Juvenile Idiopathic/Rheumatoid Arthritis

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Purpose The purpose of this course is to describe the etiology, pathophysiology, diagnostic criteria, types, characteristics, and treatment options for juvenile idiopathic arthritis (JIA).

Goals Upon completion of this course, the healthcare provider should be able to:

- Discuss the pathophysiology of JIA.
- Discuss diagnostic criteria for JIA.
- Differentiate among the 3 types of JIA.
- Describe the clinical presentation and complications of oligoarticular JIA.
- Discuss chronic iridocyclitis.
- Describe the clinical presentation and complications of systemic JIA.
- Discuss macrophage activation syndrome.
- Differentiate between seropositive and seronegative polyarticular JIA.
- Discuss 3 groups of medications used for JIA treatment.
- Discuss at least 5 additional therapies.
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Introduction

Juvenile idiopathic arthritis (JIA), also often referred to as juvenile rheumatoid arthritis (JRA) or juvenile chronic arthritis (JCA), is a chronic autoimmune inflammatory disorder affecting children and adolescents (≤16). It is the most common chronic disease affecting

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children. JIA is a significant cause of pain and impaired mobility as well as depressed growth in children.

While sharing some symptoms with adult rheumatoid arthritis **[see CE course Rheumatoid Arthritis]**, JIA is a different disease. JIA may be chronic, intermittent, or transient. JIA is now referred to as idiopathic rather than rheumatoid in much of the literature about the disease because rheumatoid factor is often not present, and the cause of the disease is unknown.

There are 3 types of JIA: Oligoarticular/Pauciarticular, systemic (Still's disease), and polyarticular. JIA is characterized by periods of exacerbation and periods of remission, which may last for months or year. In some cases, after the initial period of inflammation, there is no recurrence. About 70% of children with JIA have permanent remission by adulthood.

JIA most commonly occurs between ages 2 and 5 or 9 and 12 and is more common in females than males and Caucasians than other races. Onset after age 16 is classified as adult-onset arthritis. JIA affects joints but can also affect numerous other organs, including the eyes, heart, lungs, and liver.

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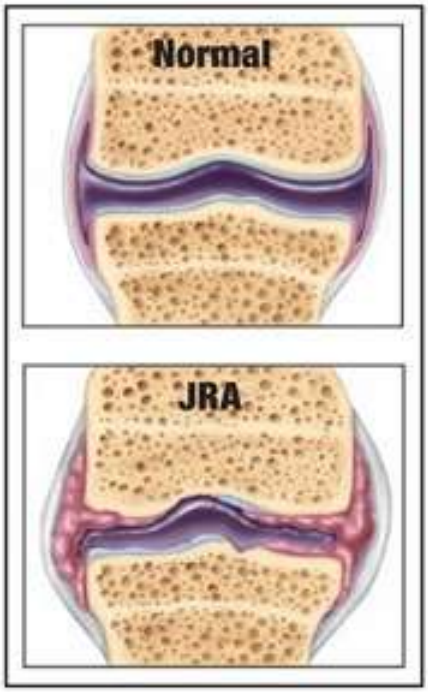
Diagnosis

There is no definitive test to diagnose JIA. Diagnosis is based on clinical presentation during the first 6 months and persistence of symptoms. Tests that may be done include:

- **Rheumatoid factor** (RF): Indicates presence of a macroglobulin type antibody that occurs in connective tissue disease. It is often positive in adult rheumatoid arthritis. RF is usually negative in about 90% of cases of JIA. RF may be positive in JIA with later onset.
- **Erythrocyte sedimentation rate (ESR):** May be elevated in some cases but is a non-specific indicator of inflammation.
- **Human leukocyte antigen (HLA) B 27**: May be positive in some types of JIA, especially oligoarticular.
- **Antinuclear antibody (ANA):** May be present in autoimmune disorders and is commonly used to diagnose systemic lupus erythematosus. ANA may be positive with some types of JIA, such as oligoarticular JIA.

Radiography may be used to diagnose joint damage and monitor progress of the disease. X-ray does not show early cartilage damage or synovitis, so CT scan, MRI, and/or ultrasound may be used for diagnosis and monitoring.

Etiology/Pathophysiology



Joint damage found in JIA is similar to that of adult rheumatoid arthritis, especially with chronic presentations. The synovium becomes inflamed and thickened. Edema of soft tissue about the joint occurs.

Proteolytic enzymes break down collagen, damaging the protective cartilage at the ends of bones, so the bone may begin to erode. Joint effusion and synovitis occurs. Pannus (highly vascular granulation tissue) forms and begins to destroy cartilage at the joint periphery. Rheumatoid nodules may occur with some forms of JIA.

