Rheumatology Nurses Society
Professional Education Series

HANDLING THE
HARD QUESTIONS:

What Patients and Their Families Are Asking Us About

JUVENILE IDIOPATHIC ARTHRITIS

The Purpose of This Document

Patients with newly diagnosed juvenile idiopathic arthritis (JIA) and their families often ask many questions about JIA and how best to manage the disease. Rheumatology nurses and advanced practice providers (nurse practitioners and physician assistants) play a key role in educating patients and families about JIA, and it is important to be able to answer questions accurately and communicate successfully. This pocket guide briefly summarizes evidence on some of the most common—and challenging—questions that patients with JIA and their families are asking providers. We hope you find the guide useful for your professional development.

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How do you know my child has JIA?

Children with JIA have arthritis for an unknown reason that lasts for more than 6 weeks and starts before 16 years of age.[1] JIA is one of the most common chronic diseases in children, and it is the most common of all childhood diseases that cause long-term inflammation. Each year in the United States, JIA is diagnosed in approximately 12 of every 100,000 children.[2]

When a child develops JIA, the immune system starts to attack the synovial membrane (i.e., the inner lining of the joints). As a result, the joint becomes inflamed and begins to swell. This is often painful. Without treatment to reduce inflammation, the joint can eventually become damaged.

There is no blood test that confirms or refutes a diagnosis of JIA. Instead, we will ask how long your child's joint(s) have been swollen, whether any other symptoms are present, and how these symptoms are affecting their day-to-day activities. A complete physical exam is performed to evaluate joint swelling, pain, stiffness, overall function, and gait. We also typically look for signs such as rash, fever, or swelling in other parts of the body. While these complementary signs are not required for a diagnosis of JIA, they are important to check during the diagnostic workup. Providers also will typically ask if your child has a history of any other events or diseases that can sometimes cause joint swelling, such as a recent injury or infection, inflammatory bowel disease (IBD), certain cancers, and other diseases that cause inflammation in the body.[1,3,4]

JIA actually includes seven different diseases, sometimes called subtypes, all of which cause joint swelling but are unique in certain ways. The subtypes of JIA include systemic arthritis, oligoarthritis, psoriatic arthritis, rheumatoid factor (RF)-positive polyarthritis, RF-negative polyarthritis, enthesitis-related arthritis, and undifferentiated arthritis.[1,5] These subtypes are diagnosed based on the number of joints involved, whether there are other symptoms outside the joints, and the results of certain tests (see Table).[5]

The frequency (prevalence) of JIA subtypes varies by ethnicity and geographic region, which may reflect differences in genetic and environmental risk factors as well as the underdiagnosis of milder subtypes of JIA in some areas of the world.[6-8] In

North America, oligoarticular JIA is most common subtype, while RF-positive polyarthritis and enthesitis are among the least common.

JIA Subtype	Brief description
Systemic arthritis	Fever for at least 3 weeks (and daily for at least 3 days), arthritis in at least one joint, and at least one of the following: red rash, swollen lymph nodes, enlarged liver and/or spleen, and/or serositis (inflammation of certain tissues in the body)
Oligoarthritis (also known as oligoarticular JIA)	Consists of arthritis in up to 4 joints in the first 6 months after onset. May be persistent or extended (meaning that additional joints are affected after 6 months).
Psoriatic arthritis	Arthritis and psoriasis, or arthritis and at least two of the following: dactylitis (swelling of a finger or toe), nail pitting, and/or psoriasis in a first-degree relative (a parent or sibling)
RF-negative polyarthritis	Arthritis of at least 5 joints in the first 6 months and a negative blood test for rheumatoid factor (RF)
RF-positive polyarthritis	Arthritis of at least 5 joints in the first 6 months and a positive blood test for RF. Much less common than RF-negative polyarthritis.
Enthesitis-related arthritis	Consists of swelling located at sites where the tendons insert into the bone.
Undifferentiated arthritis	Arthritis that fulfills criteria in at least 2 or none of the above categories

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Why did my child develop JIA? What are the chances our other kids will develop it?

It is not clear why some children develop JIA; this is an area of active research. Most experts agree that there are probably a variety of different causes of JIA.[1] In most cases, JIA occurs sporadically, meaning that it was not passed down from parents or grandparents to the child, although there are some instances where a parent does have the same type of arthritis as the child.[2] This is seen more frequently with psoriatic arthritis and enthesis-related arthritis than other subtypes of JIA.

