

You, Your Child, and ^{Pr}ACTEMRA[®] (tocilizumab)



*A guide to polyarticular and systemic
juvenile idiopathic arthritis (JIA)*

 **ACTEMRA[®]**
tocilizumab

Introduction

How to use this workbook

This workbook was developed mainly for the parents (or the caregivers) of children and teenagers who have polyarticular juvenile idiopathic arthritis (pJIA) or systemic juvenile idiopathic arthritis (sJIA).

The goals of the workbook are to:

- Help you and your child better understand JIA
- Provide a guide to treatment with ACTEMRA
- Help you and your child understand the benefits and risks associated with ACTEMRA therapy
- Offer tips on caring for a child with pJIA or sJIA
- Provide a tool (using stickers) to help you keep track of appointments with your child's doctor, for blood tests and infusion days
- Supply a list of online resources where you can find more information on pJIA and sJIA

My child has been diagnosed with (your child's doctor will check one):

☐ **pJIA** (see pages 3-19)

☐ **sJIA** (see pages 21-35)

My child's pediatric rheumatologist is:

Name: _____

Phone: _____

Address: _____

pJIA: An Overview

What is pJIA?

pJIA (or polyarticular juvenile idiopathic arthritis) is a subtype of juvenile idiopathic arthritis (JIA). This is a form of arthritis that affects children (16 years old or younger).

The word 'arthritis' comes from the Greek words 'arthron' and 'itis'.

arthron = joint

itis = inflammation

inflammation = swelling and redness, hot and often painful

Polyarticular means that many joints are inflamed. Children with pJIA have 5 or more inflamed joints within the first 6 months of having the condition.

Idiopathic means that the cause of the condition is unknown.

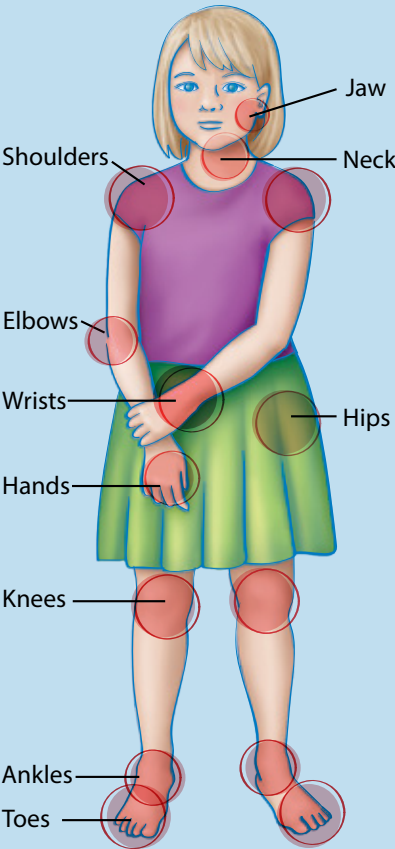
What we do know is that the inflammation is related to the body's immune system attacking healthy tissues, particularly the joints. But we just don't know why.



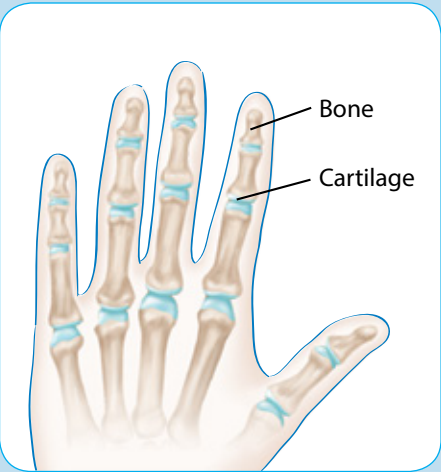
pJIA: An Overview (cont'd.)



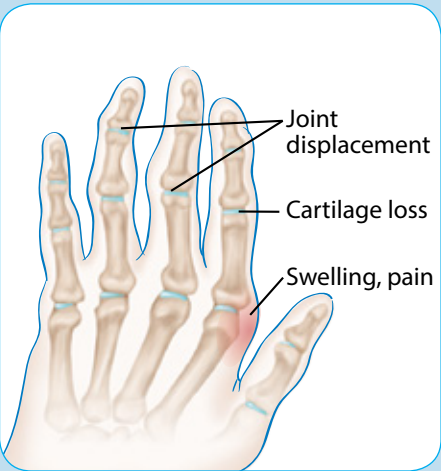
Joints that may be affected in pJIA



Healthy finger joints



Finger joints affected by pJIA



Types of pJIA and their symptoms

A protein called rheumatoid factor (RF) is found in the blood of some children with pJIA, but not others. This difference leads to two different types of pJIA.

Children with **RF-negative pJIA** do not have RF in their blood.

- This kind of pJIA can start at any age
- Symptoms may include:
 - o Joint inflammation and swelling
 - o Joint stiffness
 - o Joint pain
 - o Growth failure

Children with **RF-positive pJIA** do have RF in their blood.

- This kind of pJIA usually appears in children 10 years of age or older
- Symptoms may include:
 - o Joint inflammation and swelling
 - o Joint stiffness
 - o Joint pain
 - o Growth failure
 - o Anemia (low red blood cell count)
 - o Fatigue (tiredness), poor appetite and general unwell feeling
 - o Fever (rare)
 - o Rheumatoid nodules or bumps under the skin (rare)

How is pJIA diagnosed?

The cause of pJIA is unknown and the symptoms can be part of other illnesses such as infections. Your doctor needs to ensure that something else is not the cause of your child's joint pain and swelling.

This is why your doctor has completed a full evaluation of your child. This often takes some time. Once your doctor is sure that it is JIA, then he/she needs to determine what type. Unfortunately, there is not one single test to diagnose pJIA.

Some of the things your doctor has been looking for include:

- ✓ A complete history – past conditions, medications, tests, arthritis in the family, etc.
- ✓ Complete physical exam
- ✓ Current physical symptoms
- ✓ Changes or markers in the blood (blood tests) or urine.
A blood test will help tell whether your child has RF-negative or RF-positive pJIA
- ✓ Changes in bones, joints and/or organs (X-rays and related imaging studies)

How is pJIA treated and managed?

There are a number of different types of therapies that are used to help treat and manage pJIA.