Muscle spams occur along with the joint inflammation, impairing mobility and increasing the risk of flexion contractures. Over time ankylosis of the affected joints may occur.

Growth retardation may occur during active disease, usually with spurts of growth during periods of remission. However, with severe disease, growth may be significantly retarded with corticosteroid treatment a contributing factor. In some cases, an affected extremity may be shorter than the unaffected extremity.

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Oligoarticular/Pauciarticular

Oligoarticular JIA affects about 40 to 60% of those with JIA and is more common in females than males (5:1) with average age of onset about 2 years old. Oligoarticular JIA is often ANA positive (60%) but usually RF negative.

Oligoarticular JIA is characterized by involvement of ≤4 joints in the initial 6 months of the disease, usually the knees, ankles, and elbows (but not the hips or sacroiliac) although small joints in the fingers and toes may also be involved. Joint inflammation is often asymmetrical. Pain and stiffness, as with all types of JIA, is more pronounced in the morning after arising or after periods of inactivity

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Children with oligoarticular JIA are particularly at risk for developing anterior uveitis and chronic iridocyclitis (25 to 40%), which may lead to blindness. Iridocyclitis is characterized by misshapen pupil and conjunctivitis.

Chronic iridocyclitis, a form of uveitis related to JIA, often is present with few symptoms until the eye is severely damaged. Young children are often unaware that their vision is compromised. Onset may be years after initial onset of JIA, so children must have regular checkups throughout childhood, every 3 months for those with oligoarticular JIA and every 6 months with polyarticular.

Iridocyclitis occurs more frequently in females than males (6:1) and is most common with oligoarticular JIA, occuring in 40% of those who test ANA positive and 25% of those who test ANA negative.

Symptoms of oligoarticualr JIA usually include joint swelling and pain, and joint deformity may result in uneven bone growth. There are 2 forms:

- Persistent: Involvement of joints remains at ≤4.
- Extended: After the initial 6-month period, ≥5 joints become involved and the course is similar to polyarticular JIA.

Systemic (Still's disease)

Systemic JIA, also known as Still's disease, is characterized by onset of daily temperature spikes for at least 2 weeks, occurring primarily in the afternoon with at least one extraarticular symptom, which can include a maculopapular rash, lymphadenopathy, hepatosplenomegaly, and/or serositis (inflammation of the serous tissues of the body, including the lining of the heart, lungs, and peritoneum).

Fever, often to ≥104° F (41° C) is common and is usually associated with severe fatigue. The fever rises and falls rapidly. A faint salmon-colored rash is common and may come and go.

Because of inflammation of serous tissues, children may develop pleural effusions, pericarditis, and/or pericardial effusion. Iridocyclitis is very rare with systemic JIA, occurring in <1%.

Sore throat and general systemic symptoms, such as enlarged lymph nodes and hepatosplenomegaly usually precede joint involvement, so diagnosis may be delayed. Arthritis is severe in about 25% of cases and may be either oligoarticular (≤4 joints) or polyarticular (≥5 joints), but polyarticular involvement is most common.

Systemic JIA affects males and females equally with average age of onset about 5 years, representing about 10 to 20% of all cases of JIA.

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Typically children are RF and ANA negative. White blood counts may be markedly elevated along with elevated ESR, and anemia is common.

Incidence of symptoms	
High intermittent fever	100%
Joint inflammation, muscle pain, and chronic arthritis	100%
Skin rash	95%
Lymphadenopathy and/or hepatosplenomegaly	85%
Leukocytosis (marked)	85%
Pleuritis and/or pericarditis	60%
Severe anemia	40%
Abdominal pain	20%

The fever and systemic features tend to resolve after a few months, but the arthritis and damage to organs persists. Symptoms are often cyclic, and triggering factors may cause an exacerbation of symptoms, so each person must be aware of individual triggers, such as stress, a common trigger. Children and families need to understand the importance of continuing a medication regimen because they may believe JIA has been cured when symptoms recede for a period.

Macrophage activation syndrome

MAS is a severe, sometimes fatal, complication of systemic JIA. MAS is believed caused by the activation and uncontrolled proliferation of T lymphocytes and well-differentiated macrophages, causing widespread phagocytosis of blood cells and cytokine overproduction. Thus, the immune system becomes stimulated but ineffective.

About 6-7% of children with systemic JIA develop MAS. Onset of MAS is usually in the early stages of systemic JIA but can occur up to 14 years after initial diagnosis. Onset of symptoms is often acute and may resemble sepsis. Typical symptoms include:

- High, unremitting fever.
- Leukopenia.
- Anemia.
- Thrombocytopenia with increased signs of bleeding: purpura, bruising, and mucosal bleeding.
- Lymphadenopathy.
- Hepatosplenomegaly.
- Increased serum liver enzymes.

