

increase dramatically over the next 20 years as the population ages.¹⁶⁴

Before age 50 years, the prevalence of OA in most joints is higher in men than in women, but this changes after age 65 years. In the United States OA is second only to ischemic heart disease as a cause of work disability in men older than 50 years.¹⁰⁶

In the United States, about 6% of adults over 30 years of age have OA of the knee and 3% have OA of the hip; incidence rises with increasing age. Both incidence and prevalence are expected to rise in the coming decades as a result of the aging of America combined with more extreme sports and activities (see the following section on Etiologic and Risk Factors). OA is the most common indication for total joint replacements.

Etiologic and Risk Factors

The etiology of OA is multifactorial, including many components of biomechanics and biochemistry. Evidence is growing for the role of systemic factors such as genetics, nutrition and weight control, estrogen use, bone density, and local biomechanical factors (e.g., muscle weakness, obesity, joint laxity).¹²⁰

Serious injury and an inherited predisposition account for one half of all cases of OA in the hands and hips and knees.^{119,141,425} Smokers with knee OA sustain greater cartilage loss and have more severe knee pain than those who do not smoke, suggesting a role for tobacco in cartilage degeneration.¹⁹

There is low or no additional risk of OA from regular, moderate running, but sports that involve high-intensity, acute, direct joint impact from contact with other players do carry an increased risk of OA, especially when repetitive joint impact and twisting are combined. Football players, soccer players, hockey players, and baseball pitchers are especially at increased risk. Anterior cruciate ligament injury may predispose athletes to knee OA, especially when accompanied by meniscectomy.^{87,337}

Although the theory is as yet unproven, some experts warn that extreme sports such as snow boarding, mountain biking, and aggressive in-line skating with the increased incidence of repeated impact or injury may be risk factors for OA developing earlier in life. Much of the OA in men is attributable to occupational activities, particularly kneeling or squatting, along with heavy lifting and repetitive use of heavy machinery.^{119,120}

Generalized ligamentous laxity appears to be a predisposing factor; this may be related to the presence of estrogen receptors on the ligaments. Postmenopausal women appear to be at increased risk.⁴⁹³ Some women have a condition called *hypermobility joint syndrome* or *hypermobility syndrome* with loose, unstable joints resulting from a dominant inherited connective tissue disorder.

Hypermobility syndrome is characterized by excessive laxity of multiple joints, a condition that is separate from the generalized hypermobility associated with disorders such as Ehlers-Danlos syndrome, RA, SLE, or Marfan's syndrome. Hypermobility syndrome appears to be a systemic collagen abnormality with a decreased ratio of type I to type III collagen (see Table 6-2).⁴²⁸

Women with this syndrome may develop OA earlier than the norm. Muscle weakness in anyone can also

cause joint changes leading to OA, such as occurs with prolonged immobilization, polymyositis, multiple sclerosis, or any of the myopathies listed in Box 27-3.

There have been some studies that show a link between patellar alignment and patellofemoral OA manifested by a loss of cartilage thickness and knee pain and disability.^{214,234} Other studies suggest that malalignment is not a risk factor for OA but rather a marker of disease severity and its progression.²¹³ Additional studies will be needed to establish the normal and abnormal ranges of patellar alignment indices and their relationship to patellofemoral OA.²³⁵

Pathogenesis

The pathophysiologic events associated with OA are beginning to be understood more definitively. It is quite clear now that OA is a disorder of the whole synovial joint organ, not just "wear and tear" on the cartilage. In fact, it may be that damage to the articular cartilage is the by-product of a disease process that is centered in subchondral bone in particular. Emphasis is now on the joint as a whole rather than just the cartilage.

In recent years, the view of OA has shifted to that of both a local and systemic condition in which inflammation plays an important part in determining the symptoms and disease progression.^{118,304} The former wear-and-tear concept already mentioned has been replaced by the idea that OA is an active disease process with joint tissue destruction and aberrant repair as a result of alterations in cellular function.

Although joint cartilage is the final target of the pathologic processes, the underlying subchondral bone may be the primary etiologic agent. Treatment focused on modifying changes in the bone may alter the pathologic processes observed in the adjacent cartilage. The use of new, potent bone antiresorptive agents in clinical use will help test this hypothesis.⁴⁴⁴

Tissue changes in OA are the result of active joint remodeling processes involving an imbalance between catabolic and anabolic repair activity. People with OA may have a general tendency toward increased bone metabolic activity, especially in response to biomechanical or other stimuli such as occurs with obesity and injury.

As OA develops, loss of cartilage, hypertrophic changes in neighboring bone and joint capsule, mild synovial inflammation, and degenerative changes in the menisci, ligaments, and tendons all contribute to pain and loss of joint function, resulting in joint failure.⁴⁴⁹

In more recent years, it has been discovered that essential inflammatory cytokines such as IL-1R and TNF- α initiate this cycle of catabolic and degradative events in the cartilage, mediated by metalloproteinases, enzymes that degrade cartilage extracellular matrix as part of the normal turnover in all tissues. These enzymes have been shown to be up-regulated after joint injury.

The role of inflammation in the pathophysiology and progression of early OA is supported further by the observation that C-reactive protein levels are raised in women with early knee OA and higher levels predict those whose disease will progress. The synovium from OA joints stains positive for IL-1R and TNF- α . Nitric oxide, which exerts proinflammatory effects, is released during

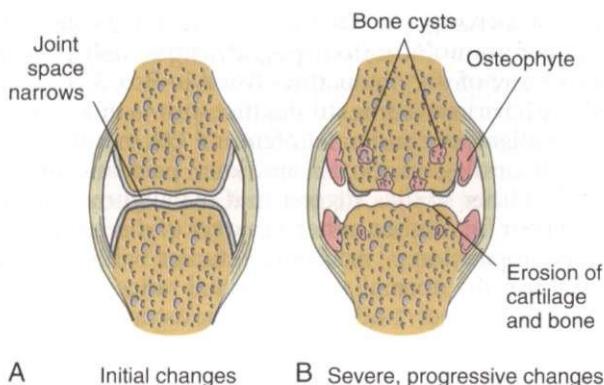


Figure 27-1

A, Early degenerative changes associated with osteoarthritis include joint space narrowing and articular cartilage erosion. **B**, Late degenerative changes associated with osteoarthritis include osteophyte formation and articular cartilage fissuring and eburnation.

inflammation. In experimental OA, nitric oxide induces chondrocyte apoptosis, thus contributing to cartilage degradation. Hence, unregulated nitric oxide production in humans plays a part in the pathophysiology of the disease.⁷⁸

Articular cartilage has an important role in joint physiology by providing a smooth, relatively friction-free surface between the bony ends making up the joint. In addition, the cartilage attenuates the mechanical load transmitted through the joint. With progressive loss of cartilage, inflammation develops, with resultant bony overgrowth, ligament laxity, and progressive muscle weakness and atrophy accompanied by joint pain.

Once the cartilage begins to break down, excessive mechanical stress begins to fall on other joint structures. Eventually, Assuring and eburnation of the cartilage (thinning and loss of the articular cartilage resulting in exposure of the subchondral bone, which becomes denser with the surface becoming worn and polished) can occur.

The joint space narrows as the cartilage thins, and sclerosis of the subchondral bone occurs as new bone is formed in response to the now excessive mechanical load. New bone also forms at the joint margins (osteophytes) (Fig. 27-1) with the end result being mechanical joint failure and varying degrees of loss of joint function.

Immobilization is another factor that can result in articular cartilage degeneration. Secondary to the lack of vascular supply, articular cartilage depends on repetitive mechanical loading and unloading for the nutritional elements to reach the chondrocytes and the cellular waste products to return to the synovial fluid and eventually to the bloodstream. This nutritional mechanism of articular cartilage is interrupted by immobilization. If the nutritional cycle is interrupted long enough, structural changes will occur.

Clinical Manifestations

The most common symptoms of OA include bony enlargement, limited range of motion, crepitus on motion, tenderness on pressure, joint effusion, malalign-

ment, and joint deformity. Inflammation is a prominent sign that plays a role in symptom generation. Soft tissue inflammation and edema are observed during acute exacerbations.¹¹⁸ The most commonly involved joints associated with this disorder are the weight-bearing joints, especially the hip and knee but also the shoulder, lumbar and cervical spine and the first carpometacarpal and metatarsophalangeal joints.

The onset of symptoms related to OA can occur insidiously or suddenly. Only a portion of people who have radiographic evidence of OA have associated pain. For most people, however, the pain complaints progress slowly and gradually. Since the cartilage is not innervated, pain is not perceived until the bone or other structures surrounding the joint are involved. The predominate cause of joint pain is attributed to a breakdown in the mechanics of movement rather than inflammation. The pain is often described as a deep ache that is worse with activity and better after rest; pain can occur at rest and at night with advanced disease.^{128,231}

Pain with activity is most likely due to enthesopathy and mechanical factors, whereas pain at rest may be caused by synovial inflammation. Night pain is a poor prognostic indicator and may occur as a result of intraosseous hypertension, which stretches periosteal pain neurons.¹⁵²

Stiffness of relatively short duration (less than 30 minutes) can occur after periods of inactivity, including sitting and sleeping. Morning stiffness, referred to as the *gel phenomenon* or "joint gelling," usually only lasts 5 to 10 minutes after awakening. Movement and activity dissipate this stiffness until the individual sits or rests for a long period of time. This differs from RA, in which the morning stiffness or gelling can last until noon or even midafternoon.

Swelling, if present, is mild and localized to the joint. Loss of flexibility is usually associated with significant disease and can occur secondary to soft tissue contractures, intraarticular loose bodies, large osteophytes, and loss of joint surface congruity.

Crepitus (audible crackling or grating sensation produced when roughened articular or extraarticular surfaces rub together during movement) may be noted on physical examination, and enlarged joint surfaces, including osteophytes, may be palpable. Although many people have physical and radiographic findings of OA, they may not have symptoms, whereas others with minimal changes observed develop significant symptoms. The reasons for this remain unknown.

For many women, OA typically develops within a few years of menopause and is often associated with mild inflammation for the first year or two that a particular joint is involved. The joints may intermittently be warm and tender. The disease is strikingly symmetric, although the degree of involvement may vary somewhat.

OA of the hands affecting the distal interphalangeal and proximal interphalangeal joints occurs most often in this group of women. The gradual loss of joint motion can assume major significance, with the person finding it difficult to grasp small objects.

After 1 or 2 years of inflammation, the joints enlarge with osteophyte (spur) formation, referred to as



Figure 27-2

Typical hand deformities in osteoarthritis. Heberden's nodes are seen on the distal interphalangeal joints, and Bouchard's nodes are at the proximal interphalangeal joints. (From Forbes CD, Jackson WF: *Color atlas and text of clinical medicine*, ed 3, London, 2003, Mosby.)

Heberden's nodes (affecting the distal interphalangeal joints) and Bouchard's nodes (affecting the proximal interphalangeal joints) (Fig. 27-2) and become unsightly. Pain may also be noted with loss of joint articular cartilage. Lateral deformities of the joints are common, with stretching of the collateral ligaments and bone resorption. This leads to overlapping of the fingers and considerable loss of functional ability.

