiopathic-arthritis-clinical-manifestations-and-diagnosis?search=sjia&source=search_result&selectedTitle=1~11&usage_type=default&display_rank=1 . Accessed June 24, 2020.

Kimura Y. Systemic juvenile idiopathic arthritis: treatment. 04/02/2019. Available at: https://www.uptodate.com/contents/systemic-juvenile-idiopathic-arthritis-treatment?search=systemic jia&source=search_result&selectedTitle=4~150&usage_type=default&display_rank=4. Accessed June 24, 2020.

Kudela H, Drynda S, Lux A, et al. Comparative study of Interleukin-18 (IL-18) serum levels in adult onset Still's disease (AOSD) and systemic onset juvenile idiopathic arthritis (sJIA) and its use as a biomarker for diagnosis and evaluation of disease activity. *BMC Rheumatol.* 2019 Feb 28;3:4.

Lachmann HJ, Kone-Paut I, Kuemmerle-Deschner JB, et al; Canakinumab in CAPS Study Group. Use of canakinumab in the cryopyrin-associated periodic syndrome. *N Engl J Med*. 2009;360(23):2416-25.

Lexi-Drugs Compendium. canakinumab (Ilaris®). 06/18/2020. [Lexicomp Online Web site]. Available at: http://online.lexi.com/lco/action/home [via subscription only]. Accessed June 24, 2020.

Livneh A, Langevitz P, Zemer D, et al. Criteria for the diagnosis of familial Mediterranean fever. *Arthritis Rheum.* 1997;40(10):1879-85.

Luthi F, Zufferey P, Hofer MF, So AK. "Adolescent-onset Still's disease": characteristics and outcome in comparison with adult-onset Still's disease. Clin Exp Rheumatol. 2002 May-Jun;20(3):427-30.

Mandl LA. Treatment of adult Still’s disease. Updated 06/11/2019. Available at: https://www.uptodate.com/contents/treatment-of-adult-stills-disease?search=adult onset still disease&source=search_result&selectedTitle=2~40&usage_type=default&display_rank=2 Accessed June 24, 2020.

National Institutes of Health (NIH). Genetic and Rare Diseases Information Center. Adult-onset Still's Disease. Updated 06/30/2020. Available at: https://rarediseases.info.nih.gov/diseases/436/adult-onset-stills-disease . Accessed December 18, 2020.

National Institutes of Health (NIH). Genetics Home Reference. Familial cold autoinflammatory syndrome. Reviewed 12/2014. Available at: https://ghr.nlm.nih.gov/condition/familial-cold-autoinflammatory-syndrome. Accessed June 24, 2020.

National Institutes of Health (NIH). Genetics Home Reference. Familial Mediterranean Fever. Reviewed 06/2014. Available at: https://ghr.nlm.nih.gov/condition/familial-mediterranean-fever. Accessed June 24, 2020.

National Institutes of Health (NIH). Genetics Home Reference. Hyper-IgD syndrome. Reviewed 06/26/17. Available at: https://rarediseases.info.nih.gov/diseases/2788/hyper-igd-syndrome. Accessed June 24, 2020.

National Institutes of Health (NIH). Genetics Home Reference. Mevalonate kinase deficiency. Reviewed 11/2018. Available at: https://ghr.nlm.nih.gov/condition/mevalonate-kinase-deficiency. Accessed June 24, 2020.

National Institutes of Health (NIH). Genetics Home Reference. Muckle-Wells syndrome. Reviewed 09/2008. Available at: https://ghr.nlm.nih.gov/condition/muckle-wells-syndrome#genes. Accessed June 24, 2020.

National Institutes of Health (NIH). Genetics Home Reference. Tumor necrosis factor receptor-associated periodic syndrome. Reviewed 02/2016. Available at: https://ghr.nlm.nih.gov/condition/tumor-necrosis-factor-receptor-associated-periodic-syndrome#genes. Accessed June 24, 2020.

National Organization of Rare Diseases (NORD). Adult-onset Still's Disease. Updated 2015. Available at: https://rarediseases.org/rare-diseases/adult-onset-stills-disease/. Accessed December 18, 2020.

National Organization of Rare Diseases (NORD). Familial Mediterranean Fever. Updated 02/2017. Available at: https://rarediseases.info.nih.gov/diseases/6421/familial-mediterranean-fever. Accessed June 24, 2020.

National Organization of Rare Diseases (NORD). Hyper-IgD syndrome. Reviewed 06/26/17. Available at: https://rarediseases.info.nih.gov/diseases/2788/hyper-igd-syndrome. Accessed June 24, 2020.

National Organization of Rare Diseases (NORD). Tumor Necrosis Factor Receptor-Associated Periodic Syndrome. Updated 2019. Available at: https://rarediseases.org/rare-diseases/tumor-necrosis-factor-receptor-associated-periodic-syndrome/. Accessed June 24, 2020.

Nirmala N, Brachat A, Feist E, et al. Gene-expression analysis of adult-onset Still's disease and systemic juvenile idiopathic arthritis is consistent with a continuum of a single disease entity. *Pediatr Rheumatol Online J.* 2015 Nov 20;13:50.