Generally, they fall into the following categories:

- Medications
- Physical therapies (e.g., exercise, physiotherapy, and occupational therapy)

Treating pJIA with ACTEMRA

ACTEMRA, also known as tocilizumab, is a drug used to treat patients with active systemic juvenile idiopathic arthritis (sJIA) and polyarticular juvenile idiopathic arthritis (pJIA) ages 2 and above.

How it works

ACTEMRA is one of a group of medicines called biologic therapies.

ACTEMRA helps keep the immune system from attacking healthy tissues in the body. Normally, the immune system leaves healthy body tissues alone. In people with certain types of arthritis (e.g., pJIA), the immune system attacks normal body tissues causing damage and inflammation, especially in the tissues of the joints.

ACTEMRA interferes with an important step in this attack by blocking a cytokine called interleukin-6 (IL-6). IL-6 is a protein that is made by the immune system and the body uses it to manage infections.

By decreasing the immune system's attack on normal tissues, ACTEMRA can reduce pain, joint inflammation and tiredness.



Important Side Effect Information

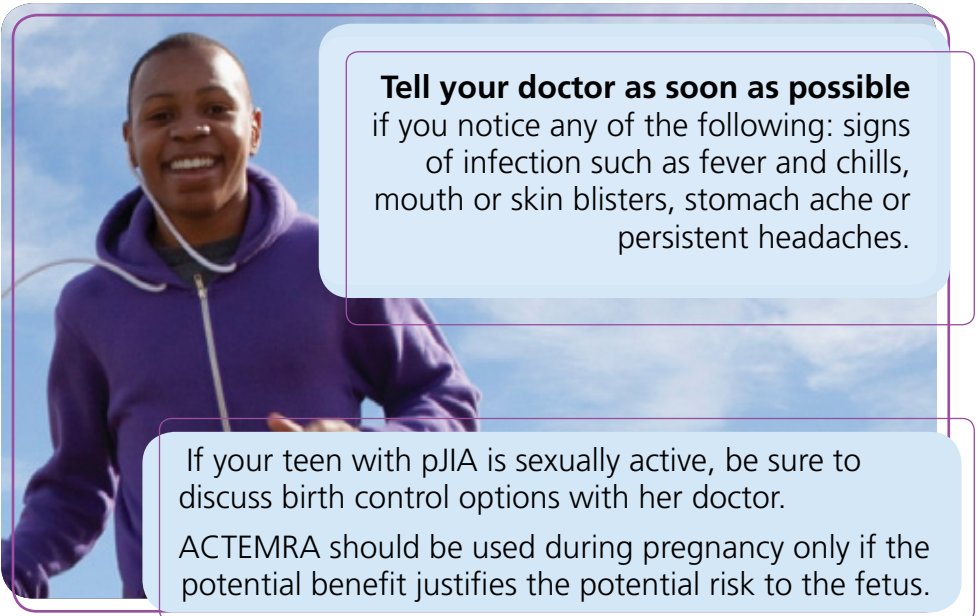
In pJIA, the most common side effects of ACTEMRA are upper respiratory tract infections, nausea, headache, dizziness, decrease in blood pressure and rash. Patients receiving ACTEMRA subcutaneously also experienced reactions at the injection site.

Possible serious side effects include serious infections, liver injury and allergic reactions. A severe skin reaction called Stevens-Johnson syndrome (SJS) and serious drug-induced liver injury, including rapid loss of liver function, inflammation of the liver and jaundice (yellowing of skin and eyes) were reported during treatment with ACTEMRA.

Since this medicine can cause dizziness, it is recommended that you do not drive or use machines until the dizziness has stopped.

Stop taking ACTEMRA and call your doctor or seek medical attention immediately if you notice any of the following:

- ✓ Difficulty with breathing, or light-headedness
- ✓ Rash, itching, hives, swelling of the lips, or other signs of an allergic reaction
- ✓ Chest pain
- ✓ Feeling dizzy or faint
- ✓ Yellowing of the skin and eyes, dark brown coloured urine, pain or swelling in the upper right side of the stomach area, or you feel very tired or confused

A young woman with short dark hair, wearing a purple hoodie, is smiling and looking towards the camera. She is holding a white stethoscope around her neck. The background is a bright blue sky with some clouds.

Tell your doctor as soon as possible if you notice any of the following: signs of infection such as fever and chills, mouth or skin blisters, stomach ache or persistent headaches.

If your teen with pJIA is sexually active, be sure to discuss birth control options with her doctor.

ACTEMRA should be used during pregnancy only if the potential benefit justifies the potential risk to the fetus.

Serious Warnings and Precautions

Serious Warnings and Precautions

Serious infections: Some serious infections have been observed with the use of ACTEMRA. These infections include: active tuberculosis (TB), bacterial, viral and fungal infections. Most patients who developed these infections were taking other drugs that lower the immune system. Hospitalization or death associated with these infections have been reported. Ensure you tell your doctor if you are taking any other medication.

Hepatotoxicity: Serious cases of drug-induced liver injury (DILI) have been observed in patients treated with ACTEMRA. Some of these cases have resulted in acute liver failure requiring a liver transplant. Treatment with ACTEMRA should not be initiated in patients with active infections or active liver disease.

Gastrointestinal perforations: GI perforations (holes in the lining of the gut) have been reported uncommonly, usually as a complication of diverticulitis (infection of the large intestine), and require immediate medical attention. If you develop fever and severe stomach pain that does not go away, seek medical attention.

Abnormalities of your lab results: Your physician will be monitoring your blood work and may notice that your levels are either too high or too low; your healthcare provider may stop your ACTEMRA treatment for a period of time or change your dose of medicine if needed because of changes in these blood test results.

Malignancies: During the clinical trials, cases of cancer have been reported very rarely in patients receiving ACTEMRA. The current number of reported cases in the ACTEMRA studies appears to be consistent with the expected number of cancer cases reported in the RA population. The role of treatment with ACTEMRA on the development of cancers is not known.

Demyelinating disorders: The impact of ACTEMRA on demyelinating disorders is not known, but multiple sclerosis and chronic inflammatory demyelinating polyneuropathy were reported rarely in clinical trials.

Allergic reactions: Serious allergic reactions, including death can happen with ACTEMRA. These reactions can occur on the first infusion, even if you have taken the premedication and can happen with future infusions of ACTEMRA. Tell your healthcare provider if you have any of the following signs of a serious allergic reaction: shortness of breath or trouble breathing, skin rash, swelling of the lips, tongue, or face, chest pain, feeling dizzy or faint.

