

Ilaris[®] (canakinumab) (Subcutaneous)

Document Number: IC-0177

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Dates Reviewed: 08/2014, 07/2015, 07/2016, 10/2016, 10/2017, 08/2018, 08/2019

I. Length of Authorization

Coverage will be provided for 12 months and may be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [Pharmacy Benefit]:

- Ilaris 150 mg: 2 vials every 28 days

B. Max Units (per dose and over time) [Medical Benefit]:

Cryopyrin-Associated Periodic Syndromes

- 150 billable units every 8 weeks (56 days)

Systemic Juvenile Idiopathic Arthritis

- 300 billable units every 4 weeks (28 days)

Tumor Necrosis Factor Receptor Associated Periodic Syndrome

- 300 billable units every 4 weeks (28 days)

Hyperimmunoglobulin D Syndrome/Mevalonate Kinase Deficiency

- 300 billable units every 4 weeks (28 days)

Familial Mediterranean Fever

- 300 billable units every 4 weeks (28 days)

III. Initial Approval Criteria

Coverage is provided in the following conditions:

- Patient has been evaluated and screened for the presence of latent TB infection prior to initiating treatment; **AND**
- Patient does not have an active infection, including clinically important localized infections; **AND**
- Must not be administered concurrently with live vaccines; **AND**

- Patient is not on concurrent therapy with other IL-1 blocking agents (e.g., anakinra, rilonacept, etc.): **AND**
- Patient is not on concurrent treatment with another TNF inhibitor, biologic response modifier or other non-biologic immunomodulating agent (i.e., apremilast, tofacitinib, baricitinib); **AND**

Cryopyrin-Associated Periodic Syndromes (CAPS) †

- Patient is over the age of 4; **AND**
- Must be used as a single agent; **AND**
- Patient has documented baseline serum levels of inflammatory proteins (C-Reactive Protein [CRP] and/or Serum Amyloid A [SAA]); **AND**
- Patient has documented laboratory evidence of a genetic mutation in the Cold-Induced Auto-inflammatory Syndrome 1 (CIAS1), also known as NLRP3; **AND**
 - Diagnosis of Familial Cold Autoinflammatory Syndrome (FCAS); **OR**
 - Diagnosis of Muckle-Wells Syndrome (MWS); **AND**
- Patient has two or more of any of the CAPS-typical symptoms:
 - urticaria-like rash
 - cold-triggered episodes
 - sensorineural hearing loss
 - musculoskeletal symptoms
 - chronic aseptic meningitis
 - skeletal abnormalities

Systemic Juvenile Idiopathic Arthritis (sJIA) †

- Patient is over the age of 2; **AND**
- Patient has active Systemic Juvenile Idiopathic Arthritis (sJIA); **AND**
- Patient has had at least a 1-month trial and failure (unless contraindicated or intolerant) of previous therapy with either oral non-steroidal anti-inflammatory drugs (NSAIDs) OR a systemic glucocorticoid (prednisone, methylprednisolone, etc.); **AND**
- Physician has assessed baseline disease severity utilizing an objective measure/tool

Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS) †

- Patient is over the age of 2; **AND**
- Patient has chronic or recurrent disease (defined as 6 or more flares per year); **AND**
- Patient has documented baseline serum levels of C-Reactive Protein (CRP)

Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD) †

- Patient is over the age of 2; **AND**

- Patient has a confirmed diagnosis based on genetic/enzymatic laboratory findings; **AND**
- Patient has a documented prior history of greater than or equal to 3 febrile acute flares within a 6 month period; **AND**
- Patient has documented baseline serum levels of C-Reactive Protein (CRP)

Familial Mediterranean Fever (FMF) †

- Patient is over the age of 2; **AND**
- Patient has failed on colchicine therapy or has a documented allergy or intolerance; **AND**
- Patient has active disease defined as at least one flare per month; **AND**
- Patient has documented baseline serum levels of C-Reactive Protein (CRP)