Some individuals experience OA of the carpometacarpal joint. With advanced disease, individuals with carpometacarpal involvement may develop joint subluxation as the metacarpal flexes and adducts, leaving the metacarpal base prominent. Axial loading (e.g., pinching) and rotation characteristically reproduce symptoms and cause crepitus. An excellent review of carpometacarpal OA is available.⁴⁹³

MEDICAL MANAGEMENT

PREVENTION. Arthritis (including OA and rheumatic conditions) is the leading cause of disability in the United States, affecting a total of over 43 million people in the United States, with an estimated prevalence of nearly 60 million by the year 2020.^{72,74} The Arthritis Foundation, Centers for Disease Control and Prevention (CDC), and *Healthy People 2010* are working together to implement the National Arthritis Action Plan to promote progress toward reaching arthritis-related national objectives for 2010.¹⁸⁵

Arthritis research is providing a growing body of knowledge about prevention as well as slowing the disease's progression and new, more effective combinations of drug and behavioral interventions. Education is a cornerstone of prevention and management for this condition. A healthy lifestyle helps prevent OA, and exercise can lessen disability if OA has developed. Moderate exercise has been shown to improve the knee cartilage glycosaminoglycan content in individuals at high risk of developing OA.⁴²⁶ Strengthening the quadriceps muscle and maintaining an appropriate body weight for height reduce risk of OA at the knee by 30%.^{119,120}

Sports officials and athletes need to work with athletic trainers, exercise physiologists, and physical therapists to evaluate and modify rules, equipment, and playing sur-



Figure 27-3

Osteoarthritis of the shoulder. There is osteophytic lipping (open arrow) from the humeral head, including new bone formation deep to the cartilage (closed arrow). (From Harris ED: *Kelley's textbook of rheumatology*, ed 7, Philadelphia, 2005, Saunders.)

faces while providing adequate training to help reduce injuries. Early diagnosis and intervention with complete rehabilitation of joint injuries can decrease the risk of subsequent OA.^{119,120} In the future, biomarkers found in joint fluid, blood, or urine that indicate changes in bone or cartilage may help identify people at risk for OA, allowing for prevention of disease progression and early intervention.

High intakes of vitamin C are associated with lower rates of OA on radiograph examination and less knee pain from OA. High levels of vitamin D protect against new and progressive OA.^{119,120}

DIAGNOSIS. OA is diagnosed by correlation of history, physical examination, radiologic findings (Figs. 27-3 and 27-4), and laboratory tests, which rule out rheumatic disease. Box 27-6 lists radiographic changes associated with OA. The history of location of symptoms, symptom duration, functional limitations, trauma, medical comorbidities, and family history helps guide the physician in making the diagnosis.

The American College of Rheumatology's guidelines for the diagnosis of knee OA include knee pain with radiographic changes of osteophyte formation and at least one of the following: age more than 50 years old, morning stiffness lasting less than 30 minutes, or crepitus on motion.¹²

Other symptoms diagnostic of OA include a locking or a "giving way" sensation in the knees, swelling, and exacerbation of symptoms with inactivity or overactivity. Walking on uneven ground and climbing stairs also aggravate knee and/or hip OA.⁵¹³

Box 27-6**OSTEOARTHRITIS: RADIOGRAPHIC FINDINGS***

- Joint space widening (early evidence)
- Subchondral bone sclerosis
- Subchondral bone cysts
- Osteophytes
- Joint space narrowing

*Listed in order of progression.

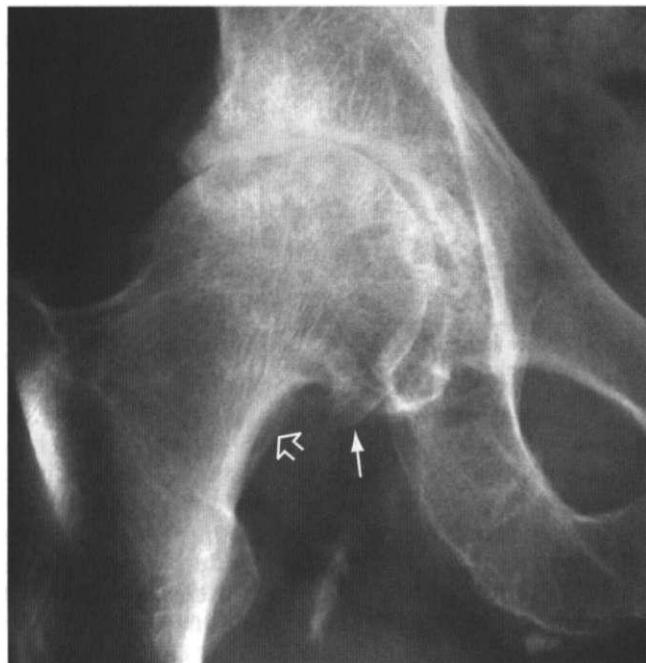


Figure 27-4

Osteoarthritis of the hip. The anteroposterior view of the hip shows complete cartilage space loss superiorly. There is osteophytic lipping from the femoral head, especially medially (arrow), and buttressing bone (open arrow) is present along the femoral neck. (From Harris ED: Kelley's textbook of rheumatology, ed 7, Philadelphia, 2005, Saunders.)

The physician also relies on findings from the physical examination, such as joint line or bony tenderness, joint effusion (not always present), quadriceps muscle atrophy, varus or valgus deformity (knee), and any abnormalities such as Heberden's nodes, a classic osteoarthritic change observed in the distal interphalangeal joints of the hands (see Fig. 27-2).

OA is classified based on clinical information and radiologic evidence. The radiographic classification used most often is the 0 to 4 grading system proposed by Kellgren and Lawrence (Box 27-7).²⁴² Grade 4 changes include large osteophytes, severe joint space narrowing, bony sclerosis, and bone exposure (Fig. 27-5).

MRI is becoming increasingly helpful in determining OA pathology because of its ability to show the condition of cartilage and the surrounding soft tissues. Laboratory evaluation may include ESR and rheumatoid factor, but generally these tests are not needed.⁵¹³

Box 27-7**KELLGREN AND LAWRENCE GRADING SYSTEM FOR THE KNEE****Grade Radiographic Findings**

1	Possible osteophytes; no joint space narrowing
2	Definite osteophytes; possible narrowing of joint space
3	Moderate multiple osteophytes; definite joint space narrowing; some sclerosis and possible deformity of bone ends
4	Large osteophytes; marked joint space narrowing; severe sclerosis and definite deformity of bone ends

Data from Kellgren J, Lawrence J: Radiologic assessment of osteoarthritis, *Ann Rheum Dis* 16(4):494-501, 1957.

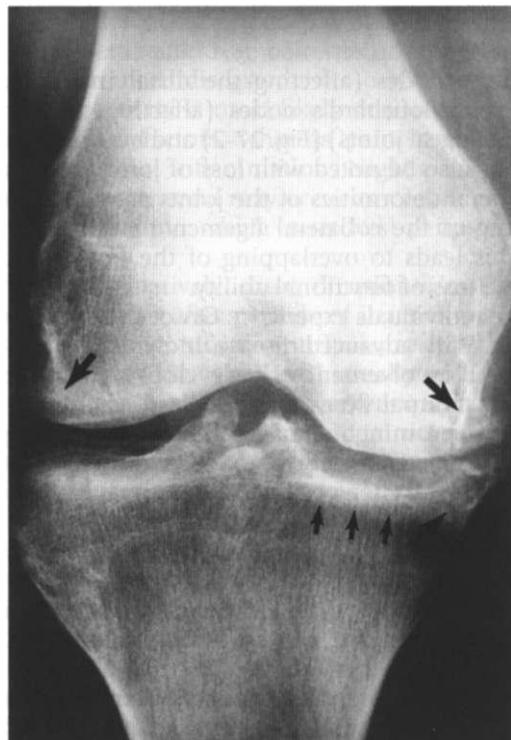


Figure 27-5

Osteoarthritis of the knee. Proliferative marginal osteophytes (larger arrows), narrowing of the medial weight-bearing joint space, and eburnation (exposure of the subchondral bone, surface becomes smooth and polished as it wears down) (smaller arrows). (From Noble J: *Textbook of primary care medicine*, ed 3, St Louis, 2001, Mosby.)

A goal of current research is to develop laboratory tests (i.e., serum, synovial, or urine biomarkers) that would help identify people who are predisposed to OA, detect the disease in its earliest stages, and assess the response to therapy.

TREATMENT. OA is managed on an individual basis, and treatment consists of a combination of nonsurgical and surgical options. Treatment is modified based on response and should begin with conservative care, including education, weight loss, exercise, orthotics and/or braces, medications, and complementary approaches.

The combination of modest weight loss and moderate exercise provides better overall improvements in function, pain, and mobility in older adults who are overweight or obese with knee OA.³³² Greater improvements in function have been observed in older obese adults with the most weight loss.³³⁸ Surgery is avoided and considered only when debilitating pain and major limitation of functions interfere with walking and daily activities or impair ability to sleep or work.²¹⁵

Pharmacotherapy. For many years, analgesics and NSAIDs have been the first-line medical treatment approach for OA. Newly discovered information about the pathophysiology of OA is paving the way for researchers to design medical therapy that targets specific sites of pathophysiologic pathways involved in the pathogenesis of OA. Medical attention has shifted from easing the pain of OA to slowing the disorder's progression and actually preventing it.

Strategies under consideration at this time include cyclooxygenase-2 (COX-2) inhibitors (new agents or "super aspirins" that act against inflammation with much less effect on the gastrointestinal system), nitric oxide synthesis inhibitors and antioxidants, chondrocyte and bone growth promoters, metalloproteinase and cytokine inhibitors, and gene therapy.⁷⁸

Research on cytokines, growth factors, and signaling pathways that was started in the 1990s is now producing new concepts for disease-modifying OA drugs (DMOADs), much like the disease-modifying drugs developed and now available for RA.^{443,513}

Antiresorptive drugs aimed at altering the increased metabolic states of the subchondral bone may have an effect in altering damage done to the overlying cartilage. This approach is based on the hypothesis that the underlying subchondral bone, either indirectly through biomechanical effects or directly via release of cytokines, is responsible for driving the release of degradative enzymes and, ultimately, the destruction of overlying cartilage.

A fairly recent nondrug, nonsurgical treatment known as *viscosupplementation* has been developed. This intervention involves direct injections into the knee of substances derived from sodium hyaluronate (hyylan G-F 20 [Synvisc], Hyalgan), a principal component of natural synovial fluid. These injections help restore some of the viscosity and elasticity of the diseased joint fluid and offer pain relief for 6 to 12 months. Viscosupplementation is used when standard conservative treatments for knee OA (e.g., medications, physical therapy, behavioral therapy) have been inadequate or ineffective.

Education. The CDC has launched a major public health initiative, called the National Arthritis Action Plan, to identify and change behaviors that may cause OA, calling for a significant change in the way this disease is treated. Medical interventions involving expensive medications, joint injections, and surgery are now suggested for use in the 10% to 30% of cases where OA will progress to severe joint damage.

Multimodal treatment should include client education and self-management. Two excellent resources for consumer education and self-management of OA are available: *The Arthritis Cure*⁴⁹⁵ and *Maximizing the Arthritis Cure*.⁴⁹⁶ More attention to psychosocial problems (e.g.,

isolation, depression) that may influence the person's perception of pain, and to exercise is recommended. The value of a well-designed exercise program including training for strength and endurance also has been recognized.^{103,119,120,514} Aquatic therapy is especially helpful as a form of moist heat and gravity-eliminated resistive exercise.

Behavioral interventions directed toward enhancing self-management are important, including prevention (see previous discussion), diet and weight control, and low-impact exercise. Complementary approaches to OA (e.g., chondroitin sulfate and glucosamine supplements, acupuncture, yoga, tai chi¹⁸⁰) are also gaining in popularity with many people.

