Review Article

Management of the Upper Extremity in Juvenile Idiopathic Arthritis

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ABSTRACT

Juvenile idiopathic arthritis (JIA) is a chronic heterogeneous condition characterized by inflammatory arthritis persisting for at least 6 weeks in children younger than 16 years without another identifiable cause. Clinical examination is the cornerstone of diagnosis, although various imaging modalities may be used to establish clinically inconspicuous disease, identify structural damage, and monitor treatment response. Serologic testing is primarily used to categorize JIA subtype and provide prognostic information on the disease course. Early diagnosis and treatment initiation are important to preserve functionality, facilitate expected skeletal growth potential, and mitigate long-term articular damage. Treatment can involve physical and occupational therapies, systemic medication, intra-articular corticosteroid injections, and surgical intervention. Disease-modifying antirheumatic drugs have been shown to be effective and safe in children, with remission rates of more than 50% within 1 year of treatment initiation. JIA typically follows a lifelong relapsing course. It is ideal to avoid surgical intervention until a child reaches skeletal maturity to prevent physeal damage. However, surgery can be important to mitigate lifelong articular and soft-tissue damage and to optimize pain management and functionality. Overall, there is a paucity of literature regarding long-term outcomes in the surgical management of JIA. This article will review the current literature on the pathophysiology, diagnosis, and management of JIA of the upper extremity.

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Juvenile idiopathic arthritis (JIA), formerly known as juvenile rheumatoid arthritis, is the most common rheumatic disease of childhood. The term refers to all forms of inflammatory arthritis of unknown etiology that present before the age of 16 years and persist for at least 6 weeks. JIA includes seven subtypes: systemic JIA, oligoarticular, polyarticular rheumatoid factor positive, polyarticular rheumatoid factor negative, psoriatic arthritis, enthesitis-related arthritis (ERA), and undifferentiated arthritis.[1] The incidence and prevalence of JIA have been estimated to be 2 to 20 cases per 100,000 individuals and 16 to 150 cases per 100,000 individuals,

respectively. JIA has a predilection for females and children of northern European descent.[1,2]

Upper extremity involvement can lead to debilitating loss of function, independence, and self-esteem. Approximately 80% of children and adolescents with JIA have hand and/or wrist involvement, and 45% and 21% have elbow and/or shoulder involvement, respectively.[3-5] Children with upper extremity involvement report lower quality of life and poorer school performance than those with isolated lower extremity involvement.[6] By 5 years after initial diagnosis, 93% of patients demonstrate radiographic evidence of disease progression. Long-term synovial inflammation can weaken structural stabilizers and can lead to joint degeneration, deformity, or instability.[7] For this reason, early diagnosis and treatment are important to slow disease progression.

Pathophysiology

JIA is a multifactorial lymphocyte-mediated autoimmune disease that targets synovial tissue.[7] The specific etiology remains largely elusive. It is believed that chronic synovial inflammation arises from an imbalance between pro- and anti-inflammatory cytokines, leading to synovial hyperplasia.[7]

Chronic synovitis can lead to irreversible joint destruction. Synovial proliferation stimulates adjacent articular cartilage damage, progressing to bone destruction at the articular margin. Consequently, further recruitment of proinflammatory cells supports a continued inflammatory cycle. The persistent inflammation increases intra-articular pressures and causes the characteristic swelling, warmth, discomfort, limited joint motion, and possible joint contracture and ankylosis. In children, articular hypertrophy can result in premature epiphyseal closure and deformity.[7] Resultant short stature and leg length discrepancy are unfortunate consequences of uncontrolled disease.[8]

Presentation

There are several patterns of JIA presentation that largely depend on disease subtype. Variation in age of onset, associated comorbidities, distribution of involved joints, presence of systemic inflammation, presence of extra-articular inflammation, and the role of family history support identification of familiar disease states. Although children often experience a similar indolent onset of inflammatory joint pain (eg, joint warmth, joint swelling, limited range of motion, stiffness with inactivity, improvement in pain, and stiffness with activity), it often takes an astute clinician to recognize this pattern in younger children. By history, it is common for children with JIA to deny joint pain and lack the ability to verbalize joint stiffness. Exploring surrogate markers for joint stiffness is needed to aid in localizing joint disability, for example, asking about limping in the morning, avoiding the use of the dominant hand, or wanting to be held after napping. The presence of notable nighttime pain, migratory arthritis, or erythematous joints should raise alarm for an alternative diagnosis.

The initial stage of approaching disease presentation begins with categorization as oligoarthritis (fewer than five joints) or polyarthritis (five or more joints) onset (Box 1). Asymmetric joint involvement is most common in younger children. Asymmetric joint involvement in a teenager or symmetric joint involvement in younger children are important features to identify, suggesting less common disease courses. Involvement of the axial spine, midfoot, or widespread enthesitis, particularly in teenage males, is uniquely seen in ERA. The presence of nail dystrophy, psoriasiform patches, dactylitis, or distal interphalangeal joint involvement is seen in patients with psoriatic JIA. Finally, the existence of systemic disease (daily fever, serositis, organomegaly, lymphadenopathy) is rare, but it is the primary feature seen in the systemic subtype.