Treatment with ACTEMRA should not be initiated in patients with active infections or active liver disease.

Taking ACTEMRA for pJIA

ACTEMRA comes in two forms. ACTEMRA is supplied as a solution for intravenous infusion (IV) or as a solution for subcutaneous injection (SC) in either a single-use pre-filled syringe, or a single use autoinjector.

Before starting treatment, make sure your doctor knows if you are taking or have recently taken any other medicines.

Note that ACTEMRA should not be taken with biological medicines for the treatment of rheumatoid arthritis (including Enbrel®, Humira®, Remicade®, RITUXAN® (rituximab), Orencia®, Kineret®, Simponi™ and Cimzia™) as it has not been studied in combination with these medicines.

If a decision has not yet been made regarding the route of administration for ACTEMRA, talk to your doctor about which option may be best for your child.



If your child will be receiving subcutaneous injections, continue reading on page 13.

ACTEMRA by Intravenous Infusion for pJIA

Preparing Your Child

Vaccinations → Be sure your child is up to date (if possible) on all recommended vaccinations, *prior to initiation of therapy* with ACTEMRA. Certain vaccines should not be given *while* receiving ACTEMRA.

Infections → Be sure your child does not have any existing infections prior to receiving ACTEMRA. Treatment with ACTEMRA could cause your child's infection to get worse.

Plenty of water → To help with IV access, be sure your child is well hydrated, but also make sure he/she goes to the bathroom before the infusion. Once started, the infusion takes about an hour.

Comfortable, loose fitting clothes → Be sure your child is comfortable, as he/she will be sitting for about an hour. Also, loose fitting clothes allow the medical staff to monitor your child's vital signs (e.g., blood pressure, heart rate, etc.) easily.



Layers → Dress your child in layers to allow for temperature control. The infusion centre may be cool or warm; or the infusion itself may make your child feel too warm or too cold. Layers of clothing allow your child to control his/her comfort.

Contact the infusion centre about what they provide for patient comfort, e.g., blankets, pillows, drinks, etc.

Be sure your child is not wearing any fragrance or perfume, as other patients may be allergic.

Bring a complete list of current medications, allergies, and emergency contact information for the infusion staff to add to your child's chart.

ACTEMRA by Intravenous Infusion for pJIA

Receiving the Infusion

ACTEMRA will be given to your child by a healthcare professional using an intravenous line. This means the medicine will be given to your child through a needle placed in a vein in his/her arm. It will take about 1 hour to give your child the full dose of medicine.

The recommended dose for children with pJIA is either 8 or 10 mg per kg of body weight depending on the child's weight. Children receive a dose of ACTEMRA every 4 weeks.

In the event of a missed dose, ask your child's doctor when to schedule the next dose.

During the Infusion

Since the infusion takes approximately an hour, it's a good idea to bring a few things to keep your child busy during this time.

Below is a list of some of the things your child might enjoy doing during the infusion:

- ☐ Read a book or comic/magazine
- ☐ Do a crossword puzzle or play a sudoku game
- ☐ Play cards
- ☐ Play hand-held games (if they are quiet or have headphones)
- ☐ Listen to music (with headphones)
- ☐ Bring a laptop computer or tablet to watch a movie, or play games (don't forget the headphones)
- ☐ Have snacks and a drink (if allowed in the infusion room)
- ☐ Draw or doodle
- ☐ Take a nap



Your child will be monitored for allergic reactions during the infusion. These signs may include shortness of breath or trouble breathing, skin rash, swelling of the lips, tongue or face, chest pain, feeling dizzy or faint.

Continue on page 14 if taking ACTEMRA by intravenous infusion

ACTEMRA by Subcutaneous Injections for pJIA

“Subcutaneous” means that it is given into the fat layer just under the skin. The recommended injection sites are the abdomen, thigh and upper arm. The sites should be rotated and injections should never be given into moles, scars, or areas that are tender, bruised, red, hard or have open sores.

Your healthcare provider should show you how to prepare and inject properly before using ACTEMRA subcutaneously for the first time. Ask your healthcare provider any questions you may have. Do not attempt to administer an injection until you are sure you understand how to inject using the pre-filled syringe or a single-use autoinjector. It is important that you follow the specific instructions for using the kind of ACTEMRA that your doctor has prescribed.

The ACTEMRA syringe or autoinjector are intended to be used by patients or caregivers who have been properly trained.

The recommended dose for children with pJIA is 162 mg either every two or three weeks depending on the child’s weight.

Keep the ACTEMRA pre-filled syringe or autoinjector and all medicines out of the reach and sight of children. Always store the them in a refrigerator at a temperature of 2–8 °C. Protect the syringe or autoinjector from freezing and from light.

Vaccinations → Be sure your child is up to date (if possible) on all recommended vaccinations, *prior to initiation of therapy with ACTEMRA*. Certain vaccines should not be given *while* receiving ACTEMRA.

Infections → Be sure your child does not have any existing infections prior to receiving ACTEMRA. Treatment with ACTEMRA could cause your child’s infection to get worse.

After taking ACTEMRA

Unwanted effects are possible with all medicines. Tell your child's doctor or nurse as soon as possible if your child does not feel well while receiving treatment with ACTEMRA.

Possible side effects

Common (at least 1 in 100 people)

- Upper respiratory tract infections like coughs and cold, pneumonia, skin infection
- Cold sores (oral herpes simplex), blisters, shingles (herpes zoster), skin infection sometimes with fever and chills
- Low white blood cell counts shown by blood tests
- High blood fats (cholesterol levels)
- Headache, dizziness
- High blood pressure
- Mouth ulceration
- Stomach pain
- Abnormal liver function tests
- Rash and itching
- Pneumonia
- Cellulitis
- Injection site reactions (with subcutaneous use)

Be sure to ask for any important post-infusion or post-injection instructions.

Uncommon (at least 1 in 1000 people)

- Diverticulitis (fever, nausea, diarrhea, constipation, stomach pain)
- Red swollen (inflamed) areas in the mouth
- High blood fat (triglyceride levels)
- Serious allergic reactions
- Pancreatitis (stomach pain, back pain, nausea, vomiting)
- Lung disease (shortness of breath, trouble breathing, cough)

Rare (less than 1 in 1,000 patients)

- Multiple Sclerosis (including blurred vision, loss of vision, eye pain, feeling dizzy, or numbness, weakness or tingling in the face, arms or legs)
- Drug-induced liver injury (loss of appetite, nausea and vomiting, fatigue, itching, dark urine, confusion, abdominal swelling and/or pain in the upper-right side of the stomach); Jaundice (yellowing of skin and eyes)

If any of these uncommon or rare side effects occur, stop taking the drug and call your doctor or pharmacist.