Complementary or Alternative Therapy. Glucosamine and chondroitin sulfate are components of cartilage taken with the hope of decreasing pain and improving function while halting the progression of disease by stopping an enzyme that is believed to break down cartilage. Glucosamine is derived from the shells of lobster, shrimp, and crabs; chondroitin sulfate is derived from cow cartilage. These are available as a nutritional supplement, sometimes referred to as a *nutraceutical*.

Long-term use of these agents may provide combined structure-modifying and symptom-modifying effects, making these potentially disease-modifying agents for OA. Most people take 1500 mg of glucosamine and 1200 mg of chondroitin daily; anyone weighing more than 200 lb should increase the dosage to 2000 mg of glucosamine and 1600 mg of chondroitin sulfate.^{190,418,419,495}

Investigators in various trials report a range of results with this treatment, making conclusions difficult. Differences in studies are too great to make comparisons or metaanalysis conclusive. Different glucosamine preparations add to the complexity of comparative studies.^{417,530} The safety of this product used over a long period of time has also been questioned.²⁹⁵

An extensive review of four metaanalyses as well as the recently published findings of the Glucosamine/Chondroitin Arthritis Intervention Trial (GAIT)⁵⁶ on the efficacy of glucosamine sulfate, glucosamine hydrochloride, and chondroitin sulfate for the treatment of OA has been published.¹⁰⁴ Results of previous studies were inconclusive because of poor research design. The GAIT study used rigorous research design to elicit cause and effect and found that glucosamine sulfate and chondroitin sulfate were not effective in reducing knee pain; when combined together, these two were more effective for individuals with moderate to severe pain.¹⁰⁴

In another metaanalysis, 20 trials were reviewed to compare the effects of chondroitin with either placebo or no treatment in adults with hip or knee OA. The evidence did not support the use of chondroitin for OA and found no clinically relevant benefit for individuals with advanced OA. According to researchers, the symptomatic benefit of chondroitin is minimal or nonexistent.^{416a}

Results of long-term studies in these two areas (behavioral interventions, complementary medicine) are expected to be published in the next few years. Preliminary results of clinical trials are available but limited.¹³³ Best available evidence suggests that acupuncture, several

herbal preparations (e.g., devil's claw root, white willow bark), and capsaicin cream are beneficial. Evidence is weak or contradictory for homeopathy, magnet therapy, tai chi, leech therapy, music therapy, yoga, imagery, and therapeutic touch. Many other treatments have not been scientifically tested.¹¹²

The Arthritis Foundation has published an excellent resource on alternative therapies²³; some experts advise people not to think of these therapies as "alternative" but rather as synergistic measures that can be integrated into a total care plan. An area of medicine called complementary and integrative medicine (CIM) has developed as more medical doctors have started incorporating these treatment methods into their overall management plans.

Surgery. Surgical intervention is considered when pain and loss of function are severe. Arthroscopic management, including lavage and debridement, abrasion arthroplasty, subchondral penetration procedures such as drilling and microfracture, and laser/thermal chondroplasty, may benefit some individuals, potentially delaying reconstructive procedures (e.g., osteotomy, joint arthrodesis or fusion, total joint replacement). Each of these procedures is under investigation for efficacy and long-term results.²¹²

To help joint replacements last longer, intense research is focusing on more wear- and corrosion-resistant materials as well as investigating how the tissue around the replacement responds. Replacement of damaged cartilage is also under investigation using one of three types of cartilage: one's own cartilage, donor cartilage, and cartilage produced by tissue engineering of progenitor cells (see the section on Tissue Engineering in Chapter 21). Medications that stimulate human cartilage matrix formation without stimulating the chondroresorption processes are also under investigation.¹⁸⁸

PROGNOSIS. OA is a major contributor to functional impairment and reduced independence in older adults (more than 65 years). It is a chronic condition with unpredictable symptoms that often cause fluctuations in pain and function.²

Mobility disability, defined as needing help walking or climbing stairs, is common for those with hip and/or knee OA. The social burden in terms of personal suffering and use of health resources is expected to increase with the increasing prevalence of obesity and the aging of the American population.²¹⁵

Although there is no known cure for OA, by following the guidelines for lifestyle changes, pain management, and self-management incorporating exercise and weight loss, affected individuals can substantially decrease the pain and dysfunction associated with OA.

SPECIAL IMPLICATIONS FOR THE THERAPIST 27-7

Osteoarthritis

PREFERRED PRACTICE PATTERNS

4E: Impaired Joint Mobility, Motor Function, Muscle Performance, and Range of Motion Associated with Localized Inflammation

4H: Impaired Joint Mobility, Motor Function, Muscle Performance, and Range of Motion Associated with Joint Arthroplasty

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

Therapists need to be aware of the potential poor correlation between the extent of radiographic degenerative changes and the presence of symptoms. The assumption that a person with significant, extensive joint degeneration will not improve should not be made until a thorough rehabilitation program has been attempted.

Conversely, it should not be assumed that someone with minor radiographic degenerative changes cannot be experiencing severe, intense pain. Therapists should rely primarily on the clinical examination findings for direction regarding the development of prognosis (including plan of care) and intervention.

Medications and Nutraceuticals

The medications commonly prescribed for OA have significant potential side effects. The NSAIDs have ulcerogenic potential, especially when taken with over-the-counter drugs. Gastric irritation can result from inhibition of prostaglandin production, which can reduce mucus and bicarbonate production and decrease local blood flow.

Peptic ulcer disease can be manifested by a multitude of complaints, including indigestion, nausea, vomiting, thoracic pain, and melena (black tarry stools). The onset of any of these complaints calls for communication with a physician.

Nutraceuticals, such as chondroitin, can also cause problems in some individuals. Chondroitin is chemically similar to blood-thinning drugs, such as heparin, warfarin (Coumadin), and even aspirin, and could cause excessive bleeding. Anyone taking these supplements with unexplained back or shoulder pain or excessive bruising or bleeding from any part of the body (nose, gums, vagina, urine, rectum) must be evaluated by a physician.

Anyone taking these supplements who also has diabetes should be aware that some studies in animals have shown that glucosamine increases blood glucose levels. Preliminary studies in humans remain inconclusive.³⁴⁹

Joint Protection

People with symptoms associated with OA must understand their role in minimizing the mechanical stresses on the involved joint or joints. The diseased joints need to be protected from excessive mechanical forces. Educating the client on how to reduce the daily wear and tear on the joint is essential. This may include the use of postural supports or a cervical collar or an exercise program to vary the stresses with which the involved joints are dealing.

Proper posture and avoidance of prolonged stressful postures, use of supports, varying of physical activities to vary the stresses (i.e., alternating biking with swimming or walking), and following through with a

flexibility and strengthening exercise program are all components under the affected individual's control.

Wearing shoes that fit properly and are appropriate for the activity may help avoid injury. Good alignment of the joints is important, especially in the knee. Evaluating the need for an assistive ambulatory device, a shock-absorbent shoe insert or heel wedge, brace, or other orthotic devices is important to unload the pressure on affected joints. A simple lateral wedge insole of 5 or 10 degrees directly reduces the knee varus torque in individuals with medial knee OA and can interrupt the OA cycle, slowing the progression of the disease and disability.²⁴⁵

Exercise and Osteoarthritis

Since the realization that structured exercise programs can improve function without exacerbating symptoms, exercise and joint protection techniques have become mainstays of treatment. In fact, attempts to alleviate pain through pharmacologic or physical modalities may not improve symptoms unless accompanied by some form of physical conditioning.³⁰⁵

The plan of care for someone with OA is dictated by the extent of the disease and the joints involved, but everyone with OA should be encouraged to continue exercising, including strength training and low-intensity aerobic components.

GENERAL CONCEPTS

Physical therapy has been shown effective in OA of the knee to reduce pain; improve physical function; increase isometric muscle strength, gait speed, and stride length; and improve quality of life.

A combination of manual physical therapy and supervised exercise provides beneficial effects still present 1 year later and delays or prevents the need for surgical intervention, with fewer joint replacements reported.^{102,131} Such supervised exercise/manual therapy programs have been shown to increase improvement and provide greater symptomatic relief compared with a similar unsupervised home exercise program.^{101,298}

Optimizing existing and potential joint function by improving flexibility and strength is important. In fact, exercise combined with self-management appears to have a similar effect to drug treatments and is generally safer.^{114,411} Low-intensity, controlled movements that do not increase pain can help individuals regain or maintain motion and flexibility.^{119,120}

The therapist can use the same guidelines for all individuals when establishing frequency, intensity, and duration by following well-known general concepts (e.g., establish *intensity* by calculating heart rate at 60% of the heart rate maximum, begin at the individual's level of *duration* anywhere from 1 to 2 minutes and build up to 30 minutes, work toward a *frequency* of five to seven times per week). Clients should be taught how to monitor and progress frequency, intensity, and duration; use good biomechanics; and avoid exacerbating musculoskeletal symptoms.

In the presence of mild joint swelling, the client should be taught to use ice before exercise and to

incorporate a program of submaximal exercise to warm up before beginning the prescribed exercise program.³²² If there is joint effusion, the surrounding muscles cannot contract maximally because of reflex inhibition caused by joint distention. Submaximal exercise for 3 or 4 minutes on a swollen joint decreases this inhibition mechanism, allowing for continued strength training. Moderate to severe joint effusion may require additional physical therapy intervention, such as electrical stimulation.

Resistance and low-intensity aerobic exercise may reduce the incidence of disability related to ADLs and prolong autonomy in adults over 60 years of age, specifically those with knee OA. The lowest ADL disability risks were found for participants with the highest compliance to the exercise program.³⁹⁵ Long-term weight training and aerobic walking programs significantly improve postural sway in older adults with OA, thereby improving static postural stability.³³³

In the case of the frail older adult, progressive resistive exercises have been shown to be safe and effective following an acute illness when monitored carefully (e.g., measuring vital signs, observing for signs of physiologic distress).⁴⁸¹ Other specific exercise recommendations and safety considerations are available,^{15,102,342,346} but the optimal type of exercise and duration for the prevention and treatment of OA remain under investigation.

Successful management of degenerative joint disease may require evaluating and treating dysfunction of other body regions. For example, someone with OA of the knee and pain on ambulation may have significant foot or ankle dysfunction. Treating joint and soft tissue hypomobility and muscle imbalances and fabricating orthoses may considerably alter the mechanical stresses on the arthritic knee.

For those individuals who do not seem to respond, progress, or improve with physical therapy intervention, a number of factors should be considered. Mode of treatment delivery is not the only parameter. Treatment compliance, mechanical characteristics (e.g., joint laxity, malalignment), and radiographic severity should be carefully considered. Research is needed to focus on predictive factors and outcomes for individual characteristics and type of exercise protocols prescribed.¹²⁵

EDUCATION

Exercise at moderate levels of training has been shown to be effective in clinical trials of older adults in improving generalized and localized pain and functional status. There is no evidence that one type of exercise is better than others. The exercise prescription must be individualized to the needs and preferences of each person.

Clients should be encouraged to exercise to the extent that they are capable. Many older adults are more likely to adhere to a program that is short and sweet (i.e., short, frequent episodes of exercise) rather than long, once-a-day programs. Educating, motivating, and providing prescriptive exercise are important

Continued.

roles the therapist plays to help maximize function and prevent significant recurrence of symptoms.

The therapist should take into account psychosocial factors, especially self-efficacy, which has been shown to be an important variable in the rehabilitation process predictive of physical function.^{126,179} Self-efficacy is defined as a person's belief about his or her ability to successfully complete a task or activity.³⁰ See further discussion of self-efficacy in Chapter 2.