Oligoarticular JIA is the most common form in North America and Europe, accounting for approximately 46% to 55% of patients with JIA.[2] Oligoarticular JIA classically manifests in a preschool age female with asymmetric arthritis most commonly involving a knee. Accelerated limb growth on the affected side may first be seen with future risk for eventual undergrowth without disease control. A subset of oligoarticular patients will be classified as "extended oligoarticular JIA" if the disease spreads to five or more joints after the first 6 months of disease. The incidence of painless chronic anterior uveitis is highest in this subset of patients, in particular, oligoarticular JIA who are also antinuclear antibody positive.[9] With the exception of uveitis, extra-articular manifestations are less common in oligoarticular JIA.[9] Oligoarticular JIA is uncommon in teenagers. Therefore, older children presenting with few arthritic joints need careful consideration for other JIA subtypes (eg, psoriatic, ERA) or etiologies of chronic arthritis, including sarcoidosis, inflammatory bowel disease, or pigmented villonodular synovitis.

Box 1. Diagnostic Criteria for Juvenile Idiopathic Arthritis[2]

1. Age of onset <16 years
2. Duration of disease >6 weeks
3. Affects ≥1 joint(s)
4. Exclusion of other forms of juvenile arthritis

Definitions of Subtypes

A. Systemic arthritis: Arthritis in 1 or more joints with or preceded by fever of at least 2-week duration that is documented to be daily for at least 3 days, and accompanied by 1 or more of the following:
   i. Evanescent (nonfixed) erythematous rash
   ii. Generalized lymph node enlargement
   iii. Hepatomegaly and/or splenomegaly
   iv. Serositis
B. Oligoarthritis: Arthritis affecting 4 or fewer joints during the first 6 months of disease
   i. Persistent: Affecting not more than 4 joints throughout the disease course
   ii. Extended: Affecting a total of more than 4 joints during the disease course
C. Polyarthritis (rheumatoid factor negative): Arthritis affecting 5 or more joints during the first 6 months of disease. A test for RF is negative
D. Polyarthritis (rheumatoid factor positive): Arthritis affecting 5 or more joints during the first 6 months of disease. 2 or more tests for RF at least 3 months apart during the first 6 months of disease are positive
E. Psoriatic arthritis: Arthritis and psoriasis, or arthritis and at least 2 of the following:
   i. Dactylitis
   ii. Nail pitting or onycholysis
   iii. Psoriasis in a first-degree relative
F. Enthesitis related arthritis: Arthritis and enthesitis, or arthritis or enthesitis with at least 2 of the following:
   i. Presence of or a history of sacroiliac joint tenderness and/or inflammatory lumbosacral pain
   ii. Presence of HLA-B27 antigen
   iii. Onset of arthritis in a male over 6 yr of age
   iv. Acute (symptomatic) anterior uveitis
   v. History of ankylosing spondylitis, enthesitis related arthritis, sacroiliitis with inflammatory bowel disease, Reiter's syndrome, or acute anterior uveitis in a first-degree relative
G. Undifferentiated arthritis: Arthritis that fulfills criteria in no category or in 2 or more of the above categories.

Polyarticular JIA accounts for approximately 14% to 24% of patients with JIA.[2] There are two forms of polyarticular JIA based on the presence (seropositive) or absence (seronegative) of rheumatoid factor. Seronegative disease often develops slowly with bimodal incidence around 1 to 3 years of age and again at 9 to 14 years of age. Seropositive disease tends to develop primarily in older children.[10] Among all children with polyarticular JIA, only 15% are seropositive, highlighting the poor negative predictive value of serologic testing during diagnostic evaluation.[10] Seronegative cases often present with large upper extremity joint involvement, including shoulders, elbows, and wrists with more reliable pharmacologic response and nonsurgical management.

Finally, systemic disease accounts for approximately 7% to 10% of patients with JIA.[2] Systemic disease is defined by extra-articular manifestations, including the presence of daily fevers, rash, lymphadenopathy, hepatosplenomegaly, serositis, and potentially life-threatening macrophage activation syndrome. Systemic JIA follows a monophasic course in some children with long-term remission off medication possible. Most have a chronic course often with resolution of the systemic features with chronic arthritis as the primary ongoing disease manifestation.[11]

Diagnosis

JIA is a clinical diagnosis made by a rheumatologist after confirming the presence of chronic arthritis, either on

physical examination or cross-sectional imaging, in the absence of mimicking processes. Orthopaedic surgeons may be one of the first clinicians to identify the characteristics, provide the referral to a pediatric rheumatologist, and help establish an essential interdisciplinary treatment team.

Diagnosis of all forms of JIA require at least 6 weeks of symptoms in one or more joints in children younger than 16 years at the onset without another identifiable cause.[1] This history of childhood onset distinguishes the diagnosis from adult rheumatoid arthritis or osteoarthritis, which may follow a different disease course.

Identification of active inflammatory arthritis (or enthesitis in the ERA subtype) is typically established on physical examination. Corresponding limb length discrepancies, contractures, and muscle atrophy support chronicity when found. Diagnostic imaging, including ultrasonography or MRI, are commonly used to establish the presence of active arthritis. In addition to visualizing joint effusions, synovial hypertrophy, and increased Doppler signal support active synovitis. MRI is ideally ordered with intravenous contrast when inflammatory arthritis is high on the differential, as contrast enhancement will increase test sensitivity. Radiograph and CT imaging are helpful for interrogating joints for structural changes and joint effusions. Considering many children present shortly after symptom onset, structural changes are fortunately absent in many at diagnosis. Consequently, radiographs and CT lack the sensitivity of MRI to discriminate between active disease and changes from prior disease activity, limiting their diagnostic value.