Ringold S, Weiss PF, Beukelman T, et al. 2013 update of the 2011 American College of Rheumatology recommendations for the treatment of juvenile idiopathic arthritis: recommendations for the medical therapy of children with systemic juvenile idiopathic arthritis and tuberculosis screening among children receiving biologic medications. *Arthritis Rheum.* 2013;65(10):2499-512.

Ruperto N, Brunner HI, Quartier P, et al; PRINTO; PRCSG. Two randomized trials of canakinumab in systemic juvenile idiopathic arthritis. *N Engl J Med.* 2012;367(25):2396-406.

Sfriso P, Bindoli S, Doria A, et al. Canakinumab for the treatment of adult-onset Still's disease. *Expert Rev Clin Immunol.* 2020 Feb;16(2):129-138.

Shinar Y, Obici L, Aksentijevich I, Bennetts B, Austrup F, Ceccherini I, et al. Guidelines for the genetic diagnosis of hereditary recurrent fevers. *Ann Rheum Dis*. 2012;71:1599-1605.

Tran T. Muckle–Wells syndrome: clinical perspectives. *Open Access Rheumatol.* 2017;9: 123-129.

Truven Health Analytics. Micromedex® DrugDex® Compendium. canakinumab (Ilaris®). 09/28/2020. Greenwood Village, CO. [Micromedex® Solutions Web site]. Available at: http://www.micromedexsolutions.com/micromedex2/librarian [via subscription only]. Accessed December 17, 2020.

US Food and Drug Administration (FDA). Center for Drug Evaluation and Research. canakinumab (Ilaris®) prescribing information and approval letter [FDA Web site]. 09/18/2020. Available at: https://www.accessdata.fda.gov/scripts/cder/daf/. Accessed December 17, 2020.

Xia X, Dai C, Zhu X, et al. Identification of a Novel NLRP12 Nonsense Mutation (Trp408X) in the Extremely Rare Disease FCAS by Exome Sequencing. *PLoS ONE.* 2016;11(6): e0156981.

Coding

Inclusion of a code in this table does not imply reimbursement. Eligibility, benefits, limitations, exclusions, precertification/referral requirements, provider contracts, and Company policies apply.

The codes listed below are updated on a regular basis, in accordance with nationally accepted coding guidelines. Therefore, this policy applies to any and all future applicable coding changes, revisions, or updates.

In order to ensure optimal reimbursement, all health care services, devices, and pharmaceuticals should be reported using the billing codes and modifiers that most accurately represent the services rendered, unless otherwise directed by the Company.

The Coding Table lists any CPT, ICD-10, and HCPCS billing codes related only to the specific policy in which they appear.

CPT Procedure Code Number(s)

N/A

ICD - 10 Procedure Code Number(s)

N/A

ICD - 10 Diagnosis Code Number(s)

E85.0	Non-neuropathic heredofamilial amyloidosis
M04.1	Periodic fever syndromes
M04.2	Cryopyrin-associated periodic syndromes
M06.1	Adult-onset Still's disease
M08.20	Juvenile rheumatoid arthritis with systemic onset, unspecified site
M08.211	Juvenile rheumatoid arthritis with systemic onset, right shoulder
M08.212	Juvenile rheumatoid arthritis with systemic onset, left shoulder
M08.221	Juvenile rheumatoid arthritis with systemic onset, right elbow
M08.222	Juvenile rheumatoid arthritis with systemic onset, left elbow
M08.231	Juvenile rheumatoid arthritis with systemic onset, right wrist
M08.232	Juvenile rheumatoid arthritis with systemic onset, left wrist
M08.241	Juvenile rheumatoid arthritis with systemic onset, right hand
M08.242	Juvenile rheumatoid arthritis with systemic onset, left hand
M08.251	Juvenile rheumatoid arthritis with systemic onset, right hip
M08.252	Juvenile rheumatoid arthritis with systemic onset, left hip
M08.261	Juvenile rheumatoid arthritis with systemic onset, right knee
M08.262	Juvenile rheumatoid arthritis with systemic onset, left knee

M08.271 Juvenile rheumatoid arthritis with systemic onset, right ankle and foot

M08.272 Juvenile rheumatoid arthritis with systemic onset, left ankle and foot

M08.28 Juvenile rheumatoid arthritis with systemic onset, vertebrae

M08.29 Juvenile rheumatoid arthritis with systemic onset, multiple sites

M08.2A Juvenile rheumatoid arthritis with systemic onset, other specified site

M1A.00X0 Idiopathic chronic gout, unspecified site, without tophus (tophi)

M1A.00X1 Idiopathic chronic gout, unspecified site, with tophus (tophi)

M1A.0110 Idiopathic chronic gout, right shoulder, without tophus (tophi)

M1A.0111 Idiopathic chronic gout, right shoulder, with tophus (tophi)

M1A.0120 Idiopathic chronic gout, left shoulder, without tophus (tophi)

M1A.0121 Idiopathic chronic gout, left shoulder, with tophus (tophi)

M1A.0210 Idiopathic chronic gout, right elbow, without tophus (tophi)

M1A.0211 Idiopathic chronic gout, right elbow, with tophus (tophi)

M1A.0220 Idiopathic chronic gout, left elbow, without tophus (tophi)

M1A.0221 Idiopathic chronic gout, left elbow, with tophus (tophi)

M1A.0310 Idiopathic chronic gout, right wrist, without tophus (tophi)

M1A.0311 Idiopathic chronic gout, right wrist, with tophus (tophi)