Blood Work and Other Tests

Below are descriptions of some of the tests your child may undergo, either at the time of diagnosis or as follow-up tests to monitor disease activity and check for side effects.

Blood tests

Complete blood cell count (CBC): This test measures

- Amounts of each type of blood cell in a sample of blood
- Level of hemoglobin in the blood (low hemoglobin = anemia)
- Abnormalities in the numbers of various kinds of white blood cells or of platelets

Erythrocyte sedimentation rate (ESR): This test does not point specifically to pJIA but indicates active inflammation in the body. These tests are almost always elevated in children with active pJIA.

C-reactive protein (CRP): This test is another measure of the level of inflammation in the body.

Rheumatoid factor (RF): This test identifies RF, a type of protein in the bloodstream. It's used to tell what type of pJIA your child may have. Only 5% of children will test positive for RF.

Liver and kidney function: These tests may be done as part of a general health screen.

Cholesterol: Your child's blood cholesterol levels may be monitored, as higher cholesterol levels are a common possible side effect of ACTEMRA therapy.

Follow-up imaging

X-rays: X-rays give information about whether or not damage has occurred in the joint.

MRI: MRI is similar to X-ray but is much more detailed and gives a better 3-dimensional view of the joint.

Bone scan: A bone scan may be necessary if the results of the workup do not support the diagnosis. A bone scan can detect inflammation in the bone and other abnormalities that do not show up well on X-ray.

Please note that other tests may be ordered at the discretion of your child's doctor.

Coping with pJIA

Family members can help your child cope with pJIA by doing the following:

- Keeping a positive outlook
- Learning as much as they can about your child's disease and its treatment
- Treating your child as normally as possible
- Trying to get the best care possible
- Working closely with your child's healthcare team
- Joining a support group
- Encouraging exercise and physical therapy for your child if necessary
- Working closely with your child's school
- Talking openly with your child

Talking to your child about pJIA

Your attitude toward pJIA can affect how your child feels about it.

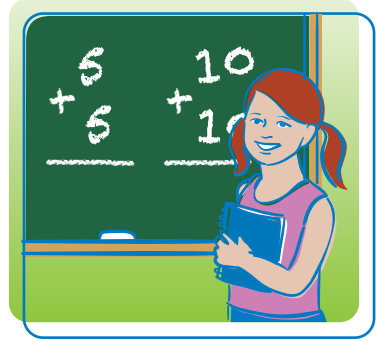
Below are some tips on communicating with your child about pJIA.

- If your child is angry, allow him/her to express his/her anger about arthritis.
- Expect your child to behave as you would any other child and expect the same responsibilities (so long as he/she is physically able).
- Avoid giving him/her special attention or treatment.
- Encourage your child to learn as much as possible about pJIA and its treatment.
- Try not to overprotect your child.
- Being as consistent as possible will help your child learn what is expected.

Be sure of your child's rights. There may be services available at the provincial or local level for children with disabilities like pJIA. Find out what services are available in your area.

Your Child at School

Depending on your child's individual needs, you may want to contact your child's school to meet with your child's teachers, the principal and/or guidance counsellors and help them learn about pJIA. Where possible, you and the school should work together to come up with creative ways to deal with your child's situation.



If your child is experiencing increased arthritis symptoms there are many ways to modify activities and schoolwork. Below is a list of some of the ways adjustments can be made to help your child at school.

- Have two sets of schoolbooks (texts), one for home and one for school.
- Give extra time to change classes.
- See if your child's classes can be close to each other to minimize walking and stair climbing.
- Arrange for activity/stretch breaks.
- Provide a larger pencil or pen or get a foam grip to help with handwriting.
- Depending on other symptoms, your child may need a different desk or chair.
- Timed written tests may need to be changed or extended.

Ask your child's physical or occupational therapist for other ideas.

Be sure to talk to your child's teacher about missing school for infusion appointments and make arrangements for catching up on any missed schoolwork or special assignments.

Other Helpful Tips

Exercise

Physical activity is very important for children with pJIA, although there may be times that activity is limited due to pain.

A well-balanced exercise program will help maintain muscle tone and range of motion and function in the joints. Ask your doctor to recommend a physical therapist to develop an appropriate exercise plan for your child.

Tip: Swimming is an excellent way to use joints and muscles without putting weight on the joints.

Tip: Warm baths or an electric blanket may help soothe sore joints.

Nutrition

Good nutrition can help fight the effects of pJIA. So it is important that your child gets the nutrients he or she needs for proper growth and development. These include protein, carbohydrate, fat, vitamins, and minerals.

Your child can eat a variety of foods as long as his or her weekly intake is balanced and varied.

Ask your doctor whether you should be supplementing your child's diet with any vitamins or minerals. For example, calcium and vitamin D are important for strong bones.



pJIA: Resources and Web

For more information about pJIA and its care, visit the following online resources:

Arthritis Society of Canada – <http://www.arthritis.ca>

Health Link BC –

<http://www.healthlinkbc.ca/kb/content/major/hw104391.html>

Arthritis Foundation – <http://www.arthritis.org>

LEAP – <http://www.leapjia.com>

NIAMS (National Institute of Arthritis and Musculoskeletal and Skin Diseases) – <http://www.niams.nih.gov>

Rheuminfo – <http://rheuminfo.com>

PRINTO (Pediatric Rheumatology International Trials Organisation)
– <http://www.printo.it>

Pediatric rheumatology clinics and/or arthritis centres in children's hospitals may also have excellent resources. Visit your nearest children's hospital resource centre in person or on the Web for more information on pJIA.

