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A Review of Juvenile Rheumatoid Arthritis

Jill C. Ballan

University of North Dakota

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A REVIEW OF JUVENILE RHEUMATOID ARTHRITIS

by

Jill C. Ballan
Bachelor of Science in Physical Therapy
University of North Dakota, 1995

An Independent Study
Submitted to the Graduate Faculty of the
Department of Physical Therapy
School of Medicine
University of North Dakota
in partial fulfillment of the requirements
for the degree of
Master of Physical Therapy

Grand Forks, North Dakota
May
1996
This Independent Study, submitted by Jill C. Ballan in partial fulfillment of the requirements for the Degree of Master of Physical Therapy from the University of North Dakota, has been read by the Faculty Preceptor, Advisor, and Chairperson of Physical Therapy under whom the work has been done and is hereby approved.

(Graduate School Advisor)

(Faculty Preceptor)

(Chairperson, Physical Therapy)
PERMISSION

Title A Review of Juvenile Rheumatoid Arthritis

Department Physical Therapy

Degree Masters of Physical Therapy

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Date 12-7-95
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ACKNOWLEDGEMENTS

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ABSTRACT

Juvenile rheumatoid arthritis (JRA) is the most common pediatric rheumatoid disease in North America and a principle cause of childhood disability. The disease course is unremitting in up to a quarter of the children with JRA, and a small percentage will grow-up with severe functional disabilities. Physical and occupational therapy, along with prescribed drug and orthopedic provisions, are important in the management of JRA. The therapists, and other medical professionals involved, work toward the following goals: decreasing joint inflammation, relieving pain, achieving or maintaining an optimal level of function, and educating the patient and family on the disease course and the required care. The focus of this study was to analyze the therapy programs currently used to treat JRA and the effects of early therapeutic intervention. The literature review indicated that therapy should begin as early and as decisively as possible to promote remission of JRA. Aquatic therapy is the method that has provided optimal results.
CHAPTER ONE

INTRODUCTION

In 1897 George Frederick Still\textsuperscript{1} published the first English-language description of chronic juvenile arthritis. Arthritis is defined as swelling or decrease in joint motion with heat, pain, or tenderness.\textsuperscript{2-4} In the United States juvenile rheumatoid arthritis (JRA) is a term used to describe a chronic condition affecting the joints, which has its onset during childhood or adolescence. The term juvenile rheumatoid arthritis suggests that it is a childhood version of adult rheumatoid arthritis (RA).\textsuperscript{2,5} However, certain expressions of JRA bear no resemblance to those of adult RA.\textsuperscript{6,7} Chronic eye inflammation is common in JRA and is not a symptom of adult RA. Wrist subluxation and radial deviation with ulnar deviation of the fingers are common wrist and hand deformities found in JRA. Wrist ulnar deviation and finger radial deviation is the pattern most often found in adults with RA. In addition, rheumatoid factor (RF), an abnormal immune complex in the blood, is found in less than 15\% of JRA patients, but is seen in 80\% to 85\% of adult RA patients.\textsuperscript{6,8} The most prevalent commonality between adult RA and JRA is chronic inflammation of the joints.

JRA is divided into three subtypes defined according to the symptoms present during the first 6 months of the disease.\textsuperscript{1,2,5,6,9,10} The three types are systemic-onset, polyarticular-onset, and pauciarticular-onset. The determination of subtype is made by the
presence of systemic systems, the results of antinuclear antibody and rheumatoid factor studies, and the type and number of joints involved\textsuperscript{9} (Table 1).

JRA is the most common childhood rheumatoid disease in North America and is a major contributor to childhood disability.\textsuperscript{3,6,11-15} JRA has an annual incidence of approximately 11 cases per 100 000 in the United States.\textsuperscript{5,9,12,16,17} A study by Gewanter and associates\textsuperscript{18} indicated a prevalence of 50 cases per 100 000 children in the United States. Cassidy reported in a study completed at the Mayo Clinic a prevalence as high as 113.4 cases per 100 000 individuals.\textsuperscript{11} JRA affects anywhere from 30 000 to 200 000 children in the United States.\textsuperscript{2,5,8-10,18} The age and sex of those children affected with JRA varies with each subtype.\textsuperscript{2,5,6,10}

The cause of JRA is unknown.\textsuperscript{4,8} It has been suggested that there is a genetic predisposition to the development of JRA. There is also evidence that infections or defects in the body's immune system, emotional trauma, or physical trauma may be responsible for onset or exacerbation of JRA.\textsuperscript{4,8}

Most children with JRA go into remission by adulthood, with three out of four having a favorable future.\textsuperscript{4,5,8,19} Twenty-five percent of the children with an unremitting disease course may suffer serious musculoskeletal deformities and functional limitations.\textsuperscript{1,19} Recently, it has become more apparent that the prediction of long-term disease activity and eventual functional status in JRA is reliant on the subtype and course experienced by the child.\textsuperscript{8} Brewer and associates\textsuperscript{8} cited several studies that indicated
Table 1. Classification of juvenile rheumatoid arthritis

<table>
<thead>
<tr>
<th>Subgroup</th>
<th>Age at Onset b</th>
<th>Sex Ratio a</th>
<th>Joints Involved b</th>
<th>Rheumatoid Factor a</th>
<th>Antinuclear Antibodies a</th>
</tr>
</thead>
<tbody>
<tr>
<td>Systemic</td>
<td>childhood</td>
<td>equal</td>
<td>multiple</td>
<td>negative</td>
<td>negative</td>
</tr>
<tr>
<td>Polyarticular RF negative</td>
<td>childhood</td>
<td>females&gt;males</td>
<td>multiple</td>
<td>negative</td>
<td>25% positive</td>
</tr>
<tr>
<td>Polyarticular RF positive</td>
<td>late childhood</td>
<td>females&gt;males</td>
<td>multiple</td>
<td>positive</td>
<td>75% positive</td>
</tr>
<tr>
<td>Pauciarticular early-onset</td>
<td>early childhood</td>
<td>females&gt;males</td>
<td>large joints</td>
<td>negative</td>
<td>negative</td>
</tr>
<tr>
<td>Pauciarticular late-onset</td>
<td>late childhood</td>
<td>males&gt;females</td>
<td>large joints</td>
<td>negative</td>
<td>negative</td>
</tr>
</tbody>
</table>