† FDA Approved Indication(s)

IV. Renewal Criteria

Coverage can be renewed based upon the following criteria:

- Patient continues to meet criteria in Section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: severe hypersensitivity reactions, serious infections (including but not limited to tuberculosis), and macrophage activation syndrome (MAS); **AND**
- Patient is receiving ongoing monitoring for presence of TB or other active infections; **AND**

Cryopyrin-Associated Periodic Syndromes

- Disease response as indicated by improvement in patient's symptoms from baseline AND improvement in serum levels of inflammatory proteins (e.g. CRP and/or SAA, etc) from baseline

Systemic Juvenile Idiopathic Arthritis

- Disease response as indicated by improvement in signs and symptoms compared to baseline such as the number of tender and swollen joint counts and/or an improvement on a disease activity scoring tool [e.g. an improvement on a composite scoring index such as Juvenile Arthritis Disease Activity Score (JADAS) or the American College of Rheumatology (ACR) Pediatric (ACR-Pedi 30) of at least 30% improvement from baseline in three of six variables]

Tumor Necrosis Factor Receptor Associated Periodic Syndrome; Hyperimmunoglobulin D Syndrome/Mevalonate Kinase Deficiency; Familial Mediterranean Fever

- Disease response as indicated by improvement in patient's symptoms from baseline AND improvement of serum levels of CRP.

V. Dosage/Administration

| Indication | Dose |
|---|--|
| Cryopyrin-Associated Periodic Syndromes | Weight greater than 40 kg <ul style="list-style-type: none">150 mg subcutaneously every 8 weeks |
| | Weight equal to 15-40 kg <ul style="list-style-type: none">2 mg/kg subcutaneously every 8 weeks. May be increased to 3 mg/kg if inadequate response. |
| Systemic Juvenile Idiopathic Arthritis | Weight is greater than or equal to 7.5 kg <ul style="list-style-type: none">4 mg/kg (with a maximum of 300mg) subcutaneously every 4 weeks. |
| All other indications | Weight greater than 40 kg <ul style="list-style-type: none">150 mg subcutaneously every 4 weeks. May increase dose to 300mg if inadequate response. |
| | Weight less than or equal to 40 kg <ul style="list-style-type: none">2 mg/kg subcutaneously every 4 weeks. May be increased to 4 mg/kg if inadequate response. |

VI. Billing Code/Availability Information

Jcode:

- J0638 – Injection, canakinumab, 1 mg : 1 billable unit = 1 mg

NDC:

- Ilaris 150 mg single-dose solution vial: 00078-0734-xx

VII. References

- Ilaris [package insert]. East Hanover, NJ; Novartis Pharmaceuticals Corporation; December 2016. Accessed July 2019.
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- Ruperto N, Brunner H, Quartier P, et al. Two Randomized Trials of Canakinumab in Systemic Juvenile Idiopathic Arthritis. *N Engl J Med* 2012; 367:2396-2406.
- Ringold, S., Weiss, P. F., Beukelman, T., DeWitt, E. M., Ilowite, N. T., Kimura, Y., Laxer, R. M., Lovell, D. J., Nigrovic, P. A., Robinson, A. B. and Vehe, R. K. (2013), 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 & Rheumatism*, 65: 2499–2512. doi: 10.1002/art.38092
- DeWitt EM, Kimura Y, Beukelman T, et al. Consensus treatment plans for new-onset systemic juvenile idiopathic arthritis. *Arthritis Care Res (Hoboken)*. 2012 Jul;64(7):1001-10.

6. Kuemmerle-Deschner JB, Ozen S, Tyrrell PN, et al. Diagnostic criteria for cryopyrin-associated periodic syndrome (CAPS). *Ann Rheum Dis*. 2017 Jun;76(6):942-947. doi: 10.1136/annrheumdis-2016-209686.
7. Terreri MT, Bernardo WM, Len CA, et al. Guidelines for the management and treatment of periodic fever syndromes: Cryopyrin-associated periodic syndromes (cryopyrinopathies - CAPS). *Rev Bras Reumatol Engl Ed*. 2016 Jan-Feb;56(1):44-51. doi: 10.1016/j.rbre.2015.08.020.