M1A.0320 Idiopathic chronic gout, left wrist, without tophus (tophi)

M1A.0321 Idiopathic chronic gout, left wrist, with tophus (tophi)

M1A.0410 Idiopathic chronic gout, right hand, without tophus (tophi)

M1A.0411 Idiopathic chronic gout, right hand, with tophus (tophi)

M1A.0420 Idiopathic chronic gout, left hand, without tophus (tophi)

M1A.0421 Idiopathic chronic gout, left hand, with tophus (tophi)

M1A.0510 Idiopathic chronic gout, right hip, without tophus (tophi)

M1A.0511 Idiopathic chronic gout, right hip, with tophus (tophi)

M1A.0520 Idiopathic chronic gout, left hip, without tophus (tophi)

M1A.0521 Idiopathic chronic gout, left hip, with tophus (tophi)

M1A.0610 Idiopathic chronic gout, right knee, without tophus (tophi)

M1A.0611 Idiopathic chronic gout, right knee, with tophus (tophi)

M1A.0620 Idiopathic chronic gout, left knee, without tophus (tophi)

M1A.0621 Idiopathic chronic gout, left knee, with tophus (tophi)

M1A.0710 Idiopathic chronic gout, right ankle and foot, without tophus (tophi)

M1A.0711 Idiopathic chronic gout, right ankle and foot, with tophus (tophi)

M1A.0720 Idiopathic chronic gout, left ankle and foot, without tophus (tophi)

M1A.0721 Idiopathic chronic gout, left ankle and foot, with tophus (tophi)

M1A.08X0 Idiopathic chronic gout, vertebrae, without tophus (tophi)

M1A.08X1 Idiopathic chronic gout, vertebrae, with tophus (tophi)
M1A.09X0 Idiopathic chronic gout, multiple sites, without tophus (tophi)
M1A.09X1 Idiopathic chronic gout, multiple sites, with tophus (tophi)
M1A.10X0 Lead-induced chronic gout, unspecified site, without tophus (tophi)
M1A.10X1 Lead-induced chronic gout, unspecified site, with tophus (tophi)
M1A.1110 Lead-induced chronic gout, right shoulder, without tophus (tophi)
M1A.1111 Lead-induced chronic gout, right shoulder, with tophus (tophi)
M1A.1120 Lead-induced chronic gout, left shoulder, without tophus (tophi)
M1A.1121 Lead-induced chronic gout, left shoulder, with tophus (tophi)
M1A.1210 Lead-induced chronic gout, right elbow, without tophus (tophi)
M1A.1211 Lead-induced chronic gout, right elbow, with tophus (tophi)
M1A.1220 Lead-induced chronic gout, left elbow, without tophus (tophi)
M1A.1221 Lead-induced chronic gout, left elbow, with tophus (tophi)
M1A.1310 Lead-induced chronic gout, right wrist, without tophus (tophi)
M1A.1311 Lead-induced chronic gout, right wrist, with tophus (tophi)
M1A.1320 Lead-induced chronic gout, left wrist, without tophus (tophi)
M1A.1321 Lead-induced chronic gout, left wrist, with tophus (tophi)
M1A.1410 Lead-induced chronic gout, right hand, without tophus (tophi)
M1A.1411 Lead-induced chronic gout, right hand, with tophus (tophi)
M1A.1420 Lead-induced chronic gout, left hand, without tophus (tophi)
M1A.1421 Lead-induced chronic gout, left hand, with tophus (tophi)
M1A.1510 Lead-induced chronic gout, right hip, without tophus (tophi)
M1A.1511 Lead-induced chronic gout, right hip, with tophus (tophi)
M1A.1520 Lead-induced chronic gout, left hip, without tophus (tophi)
M1A.1521 Lead-induced chronic gout, left hip, with tophus (tophi)
M1A.1610 Lead-induced chronic gout, right knee, without tophus (tophi)
M1A.1611 Lead-induced chronic gout, right knee, with tophus (tophi)
M1A.1620 Lead-induced chronic gout, left knee, without tophus (tophi)
M1A.1621 Lead-induced chronic gout, left knee, with tophus (tophi)
M1A.1710 Lead-induced chronic gout, right ankle and foot, without tophus (tophi)
M1A.1711 Lead-induced chronic gout, right ankle and foot, with tophus (tophi)
M1A.1720 Lead-induced chronic gout, left ankle and foot, without tophus (tophi)
M1A.1721 Lead-induced chronic gout, left ankle and foot, with tophus (tophi)
M1A.18X0 Lead-induced chronic gout, vertebrae, without tophus (tophi)
M1A.18X1 Lead-induced chronic gout, vertebrae, with tophus (tophi)
M1A.19X0 Lead-induced chronic gout, multiple sites, without tophus (tophi)