RF - rheumatoid factor


b Adapted from Chapman RW. Diagnosis of juvenile rheumatoid arthritis. J Am Board Fam Pract. 1995;8:47.
those who have no systemic symptoms, and whose disease began after the age of 5, have an encouraging long-term prognosis. The results of a study by Wallace and Levinson\textsuperscript{20} showed a .29\% to 1.1\% mortality rate in patients with JRA in Canada and the United States.\textsuperscript{21}

According to Riggs and Gall,\textsuperscript{22} "The objective of rehabilitation of the child with JRA is to minimize disabilities and to maintain or restore the patient to as normal a functional state as possible." In children the "normal functional state" is changing rapidly. Expanding psychosocial, educational and physical abilities assist the child in becoming a self-sufficient, constructive adult. Chronic illness and physical disability can significantly impact the child's movement toward adulthood. The influence of the disease on development relies on the age when illness began, the severity, the length and the outcome of the disease, the child's response to the illness, and the environment the child is in. Health care professionals responsible for the patient's care must be aware of all of the patient's needs and goals in order to devise a comprehensive treatment program. Prompt diagnosis and an immediate initiation of appropriate treatment are important.\textsuperscript{7}

The purpose of this paper is to give an overview of JRA and to discuss the various treatment approaches available. The early diagnosis and therapeutic intervention will be discussed. Also, recommendations for preferred methods of rehabilitation will be provided.
CHAPTER TWO
DISEASE COURSE AND TREATMENT

Types of JRA

Systemic

Systemic-onset JRA, also known as Still's Disease, can appear at any age but most frequently occurs in those under the age of 10.\(^2,3,6\) Before the age of 4, almost half of the patients will have had their first experience with this condition and two-thirds of the patients will be less than 6 years old when onset occurs.\(^5\) Ten percent to 20% of the patients with JRA are classified under systemic type and are found to have a nearly equal sex ratio.\(^2,10\) Generally, lab tests are negative for rheumatoid factor (RF) and antinuclear antibodies (ANA).

By definition, systemic JRA begins with a high swinging fever, along with other systemic symptoms including a maculopapular rash, enlarged lymph nodes, hepatosplenomegaly, leukocytosis, anemia, myalgia and arthralgia.\(^2,5-7,23\) Pericarditis, an inflammation of the sac around the heart, that occurs almost solely in systemic disease and pleurisy are also common. However, pleurisy and pericarditis seldom cause serious trouble.

The fever of systemic JRA has a trademark course. It is typically a swinging fever
that peaks above 39.5° C each day, usually spiking in the afternoon, followed by an abrupt fall to normal or subnormal temperatures.\textsuperscript{2,5,6} This fever continues for weeks or months at a time over the first 6 months of the illness. This often results in the child being irritable, fretful and even anorexic.\textsuperscript{2,22} Sometimes there is an evanescent, red rash, commonly called the rheumatoid rash, that accompanies the fever. This rash is generally neither painful nor itchy.

Arthralgia, myalgia and transient arthritis frequently plague the young patient, especially when the fever peaks.\textsuperscript{2,5,10} Movement is painful, often resulting in diminished movement leading to muscle wasting, and contractures. Fatigue and poor endurance also become problems.

Every person with systemic JRA eventually develops arthritis.\textsuperscript{5} Approximately 60\% of the children will develop arthritis in five or more joints (polyarticular), with 35\% of those cases remitting with little joint damage.\textsuperscript{2,6} About 25\% of patients ultimately develop destructive, severe chronic arthritis that lasts beyond the systemic manifestations. Approximately 40\% will develop arthritis in four or fewer joints (pauciarticular) with full remission for most.

Within 2 years of onset, 90\% of the children with systemic JRA are in remission, usually without any persistent involvement of the joints.\textsuperscript{22} However, active arthritis into young adulthood is evident in 10\% with major articular disabilities and general growth retardation frequently seen.\textsuperscript{5,22} To help prevent or slow the loss of joint function it is essential to have a well-planned therapeutic exercise program during the active inflammatory stage of the joints.
Polyarticular

Polyarticular-onset (rheumatoid factor positive) is the type of JRA that is similar to adult rheumatoid arthritis clinically and is considered the childhood equivalent.\(^2,5,6\) This condition starts with inflammation of five or more joints as the predominant symptom, excluding the swinging fever or rheumatoid rash. Polyarticular-onset occurs in almost 40% of all children with JRA.\(^3,6,7,10\) Patients that develop polyarticular JRA tend to be older at the time of their first experience with the disease. Children diagnosed with polyarticular JRA may test either rheumatoid factor positive or rheumatoid factor negative. The ratio of girls to boys may be as high as 4:1 for both RF positive and RF negative patients.

Children with this subtype who test positive for RF have an altered clinical picture from those who test negative for RF.\(^2,6,8,11\) The seropositive course often occurs between ages 10 to 15 and tends to run a prolonged course, similar to the adult form. Patients may experience some systemic symptoms including low-grade fever and malaise.\(^2,10\) Polyarticular JRA carries the worst prognosis with 50% to 70% entering adulthood with major articular disabilities.\(^9,22\) Fortunately, of all JRA patients, this subgroup makes up only 10%. The seronegative course may occur at any age, but the peak age of onset is 3 years. It is estimated that 25% to 40% of all JRA patients have this form of polyarticular JRA.\(^2,6,7,11\) The majority have a milder course of arthritis with a greater prospect for remission than the positive RF group.\(^5\) These individuals appear to be less vulnerable to early destructive changes with only 20% having significant articular disabilities into
adulthood.\textsuperscript{5,22}

The pattern of joint involvement in polyarticular JRA follows a similar symmetric pattern as that of adult rheumatoid arthritis.\textsuperscript{2,5,6} In order of most frequent to least involvement, these joints include: hands, wrists, feet, knees, ankles, elbows, cervical spine, temporomandibular joint, shoulders, and hips. Patients are also prone to inflammation of the tendon sheaths which can lead to the formation of ganglions. “Morning stiffness” and gelling (difficulty straightening out joints that have been in one position over an extended period of time) are characteristic of polyarticular JRA. Patients may get relief from a hot bath or with a conscious increase in active movement.\textsuperscript{5} As the inflammatory process diminishes so will these symptoms. Eye inflammation may occur, and is recognized by the following: blurred vision (or other visual loss), eye redness, eye pain, and discomfort when exposed to bright lights. These symptoms require attention from an eye specialist.