Sometimes increasing physical activity does cause increased pain, but studies show this is short-lived. The therapist can help clients get over the "pain hump" by assuring them that this response is normal and temporary.¹²⁷ Older adults may become more motivated to exercise if they understand the benefits. Teach them the overall health benefits of exercise in preventing chronic conditions such as diabetes, heart disease, osteoporosis, and cancer.

The therapist can help educate them about how exercise can help reduce their risk factors for OA as well as decrease risk for falls with a program of balance and gait training.¹⁹⁹ Use the information in this chapter on falls prevention to personalize the message and remind the client that people who exercise have a lower incidence of falls, fractures, hospitalizations, and premature death.³³⁴

SPECIFIC EXERCISE TRAINING

Muscles have been shown over and over to be the major shock-absorbing mechanism of joints, especially the knee. Eccentric muscle performance serves this shock-absorbing function, supporting the idea that rehabilitation programs should include activities to enhance eccentric function, especially of the quadriceps muscle.

The quadriceps muscle group must absorb the force and decelerate the increasing load as the weight-bearing limb stabilizes under the load. Overloading the bones' capacity to accept force may be what leads to osteoarthritic changes and/or pain in the knee.¹⁶⁵

Studies show that quadriceps weakness might be due to muscle dysfunction and not necessarily muscle atrophy. Quadriceps weakness may be present in people who have OA but do not have knee pain or muscle atrophy. Pain may be a consequence of the changes in muscle activity rather than the other way around. More study is needed to identify the level of motor units' activation and contractile properties of the muscle in OA during concentric and eccentric contractions.^{466,467}

For now, these researchers recommend that both isokinetic concentric and eccentric strength be measured in people with knee OA; exercises to strengthen both types of muscle contractions should be included in the rehabilitation program for knee OA to prevent possible injury.¹⁶⁵

In one small study, a group of 90-year-old nursing home residents increased quadriceps strength and walking speed by 174% and 48%, respectively, in 8 weeks with a weight-resistance training program, suggesting that no one is too old to benefit from exercise.¹²³

Open chain quadriceps-strengthening exercises can cause exercise-induced arthralgias in this population and generally should be avoided. Closed chain kinetic strengthening exercises (e.g., leg press, wall slide) can be effective for quadriceps strengthening and are usually well tolerated when performed correctly. Hamstring strengthening should be included when weakness is present. Research on strengthening exercises for people with hip OA is not as abundant as that for people with knee OA, but the evidence supports a similar approach.⁴⁷⁸

It is likely that the loss of joint proprioception associated with OA may contribute to gait alterations, muscle imbalances, repetitive microtrauma, loss of coordination, and, ultimately, excessive joint loading.²⁷⁷ Functional ability is also limited in individuals with OA who have poor proprioception combined with muscle weakness.⁵¹⁷ Exercises to facilitate proprioception and closed kinetic chain exercises for knee OA have both been shown to improve functional score, walking speed, and muscle strength.²⁸⁵

AQUATIC PHYSICAL THERAPY

Aquatic physical therapy is used often in the management of individuals with hip and knee OA. Studies to confirm the efficacy of this treatment and the long-term outcomes are lacking or yield inconsistent results. Compared with no intervention, a 6-week program of aquatic therapy resulted in significantly less pain with improved physical function, strength, and quality of life.

In studies with only short-term follow-up, aquatic exercise did not make the joint condition worse or result in injury.^{196,533} Compared with a gym-based resistance exercise program, aquatic therapy yields equal results.¹²⁹

Degenerative Intervertebral Disk Disease

Overview and Definition

The degenerative joint process described in the previous section applies to any synovial joint, including the facet joints of the spinal column. Degenerative joint changes are not limited to synovial joints, however, and in the spine (particularly the low lumbar segments) they commonly occur at the intervertebral disk articulations as well.²⁶²

Currently, there is no consensus on what "disk degeneration" actually is or how it should be defined or distinguished from the physiologic processes of growth, aging, healing, and adaptive remodeling. The following working definition has been proposed as a starting point: "The process of disk degeneration is an aberrant, cell-mediated response to progressive structural failure."¹

Lumbar disk degeneration begins early in life. Severe macroscopic changes are visible from the age of 30 onward. Atherosclerosis is present by middle age or even earlier for some people in the abdominal aorta.³³⁹ It is estimated that half of all Americans over the age of 40 are affected by degenerative disk disease (DDD).

Disk degeneration follows a predictable pattern. First, the nucleus in the center of the disk begins to lose its ability to absorb water. The disk becomes dehydrated. Then the nucleus becomes thick and fibrous, so that it looks much the same as the annulus. As a result, the nucleus isn't able to absorb shock as well. Routine stress and strain begin to take a toll on the structures of the spine. Tears form around the annulus. The disk weakens. It starts to collapse, and the bones of the spine compress.³²⁵ This process will be discussed in greater detail later in the Pathogenesis section.

Incidence

Lumbar DDD is a common musculoskeletal disease estimated to affect up to 5% of all adults. Although this disease and resulting condition can occur before age 20, it tends to peak in the fourth and fifth decades of life and declines after that. It has been reported that men suffer from sciatica due to disk herniation about 1.5 to 3 times more often than women, but it is not clear if this is a true difference in prevalence of DDD or can be attributed to anatomic and mechanical factors that contribute to nerve root compression.⁴

Risk Factors

The greatest risk factor for disk degeneration is genetic inheritance, which accounts for approximately 50% to 70% of the variability in disk degeneration between identical twins. Individual genes associated with disk degeneration have been identified (e.g., aggrecan, collagen type IX, matrix metalloproteinase-3 [MMP-3], vitamin D receptor).^{1,34,434}

Twin and family studies have shown that the risk of developing a lumbar disk herniation is approximately five times greater in people who have a positive family history. Mutations that alter collagen I structure (the major component of the outer annulus) may contribute to annular tears and disk ruptures. Sequence variations in the genes for collagen IX may also be linked with DDD.⁴

Age and body weight appear to be two other significant risk factors for DDD. There has been a prevailing view that DDD occurs as a result of excessive forces, particularly from the cumulative effects of repeated loading from occupational physical demands, such as manual material handling. Results of research to confirm or refute this hypothesis have been mixed; some studies find a correlation between disk degeneration and physical demands, while others do not.^{35,527} It appears that actual time spent lifting and carrying loads is less important than body weight or mass.⁵²⁷

Results of a recent study of the Finnish Twin Cohort suggest that routine (daily) physical loading of the spine may actually have a training (rather than detrimental) effect. Occupational lifting or repeated loading of the spine from physical activity may in fact benefit the disks;⁵²⁷ cyclic mechanical stress may increase the growth rate and collagen fibers of the nucleus pulposus.³¹³

Height has been suggested but not consistently found to be a risk factor in all studies; the link between DDD and obesity and smoking also remains controversial. The role of psychosocial factors has also been investigated,

Box 27-8 DISK DEGENERATION

Three Stages of Disk Degeneration²⁵¹

- Dysfunction
 - Circumferential and radial tears in the disk annulus
 - Localized synovitis and hypermobility of the facet joints
- Instability
 - Internal disruption of the disk
 - Progressive disk resorption
 - Degeneration of the facet joints with capsular laxity
 - Subluxation
 - Erosion
- Stabilization
 - Osteophytosis (bone spur formation)
 - Spinal stenosis

Events Leading to Disk Degeneration

- Impaired cellular nutrition
- Reduced cellular viability
- Cellular senescence
- Accumulation of degraded matrix macromolecules
- Fatigue failure of the matrix

Risk Factors for Disk Degeneration

- Age
- Body Mass Index (BMI)
- Heredity
- Physical loading*
- Occupational (repetitive) lifting*

*New evidence suggests that these factors may not be as strongly linked with degenerative disk disease as previously thought. Regular physical activity may benefit the disks. Age, body weight, and hereditary factors may be the greatest risk factors for degenerative disk disease.^{4,527}

and they have been found to have a positive link as risk factors for disk herniation, which can lead to further DDD. Atherosclerosis is another potential risk factor. Obstruction in the abdominal aorta, lumbar, and middle arteries can lead to ischemia in the lumbar spine, resulting in hypoxia and tissue dysfunction with eventual disk degeneration.²⁴⁰

Etiology and Pathogenesis

The intervertebral disk undergoes marked changes with age, although genetic inheritance, inadequate metabolite transport, and loading history combined with age contribute significantly to structural failure that can occur during ADLs.¹

The most significant alterations occur in the nucleus pulposus. The number of cells and the concentration of proteoglycans and water decrease. In addition, there is fragmentation of proteoglycans, which contributes to water loss. As the nucleus breaks down, the fibrocartilaginous inner annulus expands. Fissures and clefts may form within the disk, and the height of the disk decreases. This loss of disk height can contribute to the age-related condition of spinal stenosis.²¹

A number of events can contribute to the three stages of age-related disk degeneration (Box 27-8).²⁵¹ The most important event appears to be the decreased cellular

function and concentration. The progressive decline in arterial supply to the periphery of the disk and the impairment of nutrient delivery across the cartilaginous endplate contribute to the reduced nutritional supply to the cells, affecting cellular function.

In addition, the impaired cartilaginous endplate diffusion results in reduced cellular waste product removal and an increased lactic acid concentration. The resultant decreased pH level compromises cellular metabolism and biosynthesis and can lead to cell death. The reduced biosynthesis can adversely affect the biomechanical properties of the matrix over an extended period of time.

Besides the internal events affecting the general health of the intervertebral disk, repetitive external mechanical loading on the structure can lead to fatigue failure of the matrix. When enough structural breakdown occurs, what once were normal mechanical loads acting on a normal disk are now excessive loads on a compromised disk. At this point the degenerative process is accelerated.¹⁸

Collapse of the inner annulus into the nucleus is a common feature in the disks of older adults, with the anterior annulus being affected more than the posterior (Fig. 27-6). This could be caused by nucleus decompression following endplate fracture. In many older disks, the cartilage endplate becomes detached from the underlying bone, presumably because the high internal pressure that presses it against the bone in young disks has been lost.^{1190,491}

The results of the Videman et al study⁵²⁷ go against the traditional view that physical loading and repetitive spinal movements are bad for the back. The authors suggest that lifelong loading from body weight is more detrimental than work-related lifting and loading. Loading from daily physical activity may actually help offset the effects of aging. Forces across the lumbar spine vary with body weight and lifting strength; this may explain why some disks degenerate more than others. The authors also suggest that smaller disks hold up better because it is easier for nutrients to reach each cell.

Associated with intervertebral disk degeneration are spinal stenosis and degenerative spondylolisthesis, two common conditions in the older adult (more than 65 years). As the intervertebral disk loses height, the annulus may bulge circumferentially and the ligamentum flavum can buckle. Both encroach on the spinal canal, subtalar (facet) recesses, and lateral intervertebral foramina. With a loss of disk height, compressive force increases on the neural arch,⁴⁰⁵ causing OA of the facet (apophyseal) joints and osteophytes around the margins of the vertebral bodies (Fig. 27-7).⁵²⁶ In addition, concurrent osteophyte formation on the vertebral bodies or articular processes may occur, compounding the stenosis.