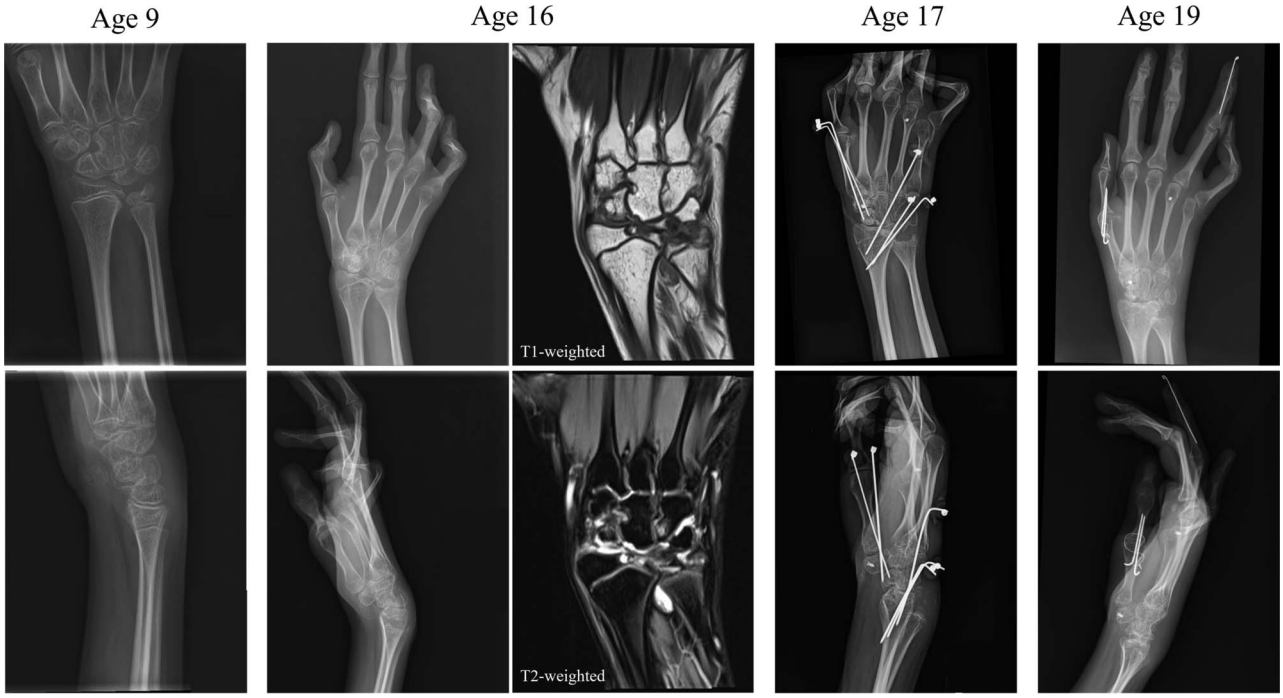
Radiographs are quite helpful to obtain a baseline in patients with wrist involvement or joint contractures as a comparator to monitor disease progression (Figure 1). Early radiographic findings in JIA include periarticular osteopenia, joint effusions, and juxtasynovial soft-tissue swelling (Figure 2).[12] Late-stage, progressive disease may demonstrate severe arthritic changes, bony ankylosis, and angular deformity. In 13 patients with polyarticular JIA, Mason et al[13] found that 46% demonstrated radiographic evidence of disease progression within 8 to 25 months of follow-up. Finally, imaging is universally employed in children with suspected sacroiliac joint involvement to confidently diagnose sacroiliac joint arthritis. There is not a widely accepted radiographic classification system to monitor damage progression in children and adolescents. Synovial biopsy is reserved for rare cases where validity of clinical diagnosis comes into question, such as treatment resistant monoarthritis.

Laboratory studies are complementary but are neither sensitive nor specific for the diagnosis of JIA. Considering the poor negative predictive value, clinicians should not be falsely reassured by negative serologies, including antinuclear antibody (ANA), rheumatoid factor, or Anti-CCP in children they suspect may have JIA. These biologic markers primarily help to classify JIA subtype and stratify risk for disease complications. In the setting of polyarticular and systemic disease, laboratory studies typically demonstrate elevated erythrocyte sedimentation rate and C-reactive protein; these laboratory values are often within normal limits in the setting of oligoarticular disease. Conversely, ANA is positive in up to 80% of children with oligoarticular diseases, whereas only 30% of children with polyarticular disease may be ANA positive. HLA-B27 positivity is useful in the diagnosis of ERA JIA. Consideration of additional serum or synovial analysis for other causes of chronic arthritis may be necessary in the appropriate clinical context. Common clinical situations include screening for Lyme disease in endemic areas or systemic lupus screening in a patient with additional disease features. Synovial fluid testing is usually limited to a cell count supporting an inflammatory process and a Gram stain and culture when infectious arthritis is a consideration. Crystal analysis is rarely indicated in the pediatric population.

Nonsurgical Management

Comprehensive treatment of JIA of the upper extremity requires a multidisciplinary treatment team, including, but not limited to, rheumatologists, surgeons, physical and occupational therapists, and mental health counselors (Figure 3). Nonsurgical treatment modalities include nonpharmacologic and pharmacologic strategies. The goal of this therapy is to alleviate symptoms, but just as importantly, to quell the inflammatory process and maintain disease inactivity. After 6 months of inactivity, patients are considered in remission (on or off medication) with many reporting absence of daily JIA symptoms. Timely control will ideally forestall articular damage and maintain functional capacity. Unfortunately, despite widely available biologic agents over the past 20 years, a large percentage of children may not achieve long-term clinical remission.[11] Considering there is no cure for JIA, patients may accrue joint damage either during persistent disease activity or during alternating periods of disease activity and inactivity.

