

# Treatment Overview for Juvenile Idiopathic Arthritis

We understand that when a child or adolescent is diagnosed with juvenile idiopathic arthritis (JIA), the entire family is affected. At CHOC, we provide specialized JIA treatment and care so that rheumatic conditions don't put childhood on pause.

JIA is the most common type of arthritis in kids and teens. It usually causes joint pain and swelling in the hands, knees, ankles, elbows, and/or wrists but can affect other body parts too. In mild cases, the joint movement may decrease. In severe cases, there can be high fever, rash, and lymph node enlargement.

Although there is no cure for JIA, there are ways to keep it under control. The main goals of JIA treatment are:

- Slow down or stop inflammation
- Reduce pain and stiffness
- Prevent joint and organ damage
- Maintain joint function and movement
- Keep JIA in remission

## Drug Treatments

There are several different medications that can be used to help treat JIA.

- **Nonsteroidal anti-inflammatory drugs (NSAIDs)** to help decrease inflammation, reduce fever and relieve pain. Some common NSAIDs include ibuprofen (Advil, Motrin) and naproxen (Naprosyn, Aleve).
- **Disease Modifying Anti-Rheumatic Drugs (DMARDs)** are slow acting anti-inflammatory drugs. They tend to be stronger than NSAIDs and have more potential for side effects. They help slow the progression of JIA by suppressing the immune system so it does not attack the joints. DMARDs may include methotrexate, leflunomide (Arava) and sulfasalazine.
- **Corticosteroids**, or steroids, are anti-inflammatory drugs. They are used only for a short amount of time to help treat an acute inflammation.
- **Biological agents** are a newer class of drugs that are designed to target very specific steps in your body that causes inflammation. They can prevent and stop the progression of joint damage. On the next page are the biological agents we carry.



## Specialty JIA Medications at CHOC

### **Abatacept (ORENCIA®)**

Medication guide: [https://packageinserts.bms.com/ppi/ppi\\_orencia.pdf](https://packageinserts.bms.com/ppi/ppi_orencia.pdf)

Orencia comes in 3 different formulations: intravenous (IV) infusion, prefilled syringe and an autoinjector.

#### ORENCIA® IV Infusion

This is given through a vein in your arm by a healthcare professional. It is approved for patients 6 years and older. It is given once a month and usually takes about 30 minutes.

#### ORENCIA® Prefilled Syringe

Medication guide: [www.orencia.com/servlet/servlet.FileDownload?file=00P1Y000010f0bTUAQ](http://www.orencia.com/servlet/servlet.FileDownload?file=00P1Y000010f0bTUAQ)

This is an injection you give yourself subcutaneously (under the skin). It is approved for patients 2 years and older. It is given once a week.

How to give medication:

1. Remove one prefilled syringe from the refrigerator 30 minutes prior to giving to allow it to warm up to room temperature.
2. Wash your hands before injecting Orencia.
3. Clean the skin at and around the injection site
  - a. Stomach (avoid 2 inches around the navel)
  - b. Front of thighs
  - c. Outer area of upper arm (if your caregiver is giving you your dose)
4. With your free hand, gently grasp or pinch a large area of the skin at the injection site and hold firmly for the entire injection procedure.
5. Insert the needle into the pinched skin at 45° angle. Push the plunger with your thumb as far as it will go.
6. Slowly lift your thumb from the plunger to activate the needle guard. After a complete injection, the needle guard will cover the needle and you may hear a “click.”
7. You may notice a spot of blood at the injection site. You can press a cotton ball or a piece of gauze over the injection site.
  - **Do not** rub the injection site
8. Put your used prefilled syringe in an FDA-cleared sharps disposal container right away after use.
  - **Do not** throw away in household trash

#### ORENCIA® ClickeJet™ Autoinjector

This is an injection you give yourself under the skin. It is given once a week. It is approved for adults with rheumatoid arthritis or psoriatic arthritis.

