Description/Etiology
Juvenile idiopathic arthritis (JIA)—formerly known as juvenile rheumatoid arthritis (JRA) and called juvenile chronic arthritis (JCA) in Europe—is a group of disorders characterized by chronic joint pain and inflammation; JIA can manifest in several forms (e.g., with only a few involved joints or with multiple joint involvement, with or without systemic manifestations; see Quick Lesson About ... Arthritis, Juvenile Idiopathic). JIA is a cause of serious physical and psychological morbidity and can cause permanent deformities, although most patients with JIA eventually enter remission with minimal functional loss or deformity. Treatment by a multidisciplinary clinician team usually includes pharmacotherapy, physical/occupational therapy, and psychosocial support for patient and family.

JIA causes pain (described as aching, sharp, burning, or uncomfortable) and fatigue. Pain prevention and management through frequent assessment and intervention is extremely important. Standardized, age-appropriate pain assessment scales are used to determine the presence, location, type, and intensity of pain. Drugs (e.g., NSAIDs), physical therapy, moist heat or cold treatments, and massage are among the treatment strategies for preventing and relieving JIA-associated pain.

JIA can also cause psychological distress to both patient and family; JIA can change a child’s mood and behavior, lower self-esteem, cause depression and anxiety, disrupt school attendance, change family relationships, and interfere with normal psychosocial development. Chronic pain and physical limitations can make the patient feel isolated from other children (e.g., siblings, classmates).

Relaxation therapy, support groups, counseling about problem-solving techniques, and other psychological interventions can help patients and families cope with the JIA diagnosis. Adequate social support from family members and friends can reduce patient pain and stress. Antidepressants (e.g., selective serotonin reuptake inhibitors [SSRIs]) can reduce depression and anxiety as well as pain.

Facts and Figures
JIA is the most common form of arthritis in children. In the U.S., estimated JIA incidence is 4–14:100,000 person-years; reported prevalence is 1.6–86.1:100,000. Approximately 86% of children with JIA report pain during routine doctor visits. On average, patients with JIA report pain on 73% of days. Among patients with disease duration > 10 years, prevalence of depression is 28%.

Risk Factors
Increased frequency/intensity of pain and fatigue can predict increased anxiety in children with JIA. Risk for depression correlates with disability and persistently-active disease.

Signs and Symptoms/Clinical Presentation
Arthritic signs and symptoms (S/S) common in children with JIA include joint stiffness (especially in the morning); limited range of motion (ROM); warm, swollen, painful
joints; back pain; and reduced function of the affected limb. Systemic JIA S/S include high fevers, evanescent salmon-pink rash on trunk and extremities, hepatosplenomegaly, lymphadenopathy, and muscle tenderness. Ocular S/S include photophobia, red eyes, and visual changes.

**Assessment**

› **Patient History**

- Ask patient/parents about presence and duration of morning stiffness
- Obtain information about child’s independence with ADLs and involvement in sports/school activity
- Ask patient to describe pain at rest

› **Physical Findings of Particular Interest**

- The pain caused by JIA may manifest as refusal to use a limb, excessive guarding of a joint, limping, regression to infantile movements, and/or irritability

› **Laboratory Tests**

- Examination of joint fluid will identify infection, if present
- MRI scan will identify soft tissue swelling, synovial hypertrophy and cartilage degeneration, if present and is also used to monitor clinical responsiveness to treatment in peripheral joints
- Serum c-reactive protein and erythrocyte sedimentation rate will identify inflammation, if present

**Treatment Goals**

› **Promote Optimal Psychologic Function and Reduce Risk of Complications**

- See the *Quick Lesson* referenced above for treatment unrelated to psychological distress or pain
- Monitor vital signs, all psychologic systems, and laboratory/other diagnostic study results; report abnormalities and treat, as ordered
- Frequently assess for pain using a facility approved assessment tool, if available;
  - Pain assessment tools can be used to assess pain presence, type, location, and severity; use developmentally appropriate tools (e.g., depending on age, language ability, ability to follow instructions)
    - The Oucher tool is comprised of 6 photographs of children’s faces with expressions ranging from neutral to severe distress; the patient is asked to choose the picture that best represents his/her current pain level
    - Children who understand quantification and are able to count to 10 are asked to rate their pain on a scale of 0 to 10
    - Self-report questionnaires can be useful in older children and adolescents to obtain information about perceived mental health and quality of life (QOL)
- Standardized cognitive/emotional assessment tools (e.g., Cognitive Emotion Regulation Questionnaire [CERQ]) can be used to obtain information about thought processes and coping strategies as they relate to living with JIA
- Administer analgesia and symptomatic relief, as ordered (e.g., NSAIDs, SSRIs to reduce both depression, anxiety, and pain)
  - Monitor treatment efficacy and for adverse drug effects; consult a drug information source for a complete list
  - Notify the treating clinician in the case of unresolved pain and request referral to a pain management clinician
- Request referral to physical/occupational therapy for evaluation and formulation of an individualized program of exercise and other strategies to prevent/alleviate pain and promote optimum function
  - Nondrug therapies include heat/cold packs, whirlpool, paraffin baths, transcutaneous electrical nerve stimulation (TENS), massage, and exercise
- Encourage family member visitation and rooming-in, as appropriate

