A GUIDE TO YOUR ILARIS® (CANAKINUMAB) TREATMENT

ILARIS®
(canakinumab)
150 mg subcutaneous injection

Funded and developed by Novartis Pharmaceuticals UK Ltd.
This booklet is intended for patients or caregivers of patients who have been prescribed Ilaris for their periodic fever syndrome.

This document does not replace the patient information leaflet that accompanies your medication

NOVARTIS

Reporting of side effects

If you get any side effects, talk to your doctor, pharmacist or nurse. This includes any possible side effects not listed in the package leaflet. You can also report side effects directly via the Yellow Card Scheme at **www.mhra.gov.uk/yellowcard**. Side effects may also be reported to Novartis on **01276 698370** or via **medinfo.uk@novartis.com**. By reporting side effects you can help provide more information on the safety of this medicine.

A GUIDE TO YOUR ILARIS® (CANAKINUMAB) TREATMENT

You have been prescribed this medicine for your periodic fever syndrome (sometimes also referred to as a hereditary recurrent fever syndrome). In this booklet, you will find information about what this medicine is and how it works, and things you should know before starting treatment.

The patient information leaflet that comes with the medicine also contains important information that you should read and keep for future reference.

Periodic fever syndromes are very rare and chronic autoinflammatory diseases that can affect both children and adults. The majority present in infancy or childhood. Periodic fever syndromes are caused by mutations (changes) that affect the immune system. The part of the immune system affected in periodic fever syndromes is called the 'innate immune system'. The innate immune system is our body's first defence against infections. Normally, the innate immune system is activated if the body is injured or there is infection or disease. This leads to an inflammatory response which helps fight off the disease and promote healing. This inflammatory response may also lead to unpleasant symptoms such as fever, but once the body has healed or fought off the infection, the inflammatory response switches off and the symptoms also disappear. In periodic fever syndromes, the innate immune system is overactive and this inflammatory response occurs by itself, without any external trigger, and for prolonged periods; this is known as auto-inflammation. This can lead to symptoms such as fever, headache, fatigue, skin rash, or painful joints and muscles.

Ilaris is used for treatment of the following periodic fever syndromes:

- Cryopyrin-associated periodic syndromes (CAPS)
- Tumour necrosis factor receptor associated periodic syndrome (TRAPS)
- Hyperimmunoglobulin D syndrome (HIDS), also known as mevalonate kinase deficiency (MKD)
- Familial Mediterranean fever (FMF)

This medicine contains the active substance canakinumab, a monoclonal antibody that belongs to a group of medicines called interleukin inhibitors. It blocks the activity of a substance called interleukin-1 beta (IL-1 beta) in the body, which is present at increased levels in inflammatory diseases, like periodic fever syndromes. By blocking the activity of IL-1 beta, this medicine may improve the symptoms of these conditions.

WHAT YOU NEED TO KNOW BEFORE YOU START TREATMENT

Do not use Ilaris

- If you are allergic to canakinumab or any of the other ingredients of this medicine (which are listed in the patient information leaflet).
- If you have, or suspect you have, an active and severe infection.

Warning and precautions

Talk to your doctor before using this medicine if any of the following applies to you:

- If you currently have an infection or if you have had repeated infections or a condition such as a known low level of white blood cells, which makes you more likely to get infections.
- If you have or have ever had tuberculosis or direct contact with a person with an active tuberculosis infection. Your doctor may check whether you have tuberculosis using a specific test.
- If you have signs of a liver disorder such as yellow skin and eyes, nausea, loss of appetite, dark-coloured urine and light-coloured stools.
- If you need to have any vaccinations. You are advised to avoid being vaccinated with a type of vaccine called a live vaccine while being treated with Ilaris (see also "Other medicines and Ilaris").