### **Adalimumab (HUMIRA®)**

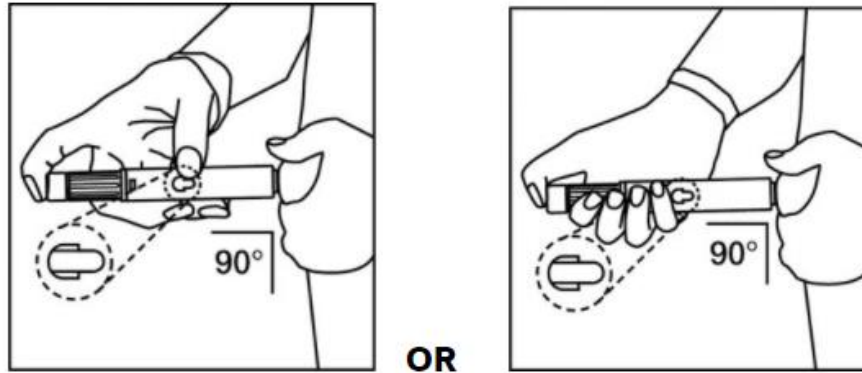
Medication guide: [www.rxabbvie.com/pdf/humira\\_medguide.pdf](http://www.rxabbvie.com/pdf/humira_medguide.pdf)

HUMIRA is a prescription medicine used alone, with methotrexate, or with certain other medicines to reduce the signs and symptoms of moderate to severe polyarticular juvenile idiopathic arthritis in children 2 years of age and older.

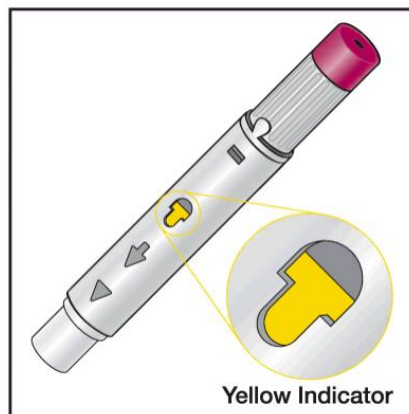
How to give medication:

1. Wash your hands before injecting Humira.
2. Clean the skin at and around the injection site.
  - a. Stomach (avoid 2 inches around the navel)
  - b. Front of thighs
3. With your free hand, gently grasp or pinch a large area of the skin at the injection site and hold firmly for the entire injection procedure.

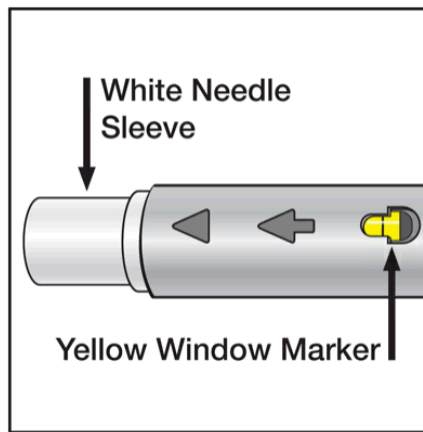
4. Position the white end of the pre-filled pen at a right angle (90 degrees) to the skin, so that you can see the window. One or more bubbles in the window is normal.
5. Holding the barrel of the pre-filled pen, press down slightly onto the injection site (holding in place without moving), but **do not** press the plum colored end until ready for injection.
6. With your index finger or your thumb, press the plum colored button on top once you are ready to begin the injection. You will hear a loud “click” as the needle is released, and your child will feel a small prick as the needle advances.



7. Keep pressing and continue to hold the pen with steady pressure for about 10 seconds to ensure a complete injection. **Do not** remove the pen while the injection is being given.
8. You will see a yellow indicator move into the window during the injection. The injection is complete when the yellow indicator stops moving.



9. Lift the pen straight up from the injection site. The white needle sleeve will move down over the needle and lock into place over the needle tip. **Do not** try to touch the needle. The white needle sleeve is there to protect you from touching the needle.



10. You may notice a spot of blood at the injection site. You can press a cotton ball or a piece of gauze over the injection site for 10 seconds.