There may be a general pattern of autoimmunity, meaning that family members are at heightened

risk for changes in the immune system that lead to diseases such as spondyloarthropathy, type 1 diabetes, rheumatoid arthritis, and JIA.[3]

Scientists also have found that variations or changes in some genes might increase a child's risk of developing JIA. Most of these genes tell the body to make a group of proteins called human leukocyte antigen (HLA) complex, which helps the immune system understand the difference between the body's own proteins and "foreign" (outsider) proteins such as bacteria and viruses.[1] Certain changes in HLA genes and other genes related to immunity seem to increase the chances that a child's immune system will lose the ability to understand this difference and will start to attack the synovial membrane of the joints, leading to JIA.

Compared with the general population, siblings of children with JIA are at increased risk for developing JIA, but their chances of developing JIA still are fairly low. In one study, about 2.5% of siblings of children with JIA also developed JIA.[4] However, in identical twins, the risk is much higher. Studies have found that if one identical twin develops JIA, the other identical twin has about a 25-40% chance of also developing JIA.[2,5,6]

There is no conclusive evidence that diet, emotional factors, exposure to pets, income level, or the neighborhood or part of the country where a family lives affects a child's risk of developing JIA.[7] There is limited evidence that breastfeeding a baby for more than 4 months

might reduce a child's risk of later developing JIA.[8] This might be because breastfeeding affects a baby's immune system and gut bacteria (sometimes called the microbiome), both of which play a role in the development of autoimmune diseases. However, more studies are needed before any firm conclusions can be drawn about the role of breastfeeding in reducing risk for JIA.

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Is my child's disease ever going to go away?

Unlike rheumatoid arthritis in adults, which lasts throughout life, children with JIA often have symptoms that come and go. For some children, symptoms eventually go away completely and do not come back.[1] For other children, arthritis and other symptoms of JIA continue into adulthood, requiring long-term treatment and management. Studies indicate that anywhere from 40-60% of children with JIA continue to experience active disease (inflammation) as adults, which sometimes persists for decades.[2-4] When JIA symptoms last into adulthood, they tend to be consistent in intensity over time but often don't get significantly better or worse.[4]

No matter how long JIA lasts in a child, it is essential to diagnose the condition quickly and

control disease activity as soon as possible. This is because children with JIA whose disease is not well controlled are at risk for developing permanent physical disabilities or organ damage.[2] For example, growth problems can occur if bones near inflamed joints grow too slowly or rapidly.[5] As a result, a child might have a small chin, one leg or arm that is longer than the other, or decreased overall growth. Without effective treatment to control disease activity, these changes can become permanent. For some children with JIA, poor disease control can also lead to permanent damage to organs such as the eyes, heart, or kidneys.[2]

Fortunately, improved treatments for JIA are enabling many children to go for long periods with little or no disease activity. However, for many children, symptoms return from time to time to a noticeable degree. A noticeable return of symptoms after a period of inactive disease is called a flare. Usually, the most common sign of a flare is worsening of joint pain and swelling.[6] However, a flare can also cause flu-like symptoms (such as fever and nausea), skin rash, blurry vision or dry eyes, a low or depressed mood, or extreme fatigue.

In some cases, the cause of a flare is obvious—medication doses might have been skipped, a child might not be sleeping well, or they might have engaged in more intense exercise than usual. In other cases, the cause may be unclear, potentially because your child's treatment regimen is no longer working to control disease activity.

Any time a flare occurs, it is important to recognize it quickly and notify your rheumatology care team so they can help you and your child get the flare under control. This will help prevent long-term health effects and improve your child's quality of life.

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What are the most common pharmacologic options for my child?

There is no cure for JIA. However, in recent years, treatment options have improved significantly. The main goal of treating JIA is to achieve and maintain remission or a state of minimal disease activity.[1-3] Controlling disease activity will reduce or eliminate swelling and pain, delay flares, improve quality of life, and help prevent joint and organ damage. To achieve these goals, it is important to start treatment soon after diagnosis. After treatment is started, our rheumatology practice will carefully monitor your child to see if they are responding to treatment and recommend changes, if needed. The frequency of subsequent provider visits will depend on the type of JIA your child has and their level of disease activity.[2]

Specific treatment regimens depend on various factors, including JIA subtype and symptom severity.[1,3] As a group, the drugs used today to control disease activity are called disease-modifying antirheumatic drugs (DMARDs). There are two main categories of DMARDs used in JIA:

1. Conventional DMARDs, such as methotrexate, sulfasalazine, and leflunomide, which control disease activity by broadly suppressing the immune system. For children with JIA, the most frequently used conventional DMARD is methotrexate. Methotrexate and other conventional DMARDs are available as oral pills or injections under the skin that can be given at home.