Degenerative spondylolisthesis is marked by anterior slippage of one vertebra over another with an intact posterior neural arch. The L4-L5 spinal segment is the most common site for this to occur (Fig. 27-8). Lytic spondylolisthesis is marked by anterior slippage of one vertebra over another with a defective posterior neural arch (Fig. 27-9). The L5-S1 spinal segment is the most common site for lytic spondylolisthesis to occur. The loss of disk height associated with degeneration allows for a buckling of the annulus and ligamentum flavum, slackening them some-

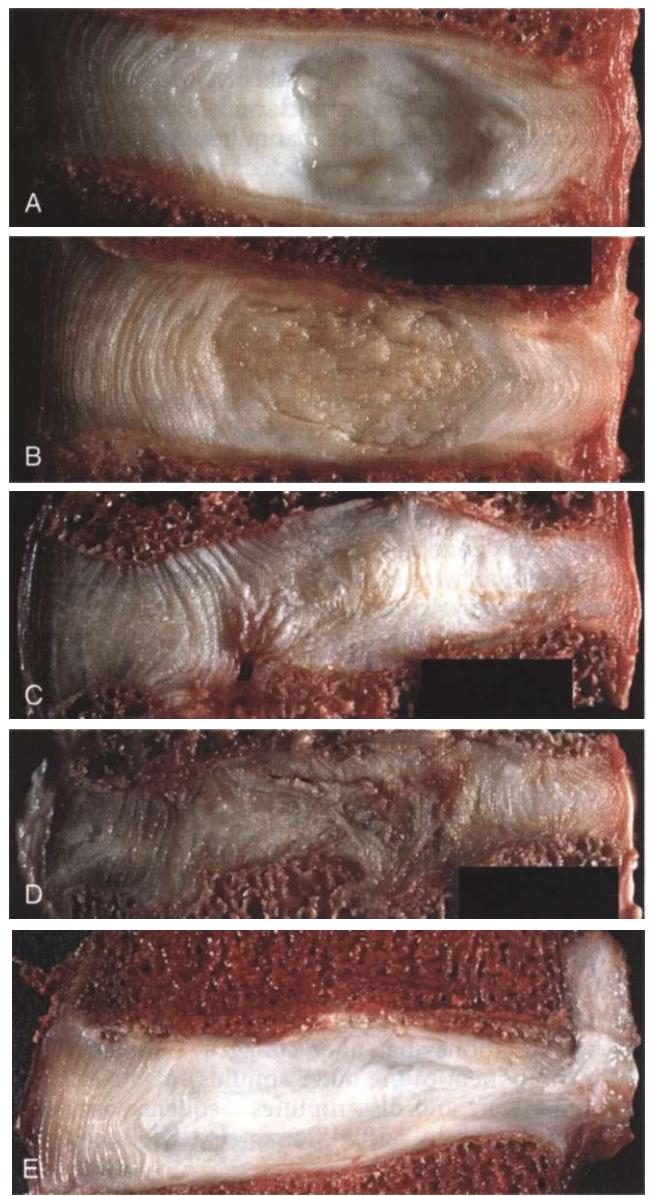


Figure 27-6

Lumbar intervertebral disks (midsagittal section; anterior on the left). **A**, Young disk in a male, 35 years old. **B**, Mature disk in a male, 47 years old. **C**, Disrupted disk in a male, 31 years old. Note the endplate and inward collapse of the inner annulus. **D**, Severely disrupted disk in male, 31 years old. Note the collapse of disk height. **E**, Disk induced to prolapse in cadaver male, 40 years old. Some nucleus pulposus has herniated through a radial fissure in the posterior anulus (right). (From Adams MA: *The biomechanics of back pain*, Edinburgh, 2002, Churchill Livingstone.)

what. This allows the vertebrae to migrate anteriorly in response to the shear forces inherent to the lumbar lordosis.¹⁸⁹

In contrast to the L5-S1 segment, the facet joint orientation at the L4-L5 segment tends to be more in the sagittal plane, so there is no structural bar to anterior slippage. The facet joint orientation at L5-S1 tends to be more in the frontal plane, making anterior migration of L5 difficult unless there is a structural defect in the posterior neural arch. Stenosis can be caused by the displacement



Figure 27-7

Radiograph of a lumbar spine (anterior on left) with severe disk narrowing, vertebral osteophytes, sclerosis of the endplates, and selective loss of horizontal trabeculae from the vertebral body. (From Adams MA: *The biomechanics of back pain*, Edinburgh, 2002, Churchill Livingstone.)

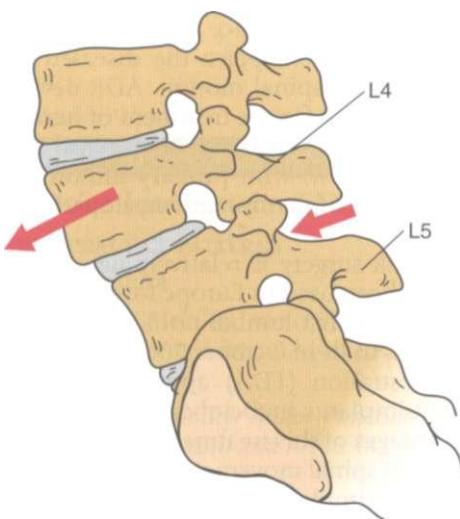


Figure 27-8

Degenerative spondylolisthesis at L4-L5. (From DeRosa C, Porterfield JA: Lumbar spine and pelvis. In Richardson JK, Iglarsh ZA, eds: *Clinical orthopaedic physical therapy*, Philadelphia, 1994, Saunders, p 144.)

of one vertebra over another as well as by the concurrent buckling of the annulus and ligamentum flavum.

Clinical Manifestations

Disk degeneration is most likely to affect the lower lumbar spine but can affect the upper lumbar spine and cervical spine as well. Low back pain is often the first symptom, but DDD is asymptomatic in up to one third of all affected individuals.^{45,228}

DDD is one of the most common causes of low back pain with radiculopathy. The intervertebral disk changes include alterations in volume, shape, structure, and composition. Although there is not a 100% correlation between the presence of DDD and pain complaints, the

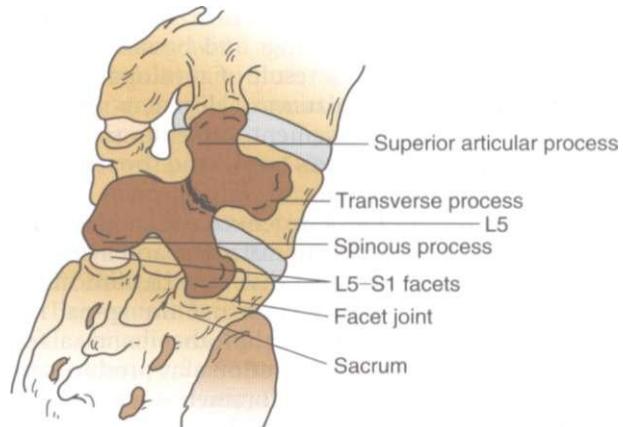


Figure 27-9

Spondylolysis or posterior arch defect, which can lead to a lytic spondylolisthesis. The Scottie dog with a collar, which is visible on the radiograph (posterior oblique view), is outlined. (From Magee D: *Orthopaedic physical assessment*, ed 2, Philadelphia, 1992, Saunders.)

structural alterations will decrease motion and alter the mechanical properties of the spine.⁵⁸

Most people with DDD have a gradual onset of increasingly severe midline lower back pain. At first, symptoms only last a few days. This type of back pain is often intermittent but recurring over the years. Each time it occurs, the pain may seem worse than the time before. Eventually the pain may spread into the buttocks or thighs, and it may take longer to subside.³²⁵

When pain is present, it tends to get worse after heavy physical activity or after a prolonged time in one position. The back may also begin to feel stiff. Resting the back eases pain. Aching of the buttock(s) and posterior thigh(s) with ambulation is common.

The symptoms of DDD should not be confused with disk herniation, which may occur as part of the process of disk degeneration but is not a constant feature and is a separate entity from DDD. Clinical presentation of disk protrusion and herniation can vary widely but can occur as a result of tears of the annulus fibrosus, causing acute back or neck pain or radicular pain if there is herniation of the nucleus pulposus.

In the lumbar spine, radiculopathy causing sciatic pain and restricted straight leg raise may occur as the sheath of the nerve root is compressed⁴⁸³ or as a result of inflammatory irritation from chemicals released by the damaged disk.

Centralization of radiating pain is characteristic of sciatica from disk protrusion or herniation. Specifically, leg pain routinely reduces or "retreats" to the lumbar midline before the disappearance of back pain. This distinctive pain pattern was first described by McKenzie.³²⁰

Other signs of disk herniation include ankle dorsiflexion weakness; great toe extensor weakness; impaired ankle reflexes; loss of light touch sensation in the medial, dorsal, and lateral aspects of the foot; and positive ipsilateral or crossed straight leg raise test. These six neurologic tests allow detection of most clinically significant nerve root compromises resulting from L4-L5 or L5-S1 disk herniations.^{42,171}

Gait abnormality, muscle weakness, sensory changes such as numbness and tingling, and bowel or bladder dysfunction can occur as a result of myelopathy associated with either disk herniation or degeneration. In the case of cervical disk involvement, difficulty swallowing, hand numbness, Lhermitte's sign, and hoarseness or voice difficulties may occur.

MRI observations of disk degeneration and herniation are poorly correlated with clinical signs and symptoms. For example, a large, extruded disk may be clinically tolerable if the spinal canal is large and the spinal nerve roots are not compressed. On the other hand, a focal, contained subligamentous herniation may produce severe symptoms if it occurs in the foramen adjacent to the dorsal root ganglion of the affected nerve.³²¹

In the case of disk degeneration associated with stenosis and spondylolisthesis, the person can be asymptomatic, even when moderate to severe changes are observed on imaging studies. Conversely, early or mild changes can be accompanied by severe pain and neurologic symptoms.

MEDICAL MANAGEMENT.

DIAGNOSIS. Diagnosis requires a thorough history, physical examination, and imaging protocol; provocative diskography (with dye injected into the suspected disk or disks) will reveal annular tears that cause pain and offer a definitive diagnosis, especially when used in conjunction with other imaging studies. The dye seen on x-ray can give information about the health of the disk(s). This test may be done when the surgeon is considering surgery, since it can help determine which disk is causing the symptoms.

The x-rays provide a measure of disk space; narrowing of the disk spaces is suggestive of the degenerative process. The presence of osteophytes affecting facet joints and any spondylolysis or spondylolisthesis can also be visualized on x-rays.

MRIs visualize the soft tissues of the body and are helpful for showing if the disks are able to absorb water and any radial fissures present inside the disk. They can also show if there are problems in other soft tissues, such as the spinal nerves.

TREATMENT. Conservative care, including NSAIDs, mild analgesics for short-term pain control, physical therapy, lifestyle changes (e.g., smoking cessation, weight loss, fitness program), and aerobic conditioning is the first line of treatment for painful DDD.³²¹ The goal is to reduce painful symptoms and enable the person to get back to normal activities as soon as possible. Bed rest is no longer the standard of care.

Surgery. Surgery to remove the disk or disk fragments (diskectomy), laminectomy, and/or spinal fusion may be considered if conservative care has been unsuccessful and/or neurologic symptoms persist. The timing of surgery can be important to avoid permanent nerve damage and footdrop.

Long-term studies show that in the absence of neurologic compromise, the outcomes for conservative care and surgical intervention are the same 10 years after the initial incident, suggesting that surgery can be postponed

indefinitely. The exceptions are when the individual is at risk for neurologic damage and when conservative care does not alter the pain, thereby compromising function and quality of life.