- **Do not** rub the injection site

### **Canakinumab (ILARIS®)**

Medication guide: [www.novartis.us/sites/www.novartis.us/files/ilaris\\_ppi.pdf](http://www.novartis.us/sites/www.novartis.us/files/ilaris_ppi.pdf)

Ilaris is a subcutaneous injection indicated for children 2 years and older. It is given once every 4 weeks by your healthcare provider.

### **Etanercept (ENBREL®)**

Medication guide: [www.pi.amgen.com/~media/amgen/repositorysites/pi-amgen-com/enbrel/enbrel\\_mg.pdf](http://www.pi.amgen.com/~media/amgen/repositorysites/pi-amgen-com/enbrel/enbrel_mg.pdf)

Enbrel is indicated in children 2 years and older. It is given subcutaneously once a week. Enbrel is available with several different devices:

#### ENBREL Mini® Cartridge with AutoTouch® Autoinjector

Not approved for use in children.

#### SureClick® Autoinjector

Not approved for use in children.

#### Prefilled Syringe

Not approved for use in children.

#### Single-Dose Vial

Medication guide: [www.pi.amgen.com/~media/amgen/repositorysites/pi-amgen-com/enbrel/enbrel\\_sdvifu.pdf](http://www.pi.amgen.com/~media/amgen/repositorysites/pi-amgen-com/enbrel/enbrel_sdvifu.pdf)

This option is available for children 2 years and older and weigh less than 68 lbs. The vials come ready to use.

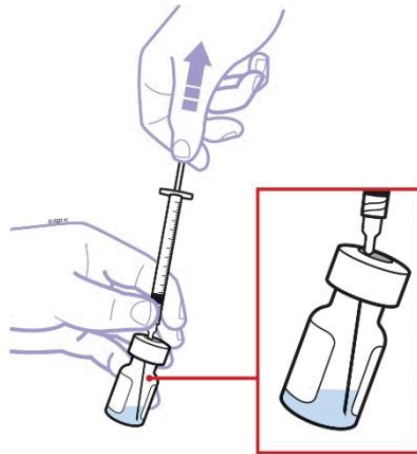
They contain no preservatives and are for **1-time use only**. Syringes and needles are required to give it.

Each vial contains 25 mg (0.5 mL).

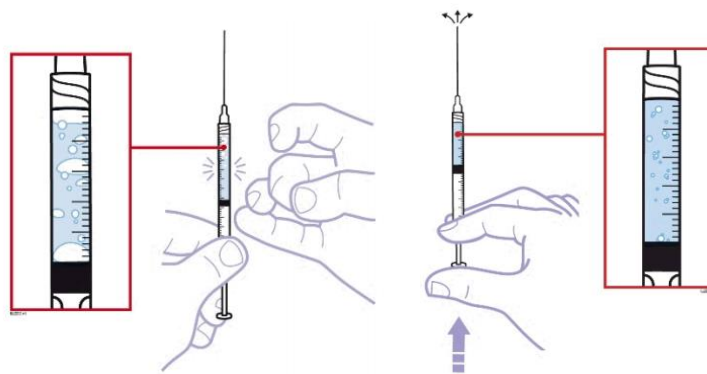
How to give medication:

1. Store Enbrel in the refrigerator between 36°F to 46°F (2°C to 8°C) in their original carton to protect from light. You may also store the Enbrel single-dose vials at room temperature between 68°F to 77°F (20°C to 25°C) for up to 14 days.
2. Remove the number of vials needed for one dose and leave at room temperature for at least 30 minutes.
  - Keep out of direct sunlight
  - Leave the green cap on until ready to give
3. Pop off the green vial cap and wipe gray stopper with an alcohol wipe.
  - If a second vial is needed to give the full dose, use a new alcohol wipe.
4. Attach a long needle to a syringe. Pull the syringe plunger back to 0.5 mL.

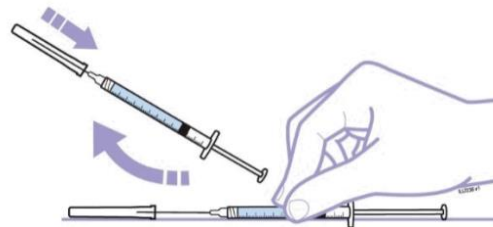
5. Hold the single-dose vial on a flat surface with 1 hand. Insert the long needle through the gray rubber stopper and slowly push 0.5 mL of air in the single-dose vial.
6. Tilt the single-dose vial to withdraw **all** medicine.