Appendix 1 – Covered Diagnosis Codes

| ICD-10 | ICD-10 Description |
|---------|---|
| M04.1 | Periodic fever syndromes |
| M04.2 | Cryopyrin-associated periodic syndromes |
| M04.9 | Autoinflammatory syndrome, unspecified |
| M08.00 | Unspecified juvenile rheumatoid arthritis of unspecified site |
| M08.011 | Unspecified juvenile rheumatoid arthritis, right shoulder |
| M08.012 | Unspecified juvenile rheumatoid arthritis, left shoulder |
| M08.019 | Unspecified juvenile rheumatoid arthritis, unspecified shoulder |
| M08.021 | Unspecified juvenile rheumatoid arthritis, right elbow |
| M08.022 | Unspecified juvenile rheumatoid arthritis, left elbow |
| M08.029 | Unspecified juvenile rheumatoid arthritis, unspecified elbow |
| M08.031 | Unspecified juvenile rheumatoid arthritis, right wrist |
| M08.032 | Unspecified juvenile rheumatoid arthritis, left wrist |
| M08.039 | Unspecified juvenile rheumatoid arthritis, unspecified wrist |
| M08.041 | Unspecified juvenile rheumatoid arthritis, right hand |
| M08.042 | Unspecified juvenile rheumatoid arthritis, left hand |
| M08.049 | Unspecified juvenile rheumatoid arthritis, unspecified hand |
| M08.051 | Unspecified juvenile rheumatoid arthritis, right hip |
| M08.052 | Unspecified juvenile rheumatoid arthritis, left hip |
| M08.059 | Unspecified juvenile rheumatoid arthritis, unspecified hip |
| M08.061 | Unspecified juvenile rheumatoid arthritis, right knee |
| M08.062 | Unspecified juvenile rheumatoid arthritis, left knee |
| M08.069 | Unspecified juvenile rheumatoid arthritis, unspecified knee |
| M08.071 | Unspecified juvenile rheumatoid arthritis, right ankle and foot |
| M08.072 | Unspecified juvenile rheumatoid arthritis, left ankle and foot |
| M08.079 | Unspecified juvenile rheumatoid arthritis, unspecified ankle and foot |
| M08.08 | Unspecified juvenile rheumatoid arthritis, vertebrae |
| M08.09 | Unspecified juvenile rheumatoid arthritis, multiple sites |
| M08.20 | Juvenile rheumatoid arthritis with systemic onset, unspecified site |
| M08.211 | Juvenile rheumatoid arthritis with systemic onset, right shoulder |