The joint cartilage can undergo degenerative changes if inappropriate stress is applied anatomically and/or functionally.\textsuperscript{5} Much of the joint destruction and disablement that occurs in polyarticular JRA can be prevented. Mechanical integrity can be maintained with a preventative program of treatment and maintaining a proper exercise regime is a prime determinant of future outcome.

\textbf{Pauciarticular}

This subgroup of individuals develops arthritis in no more than four joints during the first 6 months from onset.\textsuperscript{5,7,10,11} Pauciarticular JRA affects 40% to 50% of all children
with JRA. Two subsets of this group have been defined as young-onset and late-onset, separable by immunologic markers, sex ratios and age of onset.2

The first subset, young-onset (generally before age 6), accounts for one-third of all children with JRA.2,10 This group consists largely of girls who test positive for antinuclear antibodies.5,7 Insidious articular involvement typically affects some combination of the knees, ankles, and elbows (middle-size joints) with 74% having monarticular onset and 50% having onset affecting the knee.22 However, it is typical for the arthritis to remit in 1 to 3 years in 60% to 75% of the patients.3 Children have not been recorded as reporting this type of arthritis as painful, however the joints are swollen, warm, and often show a decrease in range of motion.2 This group has a high risk (over 50%) of developing inflammation of the anterior uveal tract, iridocyclitis, which has few symptoms and can easily go undetected. The condition can eventually lead to scarring, and without detection or treatment, approximately 20% of the cases will result in visual loss or blindness.2,6,10,22 Iridocyclitis is responsive to treatment if identified early and controlled. A slit-lamp examination can be utilized to diagnose iridocyclitis.2,8 Periodic eye examinations are recommended due to the risk in the other subgroups of JRA as well.

The late-onset subgroup is largely made up of boys (4 or 5:1 over girls) with age of onset usually between 9 and 15 years. It accounts for only 15% of the pauciarticular group.2 Late-onset pauciarticular JRA can be identified by the immunologic tissue marker called HLA-B27.2,5,10 Late-onset JRA children are predominantly affected in the lower extremity joints in an asymmetrical pattern with the hip joint being most common followed
by the knee, ankle and shoulder girdle (large joints). This arthritis is normally mild and episodic. These patients are also vulnerable to ankylosing spondylitis as well as enthesopathy (inflammation at the bony attachment of ligaments and tendons). Most late-onset patients report joint pain, stiffness and muscular aching. The late-onset group is prone to a condition of the eye as well: acute iridocyclitis. A sudden onset with severe painful eye symptoms is typical, and with proper treatment, patients in this subset tend to heal without any long-lasting residual damage. Patients in this subset can have an episodic course over many years.

Pauciarticular JRA has the best future outlook for normal joint function out of all three types of JRA. In about one-third of the patients there is a crossover, or progression, from pauciarticular to the polyarticular course, which can lead to involvement of the cervical spine, temperomandibular joint, and other joints. The arthritis in pauciarticular JRA may follow a low-grade course or may rise and fall with flare-ups. In either case, the disease is almost always benign. It is important to treat contractures as soon as possible to retard muscle imbalances and biomechanical malalignment. If this is done these children have an overall good functional prognosis.

**Diagnosis**

Arthritis is defined as swelling or limitation of motion with heat, pain, or tenderness. Pain or tenderness in itself does not qualify as arthritis. The cause is known for some types of arthritis, however, JRA is not one of these. A reliable test to determine if JRA is the proper diagnosis has not yet been developed. In order for the diagnosis of
JRA to be made, strict requirements must be met. The child must be under 16 years of age and have at least a 6 week history of continuous swelling, tenderness, warmth, or redness of one or more joints from an unknown cause. Specific subtype is determined by the pattern of joint involvement as well as general symptoms present in the first 6 months of the illness.

The normal pattern of JRA can be recognized with a review of patient’s histories. A thorough physical examination, slit-lamp eye examination, and necessary laboratory and x-ray tests are used to rule-out other possibilities. A number of conditions which resemble JRA must be ruled out before the physician settles on a diagnosis. Some examples of differential diagnoses include: other autoimmune diseases, bacterial or viral joint infections, childhood malignancies, musculoskeletal trauma, avascular necrosis, etc.

If the cause is uncertain, as in JRA, treatment must be modified to fit other possible conditions until everything else can be ruled out. Treatment consists of decreasing inflammation, suppressing symptoms of joint swelling and pain, as well as preventing mechanical disablement that has resulted from limited joint movement. JRA is a disease that can have highly successful management through drug, surgical, and therapeutic intervention even though the exact biologic mechanism is still unknown.

**Drug Treatment**

The primary function of drug therapy is to improve the patient’s condition, prognosis, and pain, which may assist in making physical therapy more effective. These conditions include pain, swelling and limited joint motion. The drug therapy for JRA
follows the same general course of that for adults. There are a variety of drugs that effectively control the disease. However, there is no one drug that safely and promptly terminates the disease process. These drugs are potentially dangerous. To achieve the desired anti-inflammatory effect they must be utilized in their toxic range. First, are the rapidly acting anti-inflammatory drugs, aspirin being the main one. Next, the slower acting anti-inflammatory drugs, such as gold, antimalarials, and penicillamine. Finally, the adrenal corticosteroids and immunosuppressive drugs, which are used only in life-threatening instances.

