

## PedsCases Podcast Scripts

This is a text version of a podcast from PedsCases.com on “**Juvenile Idiopathic Arthritis.**” These podcasts are designed to give medical students an overview of key topics in pediatrics. The audio versions are accessible on iTunes or at [www.peds-cases.com/podcasts](http://www.peds-cases.com/podcasts).

### **Juvenile Idiopathic Arthritis**

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#### Introduction

Hi, my name is Brieanne Rogers and I am a medical student at the University of Alberta, Edmonton, AB, CAN. This podcast was developed in conjunction with Dr. Janet Ellsworth, a Pediatric Rheumatologist at the Stollery Children’s Hospital, Edmonton, AB, CAN.

Today we will be discussing Juvenile Idiopathic Arthritis or JIA. Objectives of the Juvenile Idiopathic Arthritis PedsCase:

1. To learn the diagnostic criteria for JIA.
2. To distinguish between the 7 different subtypes of JIA.
3. To learn general treatments for the different subtypes of JIA.
4. To recognize serious complications of different subtypes of JIA.

Let’s start by going over some general information about JIA:

JIA is the most common chronic rheumatic disease in children, with a prevalence of 1/1000, therefore it is very important to learn about! Some key words on history with a pediatric patient that should make you think about JIA are: swollen joint(s), morning stiffness, activity decreases the pain, and joint stiffness.

The diagnosis of JIA is a clinical diagnosis: there are no diagnostic investigations that have specificity for a diagnosis of JIA. The diagnosis of JIA is determined by the following criteria:

1. Presence of arthritis (effusion of joint, or joint pain, stiffness, tenderness) in one or more joints for more than 6 weeks
2. Patient is less than 16 years
3. Exclusion of other diseases that can also have arthritis (e.g. IBD, SLE, reactive arthritis, etc.)

Once the clinical diagnosis of JIA is made, the subtype of JIA can be determined. The sub-type is assigned after 6 months of disease and does not change even if the course

of the arthritis changes. For example, if a child has oligoarthritis at presentation and 6 months, however in 1 year goes on to have 10+ joints involved, he or she will remain to be classified as an Oligoarthritis subtype. This designation can be helpful both from a prognostic and treatment perspective as the typical course and associated features of the different subtypes vary.

We will now go through the 7 subtypes of JIA, and after will go through real-life cases of each subtype so you can test your new knowledge on subtypes!

### 1) Oligoarthritis

Oligoarthritis is the most common subtype of JIA, affecting about 40% of all children with JIA. Oligoarthritis, by definition, means 1-4 joints are affected by arthritis. Large joints such as knee joints are most commonly affected in this type of arthritis. Beyond 6 months, about 1/3 of children with oligoarthritis will develop more joint involvement. These children are called oligoarthritis-extended, because their disease has extended past oligoarthritis into polyarthritis. Oligoarthritis most commonly affects girls, and children 1-3 years of age. One very important teaching point of the oligoarthritis subtype of JIA is that uveitis is common in this subtype (20-30%) and needs to be screened for. Uveitis is the inflammation of the anterior uveal tract (including iris and ciliary bodies). It is especially concerning because children with JIA-associated uveitis are most often asymptomatic; they do not have injected conjunctiva, pain, photophobia, discharge, or vision changes. Untreated, it can cause blindness. Therefore, children are referred to an ophthalmologist at diagnosis of oligoarthritis JIA and have very strict follow-up with the ophthalmologist. However, from an arthritis perspective, oligoarthritis has the best prognosis for remission and is easiest to treat! Initial treatment options for oligoarthritis include NSAIDS (such as naproxen and ibuprofen) and intra-articular steroid injections. Methotrexate and biologics are often not necessary for oligoarthritis.

### 2) RF Negative Polyarthritis

Polyarthritis, by definition, means 5 or more joints are involved. RF negative polyarthritis means that the Rheumatoid Factor investigation (often positive in adult rheumatoid arthritis) is negative and the patient has 5 or more joints involved by 6-months. ANA (Antinuclear antibodies) can be positive and ESR (erythrocyte sedimentation rate) may be increased. RF negative polyarthritis is the 2<sup>nd</sup> most common subtype of JIA, with about 25% of JIA patients affected with this subtype. Commonly, this will be a mix of large (e.g. knee) and small (e.g. PIP) joints. The joint involvement may be asymmetric or symmetric. This subtype is also more common in girls, but unlike oligoarthritis, all ages are affected. Uveitis is more common in younger children with this subtype. Most children with polyarthritis will be treated with Methotrexate (a DMARD, or disease modifying anti-rheumatic drug) early in their disease course, and may require biologic treatment as the disease progresses.