2. Biologic DMARDs, which target more specific pathways in the body that cause inflammation. Biologics can be used alone or in combination with a conventional DMARD such as methotrexate.[3] Biologics are either self-injected or given by infusion at the medical office. As of February 2022, the U.S. Food and Drug Administration (FDA) has approved seven biologics for use in children with JIA (see Table on page 16-17).[4-10]

For many children with JIA, biologics work faster than traditional DMARDs. For this reason, providers sometimes recommend a biologic as a first treatment if a child who is newly diagnosed with JIA has a high level of disease activity. However,

biologics cost more than conventional DMARDs, and some insurers will not approve their use unless methotrexate has been tried first.[11] That said, if your child has a high level of disease activity and your provider thinks a biologic should be tried first, they may be able to make a special appeal to the insurance company.

In addition to conventional and biologic DMARDs, the FDA has approved tofacitinib, an oral small-molecule DMARD, for the treatment of JIA.[12] Tofacitinib is a synthetic Janus kinase (JAK) inhibitor that has been shown to reduce JIA disease activity and flares.[13,14] It is approved for use in some patients whose JIA is not responding to biologic DMARD treatment.

Some children with JIA also benefit from taking a nonsteroidal anti-inflammatory drug (NSAID) to reduce pain and swelling. In most cases, NSAIDs are added to DMARD treatment, but they are not used as a substitute. Another option to help with joint pain and swelling, especially of larger joints such as knees, hips, ankles, and wrists, is to inject a steroid known as triamcinolone hexacetonide directly into the involved joint.[3,15] For most children, this provides relief for at least 6 months.[15] If the injections are helpful, they usually can be repeated as needed.

Table *Biologic and Small Molecule DMARDs Approved by the FDA to Treat JIA* [4-10, 12]

Agent	Class
Abatacept	Biologic selective T-cell costimulation modulator
Adalimumab	Biologic tumor necrosis factor (TNF) inhibitor
Canakinumab	Biologic interleukin-1b inhibitor
Etanercept	Biologic TNF inhibitor
Golimumab	Biologic TNF inhibitor
Secukinumab	Biologic interleukin-17A inhibitor
Tocilizumab	Biologic interleukin-6 inhibitor
Tofacitinib	Small-molecule Janus kinase (JAK) inhibitor

Route of administration	Indication
Intravenous infusion or subcutaneous injection	Moderately to severely active polyarticular JIA in patients aged 2 years or older
Subcutaneous injection	Moderately to severely active polyarticular JIA in patients aged 2 years or older, alone or together with methotrexate
Subcutaneous injection	Active systemic JIA in patients aged 2 years or older
Subcutaneous injection	Moderately to severely active polyarticular JIA in patients aged 2 years or older
Intravenous injection	Active polyarticular JIA in patients aged 2 years and older Active juvenile psoriatic arthritis in patients 2 years of age and older
Subcutaneous injection	Active enthesitis-related arthritis in patients aged 4 years and older Active juvenile psoriatic arthritis in patients 2 years of age and older
Intravenous infusion or subcutaneous injection	Active polyarticular JIA in patients aged 2 years or older Active systemic JIA in patients aged 2 years or older
Oral	Active polyarticular JIA in patients aged 2 years or older who have an inadequate response or intolerance to one or more TNF inhibitors

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Why are you recommending a medication for my child that is used to treat kids with cancer?

Several drugs used to treat cancer also have shown significant benefits in patients with autoimmune diseases.[1] Methotrexate is one of these drugs (others include rituximab and cyclophosphamide). Methotrexate is used in cancer treatment because it can stop or slow the growth of some types of tumor cells. However, methotrexate also can block certain pathways in the body that increase inflammation.[1,2] For this reason, it is used at significantly lower doses as a DMARD—a drug that treats autoimmune diseases—than in cancer patients.[2,3]

Methotrexate has received FDA approval to treat polyarticular JIA, psoriasis, adult rheumatoid arthritis, and several different types of cancer.[4] Many rheumatologists also use methotrexate off-label to treat other types of chronic autoimmune diseases such as systemic sclerosis, inflammatory bowel disease, and systemic lupus erythematosus.[5]

For children with JIA, numerous studies have shown that treatment with methotrexate can control disease activity and improve quality of life.[6-9] Methotrexate has been used for more than two decades to treat JIA, and it remains the most common DMARD used today because of its efficacy, good safety profile, and low cost.[2,5,10] It is often the first DMARD that providers use to treat polyarticular JIA and is also sometimes used to treat oligoarthritis if patients have high disease activity or are not responding to NSAIDs and joint injections.[9] Methotrexate is also used to treat systemic JIA when joint inflammation is the main symptom.