Alberta

- Alberta Children's Hospital (Calgary)
- Stollery Children's Hospital (Edmonton)

British Columbia

- BC Children's Hospital (Vancouver)

Manitoba

- Health Sciences Centre Winnipeg

Newfoundland and Labrador

- Janeway Child Health Centre (St. John's)

Nova Scotia

- IWK - Health Centre (Halifax)

Ontario

- Children's Hospital of Eastern Ontario (Ottawa)

- Children's Hospital: London Health Sciences Centre
- The Hospital for Sick Children (SickKids, Toronto)
- McMaster Children's Hospital (Hamilton)

Quebec

- Centre hospitalier affilié universitaire de Québec (CHA)
- Centre hospitalier universitaire de Sherbrooke (CHUS)
- Centre hospitalier universitaire Sainte-Justine (Montréal)
- Montreal Children's Hospital

Saskatchewan

- Royal University Hospital (Saskatoon)

Record contact information for your child's healthcare team
on pages 36-39

sJIA: An Overview

What is sJIA?

sJIA (or systemic juvenile idiopathic arthritis) is a subtype of juvenile idiopathic arthritis (or JIA). This is a form of arthritis that affects children (16 years old or younger).

The word 'arthritis' comes from the Greek words 'arthron' and 'itis'.

arthron = joint

itis = inflammation

inflammation = swelling and redness, hot and often painful

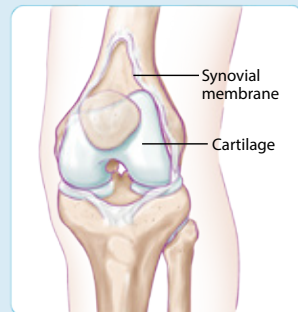
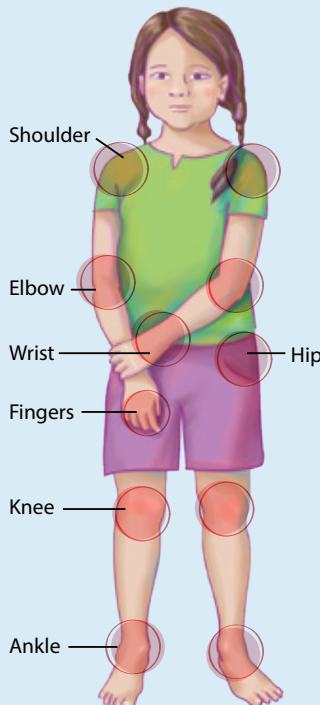
Systemic means that the condition concerns the whole body and not just a single part. Idiopathic means that the cause of the condition is unknown.

Therefore, sJIA is inflammation of the joints with symptoms that affect the whole body in a child under the age of 16. The cause of the inflammation is not known.

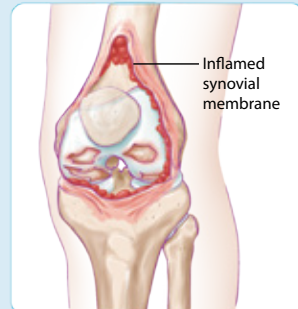
What we do know is that the inflammation is related to the body's immune system attacking healthy tissues, particularly the joints. But we just don't know why.



Joints affected by sJIA



Healthy knee joint



Knee joint affected by sJIA

sJIA: An Overview (cont'd.)

What are the signs and symptoms of sJIA?

Some common symptoms of sJIA

- Joint swelling
- Joint pain
- Whole body symptoms, such as fatigue (feeling tired all day long)

How sJIA is diagnosed?

The cause of sJIA is unknown and the symptoms can be part of other illnesses such as infections. Your doctor needs to ensure that something else is not the cause of your child's joint pain and swelling.

This is why your doctor has completed a full evaluation of your child.

This often takes some time. Once your doctor is sure that it is JIA, then he/she needs to determine what type. Unfortunately, there is not one single test to diagnose sJIA.

How is sJIA treated and managed?

There are a number of different types of therapies that are used to help treat and manage sJIA.

Generally, they fall into the following categories:

- Medications
- Physical therapies
(e.g., exercise, physiotherapy and occupational therapy)

Some of the things your doctor has been looking for include:

- ✓ A complete history – past conditions, medications, tests, arthritis in the family, etc.
- ✓ Complete physical exam
- ✓ Current physical symptoms
- ✓ Changes or markers in the blood or urine (blood tests, urinalysis, etc.)
- ✓ Changes in bones, joints and/or organs (X-rays and related imaging studies)

Treating sJIA with ACTEMRA

ACTEMRA, also known as tocilizumab, is a drug used to treat patients with active systemic juvenile idiopathic arthritis (sJIA) and polyarticular juvenile idiopathic arthritis (pJIA) ages 2 and above.

How it works

ACTEMRA is one of a group of medicines called biologic therapies.

ACTEMRA helps keep the immune system from attacking healthy tissues in the body. Normally, the immune system leaves healthy body tissues alone. In people with certain types of arthritis (e.g., sJIA), the immune system attacks normal body tissues causing damage and inflammation, especially in the tissues of the joints.

ACTEMRA interferes with an important step in this attack by blocking a cytokine called interleukin-6 (IL-6). IL-6 is a protein that is made by the immune system and the body uses it to manage infections.

By decreasing the immune system's attack on normal tissues, ACTEMRA can reduce pain, joint inflammation and tiredness leading to a better quality of life.*

*Quality of life was measured using the Health Assessment Questionnaire (HAQ) to gauge disability (dressing, grooming, eating, walking, hygiene, reach, grip, activities).

Important Side Effect Information

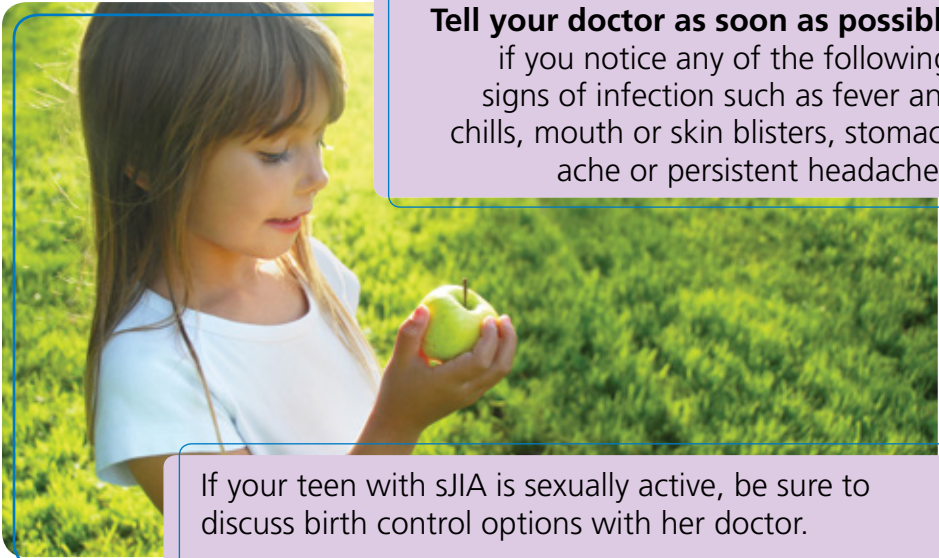
The most common side effects of ACTEMRA are upper respiratory tract infections (common cold, sinus infections) headaches, and increase in blood pressure.