### 3) RF Positive Polyarthritis

Rheumatoid Factor positive polyarthritis is very similar to adult Rheumatoid Arthritis, but only about 5% of patients with JIA have this subtype. For this subtype, 5 or more joints are involved and it is commonly a symmetric disease. Nodules on hands, elbows, etc. can be seen, as well as radiologic evidence of joint erosions. It is most common in teenagers, especially in aboriginal teenagers. Aggressive treatment is necessary. Methotrexate is often the first medication used and biologics (such as TNF-inhibitors, Etanercept or Enbrel and Adalimumab or Humira) may be added early if response to Methotrexate is poor.

#### 4) Systemic JIA

About 5-10% of patients with JIA have this subtype. This subtype of JIA has many extra-articular manifestations, and in fact, arthritis may be absent at onset in 1/3 of patients with this subtype! Children present with a daily, spiking fever to 39C or higher, a maculopapular rash which comes and goes, lymphadenopathy and hepatosplenomegaly, as well as pericardial and/or pleural effusions. The initial work-up for children with suspected systemic JIA must include careful screening for infection and malignancy. Other differential diagnoses for systemic JIA include atypical (or typical) Kawasaki's disease and SLE. Treatment with NSAIDs and steroids can be helpful for the systemic features. While some children with predominantly systemic symptoms may recover completely, about 50% develop significant polyarthritis which can be very resistant to conventional treatments like Methotrexate. Some of the newer biologic treatments such as anti IL-1 (Anakinra) and anti IL-6 (Tocilizumab) appear to be more effective in treating these children.

#### 5) Enthesitis Related JIA

Enthesitis is pain at tendon insertions to bone. Arthritis associated with enthesitis related JIA usually affects the lower extremities, including hips, midfeet and toes. About 10% of patients with JIA have this subtype of disease. This type of JIA is more common in boys and ages over 10 years old. There is a genetic component to this subtype, specifically Human Leukocyte Antigen (HLA) B27, therefore family history of HLA B27 associated diseases (e.g. ankylosing spondylitis) is important to elicit on history. Finally, in contrast to uveitis seen in other JIA subtypes, these children are more likely to develop acute, symptomatic (pain, redness, photophobia) uveitis or iritis. Treatment options include NSAIDs, Methotrexate, Sulfasalazine and biologics.

#### 6) Psoriatic Arthritis

Although psoriasis is associated with this type of JIA, the scaly plaque-like rash and the arthritis rarely occur simultaneously, and there can be a gap of up to 15 years between the 2 problems. The arthritis associated with Psoriatic Arthritis is most commonly oligoarthritis but it can develop into a polyarthritis (i.e. an oligoarthritis-extended type). Common manifestations in this subtype include psoriatic nail changes, dactylitis, and family history of psoriasis. Uveitis is again common in this subtype (about 20% of patients with this subtype) therefore screening and follow-up with an ophthalmologist is

important. NSAIDs may be prescribed initially. Continued active disease usually requires treatment with Methotrexate, which can also be helpful for the skin rash. In severe cases, biologic treatments, especially anti-TNF biologics, are used.

## 7) Undifferentiated Type

About 5-10% of children with JIA don't fit into one single subtype and would be placed in this category.

Ok, now that we went through the criteria for clinically diagnosing JIA, and the different subtypes of JIA, let's go through some case examples. After each case, think about 2 questions: 1) is this JIA? and 2) If so, what subtype of JIA is this? After each case, I will pause briefly to let you think about these two questions and then will go over the correct answers.

### Case 1

You are completing a 2-week elective at a Pediatric Rheumatology clinic, and a 3-year-old female comes in limping. Her parents say that she has been "walking funny" for a while now, maybe 6 weeks, mostly in the mornings, but doesn't complain of any pain. You complete your history and physical exam including examining the child's stance and gait. She has stiffness in her right knee for about 1 hour each morning, and as she moves, it gets better. Her parents say that her right knee has appeared swollen for about the same length of time as the limping. She is otherwise healthy and is developing normally. On exam you notice that the little girl's right leg is slightly turned outwards, she keeps most of her weight on the left leg, and the right knee has an effusion. All other joints are normal.

Is this JIA? 2. If so, what subtype is this? If not, what is this?

Ok, so this IS a case of JIA! The patient is a pediatric patient with symptoms of arthritis in her right knee for a minimum of 6 weeks and is otherwise healthy with no other diseases that can be associated with arthritis such as IBD, SLE, etc. This is a classic example of oligoarthritis: female, 1-3 years old, morning stiffness with gel phenomenon and 1-4 joints affected. The absence of pain is not uncommon in JIA, with up to 40% of children reporting no pain at the onset of their disease.