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I heard that JIA can cause eye problems. Is this true?

Yes, this is true, but these issues usually can be prevented or minimized with regular screening and prompt treatment by your child's provider. Many children with JIA are at risk for a condition called uveitis, in which the middle tissue layer in the eye wall becomes inflamed. Uveitis is the most common complication of JIA that occurs outside the joints.[1] The reason that JIA causes uveitis remains unclear, but without timely treatment, it can lead to cataracts, glaucoma, and macular degeneration, all of which threaten sight.[2] For this reason, early detection and treatment of uveitis is crucial to preserve vision.

In children with JIA, the most common type of uveitis (called chronic anterior uveitis) rarely causes symptoms in the early stages.[1] Therefore,

it is vital that our team regularly screen your child for uveitis if they are considered to be at risk for developing the condition. Children with JIA who are at highest risk for uveitis have either oligoarthritis, RF-negative polyarthritis, psoriatic arthritis, or undifferentiated arthritis; have tested positive for antinucleotide antibodies (ANA); were younger than 7 years old when diagnosed with JIA; and have had JIA for 4 years or less.[3] The American College of Rheumatology (ACR) recommends screening these children for uveitis every 3 months. For children with JIA who are at low to moderate risk for uveitis (these include those with oligoarthritis, RF-negative polyarthritis, psoriatic arthritis, or undifferentiated arthritis who do not meet the other high-risk criteria), the ACR recommends screening every 6 months.

If our team diagnoses uveitis in your child, topical steroids (glucocorticoids) are often prescribed as initial treatment. These are applied directly to the eye surface to rapidly reduce inflammation.[3] If topical steroids are needed for more than a few months, systemic (oral or injectable) treatments are usually considered.[3,4] If additional treatment is needed to control uveitis, options include methotrexate or the tumor necrosis factor (TNF) inhibitors adalimumab or infliximab. If one of these is needed, it should be started promptly to rapidly control uveitis and preserve vision.

We will continue to check your child frequently for uveitis even after it has been controlled. The ACR recommends checking monthly any time that topical steroids are being tapered (reduced in dose), every 2 months if an oral or injectable medication for uveitis is being tapered or stopped, and every 3 months if these medications are being continued at the same dose.[3] Although uveitis in children with JIA usually does not cause symptoms, it can sometimes lead to eye pain, redness, headaches, or a desire to avoid bright light (photophobia).[2] If you or your child observe these symptoms, you should immediately contact our rheumatology team.

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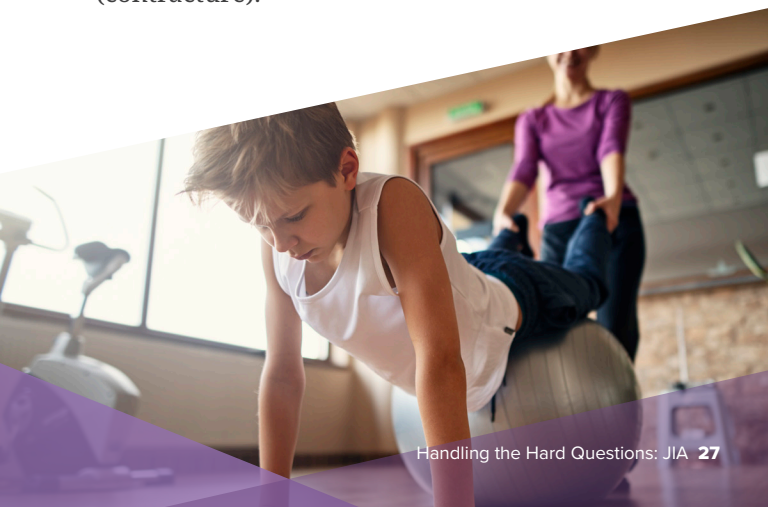
Is there anything besides medication that may help my child?