Aspirin remains a basic therapy and the first type of drug used. Aspirin does have the potential for toxic side effects, but the drug has been studied for so long that those administering it are better educated regarding what to look for and expect, therefore raising its safety margin. When aspirin does not suffice over a period of months, one of the other non-steroidal anti-inflammatory drugs (NSAIDs) may be used. There are numerous NSAIDs available, however there may be side effects that arise later on that we are not yet aware of. NSAIDs are also more expensive than aspirin. Some of the NSAIDs used are Tolectin, Naprosyn, Nalfon, Motrin, Clinoril, Indocin, and Meclomen. For children over 14 years of age any approved NSAID may be tried, and for those under 14 years of age, Tolectin, Naprosyn and Nalfon have been approved by the Pediatric Rheumatology Collaborative Study Group. Other NSAIDs are currently being studied. By trial and error, drugs are matched with the patient and, if after a one month trial period, results are not seen, then another NSAID is tried.
When the NSAIDs are not adequate, a second-line drug is administered. These are the slow-acting drugs.\textsuperscript{1,2,5,8,10} At Children's Hospital of Los Angeles, gold therapy is preferred, which is the most effective slow-acting drug available.\textsuperscript{2} Antimalarials and penicillamine have been used with JRA and adult rheumatoid arthritis, however, neither have been approved for children in the United States.\textsuperscript{2}

Oral corticosteroid use is indicated only in extreme situations, such as eye and heart damage that threatens vision or is possibly life threatening.\textsuperscript{1,2,5,8,9,24} Immunosuppressive drugs are considered to be experimental in JRA.\textsuperscript{5} These drugs are hazardous and carry the potential for severe complications. At present, use of these drugs is not generally considered.

Drugs have an important role in JRA in decreasing the inflammatory process that can be so destructive to the joints.\textsuperscript{8} Although drug therapy is necessary for the anti-inflammatory effect, it is not a specific treatment to be used alone and will not be until the cause of JRA is understood.\textsuperscript{5}

**Surgical Treatment**

No more than 1 out of 10 patients with JRA will need to be considered for the use of surgery.\textsuperscript{5} If, in the beginning stages of the disease, emphasis is given to the prevention of contractures, using basic joint physiology and maintaining muscle strength of prime joint movers, then there will be even less of a need for surgical correction of contractures and deformities.
It is uncommon for children with JRA to undergo elaborate reconstructive surgery, unless they suffer from conditions that impede their ambulatory skills and other functional limitations that do not respond to conservative therapy.\textsuperscript{5,6,19,25} Surgical procedures may include soft tissue releases, manipulation, fusions or osteotomies. Synovectomy is used extensively in Europe, but is not currently popular in the United States due to the fact that a large number of children experience remission without articular destruction.\textsuperscript{5,8,19,26} Synovectomy is now utilized only for very painful arthritis that is fairly isolated in older children. A small percentage of children with JRA will require total joint replacement. Surgery has limited use in JRA due to interference with the epiphyseal growth centers, which could result in length discrepancies between limbs.\textsuperscript{5,19,22,27,28} If surgery is to be used it must be deferred until bone growth has ceased. Recovery of motion after this type of surgery is also more difficult with children than with adults.\textsuperscript{22}
CHAPTER THREE

PHYSICAL AND OCCUPATIONAL THERAPY INVOLVEMENT

Physical and Occupational Therapy Assessment

Early and accurate diagnosis of JRA allows therapeutic, psychosocial and physical management to begin as soon as possible.\textsuperscript{20} Evaluation should be the first priority in any therapeutic program. Evaluation allows the therapist to get an objective viewpoint of the child's problems, and can direct treatment and monitor treatment effectiveness over time.\textsuperscript{8}

The long-range plan is of primary importance and takes into account the needs of the child and family. The way the child views his or her condition and therapy is critical so that appropriate guidance and instruction can be given.\textsuperscript{2} The therapist must also be familiar with basic principles of development such as developmental sequences and fundamental norms of development in order to adequately assess the child according to age-appropriate activities.

In the assessment, four areas should be covered: physical, psychosocial, educational, and home environment.\textsuperscript{22} Of these four assessment areas, the physical problems are the easiest to recognize. A thorough musculoskeletal exam is generally the basis of the therapy program.\textsuperscript{19,29-31} In order to set appropriate goals and formulate a treatment plan, there must be assessment of pain, swelling, range of motion (ROM), muscle strength, and mobility.\textsuperscript{6,22}
The first step is to obtain the patient history.\textsuperscript{6} This can be accomplished through a parent and patient interview. Attention should be focused on such issues as morning stiffness, pain and fatigue, gross motor play, participation in peer appropriate activities, functional ambulation/mobility at home and at school, and independence in activities of daily living.\textsuperscript{6,19} The therapist should also observe any equipment or exercise programs that are currently being used.\textsuperscript{6}

Next, the spine and extremities should be examined.\textsuperscript{6,8,19} Observations for the presence of effusion, heat, synovial thickening, and redness should be completed.\textsuperscript{2,6,8,19} Some difficulty has been found with determining whether chronic inflammatory joint swelling is due to scar tissue, capsular thickening, or active swelling processes.\textsuperscript{8}

Range of motion goniometric measurements of the spine and extremities should then be completed.\textsuperscript{6,19} Both passive and active ROM should be assessed with attention to the distinction between true joint limitation, muscle tightness, and increased tone.\textsuperscript{6,8}

A muscular assessment should include manual muscle tests, evaluation of atrophied muscles, and girth measurements.\textsuperscript{6,8,19} If the child is too young to undergo manual muscle testing, the use of functional activities is helpful. For example, avoidance of weight-bearing in a standing position or a decreased desire to crawl may indicate problem areas.\textsuperscript{2} Observing everyday tasks of dressing, walking, and playing may also indicate where muscle weakness lies.\textsuperscript{8}

A postural examination should include anterior, posterior and side views.\textsuperscript{6,8} The postural deviations that should be checked for are scoliosis, rounded shoulders, forward head, scapular “winging”, kyphosis, lordosis, pelvic obliquity, and any leg-length
discrepancy (common with unilateral knee arthritis).\(^6,8,19\) Other common postural deviations include femoral anteversion, genu valgum, tibial torsion, pes cavus or valgus, hallux valgus and hammertoes.\(^6,8\)

Gait should be analyzed through observation to further reveal any pain, limited ROM, or weakness that inhibits ambulation.\(^8,19,22\) The results of a study by Lechner and colleagues\(^6\) showed decreased velocity, cadence, and stride length in patients with JRA as compared to children without JRA. Observation of gait should be conducted on level surfaces, stairs, and inclines. Assistive devices used and the distance covered should be noted.