Figure 1



Hand and wrist radiographs demonstrating progressive degenerative changes in a patient with juvenile idiopathic arthritis between the ages of 9 and 19 years. Progression from early radiographic findings in juvenile idiopathic arthritis (ie, periarticular osteopenia) to findings of late-stage disease (ie, severe arthritic changes, bony ankylosis, and angular deformity) are appreciable. T1- and T2-weighted magnetic resonance imaging at age 16 years further demonstrates a mixture of characteristic early-stage (ie, joint effusion, synovitis), and late-stage findings (ie, periarticular erosions). Because of debilitating pain at the time of skeletal maturity, the patient eventually underwent multiple soft-tissue, reconstructive, and fusion surgeries in her digits and carpal bones.

Physical and Occupational Therapy

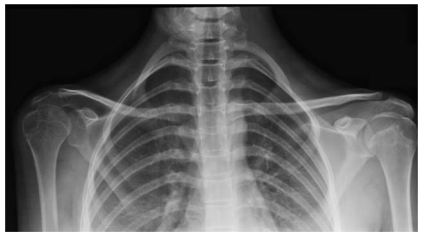
Rehabilitation with assistance from skilled physical and occupational therapists is the cornerstone of non-pharmacologic management for JIA of the upper extremity. Some children may not be able to participate in rehabilitation until achieving a reduction in disease burden pharmacologically. Standard rehabilitation focuses on overall physiologic function (such as strength and range of motion), as well as task-oriented training (such as writing).[15]

The advent of modern technologies has advanced approaches to standard rehabilitation practices for JIA of the upper extremity. One such technology is video games–based task-oriented activity training.[16] In a recent, randomized, clinical trial comparing the effects of using real versus video games–based rehabilitation techniques on activity performance in children with JIA, the children treated with video games–based task-oriented activity training demonstrated greater hand strength and functioning, pinch strength, performance, and satisfaction with self-care and productivity.[16]

Nonbiological Therapies

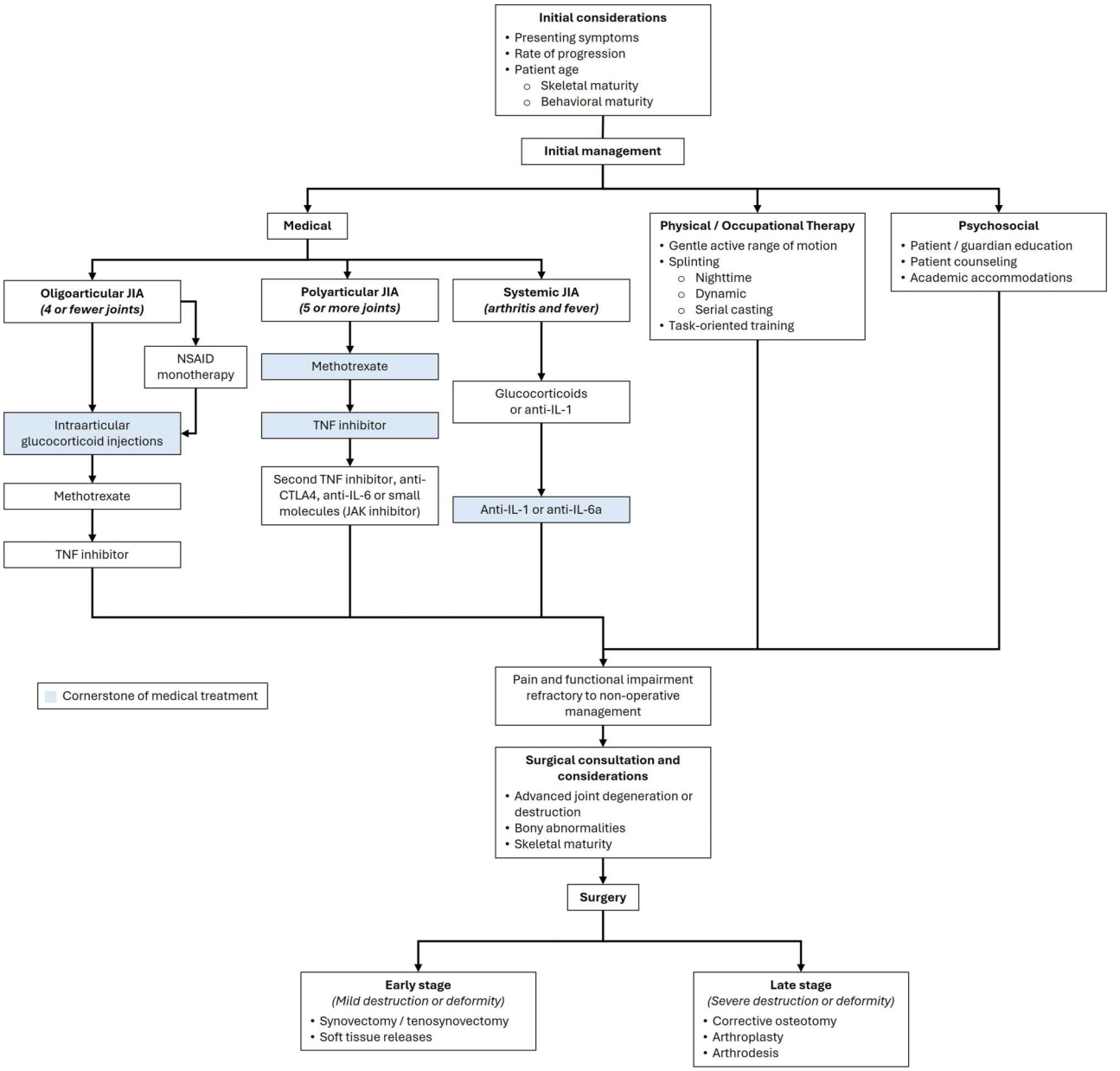
Nonsteroidal anti-inflammatory drugs, intra-articular corticosteroid injections, short courses of systemic steroids, and synthetic (nonbiologic) disease-modifying antirheumatic drugs (DMARDs), such as methotrexate, remain the first-line pharmacologic treatment modalities for JIA.