While medical treatment is key to the successful management of JIA, there are other interventions that can also help children with this disease. These include exercise, physical and occupational therapy, diet, and in some cases, certain supplements.

Physical therapy is especially important if your child has or is at risk for functional limitations (difficulty engaging in normal day-to-day activities).[1] Studies show that physical therapy and regular exercise do not make JIA symptoms worse and can improve joint pain, range of motion, muscle strength and flexibility, aerobic capacity, bone health, and quality of life.[2-6] Physical therapy may also help prevent shortening of muscles and other tissues around the joints (contracture).



Exercises for children with JIA might include weight-bearing exercises, core stability exercises (which strengthen the abdominal muscles, buttocks, and pelvic floor), and exercises that improve balance.[5] In all cases, the physical therapy program should be tailored based on your child's symptoms, level of disease activity, current level of physical functioning, and goals.

Occupational therapy is also recommended for many children with JIA.[1] Occupational therapists can help your child learn strategies to care for themselves, attend school, and play without worsening of their symptoms. Occupational therapists can also teach your child to use tools that help with daily activities, including splints and casts that are sometimes used to help manage swelling and joint pain.[7]

For children with JIA, customized foot orthotics can improve pain, walking speed, and physical functioning better than off-the-shelf shoe inserts or supportive shoes.[8] The most effective orthotics are semi-rigid but still absorb shock. A podiatrist with experience in working with children with arthritis can help evaluate your child and design the optimal orthotics for them.

Diet is another important part of managing JIA. Studies have shown that eating processed foods, sugary desserts and drinks, fried foods, red meat, and large amounts of carbohydrates can increase levels of pain and inflammation in children and adults with arthritis, particularly if their disease is active (i.e., not currently in remission).[9-11]

Children with JIA may benefit from eating less or giving up these types of foods and instead eating more unprocessed fruits, berries, vegetables, beans and lentils, nuts, olive oil, fatty fish (salmon, tuna, mackerel, and sardines), eggs, and yogurt. In studies, this type of diet was associated with improvements in pain, morning stiffness, and physical functioning in children with JIA, as well as in adults with rheumatoid arthritis.[10,12] Diet, like exercise, should be tailored based on your child's needs and preferences.

Some spices, herbs, and supplements have also been linked with improved disease activity in JIA and rheumatoid arthritis.[12] The strongest evidence supports the use of ginger powder, cinnamon powder, saffron, quercetin, and ubiquinone, as well as probiotics containing a type of bacteria called *Lactobacillus casei*, which is a beneficial part of the human intestinal tract (gut).

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What do we need to tell my child's school to get them the accommodations they may need?

Schools are legally required to provide accommodations to enable children with special needs to have equal access to the curriculum and all other resources that a school provides.[1] For children with JIA, accommodations might include the ability to miss school, arrive late, and receive other types of support so they can manage medical appointments, medication side effects, mobility challenges, and disease flares.[2] However, students are still responsible for completing schoolwork regardless of any accommodations provided to them.

To help your child receive the accommodations they need, you can work with their school to set up a

504 plan. This is a detailed list of accommodations designed for your child based on their current needs.[1] Typically, the first step in creating a 504 plan is to ask your school's guidance counselor to schedule a planning meeting with appropriate staff. Next, you should gather documents to bring to the meeting, including a written request for 504 accommodations, a letter from your child's provider(s) explaining their diagnosis, symptoms, treatment, and needs, as well as a detailed list of the accommodations you are requesting. Examples of accommodations for students with JIA are shown on pages 34-37.[2]

After the meeting, it may be helpful to request a written summary of the accommodations the school has agreed to provide and a timeline for putting these in place.[2] Your school's guidance counselor and 504 plan coordinator should then create the formal 504 plan. When you have reviewed and accepted the parameters of the plan, you should send or hand a copy to your child's teachers and other adults who work with your child at school. If you are concerned that the plan is not being followed, talk with your child's teacher first before contacting the guidance counselor, school administrator, or 504 plan coordinator.