Contact your doctor immediately

- If you have ever developed an atypical, widespread rash or skin peeling after taking Ilaris. The serious skin reactions DRESS (drug reaction with eosinophilia and systemic symptoms), has rarely been reported in association with Ilaris treatment, predominantly in patients with sJIA. Seek medical attention immediately if you notice an atypical, widespread rash, which may occur in conjunction with high body temperature and enlarged lymph nodes.

Still's disease:

- Patients with Still's disease may develop a conditional called macrophage activation syndrome (MAS), which can be life-threatening. Your doctor will monitor you for potential triggering factors of MAS that include infections and re-activation of the underlying Still's disease (flare).

POSSIBLE SIDE EFFECTS

Like all medicines, this medicine can cause side effects, although not everybody gets them. Some side effects could be serious. Tell your doctor immediately if you notice any of the side effects below:

- Fever lasting longer than 3 days or any other symptoms that might suggest a serious infection. These include shivering, chills, malaise, loss of appetite, body aches, typically in connection with a sudden onset of illness, sore throat or mouth ulcers, cough, phlegm, chest pain, difficulty breathing, ear pain, prolonged headache or localised redness, warmth or swelling of your skin or inflammation of connective tissue (cellulitis). These symptoms could be due to a serious infection, an unusual infection (opportunistic infection) or be related to low levels of white blood cells (called leukopenia or neutropenia). Your doctor may check your blood regularly if considered necessary.
- Allergic reactions with rash and itching and possibly also hives, difficulty breathing or swallowing, dizziness, unusual awareness of your heart beat (palpitations) or low blood pressure.

Please refer to the patient leaflet for further information.

OTHER SIDE EFFECTS OF ILARIS INCLUDE:

Very common May affect more than 1 in 10 people	• Infections of any kind. These can include: – Respiratory infections such as chest infection, flu, sore throat, runny nose, blocked nose, sneezing, feeling of pressure or pain in the cheeks or forehead with or without fever (pneumonia, bronchitis, influenza, sinusitis, rhinitis, pharyngitis, tonsillitis, nasopharyngitis, upper respiratory tract infection). – Other infections such as ear infection, skin infection (cellulitis), stomach pain and feeling sick (gastroenteritis) and painful and frequent urination with or without fever (urinary tract infection). • Upper abdominal pain. • Pain in joints (arthralgia). • Drop in level of white blood cells (leukopenia). • Abnormal kidney function test results (creatinine renal clearance decreased, proteinuria). • Injection site reaction (such as redness, swelling, warmth and itching).
Common May affect up to 1 in 10 people	• Candida – vaginal yeast infection (vulvovaginal candidiasis). • Feeling dizzy, spinning sensation (dizziness or vertigo). • Pain in the back or muscles. • Feeling weak or very tired (fatigue, asthenia). • Drop in level of white blood cells which help prevent infection (neutropenia). • Abnormal levels of triglycerides in your blood (lipid metabolism disorder). • Abnormal liver function test results (transaminases increased) or high level of bilirubin in the blood, with or without yellow skin and eyes (hyperbilirubinaemia).
Uncommon May affect up to 1 in 100 people	• Heartburn (gastro-oesophageal reflux disease). • Drop in level of blood cells which help prevent bleeding (platelets).

TELL YOUR DOCTOR OR YOUR CHILD'S DOCTOR IMMEDIATELY IF YOU NOTICE ANY OF THESE SYMPTOMS.

Other medicines and Ilaris

Tell your doctor, pharmacist or nurse if you are taking, have recently taken or might take any other medicines.

- Live vaccines: You are advised to avoid being vaccinated with a type of vaccine called a live vaccine while you receive this treatment. Your doctor may want to check your vaccination history and give you any vaccinations that you have missed before you start treatment with Ilaris. If you need to be given a live vaccine after starting treatment, discuss this with your doctor. A live vaccine should normally be given 3 months after your last injection of Ilaris and 3 months before the next one.
- Medicines called tumour necrosis factor (TNF) inhibitors, such as etanercept, adalimumab or infliximab: These are used mainly in rheumatic and autoimmune diseases. They should not be used with Ilaris because this may increase the risk of infections.