Before your child uses ACTEMRA, inform your doctor if your child has a history of macrophage activating syndrome (MAS), a rare but serious immune reaction in patients with sJIA.

Possible serious side effects include serious infections, liver injury and allergic reactions. A severe skin reaction called Stevens-Johnson syndrome (SJS) and serious drug-induced liver injury, including rapid loss of liver function, inflammation of the liver and jaundice (yellowing of skin and eyes) were reported during treatment with ACTEMRA.

Stop taking ACTEMRA and call your doctor or seek medical attention immediately if you notice any of the following:

- ✓ Difficulty breathing, or light-headedness
- ✓ Rash, itching, hives, swelling of the lips, or other signs of an allergic reaction
- ✓ Chest pain
- ✓ Feeling dizzy or faint
- ✓ Yellowing of the skin and eyes, dark brown coloured urine, pain or swelling in the upper right side of the stomach area, or you feel very tired or confused



Tell your doctor as soon as possible if you notice any of the following: signs of infection such as fever and chills, mouth or skin blisters, stomach ache or persistent headaches.

If your teen with sJIA is sexually active, be sure to discuss birth control options with her doctor.

ACTEMRA should be used during pregnancy only if the potential benefit justifies the potential risk to the fetus.

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Serious infections: Some serious infections have been observed with the use of ACTEMRA. These infections include: active tuberculosis (TB), bacterial, viral and fungal infections. Most patients who developed these infections were taking other drugs that lower the immune system. Hospitalization or death associated with these infections have been reported. Ensure you tell your doctor if you are taking any other medication.

Hepatotoxicity: Serious cases of drug-induced liver injury (DILI) have been observed in patients treated with ACTEMRA. Some of these cases have resulted in acute liver failure requiring a liver transplant. Treatment with ACTEMRA should not be initiated in patients with active infections or active liver disease.

Gastrointestinal perforations: GI perforations (holes in the lining of the gut) have been reported uncommonly, usually as a complication of diverticulitis (infection of the large intestine), and require immediate medical attention. If you develop fever and severe stomach pain that does not go away, seek medical attention.

Abnormalities of your lab results: Your physician will be monitoring your blood work and may notice that your levels are either too high or too low; your healthcare provider may stop your ACTEMRA treatment for a period of time or change your dose of medicine if needed because of changes in these blood test results.

Malignancies: During the clinical trials, cases of cancer have been reported very rarely in patients receiving ACTEMRA. The current number of reported cases in the ACTEMRA studies appears to be consistent with the expected number of cancer cases reported in the RA population. The role of treatment with ACTEMRA on the development of cancers is not known.

Demyelinating disorders: The impact of ACTEMRA on demyelinating disorders is not known, but multiple sclerosis and chronic inflammatory demyelinating polyneuropathy were reported rarely in clinical trials.

Allergic reactions: Serious allergic reactions, including death can happen with ACTEMRA. These reactions can occur on the first infusion, even if you have taken the premedication and can happen with future infusions of ACTEMRA. Tell your healthcare provider if you have any of the following signs of a serious allergic reaction: shortness of breath or trouble breathing, skin rash, swelling of the lips, tongue, or face, chest pain, feeling dizzy or faint.

Treatment with ACTEMRA should not be initiated in patients with active infections or active liver disease.

Taking ACTEMRA for sJIA

ACTEMRA comes in two forms. ACTEMRA is supplied as a solution for intravenous infusion (IV) or as a solution for subcutaneous injection (SC) in either a single-use pre-filled syringe, or a single use autoinjector.

Before starting treatment, make sure your doctor knows if you are taking or have recently taken any other medicines.

Note that ACTEMRA should not be taken with biological medicines for the treatment of rheumatoid arthritis (including Enbrel®, Humira®, Remicade®, RITUXAN, Oencia®, Kineret®, Simponi™ and Cimzia™) as it has not been studied in combination with these medicines.

If a decision has not yet been made regarding the route of administration for ACTEMRA, talk to your doctor about which option may be best for your child.



If your child will be receiving **subcutaneous injections**, continue reading on page 29.

ACTEMRA by Intravenous Infusion for sJIA

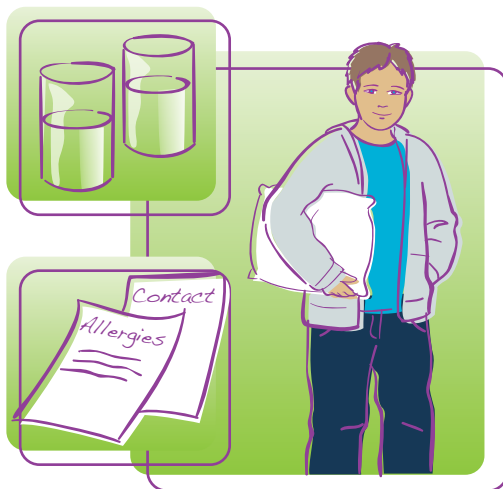
Preparing Your Child for the Infusion

Vaccinations → Be sure your child is up to date (if possible) on all recommended vaccinations, *prior to initiation of therapy* with ACTEMRA. Certain vaccines should not be given *while* receiving ACTEMRA.

Infections → Be sure your child does not have any existing infections prior to receiving ACTEMRA. Treatment with ACTEMRA could cause your child's infection to get worse.

Plenty of water → To help with IV access, be sure your child is well hydrated, but also make sure he/she goes to the bathroom before the infusion. Once started, the infusion takes about an hour.

Comfortable, loose fitting clothes → Be sure your child is comfortable, as he/she will be sitting for about an hour. Also, loose fitting clothes allow the medical staff to monitor your child's vital signs (e.g., blood pressure, heart rate, etc.) easily.



Layers → Dress your child in layers to allow for temperature control. The infusion centre may be cool or warm; or the infusion itself may make your child feel too warm or too cold. Layers of clothing allow your child to control his/her comfort.

Contact the infusion centre about what they provide for patient comfort, e.g., blankets, pillows, drinks, etc.