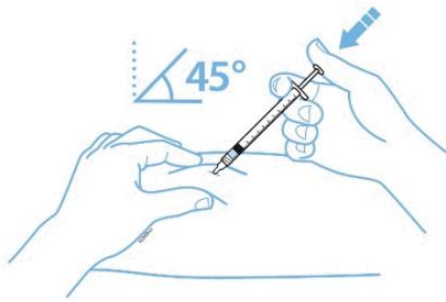
7. Remove the needle from the vial. With the needle point up, gently tap the syringe with fingers to release any air bubbles in the syringe. Push the plunger to push the air out of the syringe.



8. Push the plunger to your prescribed dose. If a second vial to make the full dose, repeat steps 5 through 7 with the same needle. Then, push the plunger to your prescribed dose.
9. Use the 1-handed scoop method to recap your needle.
  - Using 1 hand, slide the needle into the cap and scoop upwards to cap the needle. Then use your hand to secure the cap and snap into place.



10. Replace the long needle with a short needle.
11. Choose your injection site
  - Tops of thigh or stomach (avoid 2 inches around the navel)
  - Back of upper arms (if someone else is giving the injection)
12. Clean the injection site with an alcohol wipe and let dry.
13. Gently pinch skin. Insert the needle into your skin at a 45° angle and slowly push the plunger all the way down.



14. When the syringe is empty, remove the needle and syringe from your skin.
  - **Do not** recap needle.
  - **Do not** rub the injection site. If there is blood at the injection site, you can press a cotton ball over the injection site until bleeding has stopped. Apply an adhesive bandage if needed.
15. Dispose vial, syringe and needles in a sharps disposal container.

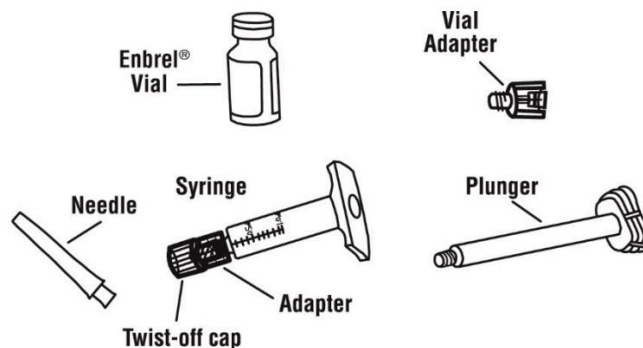
#### Multiple-Dose Vial

Medication guide: [www.pi.amgen.com/~media/amgen/repositorysites/pi-amgen-com/enbrel/enbrel\\_muvifu.ashx](http://www.pi.amgen.com/~media/amgen/repositorysites/pi-amgen-com/enbrel/enbrel_muvifu.ashx)

This option is for children who weigh less than 68 lbs. One vial contains multiple doses. There are two ways you can use to inject the medication: vial adapter method or the free-hand method. Each vial contains 25 mg and will need to be mixed with a diluent (liquid) before giving.

How to give medication:

1. Store vials in the refrigerator at 36°F to 46°F (2°C to 8°C) in the original carton to protect from light. If needed, you may store the vials and diluent syringe (dose tray) at room temperature between 68°F to 77°F (20°C to 25°C) for up to 14 days.
  - a. Once Enbrel multiple-dose vial has reached room temperature, **do not** put it back in the refrigerator.
  - b. Once the vial is mixed with the diluent the vial should be used right away or kept in the refrigerator for up to 14 days.
2. Take the Enbrel dose tray out of the refrigerator. Check to make sure the tray has:
  - One prefilled diluent syringe containing 1 mL of diluent (liquid) with attached adapted and twist-off cap
  - One plunger
  - One Enbrel vial
  - One 27 gauge ½ inch needle
  - One vial adapter



3. Leave the dose tray at room temperature for about 15 to 30 minutes before injecting to make for a more comfortable injection.
4. With clean hands, peel the paper seal off the dose tray and remove all items.