For individuals with chronic, nonradicular, diskogenic low back pain, intradiskal electrothermal therapy (IDET) has been used with success in 60% to 80% of the cases. Thermal energy directed at the disk material through a catheter uses heat to induce tissue shrinkage. IDET curbs the release of irritating substances and destroys the surrounding pain receptors and offers a less invasive alternative to surgery for the treatment of chronic and severe DDD.³²²

Other minimally invasive treatments include epidural injections and transdiskal radiofrequency therapy (called intradiskal biacuplasty); both have been used with varying degrees of success.³²³ The use of one to three epidural steroid injections for DDD has gained in popularity for individuals whose symptoms continue to limit function. A steroid combined with a long-lasting numbing agent is injected into the space around the lumbar nerve roots.

For some individuals, artificial disk replacement (ADR) is an alternative to spinal fusion. After removing what is left of the damaged or worn-out disk, the ADR device is inserted in the space between two lumbar vertebrae. The goal is to replace the diseased disk while keeping the normal spinal motion. ADR devices are not designed at this time for the treatment of herniated disks but for one or two levels of DDD. Cervical arthroplasty is also available now, with primary indications for the treatment of radiculopathy and myelopathy at one or two levels.

Artificial disk surgery is relatively new in the United States but has been used in Europe for many years. In the United States, the first lumbar artificial disk surgery was done in clinical trials in October 2001. The U.S. Food and Drug Administration (FDA) approved the use of the lumbar ADR implants in October 2004.³²⁴

The advantages of this treatment over spinal fusion are maintenance of spinal movement, disk height, and neural foramina, thus simulating a more normal spinal alignment, angulation, and mechanics. Immobilization is avoided with ADR, and early return of function is possible. There is the additional advantage of preventing adjacent-segment deterioration that can occur after spinal fusion.

There are also risks and disadvantages (e.g., infection; ossification; neurologic impairment; implant failure; and fracture, migration, or subsidence or sinking down into the bone).³²⁵ Although short-term and medium-term results are favorable, long-term results are not yet available.^{324,326}

Gene Therapy. Researchers are investigating the use of gene therapy to slow, prevent, or reverse the biochemical changes associated with disk degeneration. Alternatively, if the correct gene could be identified, it might be possible to manage an annular tear by direct repair or regeneration.³²⁷ In other areas of study, autologous chondrocyte transplantation may become a future treatment for DDD. Animal studies have shown that chondrocytes removed from damaged cartilaginous tissues maintain a capacity

to proliferate and reproduce tissue similar to normal intervertebral disk material.³²⁸

PROGNOSIS. Disk structural failure is irreversible, always progresses by physical and biologic mechanisms, and is closely associated with mechanical dysfunction and pain.¹ The potential for recovery varies based on the size of the protrusion, the size of the canal, the person's age and activity level, the extent of disk disruption, and similar parameters related to spondylolisthesis and stenosis.⁴⁹⁴ Laminectomy and/or discectomy removes the mechanical cause of pain, but the inflammatory process may cause persisting pain for a few more days or, in some cases, indefinitely as a result of fibrosis impinging on the nerve tissue.

For those with disk herniation, the type of disk herniation can predict the final outcome. Individuals with a bulging disk and normal annulus have the best result. Pain from sciatica is relieved after discectomy. Individuals with bulging disk and large annular defects have a high rate of reoperation.⁷⁰

Some, but not all, individuals with spondylolisthesis and stenosis may experience pain relief and improved function after surgery (fusion). In the older adult, recovery can be very slow; rehabilitation is a key feature in this process.

manifestations, including spinal canal stenosis. Therapists working with anyone with back or neck pain should ask about these symptoms, and if any are present, immediate communication with a physician is recommended.

Even with the local degenerative changes in the spine, stenosis may be marked primarily by lower extremity symptoms (neurogenic claudication) as opposed to back pain. The symptoms may include pain, altered sensation, or muscle weakness.

The symptoms are typically brought on by walking and are relieved by prolonged rest (sitting or lying down) or by flexion of the spine. When a person is upright and walking, the lumbar spine is in a relatively backward-bent position, which further reduces the size of the foramina and subarticular recesses. When the spine is flexed, the foramina are opened, relieving pressure on the nerves. Similar symptoms are noted with vascular claudication (tissue ischemia secondary to vascular insufficiency), except that vascular symptoms are not dependent on the position of the spine but rather on the level of activity.

Functionally, people with neurogenic claudication lack the backward-bending range of motion to tolerate walking. If the therapist can improve overall backward-bending range of motion by mobilizing the thoracic and upper lumbar regions and by stretching the hip flexors, the affected individual may be able to assume an upright posture without reaching end range of motion at the involved segments where the nerve compression is occurring. If this can be accomplished, walking tolerance should improve.

Exercise and Degenerative Disk Disease

Chronic low back pain from DDD is difficult to treat. Nonsurgical care is the first line of treatment, often involving the physical therapist. Each individual must be assessed carefully and the plan of care provided based on presenting features. Back extension exercises, abdominal strengthening, postural training, and flexibility exercises for the spine and hamstrings may be helpful.

There is some evidence supporting the efficacy of exercise therapy.^{306,521} Stability exercises have been shown effective for the treatment of DDD in at least one study. When compared with mobilization treatment, stabilization exercises improved pain and function significantly more than mobilization.²⁸⁴

More studies are needed to identify the exact type of exercise along with frequency, duration, and intensity. Predictive factors for outcome need to be identified along with identification of candidates most likely to improve with exercise therapy (or type of exercise needed for each person).

Aerobic conditioning is an important feature of the exercise program, especially to address the vascular component of this condition. Walking, swimming and/or water aerobics, and stationary bicycle are some possible choices. Each individual's lifestyle and overall physical condition will dictate the most likely course of action to prescribe or suggest.

SPECIAL IMPLICATIONS FOR THE THERAPIST 27-8

Degenerative Intervertebral Disk Disease

PREFERRED PRACTICE PATTERNS

4B: Impaired Posture

4E: Impaired Joint Mobility, Motor Function, Muscle Performance, and Range of Motion Associated with Localized Inflammation

4F: Impaired Joint Mobility, Motor Function, Muscle Performance, Range of Motion, and Reflex Integrity Associated with Spinal Disorders

4I: Impaired Joint Mobility, Motor Function, Muscle Performance, and Range of Motion Associated with Bony or Soft Tissue Surgery

5H: Impaired Motor Function, Peripheral Nerve Integrity, and Sensory Integrity Associated with Nonprogressive Disorders of the Spinal Cord (myelopathy)

Degenerative changes of the spine affecting the intervertebral disks often occur concomitantly with other spinal degenerative and osteoarthritic changes such as spinal stenosis and with the effects of vascular occlusion from atherosclerosis. There are two additional conditions associated with spinal stenosis: cauda equina syndrome and vascular/neurogenic claudication.

Cauda equina syndrome is characterized by pain in the upper sacrum, with paresthesias of the buttocks and genitalia possibly resulting in bowel or bladder incontinence or sexual dysfunction (difficulty achieving orgasm or inability to achieve or maintain an erection). Numerous conditions may lead to these

Table 27-1 Osteoarthritis and Rheumatoid Arthritis

	Osteoarthritis	Rheumatoid Arthritis
Onset	Usually begins at age 40 yr	Initially develops between ages 25 and 50 yr
Incidence ⁷³	Gradual onset over many years; affects majority of adults over age 65 yr 12% of U.S. adults; 21 million people	Onset may be sudden over several weeks to months; intermittent exacerbation and remission 1%-2% of U.S. adults; 600,000 men, and 1.5 million women; estimated prevalence rate of juvenile RA in children under 16 is between 30,000 and 50,000
Gender	Most common in men before age 45 yr; after 45 yr more common among women	Affects women 3:1 compared with men but more disabling and severe when present in men
Etiology	Etiology remains unknown; immunologic reaction with massive inflammatory response; possible genetic and environmental triggers	Multifactorial; local biomechanical factors, biochemistry, previous injury, inherited predisposition
Manifestations	Usually begins in joints on one side of the body Primarily affects hips, knees, spine, hands, feet Inflammation with redness, warmth, and swelling in 10% of cases Brief morning stiffness that is decreased by physical activity movement No systemic symptoms; possible associated trigger points	Symmetric simultaneous joint distribution Can affect any joint (large or small); predilection for upper extremities Inflammation almost always present
Associated signs and symptoms	Effusions infrequently, synovial fluid has low WBC and high viscosity ESR may be mildly to moderately increased Rheumatoid factor absent	Prolonged morning stiffness lasting 1 hr or more Systemic presentation with constitutional symptoms (e.g., fatigue, malaise, weight loss, fever) Synovial fluid has high WBC and low viscosity ESR markedly increased in the presence of an inflammatory process but not specifically diagnostic for RA Rheumatoid factor usually present but is not specific or diagnostic for RA (can be elevated in other diseases)
Laboratory values	New biomarkers under investigation (e.g., C-telopeptide, CTX, C-reactive protein, cartilage glycoprotein 39 [YKL-40])	C-reactive protein, a true indicator of systemic inflammation, strong predictor of disease outcome (RA progresses more rapidly in the presence of elevated C-reactive protein) Other biomarkers are under investigation (e.g., vascular endothelial growth factor [VEGF],* matrix metalloproteinase 3 [MMP-3])

RA, Rheumatoid arthritis; WBC, white blood cell count; ESR, erythrocyte sedimentation rate.

*VEGF is an angiogenic cytokine; its presence supports the theory that expansion of the synovial vasculature is important for the development of joint destruction in RA.

Rheumatic Diseases

Rheumatic disorders are systemic diseases encompassing over 100 different diseases divided into 10 classification categories. The pathogenesis and progression of these disorders can affect any and all body systems. The onset of joint pain and loss of function may be accompanied by fever, rash, diarrhea, scleritis, or neuritis symptoms that are not typically associated with joint or muscle conditions normally brought on by repetitive overuse or trauma.

Rheumatic disorders are also often marked by periods of exacerbation and remission. During a period of

exacerbation the therapist will often need to modify the treatment approach considerably. In addition, aggressive medical intervention (i.e., medications) may need to be initiated to prevent or minimize the tissue destruction that can occur with these disorders. Many of the rheumatic conditions are chronic and progressive, requiring long-term rehabilitation and ongoing adjustment of functional goals.

Therapists must be able to differentiate between degenerative joint disease (OA) and rheumatic joint conditions (Table 27-1). If there is any suspicion of the presence of a rheumatic disorder, immediate referral to a physician

is warranted. When someone with RA presents with systemic symptoms or if existing complaints worsen, communication with a physician is advised. An understanding of the diseases discussed in this chapter will assist the therapist regarding this clinical decision-making process.

Rheumatoid Arthritis

Overview. RA is a chronic systemic inflammatory disease presenting with a wide range of articular and extraarticular findings. Chronic polyarthritis, which perpetuates a gradual destruction of joint tissues, can result in severe deformity and disability. Systems that may be involved include the cardiovascular, pulmonary, and gastrointestinal systems. Eye lesions, infection, and osteoporosis are other potential extraarticular manifestations.

RA is a major subclassification within the category of diffuse connective tissue diseases that also includes juvenile arthritis, SLE, progressive systemic sclerosis (scleroderma), polymyositis, and dermatomyositis.¹⁶²

Incidence and Risk Factors. RA has a worldwide distribution and affects all races. Approximately 1% to 2% of the U.S. adult population (2.1 million people) has RA, which is the second most prevalent form of arthritis after OA.