Figure 2



Shoulder radiograph of a 16-year-old patient with juvenile idiopathic arthritis demonstrating characteristic periarticular osteopenia and early erosions of the right humeral head consistent with early-stages disease. The right shoulder has remained asymptomatic through age 26 years—the age at most recent follow-up.

Figure 3



Flowchart showing the general approach to the management of patients with juvenile idiopathic arthritis. Modified from Glueck and Gellman and Martini et al.[2,14] NSAIDs = nonsteroidal anti-inflammatory drugs

The choice of pharmacologic intervention depends on the extent of articular involvement. Children with polyarticular disease generally receive systemic treatment in the form of methotrexate and biologic DMARDs as initial therapy, although initial treatment in oligoarticular disease involves more localized therapies, such as intra-articular steroid injections.[17] Systemic therapy is indicated for recalcitrant or recurrent oligoarthritis.[17]

Methotrexate is the synthetic DMARD with the most quality evidence to date demonstrating efficacy in the treatment of JIA. The 2019 American College of Rheumatology and Arthritis Foundation Guideline for Treatment of JIA recommends methotrexate as the initial therapy for polyarticular JIA over nonsteroidal anti-inflammatory drug monotherapy.[18] The adverse effects must be considered when determining a pharmacologic treatment plan. Methotrexate, for instance, is associated with high rates of intolerance, manifesting as nausea and vomiting in up to 50% of children.[19,20] The incidence of methotrexate intolerance is independent of

route of delivery.[19] Children receiving oral methotrexate are more likely to report drug-induced nausea, but subcutaneous methotrexate is associated with higher rates or anticipatory nausea before and during administration. If disease inactivity is achieved on methotrexate, but intolerance limits continued administration, a transition to leflunomide or sulfasalazine can be considered.[18]

Corticosteroid Injections

The use of intra-articular corticosteroid injections in the form of triamcinolone hexacetonide has been recommended by the American College of Rheumatology for the treatment of oligoarticular JIA.[21] Triamcinolone hexacetonide is the preferred injectate over triamcinolone acetonide for its longer duration of action and more favorable adverse effect profile.[22] Efficacy as monotherapy is highest for oligoarticular JIA where it may induce a durable remission. Recalcitrant or recurrent oligoarthritis patients will require escalation to systemic therapy. The therapeutic results, however, are typically short-lived. In a study of 46 children who had received intra-articular steroids for a flare of JIA, 49% restarted systemic therapy by 6 months following their injection and 70% by 12 months post injection.[17] Interestingly, prior systemic treatment with a biologic DMARD was the only identified predicter of resuming systemic therapy.[17] Therefore, utilization of intra-articular corticosteroids in polyarthritis is reserved for the most problematic joints, typically in the upper extremities, as adjunct therapy while awaiting therapeutic response to systemic therapy.

Biological Therapies

The use of biological therapies, also called biologic DMARDs, has markedly increased over recent years due to their ability to target specific cytokines (eg, interleukin-1 [IL-1], tumor necrosis factor [TNF]), receptors (eg, IL6R, CTLA4), signaling molecules (eg, Janus kinases [JAKs]), and cells (eg, B cells) implicated in the pathophysiology of JIA.[23] TNF inhibitors (eg, adalimumab, etanercept, infliximab) are the most widely used biologic DMARDs in JIA due primarily to their efficacy, rapid onset of action, and safety profile.

Unfortunately, approximately half of children with polyarticular JIA will require escalation to a biologic DMARD to achieve disease inactivity.[24] The risk of accumulating joint damage is highest during active disease; radiographic progression in the wrist has been shown to be highest within the first year of JIA diagnosis.[25] As a result, the standard "step-up" therapy approach of first trialing a synthetic DMARD before escalation to a biologic DMARD agent for polyarticular JIA patients has been questioned. The TREAT study was a double-blinded placebo-controlled trial of 85 patients with JIA randomized to either methotrexate with placebo etanercept and placebo prednisolone or methotrexate with etanercept and a prednisolone taper at diagnosis. Disease inactivity was achieved in 40% of children in the treatment arm compared with 23% in the control arm at month 6.[26]