Functional assessment can be utilized to look at the child’s self-help skills and other activities of daily living.\(^19\) It is important to note the child’s proficiency, movement quality, speed, and endurance during various functional activities.\(^6\) The Pediatric Evaluation of Disability Inventory (PEDI) can be utilized to assess self-care, mobility, social function, and caregiver assistance and/or modifications needed.\(^6\) The Juvenile Arthritis Functional Assessment Scale (JAFAS) and Juvenile Arthritis Functional Assessment Report may also be utilized.\(^13\) However, these do not allow evaluation of children under 7 years old. That is why Singh and colleagues\(^13\) developed a self- or parent-administered functional assessment tool that covered all ages. They developed a valid, reliable and sensitive test called the Childhood Health Assessment Questionnaire (HAQ). This was modified from the Stanford HAQ. Parents serve as reliable reporters for their children.

Both physical and occupational therapy should assess the child’s splinting and
adaptive equipment needs. Joint contractures that cannot be successfully resolved through exercise are an indication for splinting. Resting splints are commonly used at night or during naps. Required equipment may include: a tub transfer seat, commode, grab bars, ambulation aids, or a wheelchair. Occupational therapists would further assess the need for any adaptive equipment required to perform other self-help activities such as eating and dressing.

In order for a child to be competent at higher levels, performance in basic activities is essential. Psychosocial development in children with JRA has not been found to significantly impact assessment and treatment. However, it has been suggested that JRA can disrupt attainment of higher levels by affecting the child’s self-concept, independence, and future goals. Hanson and King, in their study of psychosocial function, did not identify a personality specific to JRA after the onset of the disease. If problems do exist, therapy activities can enable more independence and can encourage physical participation in normal activities of his or her peer group.

Another factor that has been said to influence assessment of JRA patients is pain perception. Pain perception is affected by several things including maturation of joints and peripheral and central nervous systems. Cognitive skills, psychological mechanisms, and social learning can influence pain perception as well. Another addition to be aware of is the masking effect of anti-inflammatory and analgesic drugs. Therefore, it is important to be aware of what the child describes as being painful and to stay in tune with other indicators that may give you clues as to how accurate the patient’s pain perception is.
Pain is evaluated according to different criteria. These include pain on motion (active or passive), weight bearing, and tenderness with palpation. The factors influencing pain perception, that were mentioned earlier, should also be taken into account. In addition to those factors, a study by Hogeweg and associates studied pain threshold in JRA patients, and they found that children with JRA showed a generally diminished pain threshold as compared to healthy children, suggesting a change in the pain processing system. This was evident in children with both the active and non-active stages of the disease.

The overall evaluation is analyzed by physical and occupational therapists in order to develop appropriate treatment goals. The child’s needs should be addressed as well as increasing or maintaining the following: ROM, strength, independence in mobility and activities of daily living, and control of pain.

**Therapeutic Exercise Programs**

Physical and occupational therapy are important components of the treatment program for children with JRA. The most common physical problems are abnormalities of the musculoskeletal system. The maintenance of function and correction or prevention of deformities are two major goals of therapy. Preventing disuse atrophy, loss of ROM, and decreased endurance are other important goals of therapy. The physical and occupational therapy program must be a practical and ongoing process in order to be effective. An exercise program needs to be adapted to the child’s maturity level, developmental needs, and stage of the disease he or she is in. In the beginning stages of therapy, it is essential for the therapist to be patient and understanding, thus
gaining both the parents and the patient's confidence and cooperation. The therapy program must be individualized and properly timed to the child's stage of disease. The child in an acute stage may not tolerate exercise, but should be encouraged to avoid excessive fatigue, participate in activities of daily living, and go through gentle ROM exercises at least once a day. Heat can be applied to joints to decrease pain. A child whose disease is subacute can be treated with exercise. This includes ambulation, active ROM, and resistive strengthening exercises. Proper positioning, heat and age-appropriate play are recommended as well. The child with chronic disease who continues to have joint contractures may be treated more aggressively with progressive resistive exercise and general conditioning programs such as bicycling, swimming, or walking.

It is important to remember that children with JRA may not be able to tolerate as much exercise as their "healthy" peers. The results of a study by Giannini and Protas showed that peak VO2, highest workload completed, exercise duration and peak heart rate were significantly lower in children with JRA as compared to children without a chronic illness. The submaximum heart rates were higher for the children with JRA, suggesting a decreased aerobic capacity. Giannini and Protas also concluded that deconditioning and a lower aerobic capacity occur in the patient with JRA regardless of the severity of the articular disease. Giannini and Protas noted that intervention should occur soon after diagnosis to prevent the hypoactivity and deconditioning that is so commonly found in people with a chronic illness. Brewer and Giannini found similar results.

Children enjoy playing, but will not engage in activities that hurt. With proper
guidance from the therapist during a therapy session, free play activity may be a suitable substitute for structured exercise. The play equipment offered should encourage activity to limber up the joints, and promote strengthening of the joints, such as a bicycle or tricycle. The key is to make therapy as enjoyable as possible for the child.

**Range of Motion**

Children will not take their joints and muscles through full ROM during play and activities of daily living. This necessitates prescribed ROM exercises to overcome tightness and lost mobility. ROM will often require assistance from the therapist or parent in order to complete the motion, however the patient should participate actively as much as possible. This is called active-assisted ROM. At the end of the range the therapist may move the limb a little further, applying a stretch to the point where it begins to hurt. This stretch is contraindicated for patients in the acute stage of JRA because it may cause a painful reflex spasm in the involved joint. For patients with acute JRA, passive ROM should be used, which allows the patient to relax the joint while the therapist or parent takes it through the available range.