Be sure your child is not wearing any fragrance or perfume, as other patients may be allergic.

Bring a complete list of current medications, allergies, and emergency contact information for the infusion staff to add to your child's chart.

ACTEMRA by Intravenous Infusion for sJIA

Receiving the Infusion

ACTEMRA will be given to your child by a healthcare professional using an intravenous line. This means the medicine will be given to your child through a needle placed in a vein in his/her arm. It will take about 1 hour to give your child the full dose of medicine.

The recommended dose for children with sJIA is either 8 or 12 mg per kg of body weight depending on the child's weight. Children receive a dose of ACTEMRA every 2 weeks.

In the event of a missed dose, ask your child's doctor when to schedule the next dose.

During the Infusion

Since the infusion takes approximately an hour, it's a good idea to bring a few things to keep your child busy during this time.

Below is a list of some of the things your child might enjoy doing during the infusion:

- ☐ Read a book or comic/magazine
- ☐ Do a crossword puzzle or play a sudoku game
- ☐ Play cards
- ☐ Play hand-held games (if they are quiet or have headphones)
- ☐ Listen to music (with headphones)
- ☐ Bring a laptop computer or tablet to watch a movie, or play games (don't forget the headphones)
- ☐ Have snacks and a drink (if allowed in the infusion room)
- ☐ Draw or doodle
- ☐ Take a nap



Your child will be monitored for allergic reactions during the infusion. These signs may include shortness of breath or trouble breathing, skin rash, swelling of the lips, tongue or face, chest pain, feeling dizzy or faint.

Continue on page 30 if taking ACTEMRA by intravenous infusion

ACTEMRA by Subcutaneous Injections for sJIA

“Subcutaneous” means that it is given into the fat layer just under the skin. The recommended injection sites are the abdomen, thigh and upper arm. The sites should be rotated and injections should never be given into moles, scars, or areas that are tender, bruised, red, hard or have open sores.

Your healthcare provider should show you how to prepare and inject properly before using ACTEMRA subcutaneously for the first time. Ask your healthcare provider any questions you may have. Do not attempt to administer an injection until you are sure you understand how to inject using the pre-filled syringe or a single-use autoinjector. It is important that you follow the specific instructions for using the kind of ACTEMRA that your doctor has prescribed.

The ACTEMRA syringe or autoinjector are intended to be used by patients or caregivers who have been properly trained.

The recommended dose of ACTEMRA for patients with sJIA is:

- o 162 mg once **every two weeks** for patients **below 30 kg**
- o 162 mg once **every week** for patients ≥ 30 kg

Keep the ACTEMRA pre-filled syringe or autoinjector and all medicines out of the reach and sight of children. Always store the them in a refrigerator at a temperature of 2–8 °C. Protect the syringe or autoinjector from freezing and from light.

Vaccinations → Be sure your child is up to date (if possible) on all recommended vaccinations, *prior to initiation of therapy with ACTEMRA*. Certain vaccines should not be given *while* receiving ACTEMRA.

Infections → Be sure your child does not have any existing infections prior to receiving ACTEMRA. Treatment with ACTEMRA could cause your child’s infection to get worse.

After taking ACTEMRA

Unwanted effects are possible with all medicines. Tell your child's doctor or nurse as soon as possible if your child does not feel well while receiving treatment with ACTEMRA.

Possible side effects

Very Common (at least 1 in 10 people)

- Injection Site Reactions like erythema, pruritus, pain, and swelling (with subcutaneous use)

Common (at least 1 in 100 people)

- Upper respiratory tract infections like coughs and cold, pneumonia, skin infection
- Cold sores (oral herpes simplex), blisters, shingles (herpes zoster), skin infection sometimes with fever and chills
- Low white blood cell counts shown by blood tests
- High blood fats (cholesterol levels)
- Headache, dizziness
- High blood pressure
- Mouth ulceration
- Stomach pain
- Abnormal liver function tests
- Rash and itching
- Injection site reactions (with subcutaneous use)
- Ear infection
- Chicken pox
- Gastroenteritis (nausea, vomiting, diarrhea)
- MAS (macrophage activation syndrome)

Be sure to ask for any important post-infusion or post-injection instructions.

Uncommon (at least 1 in 1000 people)

- Diverticulitis (fever, nausea, diarrhea, constipation, stomach pain)
- Red swollen (inflamed) areas in the mouth
- High blood fat (triglyceride levels)
- Serious allergic reactions
- Pancreatitis (stomach pain, back pain, nausea, vomiting)
- Lung disease (shortness of breath, trouble breathing, cough)

Rare (less than 1 in 1,000 patients)

- Multiple Sclerosis (including blurred vision, loss of vision, eye pain, feeling dizzy, or numbness, weakness or tingling in the face, arms or legs)
- Drug-induced liver injury (loss of appetite, nausea and vomiting, fatigue, itching, dark urine, confusion, abdominal swelling and/or pain in the upper-right side of the stomach); Jaundice (yellowing of skin and eyes)

If any of these uncommon or rare side effects occur, stop taking the drug and call your doctor or pharmacist.

Blood Work and Other Tests

Below are descriptions of some of the tests your child may undergo, either at the time of diagnosis or as follow-up tests to monitor disease activity and check for side effects.

Blood tests

Complete blood cell count (CBC): This test measures

- Amounts of each type of blood cell in a sample of blood
- Level of hemoglobin in the blood (low hemoglobin = anemia)
- Abnormalities in the numbers of various kinds of white blood cells or of platelets

Erythrocyte sedimentation rate (ESR): This test does not point specifically to sJIA but indicates active inflammation in the body.

These tests are almost always elevated in children with active sJIA.

C-reactive protein (CRP): This test is another measure of the level of inflammation in the body.

Liver and kidney function: These tests may be done as part of a general health screen.

Cholesterol: Your child's blood cholesterol levels may be monitored, as higher cholesterol levels are a common possible side effect of ACTEMRA therapy.

Follow-up imaging

X-rays: X-rays give information about whether or not damage has occurred in the joint.

MRI: MRI is similar to X-ray but is much more detailed and gives a better 3-dimensional view of the joint.

Bone scan: A bone scan may be necessary if the results of the workup do not support the diagnosis. A bone scan can detect inflammation in the bone and other abnormalities that do not show up well on X-ray.

Please note that other tests may be ordered at the discretion of your child's doctor.