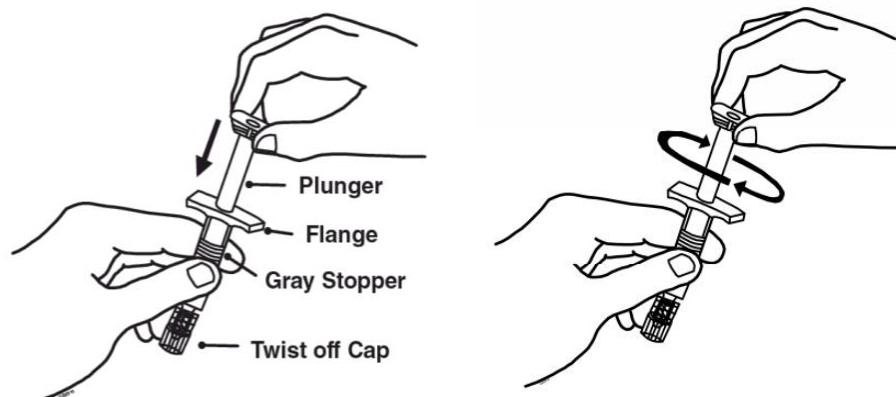


*There are 2 methods to giving the injection: vial adapter method and the free hand method.*

### **Vial adapter method**

Note: This method **should not** be used if you will be using the vial more than once.

1. Remove the pink plastic cap from the vial. **Do not** remove the gray stopper or silver metal ring around the top of the vial. Wipe the gray stopper with an alcohol wipe.
2. Open the wrapper containing the 27 gauge needle and put it aside for later use.
3. Open the wrapper that contains the vial adapter and set aside for later use. **Do not** touch the vial adapter's twist-on end or the spike inside.
4. Slide the plunger into the flange end of the syringe. Then attach the plunger to the gray rubber stopper in the syringe by turning the plunger clockwise until you feel slight resistance.



5. Remove the twist-off cap from the prefilled diluent syringe by turning counter-clockwise. Then, twist the vial adapter onto the syringe, turning clockwise until you feel a slight resistance. **Do not overtighten.**
6. Hold the vial upright. Grasp the sides of the vial adapter and place it over the top of the vial. Insert the vial adapter into the gray stopper so the plastic spike punctures into the vial.
7. Push the plunger down until all the liquid is in the vial. You may see bubbles in the vial.
8. Gently swirl (**do not** shake) the vial in circular motions to dissolve the powder. This usually takes less than 10 minutes. The solution should be clear and colorless, but bubbles may still be there.
9. Turn the vial upside down and slowly pull the plunger back to draw up your dose. Be careful not to pull the plunger completely out of the syringe.
10. Gently tap the syringe to move the air bubbles to the top of the syringe. Slowly push the plunger to remove the air bubbles.
11. Twist off the syringe from the vial adapter by turning the syringe counter-clockwise.
12. Twist on the 27 gauge needle onto the syringe

### **Free hand method**

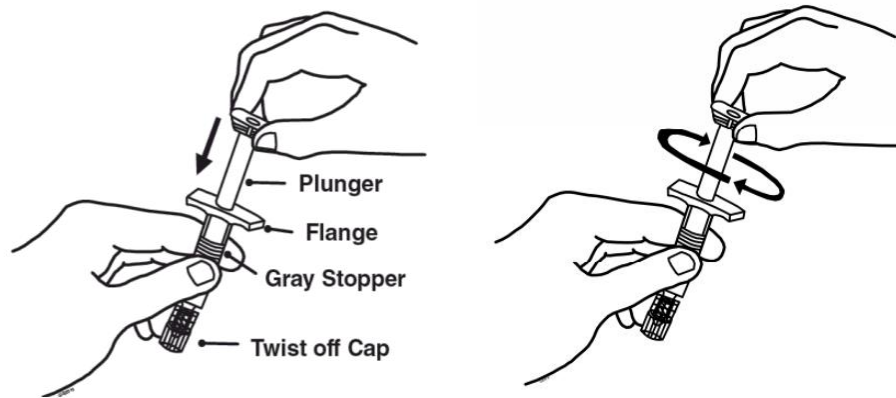
Notes:

- If using the vial for more than one dose, write the date you mixed the powder and liquid on the sticker marked "Mixing Date" and stick the sticker onto the vial.
- After withdrawing the dose from the vial store the vial in the refrigerator as soon as possible (but always within 4 hours of mixing the solution). Vial must be used within 14 days of the mixing date.
- **Do not** mix any remaining liquid in one vial with another.

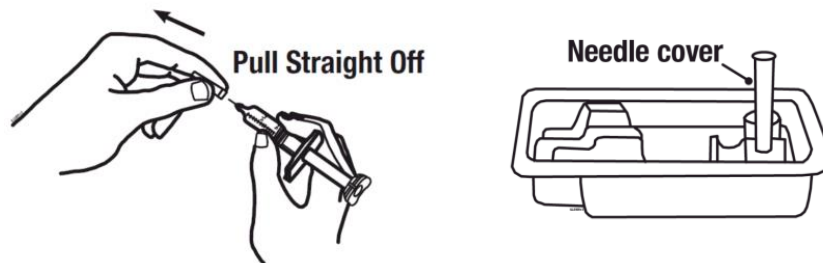
How to give medication:

1. Remove the pink plastic cap from the vial. **Do not** remove the gray stopper or silver metal ring around the top of the vial. Write the date you mix the powder and solution on the "Mixing Date" sticker and stick it on the vial.
2. Wipe the gray stopper with an alcohol wipe.
3. Open the wrapper containing the 25 gauge needle and put it aside for later use.

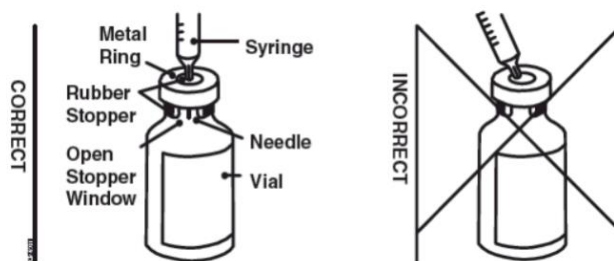
- Slide the plunger into the flange end of the syringe. Then, attach the plunger to the gray rubber stopper in the syringe by turning the plunger clockwise until you feel slight resistance.



- Remove the twist-off cap from the prefilled diluent syringe by turning counter-clockwise.
- Continue to hold the barrel of the syringe. With your free hand twist on the 25 gauge needle to the tip of the syringe until it fits snugly. Place the syringe on a flat work surface.
- Open the wrapper containing the 27 gauge needle. Set aside for later use.
- Pick up the syringe and hold the barrel of the syringe with one hand and pull the needle cover straight off. Avoid touching the needle with the cover to avoid damage to the needle. Place the needle cover with the open side up in the round space marked "N" in the dose tray.



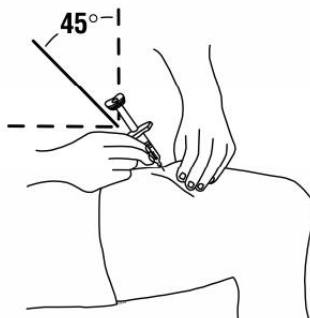
- Hold the syringe with the needle facing up and pull back the plunger to pull a small amount of air into the syringe. Then insert the needle straight down through the center ring of the gray stopper.



- Push the plunger down slowly until all the liquid in the syringe is in the vial. Leave the syringe in place and gently swirl the vial in a circular motion to dissolve the powder (usually takes less than 10 minutes).
  - Do not shake.**
- Turn the vial upside down and pull the plunger back to draw up your dose.
- Gently tap the syringe so any air bubbles rise to the top and slowly push the plunger up to remove air bubbles.
- Remove syringe and needle from vial. Keep the needle attached to the syringe and insert the needle into the needle cover that was placed in the dose tray. You should hear a "snap" once the needle cover is securely on the needle. Then untwist the needle from the syringe and dispose the needle in a sharps container.
- Twist on the 27 gauge needle onto the syringe.