› **Support Emotional Well-Being and Educate**

- Assess anxiety level and coping ability of patient (as age-appropriate) and family; provide emotional support and promote a positive self-image for patients who have experienced a dramatic change in lifestyle due to JIA functional limitations
- Educate and encourage discussion regarding JIA, risks and benefits of treatment and pain management options, changes in body function, adherence to individualized treatment regimen, importance of psychosocial support, use of cognitive techniques to reduce pain (e.g., recording thoughts in a diary when pain flares up), long-term physical therapy, and individualized prognosis
- As appropriate, request referral, to a
  - mental health clinician for counseling about living with a painful, chronic condition, adjusting to an altered body image, coping with being different from peers, experiencing depression, anxiety, and stress
–social worker for identification of local resources for support groups, in-home services, or outpatient physical/ occupational therapy

**Food for Thought**

› Researchers conducted in-depth interviews with nine parents of 2–5 years old children with JIA and reported high levels of emotional stress related to witnessing their child in pain, and that stress related to JIA negatively affected relationships between all family members, including siblings and parents (Yuwen et al., 2017)

› In a study involving 61 children with JIA, health-related quality of life (HRQOL) was dramatically improved in children who began etanercept treatment. Children in the study also reported decreases in the number of joints with tenderness, functional restrictions, pain and disease activity (Klotsche et al., 2014)

› In a 2015 study, researchers concluded that exercise treatment significantly improves musculoskeletal symptoms in patients with JIA. In addition, balance-proprioceptive exercises can be more effective than strengthening exercises for the improvement of lower extremity function (e.g., walking, balance, and climbing stairs) in patients with JIA (Baydogan et al., 2015)

› Patients with JIA from low socioeconomic backgrounds report more problems with daily activities and have lower awareness of the consequences of the disease than patients with JIA who are from high socioeconomic backgrounds, warranting special attention from a multidisciplinary team (Verstappen et al., 2015)

› Pregnant women with JIA are at increased risk for postpartum hemorrhage, thromboembolism, and complications related to anesthesia. Researchers comparing outcomes between 1,681 pregnant women with JIA and 6,724 pregnant women without JIA noted a 2- to 3-fold-increase of hemorrhage and post-anesthesia complications, and a 5-fold increase of thromboembolism among women with JIA (Ehrmann Feldman et al, 2017)

**Red Flags**

› Rumination (dwelling on negative thoughts) and catastrophizing (perceiving the disease as a terrible crisis) predict psychological maladjustment in adolescents with JIA

**What Do I Need to Tell the Patient/Patient’s Family?**

› Discuss strategies to improve sleep quality by reducing nighttime pain (e.g., waterbeds, warm nightclothes)

› Bedtime massage can reduce pain and provide sleep-inducing relaxation

› Educate about need for ongoing medical surveillance (including regular eye examination) and physical/occupational therapy

› Advise that regular exercise to maintain muscle strength and joint flexibility is important. Swimming is an excellent choice because it does not overly stress joints

› Educate about use of heat (e.g., morning warm shower/bath) to reduce stiffness

› Discuss need for a healthy diet (including calcium-rich foods) to maintain or achieve adequate body weight and promote normal growth

› Healthy siblings can feel guilt about not having JIA. Reassurance by parents that siblings are not responsible can reduce guilt

› Educate family members about importance of treating children with JIA like other children (e.g., allow to express anger about having JIA; encouraging sports participation, providing normal discipline). Encourage parents to collaborate with teachers and school administrators to provide appropriate modifications in school/sports activities to allow use of assistive devices and activity cessation if S/S occur

**References**