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- Increase triglycerides and lactic dehydrogenase (LDH).
- Hyponatremia.
- Abnormal coagulation profile.
- CNS abnormalities: Lethargy, irritability, headaches, seizures, disorientation, and coma.

With onset of MAS, some children show an improvement in arthritis symptoms and a sharp fall in the ESR. The cause of MAS is not clear, but some triggering factors have been identified: exacerbation of underlying disease, ASA or NSAID toxicity, viral infection, and a second round of therapy with gold salts or sulfasalazine or other treatments.

Treatment options: Treatment options include high doses of corticosteroids and/or IV cyclosporin A. Because cyclosporin A sometimes brings about rapid reduction in symptoms, it is now often used as the first-line drug for MAS.

Polyarticular

Polyarticular JIA involves ≥5 joints, especially the joints of the hands and fingers but may also affect the hips, knees, feet, ankles, and neck.

There are two subtypes: seropositive and seronegative.

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Seronegative Symmetric polyarthritis occurs in small and large joints, and rheumatoid nodules may be evident. Seronegative polyarticular JIA represents 20 to 30% of all cases of JIA and is severe in 10 to 15% of cases.

An identifying characteristic is a negative finding on RF testing twice at 3-month intervals. ANA is positive in about 25% of cases. Iridocyclitis is much less frequent with polyarticular JIA than with oligoarticular JIA, but occurs in 5% of those who test RF negative and <1% of those who test RF positive.

The age of onset for seronegative polyarticular JIA is 3 years with disease more common in females than males (3:1)

Seropositive Seropositive polyarticular JIA corresponds to rheumatoid arthritis in adults. Joint involvement is similar to seronegative polyarticular JIA. Seropositive disease is identified by a positive finding on RF testing twice at 3-month intervals and represents 5 to 10% of all cases of JIA and is a severe form of JIA in >50%. Rheumatoid nodules are usually evident. ANA is positive in about 75%.

The age of onset for seropositive polyarticular JIA is 12 years with disease more common in females than males (4:1).

Treatment options

While treatment protocols vary, first-line drugs (NSAIDS) are usually given first, followed by methotrexate. If methotrexate does not control symptoms, then additional medications may be given.

Medications for JIA	
First-line	NSAIDS are the usual first-line therapy for all forms of JIA and is usually the only drugs used for oligoarticular JIA. NSAIDs are often also combined with second-line drugs for more serious disease. Medications include naproxen, ibuprofen, tolmetin, diclofenac, and indomethacin. Aspirin is usually avoided because of adverse GI effects. Cox-2 inhibitors, such as celecoxib, may also be used. Adverse effects include GI disturbance (gas, nausea, vomiting, heartburn, constipation, diarrhea), and rash. NSAIDs should not be taken with salicylates.

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Second-line	Immunosuppressive agents are indicated for extended oligoarticular JIA and polyarticular JIA: • **Methotrexate** (Drug of choice)**:** Affects DNA synthesis and suppresses the immune system. Oral medication is usually given but if the child is nonresponsive to oral medications, sc or IM methotrexate may be used. Adverse effects include bone marrow suppression, GI ulcerations, alopecia, nausea, vomiting, rash, increased infection, and bladder toxiity. • **Sulfasalazine**: Usually administered with NSAIDS for anti-inflammatory effect as well as reducing lymphocyte response and inhibiting angiogenesis. Requires adequate fluid intake. Adverse effects include rash, GI upset, headache, liver function abnormalities, and anemia. Contraindicated in those with allergies to sulfa drugs or salicylates. • **Corticosteroids:** Methylprednisolone (Solu-Medrol®) may be used temporarily to provide short-term relief of symptoms while waiting for long-term relief from other drugs, such as methotrexate. Prednisone may also be used short term to relieve inflammation. PO or high-dose pulse IV corticosteroids may be used for systemic JIA. Adverse effects include cataracts, GI irritation, Cushinoid syndrome, hyperglycemia, hypertension, osteoporosis, avascular necrosis, hirsuitism, and pscyhosis.
Unresponsive or systemic disease	• **TNF blocking agents** (adalimumab, etanercept, infliximab) may be used for polyarticular JIA that is nonresponsive to other medications. Requires testing for TB prior to administration. Adverse effects include increased risk of infection. Medication must be withheld with fever. • **Interleukin-1 receptor antagonist** (anakinra): Used for antifinlammatory and immunologic response for systemic JIA that does not other drugs instead of corticosteroids. Adverse effects include increased risk of infections. Medication must be withheld with fever. • **Abatacept** (Orencia®). Used for polyarticular disease, but should not be used with TNF blocking agents or other biologics, such as anakinra. Adverse effects include increased risk of infection,