M1A.19X1 Lead-induced chronic gout, multiple sites, with tophus (tophi)
M1A.20X0 Drug-induced chronic gout, unspecified site, without tophus (tophi)
M1A.20X1 Drug-induced chronic gout, unspecified site, with tophus (tophi)
M1A.2110 Drug-induced chronic gout, right shoulder, without tophus (tophi)
M1A.2111 Drug-induced chronic gout, right shoulder, with tophus (tophi)
M1A.2120 Drug-induced chronic gout, left shoulder, without tophus (tophi)
M1A.2121 Drug-induced chronic gout, left shoulder, with tophus (tophi)
M1A.2210 Drug-induced chronic gout, right elbow, without tophus (tophi)
M1A.2211 Drug-induced chronic gout, right elbow, with tophus (tophi)
M1A.2220 Drug-induced chronic gout, left elbow, without tophus (tophi)
M1A.2221 Drug-induced chronic gout, left elbow, with tophus (tophi)
M1A.2310 Drug-induced chronic gout, right wrist, without tophus (tophi)
M1A.2311 Drug-induced chronic gout, right wrist, with tophus (tophi)
M1A.2320 Drug-induced chronic gout, left wrist, without tophus (tophi)
M1A.2321 Drug-induced chronic gout, left wrist, with tophus (tophi)
M1A.2410 Drug-induced chronic gout, right hand, without tophus (tophi)
M1A.2411 Drug-induced chronic gout, right hand, with tophus (tophi)
M1A.2420 Drug-induced chronic gout, left hand, without tophus (tophi)
M1A.2421 Drug-induced chronic gout, left hand, with tophus (tophi)
M1A.2510 Drug-induced chronic gout, right hip, without tophus (tophi)
M1A.2511 Drug-induced chronic gout, right hip, with tophus (tophi)
M1A.2520 Drug-induced chronic gout, left hip, without tophus (tophi)
M1A.2521 Drug-induced chronic gout, left hip, with tophus (tophi)
M1A.2610 Drug-induced chronic gout, right knee, without tophus (tophi)
M1A.2611 Drug-induced chronic gout, right knee, with tophus (tophi)
M1A.2620 Drug-induced chronic gout, left knee, without tophus (tophi)
M1A.2621 Drug-induced chronic gout, left knee, with tophus (tophi)
M1A.2710 Drug-induced chronic gout, right ankle and foot, without tophus (tophi)
M1A.2711 Drug-induced chronic gout, right ankle and foot, with tophus (tophi)
M1A.2720 Drug-induced chronic gout, left ankle and foot, without tophus (tophi)
M1A.2721 Drug-induced chronic gout, left ankle and foot, with tophus (tophi)
M1A.28X0 Drug-induced chronic gout, vertebrae, without tophus (tophi)
M1A.28X1 Drug-induced chronic gout, vertebrae, with tophus (tophi)
M1A.29X0 Drug-induced chronic gout, multiple sites, without tophus (tophi)
M1A.29X1 Drug-induced chronic gout, multiple sites, with tophus (tophi)
M1A.30X0 Chronic gout due to renal impairment, unspecified site, without tophus (tophi)

M1A.30X1 Chronic gout due to renal impairment, unspecified site, with tophus (tophi)
M1A.3110 Chronic gout due to renal impairment, right shoulder, without tophus (tophi)
M1A.3111 Chronic gout due to renal impairment, right shoulder, with tophus (tophi)
M1A.3120 Chronic gout due to renal impairment, left shoulder, without tophus (tophi)
M1A.3121 Chronic gout due to renal impairment, left shoulder, with tophus (tophi)
M1A.3210 Chronic gout due to renal impairment, right elbow, without tophus (tophi)
M1A.3211 Chronic gout due to renal impairment, right elbow, with tophus (tophi)
M1A.3220 Chronic gout due to renal impairment, left elbow, without tophus (tophi)
M1A.3221 Chronic gout due to renal impairment, left elbow, with tophus (tophi)
M1A.3310 Chronic gout due to renal impairment, right wrist, without tophus (tophi)
M1A.3311 Chronic gout due to renal impairment, right wrist, with tophus (tophi)
M1A.3320 Chronic gout due to renal impairment, left wrist, without tophus (tophi)
M1A.3321 Chronic gout due to renal impairment, left wrist, with tophus (tophi)
M1A.3410 Chronic gout due to renal impairment, right hand, without tophus (tophi)
M1A.3411 Chronic gout due to renal impairment, right hand, with tophus (tophi)
M1A.3420 Chronic gout due to renal impairment, left hand, without tophus (tophi)
M1A.3421 Chronic gout due to renal impairment, left hand, with tophus (tophi)
M1A.3510 Chronic gout due to renal impairment, right hip, without tophus (tophi)
M1A.3511 Chronic gout due to renal impairment, right hip, with tophus (tophi)
M1A.3520 Chronic gout due to renal impairment, left hip, without tophus (tophi)
M1A.3521 Chronic gout due to renal impairment, left hip, with tophus (tophi)
M1A.3610 Chronic gout due to renal impairment, right knee, without tophus (tophi)
M1A.3611 Chronic gout due to renal impairment, right knee, with tophus (tophi)
M1A.3620 Chronic gout due to renal impairment, left knee, without tophus (tophi)
M1A.3621 Chronic gout due to renal impairment, left knee, with tophus (tophi)
M1A.3710 Chronic gout due to renal impairment, right ankle and foot, without tophus (tophi)
M1A.3711 Chronic gout due to renal impairment, right ankle and foot, with tophus (tophi)
M1A.3720 Chronic gout due to renal impairment, left ankle and foot, without tophus (tophi)
M1A.3721 Chronic gout due to renal impairment, left ankle and foot, with tophus (tophi)
M1A.38X0 Chronic gout due to renal impairment, vertebrae, without tophus (tophi)
M1A.38X1 Chronic gout due to renal impairment, vertebrae, with tophus (tophi)
M1A.39X1 Chronic gout due to renal impairment, multiple sites, with tophus (tophi)
M1A.40X0 Other secondary chronic gout, unspecified site, without tophus (tophi)
M1A.40X1 Other secondary chronic gout, unspecified site, with tophus (tophi)
M1A.4110 Other secondary chronic gout, right shoulder, without tophus (tophi)
M1A.4111 Other secondary chronic gout, right shoulder, with tophus (tophi)