Pregnancy and breast-feeding

If you are pregnant or breast-feeding, think you may be pregnant or are planning to have a baby, ask your doctor or pharmacist for advice before taking this medicine.

- You are advised to avoid becoming pregnant and must use adequate contraception while using Ilaris and for at least 3 months after the last treatment. It is important to tell your doctor if you are pregnant, if you think you may be pregnant or are planning to have a baby. Your doctor will discuss with you the potential risks of treatment during pregnancy.
- If you received canakinumab while you were pregnant, it is important that you inform the baby's doctor or nurse before any vaccinations are given to your baby. Your baby should not receive live vaccines until at least 16 weeks after you received your last dose of canakinumab before giving birth.
- It is not known whether Ilaris passes into human milk. Your doctor will discuss with you the potential risks of taking this medicine before breast-feeding.

Driving and using machines

This treatment may give you a spinning sensation (dizziness or vertigo) or intense tiredness (asthenia). This may affect your ability to drive or use tools or machines. If you feel a spinning sensation or feel tired, do not drive or use any tools or machines until you are feeling normal again.

HOW MUCH ILARIS TO USE

Cryopyrin-associated periodic syndromes (CAPS)

The recommended starting dose of Ilaris is:

Adults and children aged 4 years or more

- 150 mg for patients who weigh more than 40 kg
- 2 mg/kg for patients who weigh between 15 kg and 40 kg
- 4 mg/kg for patients who weigh between 7.5 kg and less than 15 kg

Children aged 2 or 3 years

- 4 mg/kg for patients with body weight of 7.5 kg or more

Ilaris is injected every 8 weeks as a single dose.

If you have not responded well enough to the treatment after 7 days, your doctor may give you another dose of 150 mg or 2 mg/kg.

- If you respond well enough to the second dose, your treatment will be continued with 300 mg or 4 mg/kg every 8 weeks.
- If you do not respond well enough to the second dose, a third dose of 300 mg or 4 mg/kg may be given.
- If you respond well enough to the third dose, your treatment will be continued at 600 mg or 8 mg/kg every 8 weeks.

For children given a starting dose of 4 mg/kg who have not responded well enough after 7 days, the doctor may give a second dose of 4 mg/kg. If the child responds well enough to this, treatment may be continued with a dose of 8 mg/kg every 8 weeks.

Tumour necrosis factor receptor associated periodic syndrome (TRAPS), hyperimmunoglobulin D syndrome (HIDS)/mevalonate kinase deficiency (MKD) and familial Mediterranean fever (FMF)

The recommended starting dose of Ilaris is:

Adults and children aged 2 years or more

- 150 mg for patients who weigh more than 40 kg
- 2 mg/kg for patients who weigh between 7.5 kg and less than 40 kg

This medicine is injected every 4 weeks as a single dose.

- If you have not responded well enough to the treatment after 7 days, your doctor may give you another dose of 150 mg or 2 mg/kg.
- If you respond well enough to this, your treatment will be continued with 300 mg or 4 mg/kg every 4 weeks.

HOW LONG TO USE ILARIS FOR

CAPS, TRAPS, HIDS/MKD, or FMF:
You should continue using treatment for as long as the doctor tells you.

If you use more Ilaris than you should

If you accidentally inject more than the recommended dose, it is unlikely to be serious, but you should inform your doctor, pharmacist or nurse as soon as possible.

If you forget to use Ilaris

If you have CAPS, TRAPS, HIDS/MKD, or FMF and have forgotten to inject your medicine, inject the next dose as soon as you remember. Then talk to the doctor to discuss when you should inject the next dose. You should then continue with injections at the recommended intervals as before.

If you stop treatment

Stopping your treatment may cause your condition to get worse.
Do not stop taking Ilaris unless your doctor tells you to.

If you have any further questions on the use of this medicine, ask your doctor, pharmacist or nurse.