	such as upper respiratory infections, urinary infections, pneumonia, cellulities, diverticuliti, and acute pyelonephritis. • **Tocilizumab** (Actemra®), an interluekin-6 receptor antagonist may be used to decrease inflammatory response. This drug has been FDA approved for treatment of systemic JIA. Toclizumab may be used as monotherapy or with methotrexate. Studies showed that after 3 months of treatment with this drug, 85% had a 30% improvement in symptoms. Adverse effects include upper respiratory tract infection, mouth ulcerations, gastritis, upper abdominal pain, lipid elevation, neutropenia, thrombocytopenia, hypertension, headache, and nasopharyngitis.
Joint injection	Glucocorticoid (Triamcinolone hexacetonide) may be injected directly into a joint to reduce pain, swelling and inflammation and improve mobility.

In April 2021, the American College of Rheumatology published recommendations for treatment of JIA. Recommendations include:

- Glucocorticoid joint injections (triamcinolone hexacetonide) for active arthritis, regardless of concurrent therapy or JIA treatment group;
- TNF-α inhibitors in children with a history of arthritis in 4 or fewer joints and significant active arthritis despite 3 to 6 months of methotrexate;
- TNF-α inhibitors in children with a history of arthritis in 5 or more joints and any active arthritis after 3 to 6 months of methotrexate; and
- Anakinra in children with systemic arthritis and active fever whose treatment requires a second medication in addition to systemic glucocorticoids.

Consensus has not yet been reached about these recommendations. Some authorities believe that TNF-a inhibitors should be given before methotrexate. However, studies are ongoing to determine which treatment approach is most effective. Many drugs are very costly as well.

In addition to medications to control inflammation and reduce joint destruction, a number of other treatments are indicated.

Supportive treatments for JIA

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Exercise	Exercise is used to increase strength and mobility of joints and to prevent contractures. Typical exercise regimens include stretching, range of motion, and strengthening. During times of inflammation and acute pain, isometric exercises, which do not involve joint movement, may be best tolerated. Aquatic exercises are especially useful for children because the water reduces resistance. Children should be encouraged to engage in normal activities of living, and staying active improves psychological wellbeing.
Splinting, positioning	Splinting of joints (such as the knees, wrists, and hands) during periods of rest may help reduce pain and prevent or reduce flexion deformity. Children should be positioned without support under the knees and a low or no pillow to maintain extension of the neck, hips, and knees.
Heat	Moist heat, such as in a bathtub at 100°F (37.8°C) for 20 minutes may help relieve pain and stiffness. Moist hot packs may be applied to local areas, such as the wrist. Usually a wet towel is immersed in hot water and then wrung out or a damp towel heated in the microwave. The towel is applied to the affected area and covered with plastic wrap for 20 minutes. For morning pain and stiffness, an electric blanket may be set on a timer to turn on 10 minutes before the child arises.
Assistive devices	Children should be encouraged to remain as independent as possible, but this may require modifications in routines, clothing, and equipment. Elevated toilet seats, grab bars, tongs, and self-adhering fasteners (such as Velco®) or stretch laces on shoes may be helpful.
Nutrition	Nutritional status should be monitored frequently. Children whose mobility is impaired may be prone to weight gain, further stressing their joints, so diets should be nutritious and balanced and appropriate for the needs of the child.

Arthrocentesis	Joint aspiration may be done to aid in diagnosis and to decrease pain and swelling of the joint by removing excess fluid.

Summary

Juvenile idiopathic arthritis (JIA) is the most common chronic disease in children with onset usually between ages 2 and 5 or 9 and 12. There is no definitive test for JIA although a number of tests may be used to help determine the type, including tests for rheumatoid factor (RF), erythrocyte sedimentation rate (ESR), human leukocyte antigen (HLA) B 27, and antinuclear antibody (ANA). JIA results in inflammation, scarring, and destruction of joints. There are 3 primary types of JIA:

- Oligoarticular/Pauciarticular: Affects ≤4 joints, especially the knees, ankles, and elbows, and increases risk of chronic iridocyclitis.
- Systemic: Affects ≤4 joints or ≥5 joints and includes systemic manifestations, with fever, rash, and inflammation of serous lining of the heart, lungs, and peritoneum and hepatosplenomegaly.
- Polyarticular: Affects ≥5 joints, especially the hands and fingers.

While treatment approaches may vary, the usual first-line medications are NSAIDS, followed by methotrexate and then other drugs if methotrexate is not effective. Other treatments may include glucocorticoid injection to joint, joint aspirations, splinting and positioning, exercise, nutritional assessment, heat, and use of assistive devices.

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