Age and female gender are the two primary risk factors associated with RA. Although the onset of the disorder can occur at any age, the peak onset is usually between 20 and 50 years; with the aging of America, the prevalence of RA is expected to rise. Women are affected two to three times more frequently than men; although it is less common, children can also develop the disorder (see the section on Juvenile Idiopathic Arthritis in this chapter).

Pregnancy and oral contraceptives appear to influence the incidence and severity of the disease. The incidence of RA in women who have borne a child is lower, and oral contraceptives diminish the incidence of severe arthritis. A nulliparous woman who does not use oral contraceptives has a fourfold increased risk of developing RA.^{72,182,470}

Prophylactic administration of recombinant hepatitis B vaccine may trigger the development of RA in previously healthy people. Vaccine-related arthritis may be linked to genetic factors, such as the presence of MHC class II molecules.⁴⁰⁷ An association between autoimmune thyroid diseases and rheumatic diseases has been established, although its precise mechanism is unclear. For example, RA often occurs in association with Graves' disease and Hashimoto's thyroiditis. In these individuals there is a significant presence of antithyroid autoantibodies.³¹²

Drinking decaffeinated coffee (four or more cups per day) may be an independent risk factor among older women, especially in the presence of seropositive disease. The mechanism for this to occur remains unknown.³³⁶

Etiologic Factors. Little is known about the exact causes of RA, except that joint inflammation is a consequence of massive infiltration of immune cells, especially T lymphocytes, into the synovial fluid. Genetic predisposition and environmental triggers, such as a bacteria (e.g.,

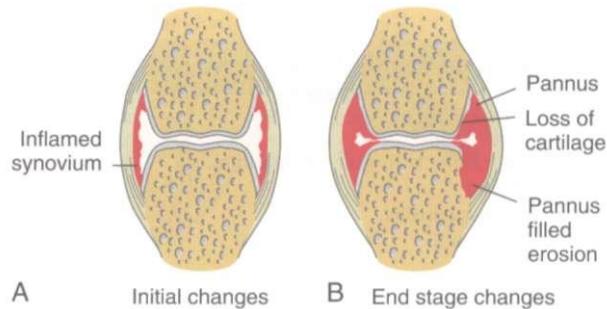


Figure 27-10

A, Early synovial changes associated with rheumatoid arthritis. **B**, Late joint changes associated with rheumatoid arthritis, including pannus formation and articular cartilage eburnation.^{116,204}

Mycoplasma fermentans), are both considered possible etiologic factors in the stimulation of T cells.^{116,204}

Pathogenesis. RA is considered an autoimmune disease, with inflammation and destruction targeted at the joint capsule (articular) and elsewhere throughout the body (extraarticular). Approximately 80% of people with RA are rheumatoid factor positive.¹⁷⁷

Rheumatoid factors are autoantibodies that react with immunoglobulin antibodies found in the blood. Rheumatoid factor has also been found in the synovial fluid and synovial membranes of those with the disease. It is hypothesized that the interaction between rheumatoid factor and the immunoglobulin triggers events that initiate an inflammatory reaction.

RA begins attacking the joint in the synovium. The normal synovial membrane consists of loose connective tissue that contains blood vessels and is covered by a layer of synovial lining consisting of macrophages and synoviocytes and is only minimally infiltrated by lymphocytes. In RA, the cells of the synovial lining multiply, there is an influx of leukocytes from the peripheral circulation, and the synovium becomes edematous. The synovial lining thickens, resulting in the clinical synovitis seen so often.

These changes can result in the development of thickened synovium, a destructive vascular granulation tissue called *pannus*. The inflammatory cells found within the pannus are destructive, preventing the synovium from performing its two primary functions: lubricating the joint and providing nutrients to the avascular articular cartilage. As this tissue proliferates, encroaching on the joint space at the margins where the hyaline cartilage and synovial lining do not adequately cover the bone, it dissolves collagen, cartilage, subchondral bone, and other periarticular tissues in its path (Fig. 27-10).

Although the cause of RA remains unknown, recent advances in molecular techniques have allowed for identification of distinct cell subtypes, surface markers, and products that may initiate and propagate the inflammatory and destructive components of the disease. Two cytokines, TNF- α and IL-1, appear to play a major role in the pathophysiological process of RA.⁶⁶

The dominant cytokine, TNF, stimulates production of other cytokines from the interleukin family, thus prompting a massive inflammatory response. As the attracted

Box 27-9**ARTICULAR AND EXTRAARTICULAR MANIFESTATIONS OF RHEUMATOID ARTHRITIS****Cardiac**

- Conduction defects (usually asymptomatic)
- Pericarditis
- Interstitial myocarditis
- Coronary arteritis
- Vasculitis
- Aortitis

Hematologic

- Anemia of chronic disease
- Felty's syndrome (splenomegaly, neutropenia)
- Lymphoma, leukemia

Musculoskeletal

- Osteopenia, osteoporosis (associated fractures)
- Joint pain (reflects severity of synovitis; may not be present at rest)
- Joint stiffness (present in most cases, especially after inactivity; duration reflects degree of synovial inflammation; improves with physical activity)
- Joint contracture (extension of involved joints most commonly affected)
- Swelling (synovial tissue)
- Muscle atrophy (hands, feet; occurs rapidly in severe disease)
- Muscle weakness; often out of proportion to the degree of muscular atrophy
- Tenosynovitis, tendonitis, tendon triggering, tendon rupture
- Joint deformity

Neurologic

- Compression neuropathies; nerve entrapment syndromes (e.g., carpal tunnel syndrome, tarsal tunnel syndrome)
- Polyneuropathy
- Peripheral neuropathy (mononeuritis multiplex, stocking-glove peripheral neuropathy)
- Myelopathy; subluxation or instability of C1-C2
- Lhermitte's sign (upper extremity paresthesias that increase with neck flexion)

Integumentary

- Nodulosis (see Fig. 27-13; subcutaneous nodules, especially over olecranon and proximal ulna, extensor surfaces of fingers, Achilles tendon ["pump bumps"])

- Nodules can occur in tendon, bone, sclerae, over pinna or ear, and in visceral organs, especially lung
- Palmar erythema (identical to changes found in liver disease and pregnancy; persists even in remission)
- Sweet's syndrome
- Vasculitis

Ocular

- Episcleritis (inflammation of the superficial sclera and conjunctiva)
- Scleritis (inflammation of the sclera)
- Sicca syndrome (dry eyes)

Psychologic

- Depression (common); other mood disorders

Pulmonary

- Effusions
- Interstitial pneumonia
- Interstitial fibrosis
- Nodules (rheumatoid nodulosis)
- Pleurisy, pleuritis
- Empyema
- Pulmonary hypertension

Renal

- Interstitial nephritis, nephritic syndrome
- Vasculitis

Vascular

- Skin changes (rash, ulcers, purpura, bullae)
- Infarctions (brain, viscera, nail folds; see Fig. 27-14)
- Digital gangrene
- Medium-vessel arteritis
- Small-vessel vasculitis

Other

- Unexplained weight loss, anorexia
- Malaise, fatigue
- Lymphadenopathy (lymph node enlargement; more common in men)
- Colon cancer

leukocytes, monocytes, and lymphocytes phagocytose the immune complexes. TNF stimulates the secretion of matrix metalloproteinases (protein-degrading enzymes that lyse the cartilage and destroy the joint), leading to articular cartilage destruction and synovial hyperplasia with local tenderness, swelling, and intense joint pain. TNF also inhibits bone formation and induces bone resorption.

Fortunately, a new member of the tumor necrosis family has been identified: osteoprotegerin (OPG). This protein plays a key role in the physiologic regulation of osteoclastic bone resorption, counteracting the destruction and bone degradation caused by the cytokine-induced inflammatory process. OPG might represent an effective therapeutic option for diseases such as RA associated with excessive osteoclastic activity.

The result of the synovial changes that occur in RA can be irreversible joint instability, joint deformity, or ankylosis (adhesions and fibrous or bony fusion of the joint). Joint destruction eventually leads to laxity of the tendons and ligaments, which contributes to the altered biomechanics and deformities frequently observed.¹¹⁰ The wide range of extraarticular problems is also probably a result of local inflammatory injury induced by the immune complexes traveling through the circulatory system.

Clinical Manifestations. RA is a systemic disease typically manifested by articular and extraarticular complaints (Box 27-9; see also the section on Rheumatoid Arthritis in Chapter 5). The symptoms usually begin insidiously and progress slowly as the disease process moves from cartilage degradation to ligamentous laxity and, finally, synovial expansion with erosion. Complaints of fatigue,

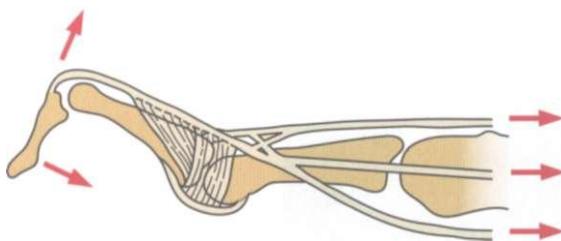


Figure 27-11

Swan-neck deformity. [From Jacobs JL: Hand and wrist. In Richardson JK, Iglesias ZA, eds: *Clinical orthopaedic physical therapy*, Philadelphia, 1994, Saunders, p 309.]

weight loss, weakness, and general, diffuse musculoskeletal pain are often the initial presentation. Deconditioning and depression are common complications of this disease.

The course of RA can vary considerably from mild to severely disabling and is difficult to predict, but it appears that adults with RA today have less severe symptoms and less functional disability than even a decade ago. This positive trend and more favorable course of disease may be attributed to earlier diagnosis with a shorter duration of symptoms at the time of diagnosis and more aggressive use of drug therapy.⁵³⁷

Joint. The musculoskeletal symptoms gradually localize to specific joints. Multiple joints are usually involved, with symmetric, bilateral presentation. The most frequently involved joints are the wrist, knee, and joints of the fingers, hands, and feet, although RA can affect any joint, including the temporomandibular joints. The metacarpophalangeal and proximal interphalangeal joints of the hand are involved early. The involved joints can be edematous, warm, painful, and stiff. After periods of rest (e.g., prolonged sitting, sleeping) intense joint pain and stiffness may last 30 minutes to several hours as activity is initiated.

As the disease progresses, joint deformity can occur, including subluxation. Deformities in the fingers are common, including ulnar deviation, swan-neck deformity, and boutonniere deformity. The ulnar deviation occurs as the extensor tendons slip to the ulnar aspect of the metacarpal head. Hyperextension of the proximal interphalangeal joint and partial flexion of the distal interphalangeal joint make up the swan-neck deformity (Fig. 27-11). The boutonniere deformity is marked by flexion of the proximal interphalangeal joint and hyperextension of the distal interphalangeal joint (Fig. 27-12).

Soft Tissue. Soft tissue manifestations of RA can include synovitis, bursitis, tendinitis, fasciitis, neuritis, and vasculitis. These problems are often overlooked but can be very debilitating. Soft tissue imbalance combined with joint involvement can result in significant deformity, especially in the hands and feet.