Once the children are able to do all the work (active ROM) they may be taught a regular program that encourages independence and confidence in their ability to improve movement range. In the chronic JRA patient, completing one ROM program per day is sufficient. However, with the acute and subacute patient, two times a day may be needed. A reliable guide for prescribing ROM exercises is pain duration. Pain or discomfort should not last for more than 1 hour after exercise. Range of motion exercises should be done at the best time of day for the patient.

**Strength**

"The strength of a normal muscle decreases at a rate of 3% to 5% per day during
inactivity."\(^2\) Atrophy is a frequent complication when pain interferes with ambulation or other activities of daily living. Maintaining joint strength assists in providing optimal function.\(^19\)

As soon as the patient's condition permits resistive strengthening exercises should be added.\(^2,5,8,22\) If the joints are too severely involved and cannot tolerate active free exercise, isometric strengthening exercise may be used. Motion usually causes pain, and isometric exercise eliminates motion. Isometric exercise is done by bringing the joint to its full range, contracting the muscles around the joint while holding it in place for a count of six or seven, and then relaxing the muscles. This can be repeated 10 to 15 times (or to the point of fatigue), twice a day, for whatever muscle groups need strengthening.\(^2,5\) Isometric strengthening can be done while wearing a splint, and is very effective for weight-bearing muscles.

With less painful or pain-free joints progressive resistive and/or active-assistive exercises may be performed.\(^2\) Progressive resistive and active-assistive exercises use isotonic strengthening methods through active ROM. Progressive resistive exercise uses graded resistance throughout the ROM. Active-assistive exercise uses a counterbalance weight mechanism to assist in completing the ROM.

**Endurance and Mobility**

Improving endurance and mobility is another goal of exercise for children with JRA.\(^6,8,19\) Many children with JRA have learned to have their parents provide assistance in many ways. Children can become manipulative to continue receiving this attention. JRA patients may also become so deconditioned that they cannot endure normal everyday activities. Age-appropriate mobility and transfer assessments are essential.\(^6\)

Gait training should try to decrease or eliminate observed deviations that are
common in JRA patients, such as decreased velocity, cadence and stride length. Gait training is critical in achieving their physical performance goals. Care must be taken in initiating gait training in the subacute or chronic inactive stages of JRA. The reason for caution is that patients with JRA may be protecting painful or previously painful joints through gait deviation. Gait training may include such things as postural training and weight-bearing activities.

Too much exercise can temporarily make arthritic inflammation worse. It is important to note signs of fatigue and pain which are the body’s signals that it has had enough. The therapy program should be altered as needed with consideration given to the cost and time required by the patients and their parents.

Aquatic Therapy

Many authors agree that swimming or pool therapy is one of the best possible exercises for children with chronic arthritis. Brewer and associates stated, “For more than twenty years the authors have observed that those children who have had uniformly the best results with regard to joint motion and muscle strength maintenance are those who managed to swim daily or almost daily.” There are many reasons why swimming is looked upon so favorably.

The heated pool can be an adjunct to pain relief as well as promoting muscle relaxation and reducing joint stiffness. Buoyancy can assist the movement of the limbs and allow greater mobility with decreased stress on the weight-bearing joints. The water provides moderate resistance, thus allowing muscular strengthening without traumatizing the joints.

An exercise program that aims to increase a child’s active ROM, endurance, and strength can easily be done in the pool. Individual and group activities, as well as play
sessions, can be arranged. These activities often provide a fun and sociable way of completing exercises. Hip and knee movements can be performed with less strain and assist in ambulation training and mobilizations.

Time in the pool should be limited to 1 hour with continuous activity. Five to 10 minutes of lap swimming (until fatigued) is suggested. This can even be done by non-swimmers with the use of a “Styrofoam ski belt” or “bubble” to provide buoyancy. Simple exercises and play sessions can be incorporated. If a pool is not available, a bathtub can be used as a substitute, however, this does no allow as much mobility. There are also some YMCA and public school locations that have special pool times arranged for individuals with disabilities.

**Therapeutic Modalities**

There are a variety of therapeutic modalities that can be used to help stiffness, muscle spasm, and other joint involvement in the active disease state. The use of heat is common, with many different forms available.

Hot packs may be utilized if only a few joints require attention. Hot packs are especially useful prior to exercise of a specific joint or application of a splint. Hot tubs, whirlpools, or heated swimming pools are some other forms of moist heat. These allow active and active-assisted ROM to be carried out. Maintaining the body’s warmth overnight, through use of a sleeping bag or sleeper pajamas, is effective in reducing pain and morning stiffness. A paraffin bath may be indicated for the hands and feet. Deep heating techniques, such as diathermy and ultra sound, are limited in use and generally are not recommended for children with JRA. There are concerns about affecting growth of the epiphyseal plate and increasing joint inflammation.

The application of cold, in the form of ice packs or ice massage, can also decrease
Most children prefer the use of heat to cold. Rhodes\textsuperscript{6} also suggested the use of transcutaneous electrical nerve stimulation (TENS) on one or two painful joints to assist in maintaining joint ROM.

It is important to remember that modalities are only aids to therapeutic exercise and are not a specific treatment.\textsuperscript{5,19} Therefore, they should not be used in an isolated manner as a treatment for inflamed joints.

- **Splints and Braces**

Splints and braces are important in the treatment of children with JRA, especially when a joint is acutely inflamed and painful.\textsuperscript{5,19,23} Splints are indicated to prevent deformities from contractures, to provide rest, and to eventually improve function of the joint.\textsuperscript{5,10,19,22,23} It is important for the patient and parents to be aware of the consequences of discontinuing exercise. Once a joint loses its ROM or functional position it is difficult, and maybe impossible, to reverse or correct the deformity.\textsuperscript{22}

Splints work by putting pressure on either side of an inflamed joint in order to remove mechanical stresses on that joint.\textsuperscript{5} Splints also maintain joints in a good functional position.\textsuperscript{4} Maintaining a functional position is especially important when children sleep. Children tend to go into a position of flexion or comfort while sleeping.\textsuperscript{5} Flexed posture often leads to the development of flexion contractures, which is why splints are often worn at night or while sleeping.\textsuperscript{1,4,5,10,22}

Three types of splints, named according to their function, are used: resting splints, corrective splints, and functional splints.\textsuperscript{7,19} The resting splint provides rest to an inflamed joint while holding it in a functional position. Corrective splints place a joint in its end range of motion to help increase the joints range of motion. Young children are sometimes unable to tolerate corrective splints and may be surgical candidates instead.\textsuperscript{5}
The design of functional splints is to protect and support a joint throughout activities of daily living. Occupational therapists are helpful in designing and fixing splints and braces used for children with JRA.