HOW TO STORE ILARIS

- Keep this medicine out of sight and reach of children.
- Do not use this medicine after the expiry date which is stated on the label and carton.
  The expiry date refers to the last day of that month.
- Store in a refrigerator (2°C to 8°C). Do not freeze.
- Keep the vial in the outer carton in order to protect from light.
- The solution should be used immediately after first piercing the vial stopper to prepare the injection.
- Do not use this medicine if you notice that the solution is not clear to opalescent or contains particles.
- Any unused medicine must be discarded after withdrawal of the dose.
- Do not throw away any medicines via wastewater or household waste. Ask your pharmacist how to throw away medicines you no longer use. These measures will help protect the environment.

INJECTING YOURSELF OR INJECTING A PATIENT WITH ILARIS

If you are a patient with CAPS, TRAPS, HIDS/MKD, or FMF or a caregiver of a patient with one of these conditions, you may administer injections yourself after proper training in the correct injection technique.

- The patient or caregiver and the doctor should decide together who will administer the injections.
- The doctor or nurse will demonstrate how to administer the injections.
- Do not try to administer an injection yourself if you have not been properly trained or if you are not sure how to do it.
- Ilaris 150 mg/ml solution for injection is supplied in a single-use vial for individual use.
- Never re-use the leftover solution.

IF YOU HAVE ANY FURTHER QUESTIONS ON THE USE OF ILARIS, ASK YOUR DOCTOR, PHARMACIST OR NURSE.

INSTRUCTIONS FOR USE OF ILARIS SOLUTION FOR INJECTION

- Read all the way through these instructions before injecting.
- It is important not to try to inject yourself until you have been trained by your healthcare professional.
- See also section 'Injecting yourself or injecting a patient with Ilaris'.

Essential preparation

- Find a clean place in which to prepare and give yourself the injection.
- Wash your hands with soap and water, then dry them on a clean towel.
- After removing the vial from the refrigerator, check the expiry date on the vial. Do not use after the expiry date which is stated on the label and carton. The expiry date refers to the last day of that month.
- Let the vial stand unopened for 10 minutes to bring the contents to room temperature. Do not try to heat the vial. Let it warm up on its own.
- Always use new, unopened needles and syringes. Do not touch the needles or the top of the vial.

Gather together the necessary items

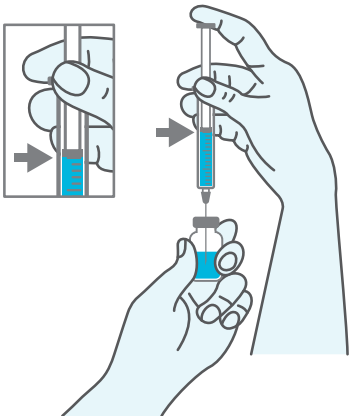
Included in the pack

- one vial of Ilaris solution for injection (keep refrigerated).

Not included in the pack

- one 1.0 ml syringe.
- one needle (such as 18 G or 21 G x 2 inch or similar, as available on the market) to draw up the solution from the vial ('withdrawal needle').
- one 27 G x 0.5 inch (or similar, as available on the market) needle for injecting ('injection needle').
- alcohol swabs.
- clean, dry cotton swabs.
- an adhesive plaster.
- a proper disposal container for used needles, syringe and vial (sharps container).

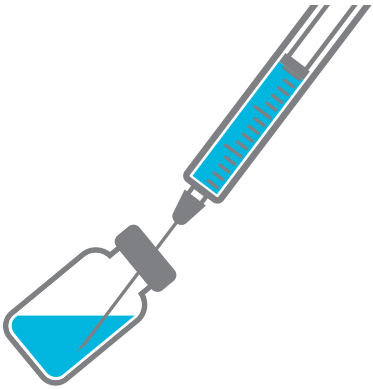
Preparing the injection



1. Take off the protective cap from the Ilaris vial. Do not touch the vial stopper. Clean the rubber stopper of the vial with an alcohol swab.

