BlueCross BlueShield of Arizona
An Independent Licensee of the Blue Cross and Blue Shield Association

MEDICAL COVERAGE GUIDELINES
SECTION: DRUGS

ORIGINAL EFFECTIVE DATE: 07/23/13
LAST REVIEW DATE: 07/09/14
LAST CRITERIA REVISION DATE:
ARCHIVE DATE:

ILARIS® (canakinumab) INJECTION

Coverage for services, procedures, medical devices and drugs are dependent upon benefit eligibility as outlined in the member's specific benefit plan. This Medical Coverage Guideline must be read in its entirety to determine coverage eligibility, if any.

The section identified as "Description" defines or describes a service, procedure, medical device or drug and is in no way intended as a statement of medical necessity and/or coverage.

The section identified as "Criteria" defines criteria to determine whether a service, procedure, medical device or drug is considered medically necessary or experimental or investigational.

State or federal mandates, e.g., FEP program, may dictate that any drug, device or biological product approved by the U.S. Food and Drug Administration (FDA) may not be considered experimental or investigational and thus the drug, device or biological product may be assessed only on the basis of medical necessity.

Medical Coverage Guidelines are subject to change as new information becomes available.

For purposes of this Medical Coverage Guideline, the terms "experimental" and "investigational" are considered to be interchangeable.

Description:

Ilaris is an interleukin-1β blocker, also known as IL-1β, that works by specifically addressing the inflammation that results from overproduction of IL-1β.

In June 2009, Ilaris was given initial FDA approval for the treatment of Familial Cold Autoinflammatory Syndrome (FACS) and Muckle-Wells Syndrome (MWS), two conditions in a group of rare diseases called Cryopyrin-Associated Periodic Syndromes (CAPS) for adults and children 4 years of age and older. In May 2013 the FDA added Active Systemic Juvenile Idiopathic Arthritis (SJIA) as an indication for Ilaris.

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Definitions:

Cryopyrin-associated periodic syndrome (CAPS) is a spectrum of autoinflammatory syndromes.

Familial Cold Autoinflammatory Syndrome (FACS) is generally caused by mutations in a gene called NLRP3. NLRP3 mutations cause increased activity of cryopyrin, a protein that regulates inflammation in the body. Increased cryopyrin activity causes overproduction of IL-1β .Symptoms of FACS include episodes of fever, skin rash and joint pain after exposure to cold temperatures.

Muckle-Wells Syndrome (MWS) occurs when a mutation in the CIAS1 gene leads to increased activity of the protein cryopyrin. This increased activity of cryopyrin leads to an increase in IL-1β. Symptoms of MWS include episodic fever, chills and painful joints.

Active Systemic Juvenile Idiopathic Arthritis (SJIA) is a type of Juvenile Idiopathic Arthritis (JIA) that generally affects both large and small joints. Symptoms include periodic fevers and rashes.

Adult: Age 18 years and older

Criteria:

See Resources section for FDA-approved dosage.

- FDA-approved dosage of Ilaris is considered ***medically necessary*** for treatment of adults and children 4 years of age and older with Cryopyrin-Associated Periodic Syndromes (CAPS) including, Familial Cold Autoinflammatory Syndrome (FCAS) and Muckle-Weiss Syndrome (MWS) with documentation of the **ALL** following:

  1. No active or latent infection present (e.g. TB, HIV, Hepatitis B, and Hepatitis C)
  2. Not scheduled to receive any live vaccines
  3. Not concurrently on any TNF-inhibitors or Interleukin-1 blocking agents
  4. Not currently pregnant and/or breastfeeding

- FDA-approved dosage of Ilaris is considered ***medically necessary*** for treatment of individuals 2 years of age and older with active systemic juvenile idiopathic arthritis (SIJA) with documentation of **ALL** the following:

  1. No active or latent infection present (e.g. TB, HIV, Hepatitis B, and Hepatitis C)
  2. Not scheduled to receive any live vaccines
  3. Not concurrently on any TNF-inhibitors or Interleukin-1 blocking agents
  4. Not currently pregnant and/or breastfeeding

ILARIS (canakinumab) INJECTION (cont.)

Criteria: (cont.)

- Ilaris for all other indications not previously listed is considered ***experimental or investigational*** based upon:

1. Lack of final approval from the Food and Drug Administration, and
2. Insufficient scientific evidence to permit conclusions concerning the effect on health outcomes, and
3. Insufficient evidence to support improvement of the net health outcome, and
4. Insufficient evidence to support improvement of the net health outcome as much as, or more than, established alternatives, and
5. Insufficient evidence to support improvement outside the investigational setting.

Resources:

Ilaris Package Insert

- FDA-approved indication and dosage:

Indication	Recommended Dose
Cryopyrin-Associated Periodic Syndromes	150 mg for CAPS patients with body weight greater than 40 kg and 2 mg/kg for CAPS patients with body weight greater than or equal to 15 kg and less than or equal to 40 kg. For children 15 to 40 kg with an inadequate response, the dose can be increased to 3 mg/kg. Administer subcutaneously every 8 weeks.
Systemic Juvenile Idiopathic Arthritis (SJIA)	4 mg/kg (with a maximum of 300mg) for patients with a body weight greater than or equal to 7.5kg. Administer subcutaneously every 4 weeks.