M1A.4120	Other secondary chronic gout, left shoulder, without tophus (tophi)
M1A.4121	Other secondary chronic gout, left shoulder, with tophus (tophi)
M1A.4210	Other secondary chronic gout, right elbow, without tophus (tophi)
M1A.4211	Other secondary chronic gout, right elbow, with tophus (tophi)
M1A.4220	Other secondary chronic gout, left elbow, without tophus (tophi)
M1A.4221	Other secondary chronic gout, left elbow, with tophus (tophi)
M1A.4310	Other secondary chronic gout, right wrist, without tophus (tophi)
M1A.4311	Other secondary chronic gout, right wrist, with tophus (tophi)
M1A.4320	Other secondary chronic gout, left wrist, without tophus (tophi)
M1A.4321	Other secondary chronic gout, left wrist, with tophus (tophi)
M1A.4410	Other secondary chronic gout, right hand, without tophus (tophi)
M1A.4411	Other secondary chronic gout, right hand, with tophus (tophi)
M1A.4420	Other secondary chronic gout, left hand, without tophus (tophi)
M1A.4421	Other secondary chronic gout, left hand, with tophus (tophi)
M1A.4510	Other secondary chronic gout, right hip, without tophus (tophi)
M1A.4511	Other secondary chronic gout, right hip, with tophus (tophi)
M1A.4520	Other secondary chronic gout, left hip, without tophus (tophi)
M1A.4521	Other secondary chronic gout, left hip, with tophus (tophi)
M1A.4610	Other secondary chronic gout, right knee, without tophus (tophi)
M1A.4611	Other secondary chronic gout, right knee, with tophus (tophi)
M1A.4620	Other secondary chronic gout, left knee, without tophus (tophi)
M1A.4621	Other secondary chronic gout, left knee, with tophus (tophi)
M1A.4710	Other secondary chronic gout, right ankle and foot, without tophus (tophi)
M1A.4711	Other secondary chronic gout, right ankle and foot, with tophus (tophi)
M1A.4720	Other secondary chronic gout, left ankle and foot, without tophus (tophi)
M1A.4721	Other secondary chronic gout, left ankle and foot, with tophus (tophi)
M1A.48X0	Other secondary chronic gout, vertebrae, without tophus (tophi)
M1A.48X1	Other secondary chronic gout, vertebrae, with tophus (tophi)
M1A.49X0	Other secondary chronic gout, multiple sites, without tophus (tophi)
M1A.49X1	Other secondary chronic gout, multiple sites, with tophus (tophi)
M1A.9XX0	Chronic gout, unspecified, without tophus (tophi)
M1A.9XX1	Chronic gout, unspecified, with tophus (tophi)

HCPCS Level II Code Number(s)

J0638 Injection, canakinumab, 1 mg

Revenue Code Number(s)

N/A

Policy History

Revisions From MA08.101c:

03/28/2025	This version of the policy will become effective 03/28/2025. This policy has been updated to communicate the Company's coverage position for the treatment of gout flares with canakinumab (Ilaris). The following ICD-10 codes were added to this policy: M08.2A Juvenile rheumatoid arthritis with systemic onset, other specified site M1A.00X0 Idiopathic chronic gout, unspecified site, without tophus (tophi) M1A.00X1 Idiopathic chronic gout, unspecified site, with tophus (tophi) M1A.0110 Idiopathic chronic gout, right shoulder, without tophus (tophi) M1A.0111 Idiopathic chronic gout, right shoulder, with tophus (tophi) M1A.0120 Idiopathic chronic gout, left shoulder, without tophus (tophi) M1A.0121 Idiopathic chronic gout, left shoulder, with tophus (tophi) M1A.0210 Idiopathic chronic gout, right elbow, without tophus (tophi) M1A.0211 Idiopathic chronic gout, right elbow, with tophus (tophi) M1A.0220 Idiopathic chronic gout, left elbow, without tophus (tophi) M1A.0221 Idiopathic chronic gout, left elbow, with tophus (tophi) M1A.0310 Idiopathic chronic gout, right wrist, without tophus (tophi) M1A.0311 Idiopathic chronic gout, right wrist, with tophus (tophi) M1A.0320 Idiopathic chronic gout, left wrist, without tophus (tophi) M1A.0321 Idiopathic chronic gout, left wrist, with tophus (tophi) M1A.0410 Idiopathic chronic gout, right hand, without tophus (tophi) M1A.0411 Idiopathic chronic gout, right hand, with tophus (tophi) M1A.0420 Idiopathic chronic gout, left hand, without tophus (tophi) M1A.0421 Idiopathic chronic gout, left hand, with tophus (tophi) M1A.0510 Idiopathic chronic gout, right hip, without tophus (tophi) M1A.0511 Idiopathic chronic gout, right hip, with tophus (tophi) M1A.0520 Idiopathic chronic gout, left hip, without tophus (tophi) M1A.0521 Idiopathic chronic gout, left hip, with tophus (tophi) M1A.0610 Idiopathic chronic gout, right knee, without tophus (tophi) M1A.0611 Idiopathic chronic gout, right knee, with tophus (tophi) M1A.0620 Idiopathic chronic gout, left knee, without tophus (tophi) M1A.0621 Idiopathic chronic gout, left knee, with tophus (tophi) M1A.0710 Idiopathic chronic gout, right ankle and foot, without tophus (tophi) M1A.0711 Idiopathic chronic gout, right ankle and foot, with tophus (tophi) M1A.0720 Idiopathic chronic gout, left ankle and foot, without tophus (tophi) M1A.0721 Idiopathic chronic gout, left ankle and foot, with tophus (tophi) M1A.08X0 Idiopathic chronic gout, vertebrae, without tophus (tophi) M1A.08X1 Idiopathic chronic gout, vertebrae, with tophus (tophi) M1A.09X0 Idiopathic chronic gout, multiple sites, without tophus (tophi) M1A.09X1 Idiopathic chronic gout, multiple sites, with tophus (tophi) M1A.10X0 Lead-induced chronic gout, unspecified site, without tophus (tophi) M1A.10X1 Lead-induced chronic gout, unspecified site, with tophus (tophi) M1A.1110 Lead-induced chronic gout, right shoulder, without tophus (tophi) M1A.1111 Lead-induced chronic gout, right shoulder, with tophus (tophi) M1A.1120 Lead-induced chronic gout, left shoulder, without tophus (tophi) M1A.1121 Lead-induced chronic gout, left shoulder, with tophus (tophi) M1A.1210 Lead-induced chronic gout, right elbow, without tophus (tophi) M1A.1211 Lead-induced chronic gout, right elbow, with tophus (tophi) M1A.1220 Lead-induced chronic gout, left elbow, without tophus (tophi) M1A.1221 Lead-induced chronic gout, left elbow, with tophus (tophi) M1A.1310 Lead-induced chronic gout, right wrist, without tophus (tophi) M1A.1311 Lead-induced chronic gout, right wrist, with tophus (tophi) M1A.1320 Lead-induced chronic gout, left wrist, without tophus (tophi)

	M1A.1321	Lead-induced chronic gout, left wrist, with tophus (tophi)
	M1A.1410	Lead-induced chronic gout, right hand, without tophus (tophi)
	M1A.1411	Lead-induced chronic gout, right hand, with tophus (tophi)
	M1A.1420	Lead-induced chronic gout, left hand, without tophus (tophi)
	M1A.1421	Lead-induced chronic gout, left hand, with tophus (tophi)
	M1A.1510	Lead-induced chronic gout, right hip, without tophus (tophi)
	M1A.1511	Lead-induced chronic gout, right hip, with tophus (tophi)
	M1A.1520	Lead-induced chronic gout, left hip, without tophus (tophi)
	M1A.1521	Lead-induced chronic gout, left hip, with tophus (tophi)
	M1A.1610	Lead-induced chronic gout, right knee, without tophus (tophi)
	M1A.1611	Lead-induced chronic gout, right knee, with tophus (tophi)
	M1A.1620	Lead-induced chronic gout, left knee, without tophus (tophi)
	M1A.1621	Lead-induced chronic gout, left knee, with tophus (tophi)
	M1A.1710	Lead-induced chronic gout, right ankle and foot, without tophus (tophi)
	M1A.1711	Lead-induced chronic gout, right ankle and foot, with tophus (tophi)
	M1A.1720	Lead-induced chronic gout, left ankle and foot, without tophus (tophi)
	M1A.1721	Lead-induced chronic gout, left ankle and foot, with tophus (tophi)
	M1A.18X0	Lead-induced chronic gout, vertebrae, without tophus (tophi)
	M1A.18X1	Lead-induced chronic gout, vertebrae, with tophus (tophi)
	M1A.19X0	Lead-induced chronic gout, multiple sites, without tophus (tophi)
	M1A.19X1	Lead-induced chronic gout, multiple sites, with tophus (tophi)
	M1A.20X0	Drug-induced chronic gout, unspecified site, without tophus (tophi)
	M1A.20X1	Drug-induced chronic gout, unspecified site, with tophus (tophi)
	M1A.2110	Drug-induced chronic gout, right shoulder, without tophus (tophi)
	M1A.2111	Drug-induced chronic gout, right shoulder, with tophus (tophi)
	M1A.2120	Drug-induced chronic gout, left shoulder, without tophus (tophi)
	M1A.2121	Drug-induced chronic gout, left shoulder, with tophus (tophi)
	M1A.2210	Drug-induced chronic gout, right elbow, without tophus (tophi)
	M1A.2211	Drug-induced chronic gout, right elbow, with tophus (tophi)
	M1A.2220	Drug-induced chronic gout, left elbow, without tophus (tophi)
	M1A.2221	Drug-induced chronic gout, left elbow, with tophus (tophi)
	M1A.2310	Drug-induced chronic gout, right wrist, without tophus (tophi)
	M1A.2311	Drug-induced chronic gout, right wrist, with tophus (tophi)
	M1A.2320	Drug-induced chronic gout, left wrist, without tophus (tophi)
	M1A.2321	Drug-induced chronic gout, left wrist, with tophus (tophi)
	M1A.2410	Drug-induced chronic gout, right hand, without tophus (tophi)
	M1A.2411	Drug-induced chronic gout, right hand, with tophus (tophi)
	M1A.2420	Drug-induced chronic gout, left hand, without tophus (tophi)
	M1A.2421	Drug-induced chronic gout, left hand, with tophus (tophi)
	M1A.2510	Drug-induced chronic gout, right hip, without tophus (tophi)
	M1A.2511	Drug-induced chronic gout, right hip, with tophus (tophi)
	M1A.2520	Drug-induced chronic gout, left hip, without tophus (tophi)
	M1A.2521	Drug-induced chronic gout, left hip, with tophus (tophi)
	M1A.2610	Drug-induced chronic gout, right knee, without tophus (tophi)
	M1A.2611	Drug-induced chronic gout, right knee, with tophus (tophi)
	M1A.2620	Drug-induced chronic gout, left knee, without tophus (tophi)
	M1A.2621	Drug-induced chronic gout, left knee, with tophus (tophi)
	M1A.2710	Drug-induced chronic gout, right ankle and foot, without tophus (tophi)
	M1A.2711	Drug-induced chronic gout, right ankle and foot, with tophus (tophi)
	M1A.2720	Drug-induced chronic gout, left ankle and foot, without tophus (tophi)
	M1A.2721	Drug-induced chronic gout, left ankle and foot, with tophus (tophi)
	M1A.28X0	Drug-induced chronic gout, vertebrae, without tophus (tophi)
	M1A.28X1	Drug-induced chronic gout, vertebrae, with tophus (tophi)
	M1A.29X0	Drug-induced chronic gout, multiple sites, without tophus (tophi)
	M1A.29X1	Drug-induced chronic gout, multiple sites, with tophus (tophi)
	M1A.30X0	Chronic gout due to renal impairment, unspecified site, without tophus (tophi)
	M1A.30X1	Chronic gout due to renal impairment, unspecified site, with tophus (tophi)
	M1A.3110	Chronic gout due to renal impairment, right shoulder, without tophus (tophi)
	M1A.3111	Chronic gout due to renal impairment, right shoulder, with tophus (tophi)
	M1A.3120	Chronic gout due to renal impairment, left shoulder, without tophus (tophi)
	M1A.3121	Chronic gout due to renal impairment, left shoulder, with tophus (tophi)