15. If there is enough solution left the vial for another dose, refrigerate the vial. If there is not enough to get a full dose out of the vial, discard the vial.
16. Once you have your dose drawn up, choose your injection site and wipe it with an alcohol swab
  - a. Front of the middle thighs
  - b. Stomach area (avoid 2 inches around navel)
  - c. Outer area of the upper arms if someone else is giving you the injection
17. Pull the cover straight off the needle. With one hand, gently pinch the area of skin and hold it firmly. With the other hand hold the syringe and insert the needle into the skin at a 45° angle.



18. When the needle is completely in, let go of the skin and hold the base of the syringe to stabilize it. Then push the plunger at a slow, steady rate until all the medicine is injected.
19. Pull the needle out of the skin and dispose the needle and syringe in a sharps container.
  - **Do not** rub the injection site. If there is blood at the injection site, you can press a cotton ball over the injection site until bleeding has stopped. Apply an adhesive bandage if needed.

### **Tocilizumab (ACTEMRA®)**

Medication guide: [www.gene.com/download/pdf/actemra\\_medguide.pdf](http://www.gene.com/download/pdf/actemra_medguide.pdf)

Actemra can be given as in intravenous (IV) infusion or a subcutaneous injection. As an IV infusion, it is given every 2 weeks for systemic juvenile idiopathic arthritis (SJIA), or every 4 weeks for polyarticular juvenile idiopathic arthritis (PJIA). Actemra as an IV infusion is given by a healthcare professional and takes about 1 hour. As a subcutaneous injection, this medication comes in a single dosed prefilled syringe or in single dose autoinjector. For SJIA, this medicine is given once a week or once every 2 weeks. For PJIA this medicine is given every 2 weeks or once every 3 weeks. Dose is based on weight but may change depending on response to treatment. Syringes need to be kept in the refrigerator at 2°C to 8°C (36°F to 46°F)

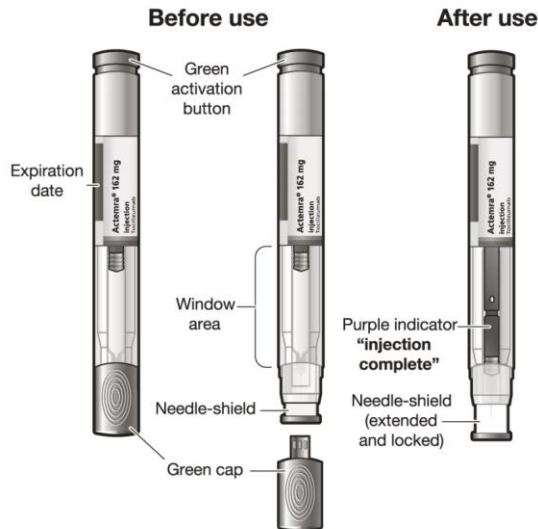
How to give medication: [www.gene.com/download/pdf/actemra\\_ifu.pdf](http://www.gene.com/download/pdf/actemra_ifu.pdf)

#### Single-dose Prefilled Syringe

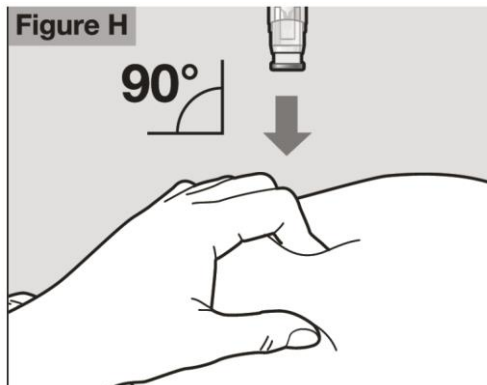
1. Remove 1 single use prefilled syringe from the refrigerator and let it warm for 30 minutes to reach room temperature.
2. Clean the injection site with an alcohol swab
  - a. Front of your thighs
  - b. Stomach area (avoid 2 inches around the navel)
  - c. Outer area of the upper arms if someone is giving you the injection
3. Hold the prefilled syringe with one hand and pull the needle cap off with your other hand.
4. There may be small bubbles in the prefilled syringe. You do not need to remove it.
5. Use the prefilled syringe within 5 minutes of needle cap removal, otherwise the syringe should be thrown away in a sharps container and a new syringe should be used.
6. Pinch the area of the skin you cleaned with one hand. With the other hand insert the needle into the skin at an angle between 45° and 90°.
7. Keep the syringe in position and let go of the pinched skin.
8. Slowly push the plunger down to inject all the medicine.