**Functional Activities**

Children with JRA should be encouraged to participate in age-appropriate recreational activities. It is important to monitor and limit activities that cause pain or discomfort. Play and recreation can be used to reinforce exercise goals. Activity through recreation may improve strength and ROM, as well as provide enjoyment and a sense of well-being for a child. As children play they acquire balance, coordination, perceptual skills, and self-confidence.

Pleasant activities, such as bicycle or tricycle riding, walking, golfing, and swimming may be valuable catalysts to therapy. Pull-apart toys, clay, coloring, and Ping-Pong are some excellent activities to increase ROM and strength of the upper extremities. Contact sports or any activity that causes an impact to the joints or body should be avoided. Some examples of impact activities are jumping rope, football, baseball, volleyball, basketball, trampoline jumping, and gymnastics. These activities may cause increased joint inflammation, injury, or fracture and could result in limited mobility. Activities that require holding a joint in one position for an extended period of time, such as piano playing, should be avoided also.

Independence in age-appropriate activities of daily living is encouraged in children with JRA. Self-care activities, in conjunction with recreational activities, can help maintain and/or improve strength, ROM, and functional independence. Advice on the way to most efficiently perform an activity is sufficient for children that are not severely involved. Carrying items close to your body, using your palms to open door knobs, and
wearing low-heeled shoes are some helpful tips. In order for the more involved children to be independent, assistive devices or modification of the environment may be required. Some necessary supplies may be adaptive utensils, bath seats, dressing sticks, button fasteners, and Velcro closures. These assistive devices, modifications, and suggestions all integrate the principle of protecting the joints of children with JRA.
CHAPTER FOUR

FAMILY AND SCHOOL INCLUSION

Patient Education

The extent to which the patient understands his or her illness has a significant impact on the effectiveness of a treatment program. The patient’s knowledge and willingness to accept the disease will gradually develop with age and maturity. However, this baseline knowledge does not eliminate the need for continuing education. Patient education should begin with an explanation of the disease process in terms a patient or parent can understand. A review of the physical, social, and psychological challenges JRA poses are necessary, as well as the appropriate balance between rest and activity. In addition to explaining suitable physical activities, education on any drugs the child may be taking and possible side effects should be covered. Physicians and therapists need to explain to the patient and parents how necessary a complete treatment plan is for a healthy future. If the patient and parents do not understand the importance of the treatment plan then all plans for a healthy future may not be beneficial. Counseling and support groups may prove to be advantageous for both the patient and parent when any problems or conflicts arise.

Preschool children have little independence and rely on their parents for almost
everything. For children with JRA, this reliance on parents can lead to feelings of inadequacy and helplessness if approval for something such as a self-care task is not recognized by the parents.\(^5\) The process of mastering self-care and recognizing such an accomplishment leads to the development of self-esteem and encourages self-sufficiency.\(^2,5,10\) School age children with JRA can develop a clear understanding of the disease and may then accept the limitations it holds. These children are active and accountable participants in their therapy.\(^5\) However, this fervent involvement is not without education. Body image, appearance, self-control, academic competence, family support, and functional effectiveness are issues that need to be addressed.\(^2\) For teenagers with JRA, vocational counseling may be needed.\(^10\) Healthy development relies on such experiences as going to school and partaking in activities with other children.\(^5\) Efforts should be made so symptoms of JRA do not prevent children from being involved in these activities.

Patient cooperation, or compliance, is another essential element of a successful treatment program.\(^8\) It is critical that physical therapists establish rapport with the patient and parent and comprehend the importance of an outlined program. Therapists should also monitor progress, alter physical therapy goals as needed, and be realistic in their expectations.\(^8\) Compliance can be hindered by the child's desire for control and the influence of interruptions in the family's routine. If the child with JRA has a difficult time comprehending the need for ongoing exercise then non-compliance may result.\(^2\) Non-compliance can also be associated with psychological denial and/or parents' understanding of exercise effectiveness. A comprehensive treatment program requires patient education,
together with the awareness of family lifestyle and their ability to accept JRA as a reality.\textsuperscript{2,5}

**Parent and Family Education**

Chronic illness can be difficult for a family to deal with, especially when the one affected is a child.\textsuperscript{4} The family is of principle importance in childhood and should be included in the child's therapy.\textsuperscript{22} Matters such as education, beliefs concerning the disease, values, family schedules, discipline for the child, and sibling involvement should be included in therapy also.\textsuperscript{2}

The family’s first encounter with the child’s chronic illness may be when the physician affirms the diagnosis of JRA.\textsuperscript{5} At that instant, parents encounter a developmental circumstance that was not expected. Parents are faced with working through the feelings of shock, disbelief, anger, and guilt. As they become more knowledgeable about the disease, they may become more accepting as well. With knowledge of the illness comes the understanding that both physical and psychosocial disabilities may result from JRA.\textsuperscript{5,22} Riggs and Gall\textsuperscript{22} suggested that families need to learn the importance of living in the present and not being distracted by the unknown. Focus by the family, on the disease process and the child, is necessary to provide the proper supervision and assistance to the patient. Maintaining the essential home environment for normal child upbringing is important also.\textsuperscript{22} The patient with JRA may benefit from the family’s positive attitude.\textsuperscript{5} A positive outlook allows the child to move ahead developmentally and to make progress in the treatment program. When parents are overprotective, the situation may result in the child’s diminished levels of self-esteem.
According to Williams, the results of being overprotective are worse than any consequence of arthritic impairment.