	M1A.3210	Chronic gout due to renal impairment, right elbow, without tophus (tophi)
	M1A.3211	Chronic gout due to renal impairment, right elbow, with tophus (tophi)
	M1A.3220	Chronic gout due to renal impairment, left elbow, without tophus (tophi)
	M1A.3221	Chronic gout due to renal impairment, left elbow, with tophus (tophi)
	M1A.3310	Chronic gout due to renal impairment, right wrist, without tophus (tophi)
	M1A.3311	Chronic gout due to renal impairment, right wrist, with tophus (tophi)
	M1A.3320	Chronic gout due to renal impairment, left wrist, without tophus (tophi)
	M1A.3321	Chronic gout due to renal impairment, left wrist, with tophus (tophi)
	M1A.3410	Chronic gout due to renal impairment, right hand, without tophus (tophi)
	M1A.3411	Chronic gout due to renal impairment, right hand, with tophus (tophi)
	M1A.3420	Chronic gout due to renal impairment, left hand, without tophus (tophi)
	M1A.3421	Chronic gout due to renal impairment, left hand, with tophus (tophi)
	M1A.3510	Chronic gout due to renal impairment, right hip, without tophus (tophi)
	M1A.3511	Chronic gout due to renal impairment, right hip, with tophus (tophi)
	M1A.3520	Chronic gout due to renal impairment, left hip, without tophus (tophi)
	M1A.3521	Chronic gout due to renal impairment, left hip, with tophus (tophi)
	M1A.3610	Chronic gout due to renal impairment, right knee, without tophus (tophi)
	M1A.3611	Chronic gout due to renal impairment, right knee, with tophus (tophi)
	M1A.3620	Chronic gout due to renal impairment, left knee, without tophus (tophi)
	M1A.3621	Chronic gout due to renal impairment, left knee, with tophus (tophi)
	M1A.3710	Chronic gout due to renal impairment, right ankle and foot, without tophus (tophi)
	M1A.3711	Chronic gout due to renal impairment, right ankle and foot, with tophus (tophi)
	M1A.3720	Chronic gout due to renal impairment, left ankle and foot, without tophus (tophi)
	M1A.3721	Chronic gout due to renal impairment, left ankle and foot, with tophus (tophi)
	M1A.38X0	Chronic gout due to renal impairment, vertebrae, without tophus (tophi)
	M1A.38X1	Chronic gout due to renal impairment, vertebrae, with tophus (tophi)
	M1A.39X1	Chronic gout due to renal impairment, multiple sites, with tophus (tophi)
	M1A.40X0	Other secondary chronic gout, unspecified site, without tophus (tophi)
	M1A.40X1	Other secondary chronic gout, unspecified site, with tophus (tophi)
	M1A.4110	Other secondary chronic gout, right shoulder, without tophus (tophi)
	M1A.4111	Other secondary chronic gout, right shoulder, with tophus (tophi)
	M1A.4120	Other secondary chronic gout, left shoulder, without tophus (tophi)
	M1A.4121	Other secondary chronic gout, left shoulder, with tophus (tophi)
	M1A.4210	Other secondary chronic gout, right elbow, without tophus (tophi)
	M1A.4211	Other secondary chronic gout, right elbow, with tophus (tophi)
	M1A.4220	Other secondary chronic gout, left elbow, without tophus (tophi)
	M1A.4221	Other secondary chronic gout, left elbow, with tophus (tophi)
	M1A.4310	Other secondary chronic gout, right wrist, without tophus (tophi)
	M1A.4311	Other secondary chronic gout, right wrist, with tophus (tophi)
	M1A.4320	Other secondary chronic gout, left wrist, without tophus (tophi)
	M1A.4321	Other secondary chronic gout, left wrist, with tophus (tophi)
	M1A.4410	Other secondary chronic gout, right hand, without tophus (tophi)
	M1A.4411	Other secondary chronic gout, right hand, with tophus (tophi)
	M1A.4420	Other secondary chronic gout, left hand, without tophus (tophi)
	M1A.4421	Other secondary chronic gout, left hand, with tophus (tophi)
	M1A.4510	Other secondary chronic gout, right hip, without tophus (tophi)
	M1A.4511	Other secondary chronic gout, right hip, with tophus (tophi)
	M1A.4520	Other secondary chronic gout, left hip, without tophus (tophi)
	M1A.4521	Other secondary chronic gout, left hip, with tophus (tophi)
	M1A.4610	Other secondary chronic gout, right knee, without tophus (tophi)
	M1A.4611	Other secondary chronic gout, right knee, with tophus (tophi)
	M1A.4620	Other secondary chronic gout, left knee, without tophus (tophi)
	M1A.4621	Other secondary chronic gout, left knee, with tophus (tophi)
	M1A.4710	Other secondary chronic gout, right ankle and foot, without tophus (tophi)
	M1A.4711	Other secondary chronic gout, right ankle and foot, with tophus (tophi)
	M1A.4720	Other secondary chronic gout, left ankle and foot, without tophus (tophi)
	M1A.4721	Other secondary chronic gout, left ankle and foot, with tophus (tophi)
	M1A.48X0	Other secondary chronic gout, vertebrae, without tophus (tophi)
	M1A.48X1	Other secondary chronic gout, vertebrae, with tophus (tophi)
	M1A.49X0	Other secondary chronic gout, multiple sites, without tophus (tophi)