A recent Canadian study reported on differences in treatment and outcomes among children diagnosed with JIA between 2005 and 2010 compared with children diagnosed between 2017 and 2021. One study outcome reported on cumulative exposure to a biologic agent and disease inactivity within 70 weeks of diagnosis. Comparing 228 seronegative polyarticular JIA patients in the early group to 131 in the later group, the percentage of patients receiving a biologic DMARD increased from 5% to 43% with attainment of inactive disease increasing from 53% to 85%.[27]

Although many children achieve disease remission on systemic therapy, there is a relatively high rate of relapse after discontinuation—varying by subtype.[11,27] The benefit of biologic agents are felt to outweigh associated risks. Because of increased immune suppression over nonbiologic DMARDs, incidence of infection is the most reported adverse event. Fortunately, serious adverse events are rare in robust pharmacosurveillance registries.[28] TNF inhibitors carry a black box warning about an increased risk for cancer. However, it is known that children with chronic inflammatory diseases experience higher rates of malignancy. After adjusting for this, TNF inhibitors do not appear to increase this risk markedly further.[29]

Surgical Management in the Upper Extremity and Joint-specific Considerations

Surgery is indicated for JIA when nonsurgical management alone can no longer effectively manage symptoms and disease sequelae. The two primary goals of surgical management are delay of joint degeneration and symptom alleviation.[30] Broadly, options for surgical treatment include tenosynovectomy, soft-tissue release, joint reconstruction, corrective osteotomies, and arthrodesis.

It is ideal to delay reconstructive surgery and arthrodesis until skeletal maturity.

When preoperative planning, it is critical to ensure that the arthritis is inactive before surgery. The determination of JIA inactivity is based on the absence of clinical symptoms (eg, pain, stiffness, swelling) with a reassuring clinical examination and normal inflammatory markers (ie, C-reactive protein and erythrocyte sedimentation rate). If there is any doubt, an MRI or ultrasonography should be obtained.

It is common that patients with JIA are taking antirheumatic medications in the perioperative setting. To balance the increased risk of infection with the risk of disease flares, guidelines for the perioperative management of these medications in JIA of the upper extremity have been extrapolated from the total hip and knee arthroplasty literature.[31] Methotrexate, and other DMARDs, may be continued through the time of surgery. Most biologics should be held for one full cycle before surgery based on the drug-specific dosing schedule; JAK inhibitors should be held 3 days before surgery. Guidelines recommend restarting antirheumatic medications after the surgical incision has healed, in the absence of signs of infection (ie, swelling, erythema, drainage)—typically around 14 days following surgery.

The following section discusses joint-specific considerations in surgical management.

Wrist

Upon initial presentation during childhood, approximately 36% of patients with JIA have wrist involvement; 80% of patients develop wrist symptoms overtime.[5] In their study of 152 school-age children with JIA, 54% reported difficulties at school related to their wrist and/or hand involvement.[3] Children with JIA were less able to sustain handwriting for longer than 5 minutes compared with their peers due to associated pain and lower stamina.[32]

The wrist is the second most common location for growth abnormality in children with JIA, with reported rates up to 50%.[5] The most common deformities of the wrist include ulnar deviation (29%) and flexion contracture (29%; Figure 4).[4] Deformity has been most strongly associated with older age at presentation, longer duration of disease at presentation, and polyarticular disease.[4] Radiographically, these abnormalities appear as premature carpal ossification, intercarpal space narrowing, and early ulnar epiphysis fusion with resultant ulnar shortening.[4]

Following nonsurgical management, open or arthroscopic synovectomy can be considered. Synovectomy of the wrist and metacarpophalangeal joints are among the most common procedures performed in patients with JIA. Indications for synovectomy in JIA include absent or minimal articular changes on plain radiograph, with moderate pain, loss of motion, and contracture recalcitrant to nonsurgical treatment modalities. Contraindications to synovectomy include severe articular cartilage destruction, multiple joint involvement, and active inflammatory or systemic disease. Better outcomes following synovectomy in JIA of the wrist are associated with earlier treatment, oligoarticular disease, and the absence of systemic inflammation.[33]

Thorough synovectomy should include the radiocarpal joint, ulnar head, and all intercarpal joints. Studies have noted an overall mean improvement in pain, grip strength, and functionality; however, synovectomy has been associated with decreased range of motion through the wrist and radiographic evidence of progressive joint degeneration over time.[30] As an epiphyseal-sparing surgery, synovectomy can be considered if necessary in skeletally immature patients.