It is the patient and parents' responsibility to translate what the medical team advises and to turn it into action. Constant communication with the parents about their child is necessary. The parents must have a clear idea of the drugs administered and the reasons they are prescribed. Parents must also understand objectives and goals of exercise and splinting programs. Parents and patients will be the first to distinguish when goals are not being met, and it is their responsibility to communicate this to the physician. Parents must also be aware of how important it is to develop self-esteem, self-image, and increasing independence in their child with JRA. To some parents and families this may all seem overwhelming. It may be helpful to keep a notebook or journal, or to go to a health education class or arthritis discussion group for reinforcement and support. It is important for families to keep in mind that the illness must be looked after, but should not override normal daily activities.

The requirement for daily therapeutic exercise leads to parents being instructed in a home program. A successful home program should be geared toward the child's age-appropriate developmental activities and should be adjusted into the family's daily schedule. To allow for a smoother adjustment and increased compliance, the following suggestions may be considered by the therapist. First, the therapist should understand the child's and parents' perceptions of the disease and should discuss their reactions to the illness. Second, the therapist should be aware of the family's life-style. This awareness will assist in choosing priorities for treatment and in helping families plan and adapt to a
new schedule. Next, the therapist should emphasize disciplining the child with JRA as one would discipline other siblings. Generally, parents are understandably troubled by the pain their child is experiencing. However, they must learn that it can be harmful for a child with JRA to experience gain as a result of complaining about joint pain.\textsuperscript{2,4,5} Parents also need to remember the siblings.\textsuperscript{2} The therapist should involve brothers and sisters in the therapy session. Helping the family to find ways to have fun together is important. Finally, the therapist should point out the need for parents to recognize their needs. Emotional disturbances and marital conflict are frequent among parents of children with chronic illnesses. Awareness of and correction of any marital problems are critical for maintaining a healthy home environment.\textsuperscript{2,22}

Parents acknowledged that psychological problems are more prevalent among children with a chronic illness as compared to their healthy peers.\textsuperscript{5,37,38} It may be helpful for parents to be aware of a study conducted by McAnarney and associates.\textsuperscript{39} In this study, it was concluded that children with disabilities resulting from JRA have fewer psychosocial problems than those with no resultant disabilities. McAnarney and associates theorized that parents of non-disabled children failed to recognize the impact the disease has on the child’s life. This is why it is necessary for the general public to give strong support to all children dealing with a chronic illness, whether physically disabled or not, and to guide them toward healthy psychosocial development.\textsuperscript{5}

One aspect of successful management of JRA is the parents ability to provide prescribed care in the home.\textsuperscript{40} Health care professionals often take for granted that parents will do what they are told and follow through with the treatment program. Most
parents do not feel prepared to do this. In addition to their everyday activities, parents must administer exercises that can be painful, apply splints to various joints, dispense often distasteful medications, and make trips to visit the physical therapist and other health professionals. The parents' world and scope of reality are altered. Jerrett concluded that parents are adapting by developing expertise in the management of their child's illness and are assuming responsibility for caregiving. Parents are challenging the label of being passive recipients of 'expert' advice and are listening to and valuing what the experts have to say. However, parents are also relying on their expertise based on what they have lived through.

School Involvement

Formal schooling is one of the most important aspects of childhood for cognitive and psychosocial development. Children with JRA are no exception. Planning for the educational needs of the student with JRA is, at times, difficult. Modifications of the school environment and classroom activities may be necessary. Essential adjustments can usually be made with a little imagination and some good communication between schools, teachers, and parents.

Clear, direct, and timely requests for school adaptations usually result in compliance. School problems may include difficulty with the following tasks: speed and amount of writing, performing fine motor activities (i.e. using scissors), carrying books, traveling a great distance for class changes in a short amount of time, stair climbing, staying in one position, keeping up in physical education class, getting dressed and undressed, and using eating utensils. Communication with the teacher
regarding such difficulties can greatly enhance the child's involvement into his or her normal school. Many children will need only minor adjustments. However, some of the more severely involved children will necessitate the inclusion of a health care professional in making modifications.

Teachers may have some difficulty understanding JRA. Difficulty in understanding the disease arises because children with JRA may show few visible signs of the disease, with all the pain and discomfort on the inside. Also, the course of JRA is unpredictable. Symptoms change from day to day. Any problems that teachers come across (learning and/or social) should be made known to the parents and physician so they can be dealt with. Necessary discipline should not be avoided just because the child has JRA. The children should be treated normally in the classroom and should not be overprotected by the teachers. The Arthritis Foundation publishes a brochure which parents can give to teachers, called When Your Student has Arthritis - A Guide for Teachers. The brochure provides guidelines to help educate teachers about juvenile arthritis and to give them tips on how to accommodate for the illness.
CHAPTER FIVE
CONCLUSION

The treatment of children with JRA is a long and strenuous task. Treatment involves several years of exercise, splinting, observation, and medication. A pyramid of treatment is formed with medication at the base, patient and family education, physical therapy, occupational therapy, and family and school support. Present and future treatment is following a trend that is dismantling, or modifying, the therapeutic pyramid. Medical management is now treating JRA as early and as decisively as possible in order to promote remission of JRA, so that function and quality of life are preserved. Encouragement from the medical team, as well as friends and relatives, will help greatly in attempts for children with JRA to achieve a normal life.

Physical therapists play a key role in the management and care of JRA patients throughout the disease course. It is a therapist’s job to “integrate the biomechanical and developmental tasks and appreciate the effects of a chronic disease on the child at each stage of the child’s development as well as globally in response to disease development throughout the course of the illness.” It is the job of both physical and occupational therapists to maintain function and correct or prevent deformities in the child with JRA. An exercise program must be designed to fit the individual needs of each child. Aquatic therapy is one form of exercise that many authors found to be the most beneficial for children with JRA.

Further study needs to be done in the area of diagnosing JRA. A diagnostic test for JRA would aid in earlier diagnosis of the illness and would allow for treatment to begin as soon as possible. Early diagnosis would hopefully assist the medical team in
reaching the long-term goal of helping the patient with JRA experience a "normal" childhood.
REFERENCES