9. Keep pressing down on the plunger while you take the needle out of the skin. Then release the plunger to allow the needle shield to cover the needle.
- **Do not** rub the injection site. If there is blood at the injection site, you can press a cotton ball over the injection site until bleeding has stopped.
- Apply an adhesive bandage if needed.

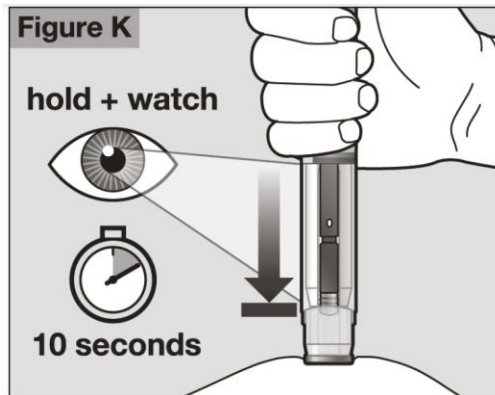
Single-dose Prefilled Autoinjector (ACTPen®)



1. Take one autoinjector from the refrigerator and let it sit for 45 minutes to allow it to reach room temperature.
2. Clean the injection site with an alcohol swab
  - a. Front of your thighs
  - b. Stomach area (avoid 2 inches around the navel)
- c. Outer area of the upper arms if someone is giving you the injection
3. Twist and pull off the green cap. Once the cap is off the autoinjector should be used within 3 minutes. If it has been more than 3 minutes, the autoinjector should be disposed of and a new one should be used.
4. Pinch the area of skin you cleaned with one hand. Then place the needle shield of the autoinjector against the skin at a 90° angle. Continue to keep the skin gently pinched throughout the injection.



5. Press the autoinjector firmly against the skin until the needle shield is completely pushed in. This unlocks the green activation button.
6. Press the green activation button to start the injection. You should hear a “click” sound. Continue pressing the green button and continue holding the autoinjector firmly against your skin.
7. The purple indicator will move along the window area during the injection. Once it stops moving the full dose is given. This may take up to 10 seconds.



8. When the purple indicator stops moving, release the green button. Lift the autoinjector straight off the injection site at a 90° angle to remove the needle from the skin.
  - **Do not** rub the injection site. If there is blood at the injection site, you can press a cotton ball over the injection site until bleeding has stopped.
  - Apply an adhesive bandage if needed.

### Tofacitinib (XELJANZ® tablets and solution)

Medication guide: <http://labeling.pfizer.com/ShowLabeling.aspx?id=959&section=MedGuide>

This medicine is approved in children 2 years and older. It comes in two forms:

- **The solution** is 1 mg/mL and is grape flavored. It is clear and colorless. The medicine is taken 2 times a day with or without food. The solution should be stored at room temperature between 20°C to 25°C (68°F to 77°F). Always keep Xeljanz in its original bottle to protect it from light.
- **The tablet** is a white pill that is taken two times a day with or without food.

### Patient Resources

CHOC Pediatric Rheumatology: [www.choc.org/programs-services/rheumatology/](http://www.choc.org/programs-services/rheumatology/)

Arthritis Foundation: [www.arthritis.org/diseases/juvenile-arthritis](http://www.arthritis.org/diseases/juvenile-arthritis)

KidsHealth: <https://kidshealth.org/en/parents/jra.html>