| ICD-10 | ICD-10 Description |
|---------|---|
| M08.212 | Juvenile rheumatoid arthritis with systemic onset, left shoulder |
| M08.219 | Juvenile rheumatoid arthritis with systemic onset, unspecified shoulder |
| M08.221 | Juvenile rheumatoid arthritis with systemic onset, right elbow |
| M08.222 | Juvenile rheumatoid arthritis with systemic onset, left elbow |
| M08.229 | Juvenile rheumatoid arthritis with systemic onset, unspecified elbow |
| M08.231 | Juvenile rheumatoid arthritis with systemic onset, right wrist |
| M08.232 | Juvenile rheumatoid arthritis with systemic onset, left wrist |
| M08.239 | Juvenile rheumatoid arthritis with systemic onset, unspecified wrist |
| M08.241 | Juvenile rheumatoid arthritis with systemic onset, right hand |
| M08.242 | Juvenile rheumatoid arthritis with systemic onset, left hand |
| M08.249 | Juvenile rheumatoid arthritis with systemic onset, unspecified hand |
| M08.251 | Juvenile rheumatoid arthritis with systemic onset, right hip |
| M08.252 | Juvenile rheumatoid arthritis with systemic onset, left hip |
| M08.259 | Juvenile rheumatoid arthritis with systemic onset, unspecified hip |
| M08.261 | Juvenile rheumatoid arthritis with systemic onset, right knee |
| M08.262 | Juvenile rheumatoid arthritis with systemic onset, left knee |
| M08.269 | Juvenile rheumatoid arthritis with systemic onset, unspecified 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 |
| M08.279 | Juvenile rheumatoid arthritis with systemic onset, unspecified ankle and foot |
| M08.28 | Juvenile rheumatoid arthritis with systemic onset, vertebrae |
| M08.29 | Juvenile rheumatoid arthritis with systemic onset, multiple sites |
| M08.3 | Juvenile rheumatoid polyarthritis (seronegative) |
| M08.40 | Pauciarticular juvenile rheumatoid arthritis, unspecified site |
| M08.411 | Pauciarticular juvenile rheumatoid arthritis, right shoulder |
| M08.412 | Pauciarticular juvenile rheumatoid arthritis, left shoulder |
| M08.419 | Pauciarticular juvenile rheumatoid arthritis, unspecified shoulder |
| M08.421 | Pauciarticular juvenile rheumatoid arthritis, right elbow |
| M08.422 | Pauciarticular juvenile rheumatoid arthritis, left elbow |
| M08.429 | Pauciarticular juvenile rheumatoid arthritis, unspecified elbow |
| M08.431 | Pauciarticular juvenile rheumatoid arthritis, right wrist |
| M08.432 | Pauciarticular juvenile rheumatoid arthritis, left wrist |
| M08.439 | Pauciarticular juvenile rheumatoid arthritis, unspecified wrist |
| M08.441 | Pauciarticular juvenile rheumatoid arthritis, right hand |
| M08.442 | Pauciarticular juvenile rheumatoid arthritis, left hand |
| M08.449 | Pauciarticular juvenile rheumatoid arthritis, unspecified hand |
| M08.451 | Pauciarticular juvenile rheumatoid arthritis, right hip |
| M08.452 | Pauciarticular juvenile rheumatoid arthritis, left hip |

| ICD-10 | ICD-10 Description |
|---------|--|
| M08.459 | Pauciarticular juvenile rheumatoid arthritis, unspecified hip |
| M08.461 | Pauciarticular juvenile rheumatoid arthritis, right knee |
| M08.462 | Pauciarticular juvenile rheumatoid arthritis, left knee |
| M08.469 | Pauciarticular juvenile rheumatoid arthritis, unspecified knee |
| M08.471 | Pauciarticular juvenile rheumatoid arthritis, right ankle and foot |
| M08.472 | Pauciarticular juvenile rheumatoid arthritis, left ankle and foot |
| M08.479 | Pauciarticular juvenile rheumatoid arthritis, unspecified ankle and foot |
| M08.48 | Pauciarticular juvenile rheumatoid arthritis, vertebrae |
| M08.80 | Other juvenile arthritis, unspecified site |
| M08.811 | Other juvenile arthritis, right shoulder |
| M08.812 | Other juvenile arthritis, left shoulder |
| M08.819 | Other juvenile arthritis, unspecified shoulder |
| M08.821 | Other juvenile arthritis, right elbow |
| M08.822 | Other juvenile arthritis, left elbow |
| M08.829 | Other juvenile arthritis, unspecified elbow |
| M08.831 | Other juvenile arthritis, right wrist |
| M08.832 | Other juvenile arthritis, left wrist |
| M08.839 | Other juvenile arthritis, unspecified wrist |
| M08.841 | Other juvenile arthritis, right hand |
| M08.842 | Other juvenile arthritis, left hand |
| M08.849 | Other juvenile arthritis, unspecified hand |
| M08.851 | Other juvenile arthritis, right hip |
| M08.852 | Other juvenile arthritis, left hip |
| M08.859 | Other juvenile arthritis, unspecified hip |
| M08.861 | Other juvenile arthritis, right knee |
| M08.862 | Other juvenile arthritis, left knee |
| M08.869 | Other juvenile arthritis, unspecified knee |
| M08.871 | Other juvenile arthritis, right ankle and foot |
| M08.872 | Other juvenile arthritis, left ankle and foot |
| M08.879 | Other juvenile arthritis, unspecified ankle and foot |
| M08.88 | Other juvenile arthritis, other specified site |
| M08.89 | Other juvenile arthritis, multiple sites |
| M08.90 | Juvenile arthritis, unspecified, unspecified site |
| M08.911 | Juvenile arthritis, unspecified, right shoulder |
| M08.912 | Juvenile arthritis, unspecified, left shoulder |
| M08.919 | Juvenile arthritis, unspecified, unspecified shoulder |
| M08.921 | Juvenile arthritis, unspecified, right elbow |
| M08.922 | Juvenile arthritis, unspecified, left elbow |