	M1A.49X1 Other secondary chronic gout, multiple sites, with tophus (tophi) M1A.9XX0 Chronic gout, unspecified, without tophus (tophi) M1A.9XX1 Chronic gout, unspecified, with tophus (tophi)

Revisions From MA08.101b:

01/01/2024	Effective 01/01/2024 this policy applies to New Jersey Medicare Advantage (MA) lines of business.
05/08/2023	This version of the policy will become effective 05/08/2023. The following ICD-10 CM codes have been deleted from this policy, due to unspecified laterality: M08.219 Juvenile rheumatoid arthritis with systemic onset, unspecified shoulder M08.229 Juvenile rheumatoid arthritis with systemic onset, unspecified elbow M08.239 Juvenile rheumatoid arthritis with systemic onset, unspecified wrist M08.249 Juvenile rheumatoid arthritis with systemic onset, unspecified hand M08.259 Juvenile rheumatoid arthritis with systemic onset, unspecified hip M08.269 Juvenile rheumatoid arthritis with systemic onset, unspecified knee M08.279 Juvenile rheumatoid arthritis with systemic onset, unspecified ankle and foot

Revisions From MA08.101b:

05/07/2024	This policy has been reissued in accordance with the Company's annual review process.
05/08/2023	This version of the policy will become effective 05/08/2023. The following ICD-10 CM codes have been **deleted** from this policy, due to unspecified laterality: M08.219 Juvenile rheumatoid arthritis with systemic onset, unspecified shoulder M08.229 Juvenile rheumatoid arthritis with systemic onset, unspecified elbow M08.239 Juvenile rheumatoid arthritis with systemic onset, unspecified wrist M08.249 Juvenile rheumatoid arthritis with systemic onset, unspecified hand M08.259 Juvenile rheumatoid arthritis with systemic onset, unspecified hip M08.269 Juvenile rheumatoid arthritis with systemic onset, unspecified knee M08.279 Juvenile rheumatoid arthritis with systemic onset, unspecified ankle and foot

Revisions From MA08.101a:

3/22/2023	This policy has been reissued in accordance with the Company's annual review process.
03/23/2022	This policy has been reissued in accordance with the Company's annual review process.
02/15/2021	This policy was updated to communicate the Company's coverage position of canakinumab (Ilaris®) for the treatment of Adult-Onset Still's Disease (AOSD). The following ICD-10 codes were **added** to this policy: E85.0 Non-neuropathic heredofamilial amyloidosis M06.1 Adult-onset Still's disease

Revisions From MA08.101:

05/20/2020	This policy has been reissued in accordance with the Company's annual review process.
01/28/2019	This new policy has been issued to communicate the Company’s coverage position.

Version Effective Date:
03/28/2025
Version Issued Date:
03/28/2025
Version Reissued Date:

N/A