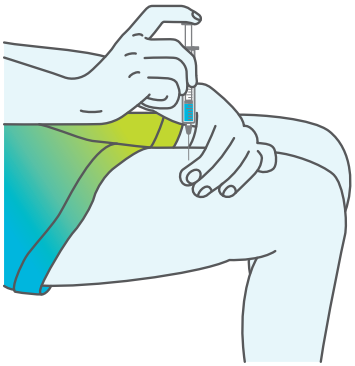
   Open the wrappers containing the syringe and the withdrawal needle.

   - Put the withdrawal needle on the syringe.
   - Take off the cap from the withdrawal needle.
   - Push the withdrawal needle into the vial of Ilaris solution through the centre of the rubber stopper.

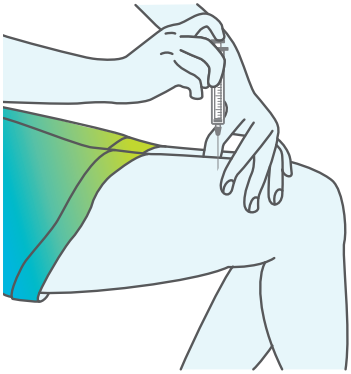


2. Tip the vial to ensure that the required amount of solution can be drawn into the syringe. NOTE: The required amount depends on the dose to be administered. Your healthcare provider will instruct you on the right amount for you.
3. Slowly pull the syringe plunger up to the correct mark (amount to be given as per healthcare provider's instructions), filling the syringe with Ilaris solution. If there are air bubbles in the syringe, remove bubbles as instructed by your healthcare provider. Ensure that the correct amount of solution is in the syringe.
4. Remove the syringe and withdrawal needle from the vial. (There may be solution remaining in the vial.) Recap the withdrawal needle as instructed by your healthcare provider or pharmacist. Remove the withdrawal needle from the syringe and place it in the sharps container.
5. Open the wrapper containing the injection needle and attach the needle to the syringe. Immediately proceed to administering the injection.

Giving the injection



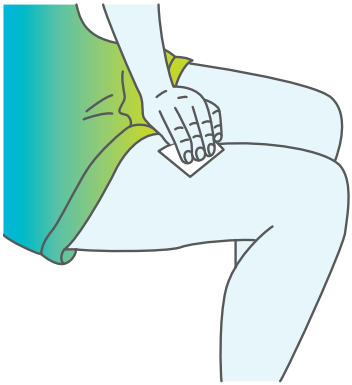
6. Choose an injection site on the upper thigh, abdomen, upper arm or buttocks. Do not use an area that has a rash or broken skin, or is bruised or lumpy. Do not inject into scar tissue as this may mean you do not get all of your medicine. Avoid injecting into a vein.
7. Clean the injection site with a new alcohol swab. Allow the area to dry. Uncap the injection needle.
8. Gently pinch the skin up at the injection site. Hold the syringe at a 90-degree angle and in a single, smooth motion, push the needle straight down completely into the skin.



9. Keep the needle all the way in the skin while slowly pushing the syringe plunger down until the barrel is empty. Release the pinched skin and pull the needle straight out. Dispose of the needle and syringe in the sharps container without recapping or removing the needle.

It is recommended to select a different injection site each time the product is injected to avoid soreness.

After the injection



10. Do not rub the injection area. If bleeding occurs, apply a clean, dry cotton swab over the area, and press gently for 1 to 2 minutes, or until bleeding stops. Then apply an adhesive plaster.

11. Safely dispose of needles and syringe in the sharps container or as directed by your healthcare provider or pharmacist. Never re-use syringes or needles.

12. Properly dispose of vials containing remaining Ilaris solution (if any) as directed by your healthcare provider or pharmacist. Any unused product or waste material should be disposed of in accordance with local requirements. Never re-use the leftover solution.

Keep the sharps container out of reach of children.

Dispose of the sharps container as directed by your healthcare provider or pharmacist.

Notes