RATIONAL FOR INCLUSION IN PA PROGRAM

Background
Ilaris is a potent and selective interleukin-1β blocker that works by attaching itself to interleukin-1β for a sustained period, neutralizing it and blocking inflammation and related symptoms. Ilaris is used in the treatment of Cryopyrin Associated Periodic Syndrome (CAPS), a group of rare genetic diseases that include Familial Cold Auto-inflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS). Additionally, Ilaris is indicated for the treatment of other periodic fever syndromes including Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS), Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD), and Familial Mediterranean Fever (FMF). Ilaris is also used in the treatment of Systemic Juvenile Idiopathic Arthritis (SJIA), a severe autoinflammatory disease, driven by innate immunity by means of pro-inflammatory cytokines such as interleukin-1β. This drug is to be injected by a healthcare provider just below the skin (subcutaneous) (1).

Regulatory Status
FDA-approved indication: Ilaris is an interleukin-1β blocker indicated for the treatment of: (1)
Periodic Fever Syndromes:
1. Cryopyrin-Associated Periodic Syndromes (CAPS), in adults and children 4 years of age and older including:
   - Familial Cold Autoinflammatory Syndrome (FCAS)
   - Muckle-Wells Syndrome (MWS)
2. Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS) in adult and pediatric patients
3. Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD) in adult and pediatric patients
4. Familial Mediterranean fever (FMF) in adult and pediatric patients
5. Active systemic juvenile idiopathic arthritis (SJIA) in patients aged 2 years and older.

Ilaris has been associated with an increased risk of serious infections. Physicians should exercise caution when administering Ilaris to patients with infections, a history of recurring infections or underlying conditions which may predispose them to infections. Discontinue treatment with Ilaris if a patient develops a serious infection. Do not administer Ilaris to patients during an active infection
requiring medical intervention (1).

An increased incidence of serious infections and an increased risk of neutropenia (low neutrophils, a type of white blood cells that help fight infections) have been associated with administration of another IL-1 blocker in combination with TNF inhibitors. Co-administration of Ilaris with TNF inhibitors is not recommended because this may increase the risk of serious infections. Drugs that affect the immune system by blocking TNF have been associated with an increased risk of new tuberculosis and reactivation of latent tuberculosis (TB). It is possible that use of IL-1 inhibitors such as Ilaris increases the risk of reactivation of tuberculosis or of opportunistic infections. (1).

Live vaccines should not be given concurrently with Ilaris. Prior to initiation of therapy with Ilaris, patients should receive all recommended vaccinations as IL-1 blockade may interfere with immune response to infections (1).

The safety and efficacy of Ilaris in SJIA, TRAPS, HIDS/MKD, and FMF patients under 2 years of age and in CAPS patients under 4 years of age have not been established (1).

Summary
Ilaris is an interleukin-1β blocker indicated for the treatment of active systemic juvenile idiopathic arthritis (SJIA), Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS), Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD), and Familial Mediterranean Fever (FMF) and in patients aged 2 years and older and 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). Ilaris has been associated with an increased risk of serious infections. Do not administer Ilaris to patients during an active infection requiring medical intervention. The safety and efficacy of Ilaris in Active Systemic Juvenile Idiopathic Arthritis (SJIA), Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS), Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD), and Familial Mediterranean Fever (FMF) patients under 2 years of age and in Cryopyrin-Associated Periodic Syndromes (CAPS) patients under 4 years of age have not been established (1).

Prior authorization is required to ensure the safe, clinically appropriate and cost effective use of
Ilaris while maintaining optimal therapeutic outcomes.

References