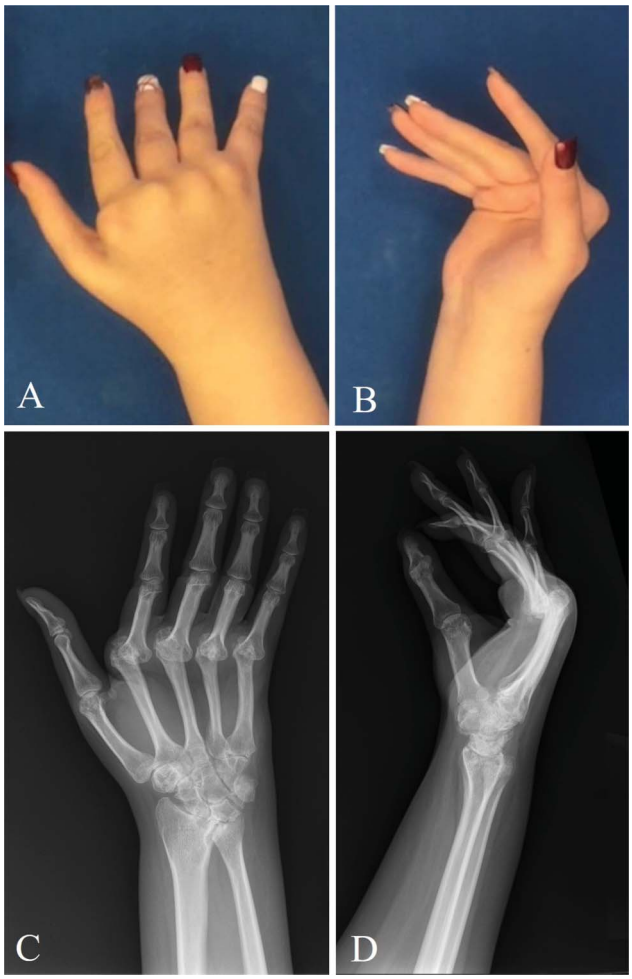
In addition to synovitis, JIA may lead to hypermobility secondary to increased ligamentous laxity and degeneration. Wharton et al[34] reported on a cohort of 18 children and adolescents who underwent arthroscopic thermal capsular shrinkage for midcarpal instability in the setting of JIA. Compared with controls who underwent arthroscopic synovectomy alone, capsular shrinkage was associated with greater improvement in pain scores at rest and with loading, as well as improved stability.[34] No complications were reported. The authors concluded that arthroscopic capsular shrinkage appears to be a safe and effective procedure for palmar midcarpal instability in children and adolescents with JIA.[34]

Dorsal osteotomies and ulnar lengthening can also be performed to address deformity of the radioulnar joint to reduce the risk of early carpal subluxation and dislocation. There is limited literature regarding total wrist arthroplasty or arthrodesis for JIA. Current studies are limited to case reports or case series that report aggregate outcomes in children with a conglomeration of wrist pathologies.[35,36]

Metacarpophalangeal Joints

Involvement of the MCP joints has been reported in approximately 50% of patients with JIA.[5] Orthoses for

Figure 4



A, **B**, Clinical photographs demonstrating characteristic ulnar deviation of the metacarpophalangeal joints in an 18-year-old with juvenile idiopathic arthritis. **C**, Anterior-posterior and (**D**) lateral radiographs demonstrating arthritic changes and notable metacarpophalangeal subluxation.

the wrist and/or hand are common in the nonsurgical management of JIA. There is controversy regarding the type of support used in the setting of inflammatory arthropathies. Static orthoses have been shown to be effective for soft-tissue rest during active inflammatory flares. However, the literature is sparse regarding the efficacy of static orthoses in preventing deformity or adjacent joint disease.

Synovectomy and release of collateral ligaments may be indicated for pain and stiffness. The condition of the articular cartilage is an indicator of good outcomes following synovectomy.

Arthroplasty has been used to improve function in the setting of MCP subluxation (Figure 5). Paul et al reported two cases of children with severe progressive JIA who underwent MCP joint arthroplasty at ages 21 and 14 years, respectively. One case used pyrocarbon implants, and another case used a silicone-based implants.[37] At 7-year follow-up, both patients had pain relief and improved hand function overall, but silicone arthroplasty performed better radiographically than pyrocarbon.[37]

Interphalangeal Joints

Approximately 49% and 19% of patients with JIA have involvement of the proximal interphalangeal (PIP) and distal interphalangeal (DIP) joints, respectively.[5] Swelling and inflammation within the flexor tendon sheath is typical upon presentation. Medications and steroid injections can reduce inflammatory progression and pain; however, flexion contractures are common, especially at the PIP joint.

Splinting and serial casting can be performed to attempt tissue elongation and restoration of extension in the setting of flexion contractures. In a retrospective study of 49 fingers from children and adults with JIA and adult rheumatoid arthritis treated with serial casting for PIP joint flexion contractures, serial casting was associated with a 27° improvement in extension and varied in magnitude depending on the initial magnitude of contracture.[38]

Flexor synovectomy, arthroplasty, or arthrodesis may be considered for recurrent triggering or severe deformity. However, outcome data in the JIA population are limited.