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Table *Examples of 504 Accommodations for Students with JIA*[3]

Absence from school or arriving late:

- Student can miss school or arrive late, without penalty, when medically necessary
- Student can make up quizzes and tests, without penalty, when medically necessary

Assignments, homework, and tests:

- Student is assigned a notetaker when needed
- Assignments can be turned in late, without penalty, if student needs extra time because of flares or medical appointments
- Student can submit verbal, rather than written, answers to class assignments, homework assignments, and tests
- Student can dictate homework to a parent who writes or types the answers
- When student is absent, the teacher will send assignments to the parents or ask them to pick them up
- Student receives extra time to take tests

Books and Materials:

- Student is given two sets of books, one for school and one for home
- Student is allowed to use ergonomic school supplies and other tools that help them function at school, such as large grips for pencils or pens, ergonomic binders, an audio recorder, a rolling backpack, and a tablet or laptop

Breaks and leaving class:

- Student is allowed to take extra breaks to stretch and move
- Student is allowed a rest period during the school day

Breaks and leaving class:

- Student is given a permanent hall pass so they can leave class to take medications or see the nurse when needed

Medical needs:

- Student can wear a mask and keep hand sanitizer at their desk to reduce the chances of getting an infection
- To reduce risk of infection, student is given a space near the nurse's office (to rest and take medications) that is not used for "sick children"
- Student is allowed to have a water bottle and snacks during the day if needed to take medications

Physical education (PE):

- Student is excused from PE, or can avoid or modify certain activities when medically necessary

Seating:

- Student is allowed to sit in a chair when others are sitting on the floor
- Student is given a seat in the classroom that provides maximum legroom and permits standing when needed
- Student is allowed to use cushions for chairs and a footstool if feet do not reach the floor

Moving Around:

- Student is allowed extra time to move between classes
- Student is allowed to use elevators and accessible ramps
- Student is assigned an accessible locker
- Student is assigned an accessible place on the school bus

Are there any specific activities or sports my child should avoid?

Children with JIA can participate in many sports and other physical activities as long their JIA is well controlled.[1] Participating in sports does not appear to worsen disease activity.[2] When your child's disease is active, however, high-impact and weight-bearing sports should be avoided. At these times, it is best to switch to low-impact activities such as walking, swimming, and aquatic exercise.

It may also be best to choose sports and other physical activities that do not place a great deal of strain on joints that have been affected by JIA. For example, if your child's neck or spine is involved, it is best to avoid high-impact sports such as tackle football, wrestling, or hockey.[1]

If one or both shoulders are involved, it is advisable to avoid sports such as tennis or basketball.

It is important for children with JIA to learn to monitor themselves during activity and take breaks during practice if needed.[1] In addition, if your child's joints hurt after every practice, that may be a sign to take a break and switch to lower-impact activities, at least for a time.

You also may wish to help your child choose a team that is less competitive, or that places children on teams based on their size instead of their age.[1] If your child played a weight-bearing, physically challenging sport prior to their JIA diagnosis and wants to return to this activity after their disease is controlled, it can be helpful to work with a pediatric exercise specialist or a physical therapist who can design a conditioning program that helps your child return safely to the sport.[3]

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When do we have to think about transitioning to an adult rheumatology practice?

In most cases, the transition to an adult rheumatology practice happens when patients are between 18 and 22 years old.[1] However, the transition might need to happen sooner if your child decides to move away from home for school or a job. Our pediatric rheumatology practice will time the transition based on your child's unique needs and our policy regarding the age of patients typically seen by our team of providers.[2]

Adult rheumatology practices tend to function differently than pediatric practices.[3] Visits are often shorter and focus more exclusively on disease and treatment instead of including discussions about self-care and life goals. Adult practices also give

patients less leeway if they miss or arrive late to appointments, and patients are expected to take medications and refill prescriptions without help or reminders. Adult practices also usually do not have on-site services such as social workers, counselors, or physical or occupational therapists. To help your child adjust to these changes, they should be encouraged throughout adolescence to develop skills involving communication, decision-making, and self-care.[1] These skills will help them effectively manage their disease and advocate for themselves.

Before your child transitions to an adult rheumatology practice, our team will assess their readiness for transition by looking at how well they can manage their disease without help from parents or guardians.[1,4] When your child is ready to transition, we will create a medical summary to share with their new, adult rheumatology practice. A medical summary states what type of JIA your child has, current level of disease activity, current and past treatments and side effects, history of flares and other complications, and current members of your child's care team, along with any other information that will help make the transition smoother.

Our practice will do our best to help identify adult rheumatology practices that might be a good fit for your child.[1] Depending upon the unique circumstances of your child's disease, we can also help identify providers in other specialties, such as social workers, who can help young adults set and reach educational and employment goals.

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