For a bonus question, what do you need to make sure you arrange with the parents in terms of referral and follow-up, and why? Ophthalmology referral! Yes! Uveitis is common in this age group and subtype, therefore you need to make sure that ophthalmology sees the child for screening and follow-up!

Great job, now onto the next case.

### Case 2

You are still on your rheumatology elective, and a 12-year-old female comes into the clinic with her mom. She tells you that she has been feeling awful lately and missed a

lot of school these past couple months. She says it is very hard for her to get out of bed and she can barely move in the morning. She is very stiff and achy. You complete your history and physical exam, including examining the child's stance and gait. She has had stiff, swollen joints for 2 months, and her stiffness is worse in the morning. She has missed about a total of 2 weeks of school and is having trouble catching up. She is otherwise healthy except for eczema and a few allergies, and loves math and swimming. She has stiff joints and effusions on 3 PIPs on the right hand, all MCPs on the left hand, and both knees and ankle joints. Luckily her family doctor sent off a few investigations before referring her to you, and she is RF negative.

Is this JIA? 2. If so, what subtype is this? If not, what is this?

Yes, this is JIA! The patient is a pediatric patient with symptoms of arthritis in multiple joints for a minimum of 6 weeks and is otherwise healthy. This is a RF negative polyarthritis subtype. This 12-year-old female has more than 4 affected joints and is RF negative. Remember that RF negative polyarthritis often has a mix of small and large joints, can be symmetric or asymmetric, is more common in girls and also has a risk for uveitis so screening and follow-up with an ophthalmologist needs to be arranged.

### Case 3

You get a phone call while at the Rheumatology clinic from a family doctor who is calling for advice on a patient he has in his office. He said that he has an 8-year-old female who has had high (38-40 degree C) fevers for the past 5 days, mouth sores, a macular rash, joint pain and cervical chain lymphadenopathy.

Is this JIA? 2. If so, what subtype is this? If not, what is this? Bonus question: what advice should you give the family doc?

While this could be systemic JIA with the features of: high spiking fevers, rash, joint pain and lymphadenopathy, you should first consider other diagnoses such as infections and Kawasaki Disease. The most appropriate advice you could give the family doc is to refer the patient to the pediatric ER to be checked out and they can consult rheumatology if necessary.

### Case 4

The last child that you see on your last day of your pediatric Rheumatology elective, is a 10-year-old boy with a limp. You walk into the room with 100 questions because you remember from listening to the "Evaluation of a Limp" PedsCase Podcast that, the differential for this presentation is vast. You complete the history and physical exam and you summarize the pertinent positives and negatives: The boy has been limping for about 2 months, states that most of the pain is in his left ankle, never has pain waking him at night, pain is alleviated with ibuprofen and there is no known trauma to the area. He is otherwise healthy. He has a family history of ankylosing spondylitis, hypothyroidism, diabetes and HTN. On exam you notice a scaly, plaque type rash on

his extensor surfaces (backs of elbows and knees), and that he has swelling and decreased ROM of his left ankle, as well as pain at the insertion of the Achilles tendon.

Is this JIA? 2. If so, what subtype is this? If not, what is this?

Yes! This is JIA. This is a bit of a tougher case, but the subtype is Undifferentiated JIA. This boy has features of more than 1 type of JIA: he most likely has enthesitis (Achilles insertion), arthritis in his left ankle, has a family history of HLA B27 associated diseases, but also has psoriasis.

#### Summary and Important Points:

1. JIA is the most common chronic rheumatic disease in children, with a prevalence of 1/1000.
2. Some key words on history with a pediatric patient that should make you think about JIA are: morning stiffness, pain and stiffness that decrease with activity, joint swelling/effusions, etc.
3. The diagnosis of JIA is a clinical diagnosis. There are no diagnostic investigations for JIA. The diagnosis of JIA in general is determined by the following criteria: Presence of arthritis (effusion of joint, joint pain, stiffness, tenderness) in one or more joints for more than 6 weeks, age of patient is less than 16 years, and exclusion of other diseases that can also have arthritis (e.g. IBD, SLE, reactive arthritis, etc.).
4. There are 7 types of JIA which are determined by their presentation: Oligoarthritis, RF negative Polyarthritis, RF Positive Polyarthritis, Systemic JIA, Enthesitis Related JIA, Psoriatic JIA, and Undifferentiated type JIA.
5. All patients with JIA need referral to ophthalmology to screen and follow for uveitis- however the risk is highest in younger patients with oligoarthritis, RF negative polyarthritis and psoriatic arthritis.

#### References

References available on request.