Coping with sJIA

Family members can help your child cope with sJIA by doing the following:

- Keeping a positive outlook
- Learning as much as they can about your child's disease and its treatment
- Treating your child as normally as possible
- Trying to get the best care possible
- Working closely with your child's healthcare team
- Joining a support group
- Encouraging exercise and physical therapy for your child if necessary
- Working closely with your child's school
- Talking openly with your child

Talking to your child about sJIA

Your attitude toward sJIA can affect how your child feels about it.

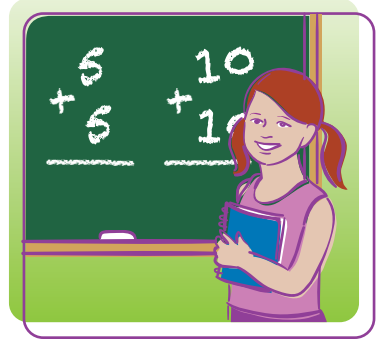
Below are some tips on communicating with your child about sJIA.

- If your child is angry, allow him/her to express his/her anger about arthritis.
- Expect your child to behave as you would any other child and expect the same responsibilities (so long as he/she is physically able).
- Avoid giving him/her special attention or treatment.
- Encourage your child to learn as much as possible about sJIA and its treatment.
- Try not to overprotect your child.
- Being as consistent as possible will help your child learn what is expected.

Be sure of your child's rights. There may be services available at the provincial or local level for children with disabilities like sJIA. Find out what services are available in your area.

Your Child at School

Depending on your child's individual needs, you may want to contact your child's school to meet with your child's teachers, the principal and/or guidance counsellors and help them learn about sJIA. Where possible, you and the school should work together to come up with creative ways to deal with your child's situation.



If your child is experiencing increased arthritis symptoms there are many ways to modify activities and schoolwork. Below is a list of some of the ways adjustments can be made to help your child at school.

- Have two sets of schoolbooks (texts), one for home and one for school.
- Give extra time to change classes.
- See if your child's classes can be close to each other to minimize walking and stair climbing.
- Arrange for activity/stretch breaks.
- Provide a larger pencil or pen or get a foam grip to help with handwriting.
- Depending on other symptoms, your child may need a different desk or chair.
- Timed written tests may need to be changed or extended.

Ask your child's physical or occupational therapist for other ideas.

Be sure to talk to your child's teacher about missing school for infusion appointments and make arrangements for catching up on any missed schoolwork or special assignments.

Other Helpful Tips

Exercise

Physical activity is very important for children with sJIA, although there may be times that activity is limited due to pain.

A well-balanced exercise program will help maintain muscle tone and range of motion and function in the joints. Ask your doctor to recommend a physical therapist to develop an appropriate exercise plan for your child.

Tip: Swimming is an excellent way to use joints and muscles without putting weight on the joints.

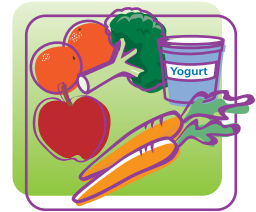
Tip: Warm baths or an electric blanket may help soothe sore joints.

Nutrition

Good nutrition can help fight the effects of sJIA. So it is important that your child gets the nutrients he or she needs for proper growth and development. These include protein, carbohydrate, fat, vitamins, and minerals.

Your child can eat a variety of foods as long as his or her weekly intake is balanced and varied.

Ask your doctor whether you should be supplementing your child's diet with any vitamins or minerals. For example, calcium and vitamin D are important for strong bones.



sJIA Resources and Web

For more information about sJIA and its care, visit the following online resources:

Arthritis Society of Canada – <http://www.arthritis.ca>

Health Link BC –

<http://www.healthlinkbc.ca/kb/content/major/hw104391.html>

Arthritis Foundation – <http://www.arthritis.org>

LEAP – <http://www.leapjia.com>

NIAMS (National Institute of Arthritis and Musculoskeletal and Skin Diseases) – <http://www.niams.nih.gov>

Rheuminfo – <http://rheuminfo.com>

PRINTO (Pediatric Rheumatology International Trials Organisation) – <http://www.printo.it>

Pediatric rheumatology clinics and/or arthritis centres in children's hospitals may also have excellent resources. Visit your nearest children's hospital resource centre in person or on the Web for more information on sJIA.

Alberta

- Alberta Children's Hospital (Calgary)
- Stollery Children's Hospital (Edmonton)

British Columbia

- BC Children's Hospital (Vancouver)

Manitoba

- Health Sciences Centre Winnipeg

Newfoundland and Labrador

- Janeway Child Health Centre (St. John's)

Nova Scotia

- IWK - Health Centre (Halifax)

Ontario

- Children's Hospital of Eastern Ontario (Ottawa)

- Children's Hospital: London Health Sciences Centre

- The Hospital for Sick Children (SickKids, Toronto)

- McMaster Children's Hospital (Hamilton)

Quebec

- Centre hospitalier affilié universitaire de Québec (CHA)

- Centre hospitalier universitaire de Sherbrooke(CHUS)

- Centre hospitalier universitaire Sainte-Justine (Montréal)

- Montreal Children's Hospital

Saskatchewan

- Royal University Hospital (Saskatoon)

Your Healthcare Team

Below is a contact form for you to record the contact information of the various members of your child's healthcare team. Your child's team will be made of some of these professionals, depending on your child's individual needs.

Pediatric rheumatologist

Name: _____ Phone: _____

Address: _____

Nurse / Nurse practitioner

Name: _____ Phone: _____

Address: _____

Physical and occupational therapist

Name: _____ Phone: _____

Address: _____

Social worker or psychologist

Name: _____ Phone: _____

Address: _____

Your family doctor

Name: _____ Phone: _____

Address: _____

Pediatrician

Name: _____ Phone: _____

Address: _____

Ophthalmologist/Optometrlist

Name: _____ Phone: _____

Address: _____

Dentist/Orthodontist

Name: _____ Phone: _____

Address: _____

Your Healthcare Team (cont'd.)

Dietitian or nutritionist

Name: _____ Phone: _____

Address: _____

School contact

Name: _____ Phone: _____

Address: _____

Other

Name: _____ Phone: _____

Address: _____

Notes

Acknowledgements

We would like to express our appreciation to the committee of rheumatology experts who provided valuable and constructive recommendations during the planning and development of this material.

If you require this information in an accessible format,
please contact Roche at 1-800-561-1759.

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