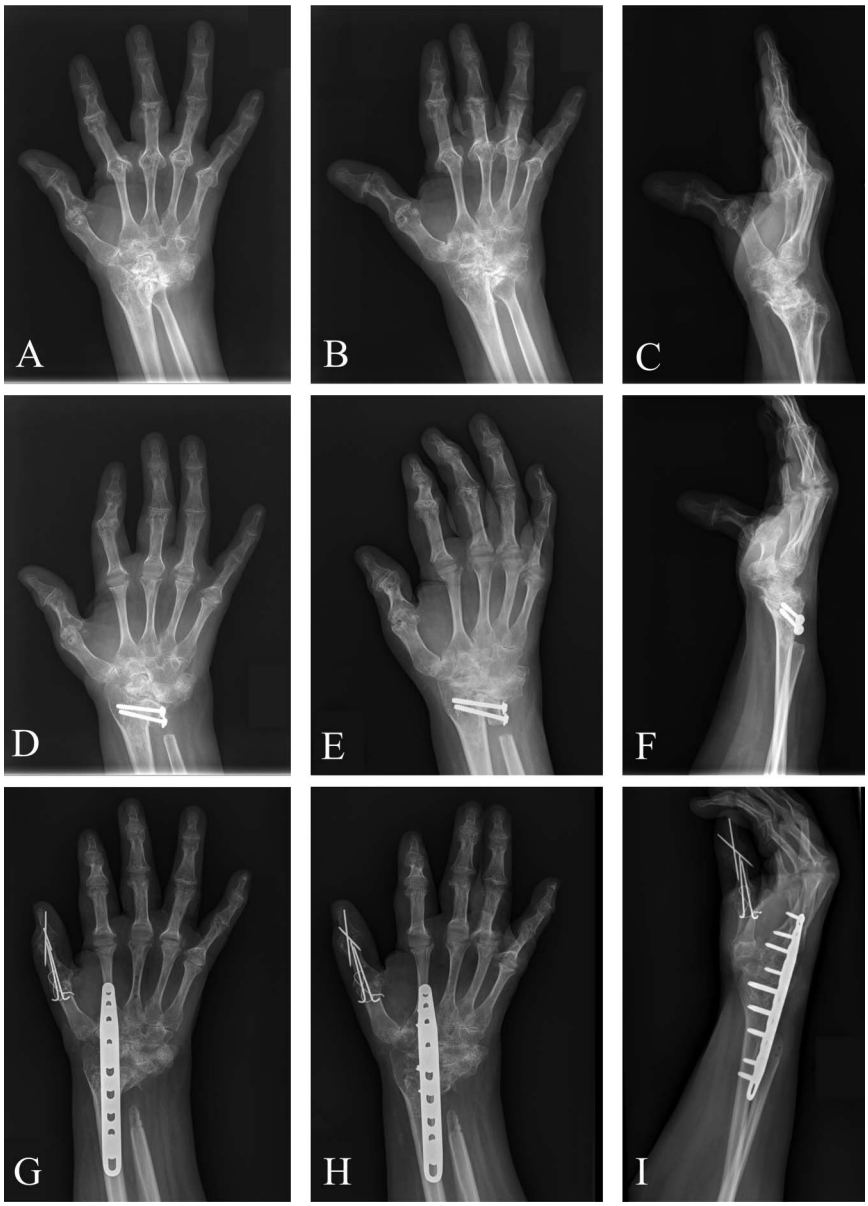
Elbow

Elbow involvement in JIA typically presents as pain and stiffness, characterized initially by diminished extension with a subsequent decline in flexion, pronation, and supination. Symptoms are strongly associated with progressive destruction and deformity of the radiohumeral joint.

After maximizing nonsurgical management, synovectomy with radial head excision can be performed in the setting of persistent synovitis and destruction of the radiocapitellar and proximal radioulnar joint with favorable results.

Although technically challenging and contraindicated in young patients, total elbow arthroplasty (TEA) has demonstrated favorable results in adults with advanced JIA.[39] Baghdadi et al reported the outcomes in 29 elbows with mean age at surgery of 37 years and at mean 10.5-year follow-up after primary semiconstrained TEA. Twenty-two elbows (76%) had subjective functional satisfaction and 18 (62%) were graded as excellent or good

Figure 5



Hand radiographs from a 35-year-old patienwith severe deformity in the setting of juvenile idiopathic arthritis. **A**–**C**, Preoperative radiographs demonstrate diffuse chronic inflammatory arthritis involving her wrist and digits. She initially underwent (**D**–**F**) a Sauve-Kapandji procedure and index through small finger metacarpophalangeal arthroplasties with use of silicone implants, and subsequent (**G**–**I**) total wrist arthrodesis, first metacarpophalangeal arthrodesis, and index and long finger proximal interphalangeal joint silicone arthroplasties. Anterior-posterior (left), oblique (middle), and lateral views (right).

results based on mean Mayo Elbow Performance Score. Revision surgery was performed on 28% of the elbows; the rate of TEA survival from revision was 96.4% and 79.9% at 5 and 10 years, respectively.[39]

Shoulder

Shoulder involvement in patients with JIA increases from fewer than 5% at initial presentation to approximately 21% with disease progression into adulthood.[5] Compared with the general population, individuals with JIA have an increased propensity for glenoid erosion.

There is limited evidence for the use of nonsalvage procedures, such as shoulder arthroscopy, in the management of JIA of the shoulder. Fortunately, symptomatic shoulder involvement typically occurs later into adulthood when arthroplasty is more appropriate. Options for shoulder arthroplasty—stemmed versus

resurfacing hemiarthroplasty, anatomic versus reverse total shoulder arthroplasty—and outcomes in patients with JIA are consistent with those of the general population.[40] Notably, in patients with JIA, total shoulder arthroplasty compared with hemiarthroplasty has been associated with better pain relief, patient-reported outcomes, and durability (90% vs. 72% 30-year implant survivability).[40]

Summary

JIA is a complex chronic inflammatory disease presenting in children before their 16th birthday. Upper extremity involvement is associated with pain and functional limitation. Diagnosis is based on clinical presentation. Early diagnosis and initiation of treatment are essential to prevent progressive joint degeneration and long-term dysfunction. Lifelong multidisciplinary care is important in most cases to preserve functionality and mitigate discomfort. Treatment involves pharmacologic intervention, physical and occupational therapies, and surgery. Increasing pharmacologic options have drastically improved the ability to achieve disease inactivity along with a generally favorable safety profile. Although surgery should be avoided in skeletally immature patients, surgery should be performed when necessary to avoid lifelong articular and soft-tissue damage and improve patient quality of life. Overall, there is a paucity of literature regarding long-term outcomes in the surgical management JIA in children and adolescents.

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