| ICD-10 | ICD-10 Description |
|---------|---|
| M08.929 | Juvenile arthritis, unspecified, unspecified elbow |
| M08.931 | Juvenile arthritis, unspecified, right wrist |
| M08.932 | Juvenile arthritis, unspecified, left wrist |
| M08.939 | Juvenile arthritis, unspecified, unspecified wrist |
| M08.941 | Juvenile arthritis, unspecified, right hand |
| M08.942 | Juvenile arthritis, unspecified, left hand |
| M08.949 | Juvenile arthritis, unspecified, unspecified hand |
| M08.951 | Juvenile arthritis, unspecified, right hip |
| M08.952 | Juvenile arthritis, unspecified, left hip |
| M08.959 | Juvenile arthritis, unspecified, unspecified hip |
| M08.961 | Juvenile arthritis, unspecified, right knee |
| M08.962 | Juvenile arthritis, unspecified, left knee |
| M08.969 | Juvenile arthritis, unspecified, unspecified knee |
| M08.971 | Juvenile arthritis, unspecified, right ankle and foot |
| M08.972 | Juvenile arthritis, unspecified, left ankle and foot |
| M08.979 | Juvenile arthritis, unspecified, unspecified ankle and foot |
| M08.98 | Juvenile arthritis, unspecified, vertebrae |
| M08.99 | Juvenile arthritis, unspecified, multiple sites |

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: <http://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx>. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD): N/A

| Medicare Part B Administrative Contractor (MAC) Jurisdictions | | |
|---|--|---|
| Jurisdiction | Applicable State/US Territory | Contractor |
| E (1) | CA, HI, NV, AS, GU, CNMI | Noridian Healthcare Solutions, LLC |
| F (2 & 3) | AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ | Noridian Healthcare Solutions, LLC |
| 5 | KS, NE, IA, MO | Wisconsin Physicians Service Insurance Corp (WPS) |
| 6 | MN, WI, IL | National Government Services, Inc. (NGS) |
| H (4 & 7) | LA, AR, MS, TX, OK, CO, NM | Novitas Solutions, Inc. |
| 8 | MI, IN | Wisconsin Physicians Service Insurance Corp (WPS) |
| N (9) | FL, PR, VI | First Coast Service Options, Inc. |

| Medicare Part B Administrative Contractor (MAC) Jurisdictions | | |
|---|---|--|
| Jurisdiction | Applicable State/US Territory | Contractor |
| J (10) | TN, GA, AL | Palmetto GBA, LLC |
| M (11) | NC, SC, WV, VA (excluding below) | Palmetto GBA, LLC |
| L (12) | DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA) | Novitas Solutions, Inc. |
| K (13 & 14) | NY, CT, MA, RI, VT, ME, NH | National Government Services, Inc. (NGS) |
| 15 | KY, OH | CGS Administrators, LLC |