Spine. Early involvement of the spinal column is common and typically limited to the cervical spine, with deep, aching neck pain radiating into the occipital, retro-orbital, or temporal areas reported in 40% to 88% of persons.^{44,248,420} Neck movement precipitates or aggravates neck pain; facial and ear pain and occipital head-

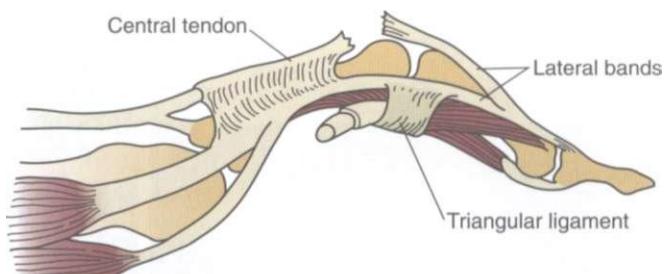


Figure 27-12

Boutonniere deformity. [From Jacobs JL: Hand and wrist. In Richardson JK, Iglesias ZA, eds: *Clinical orthopaedic physical therapy*, Philadelphia, 1994, Saunders, p 664.]

aches occur frequently with active disease from irritation of the C2 nerve root supply to the spinal trigeminal tract, greater auricular nerve, or greater occipital nerve.⁴²⁴

There is a potential for atlantoaxial subluxation (usually anterior) and brainstem or spinal cord compression. The upper cervical spine is affected most commonly because the occiput-C1 and the C1-C2 articulations are purely synovial and are thus primary targets for rheumatoid involvement. In addition, because the C1 and C2 facets are oriented in the axial plane, there is no bony interlocking to prevent subluxation in the face of ligamentous destruction.⁴²⁰

The natural history of cervical instability in people with RA is variable, and only some develop neurologic deficits.¹⁰⁸ Symptoms of C1-C2 subluxation include a sensation of the head falling forward with neck flexion, loss of consciousness or syncope, dysphagia, vertigo, seizures, hemiplegia, dysarthria, nystagmus, and peripheral paresthesias and loss of dexterity of the hands.⁴⁹ Urinary retention and later incontinence are symptoms of more severe involvement. Sleep apnea may be caused by brainstem compression associated with atlantoaxial impaction.³⁹⁶

There may be a positive Lhermitte sign with shocklike sensations of the torso or extremities with neck flexion. Atlantoaxial instability may result in vertebrobasilar insufficiency with visual disturbances, loss of equilibrium, vertigo, tinnitus, and dysphagia. These symptoms can also be caused by mechanical compression of the cervicomedullary junction or brainstem.⁴²⁰ Asymmetrical destruction of the lateral atlantoaxial joints may result in a clinical presentation of head tilt down and to one side. When the neck is flexed, the spinous process of the axis may be prominent.

Pain associated with RA in the subaxial segments of the cervical spine is located in the lateral aspects of the neck and clavicles (C3-C4) and over the shoulders (C5-C6). Neurologic symptoms include burning paresthesias and numbness, which may be attributed to carpal tunnel syndrome, delaying the diagnosis of cervical myelopathy.

Cutaneous. The visible rheumatoid nodule is a characteristic skin finding in RA, occurring in approximately 25% of all cases. These granulomatous lesions usually occur in areas of repeated mechanical pressure, such as over the extensor surface of the elbow, Achilles tendon,

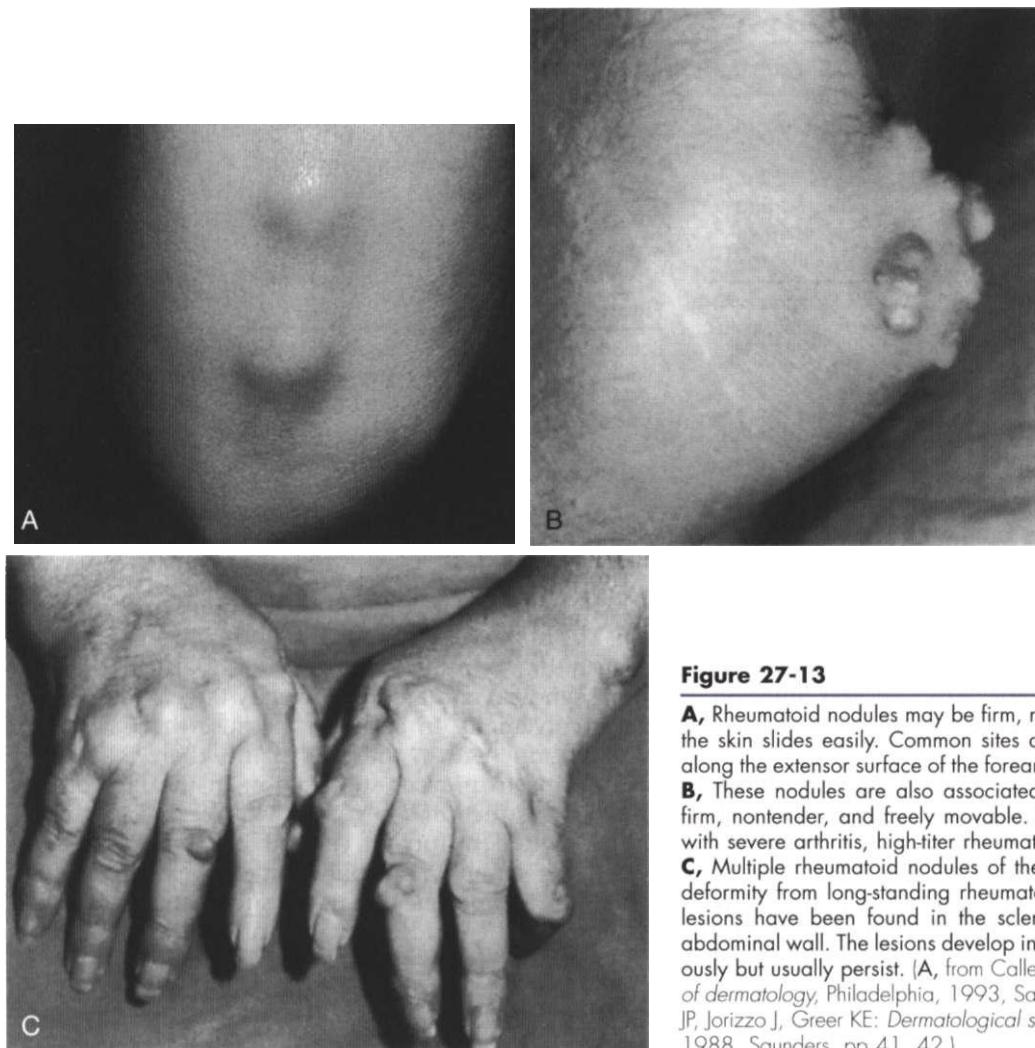


Figure 27-13

A, Rheumatoid nodules may be firm, raised, nontender bumps over which the skin slides easily. Common sites are in the olecranon bursa (elbow), along the extensor surface of the forearm, and behind the heel (calcaneus). **B**, These nodules are also associated with rheumatoid arthritis and are firm, nontender, and freely movable. These are most common in people with severe arthritis, high-titer rheumatoid factor, or rheumatoid vasculitis. **C**, Multiple rheumatoid nodules of the digits with typical ulnar deviation deformity from long-standing rheumatoid arthritis. Histologically identical lesions have been found in the sclera (eye), larynx, heart, lungs, and abdominal wall. The lesions develop insidiously and may regress spontaneously but usually persist. (A, from Callen JP, Greer KE, Hood AF: *Color atlas of dermatology*, Philadelphia, 1993, Saunders, p 130. B and C, from Callen JP, Jorizzo J, Greer KE: *Dermatological signs of internal disease*, Philadelphia, 1988, Saunders, pp 41, 42.)

and extensor surface of the fingers (Fig. 27-13). Nodules are usually asymptomatic, but they can become tender or cause skin breakdown and become infected. Nodules that cannot be seen visibly can also occur in the heart, lungs, and gastrointestinal tract, causing serious problems such as heart arrhythmias and respiratory failure.

Neurologic. One third of adults with RA have cervical spine involvement leading to compressive cervical myelopathy presented as neck pain and stiffness, Lhermitte's sign, weakness of the upper or lower extremities, hyperactive distal tendon reflexes, and presence of the Babinski's sign. In severe cases, urinary and fecal incontinence and paralysis can occur.¹³⁵

Chronic inflammation of the atlantoaxial joint can lead to laxity of the transverse ligament, which normally keeps the dens closely abutted against the anterior arch of the atlas. With loss of integrity of the ligament, the dens moves backward and presses against the spinal cord during forward neck flexion. The individual experiences a shocklike sensation and numbness down the arms with forward flexion of the neck (Lhermitte's sign, previously mentioned). Arthritic changes with erosive involvement of the lower cervical spine facet (zygapophyseal) joints

can also lead to compressive myelopathy or radiculopathy.

Peripheral neuropathies are common as the nerves become compressed by inflamed synovia in tight compartments. Pain, dysesthesias, motor loss, and muscle atrophy can occur, leading to dysfunction and disability. Rheumatoid vasculitis involving medium-sized arteries to the muscles can lead to mononeuritis multiplex, while small-vessel vasculitis causes stocking-glove peripheral neuropathy.¹³⁵

Extraarticular. The extraarticular manifestations are numerous and affect men and women equally (see Box 27-9 and Fig. 27-14). Many of these manifestations impair cardiopulmonary function, restrict activity, decrease endurance, and are disabling; some are life-threatening. They could easily hamper rehabilitation efforts, delaying or preventing progress. See Chapters 12 and 15 for descriptions of the cardiovascular and pulmonary manifestations, respectively.

Sjogren's syndrome (discussed in this chapter) is marked by lymphocytic and plasma cell infiltration of the lacrimal and parotid glands. This can result in diminished salivary and lacrimal secretions. Felty's syndrome is



Figure 27-14

Vasculitis splinter infarction around the finger of a person with systemic vasculitis associated with rheumatoid arthritis (extraarticular manifestation). Clinical features are diverse, because virtually any blood vessel anywhere in the body can be affected. [From Moots RJ, Bacon PA: Extraarticular manifestations of rheumatoid arthritis, *J Musculoskelet Med* 11:10-23, 1994.]

marked by splenomegaly and leukopenia. Mood disorders, especially depression, are common (see the section on Depression in Chapter 3).¹⁰

Individuals with RA are also at increased risk for severe infection, including tuberculosis, requiring hospitalization.^{69,107,139} There is also a greater risk of cardiovascular and cerebrovascular morbidity and mortality among adults with RA compared to adults with OA.

The increased risk of myocardial infarction, congestive heart failure, and cerebrovascular accident is not explained by traditional cardiovascular risk factors, but the mechanism for this association is unknown at this time. Altered immunologic function may possibly explain the increased association, but other factors, such as the new biotherapies for RA (e.g., TNF- α blockers), may be at work as well.^{107,372,544}

MEDICAL MANAGEMENT

PREVENTION. As mentioned in the section on OA, there is a need to implement interventions such as supervised exercise programs, weight loss, and self-education courses such as the Arthritis Self-Help Course,²⁴ which have been shown to reduce pain and physician visits.^{24,291}

DIAGNOSIS. In the early stages of RA the diagnosis can be difficult because of the gradual, subtle onset of the complaints. The symptoms may wax and wane, delaying the visit to a physician's office. Early diagnosis can help prevent or reduce erosive and irreversible joint damage as well as reduce morbidity and mortality associated with this chronic disease. The diagnosis is ultimately based on a combination of history, physical examination, imaging studies, and laboratory tests, with careful exclusion of other disorders.

Table 27-2 lists the diagnostic criteria for RA proposed by the American College of Rheumatology. Although the criteria require that signs and symptoms be present for at least 6 weeks before a definitive diagnosis can be made and this period of time represents a delay in diagnosis, the truth is that many individuals suffer symptoms much longer than this before seeing a physician. At least half of all adults with RA are not referred for rheumatologic consultation until they have had their disease for at least 6 months (sometimes more than 1 year).³⁵⁴

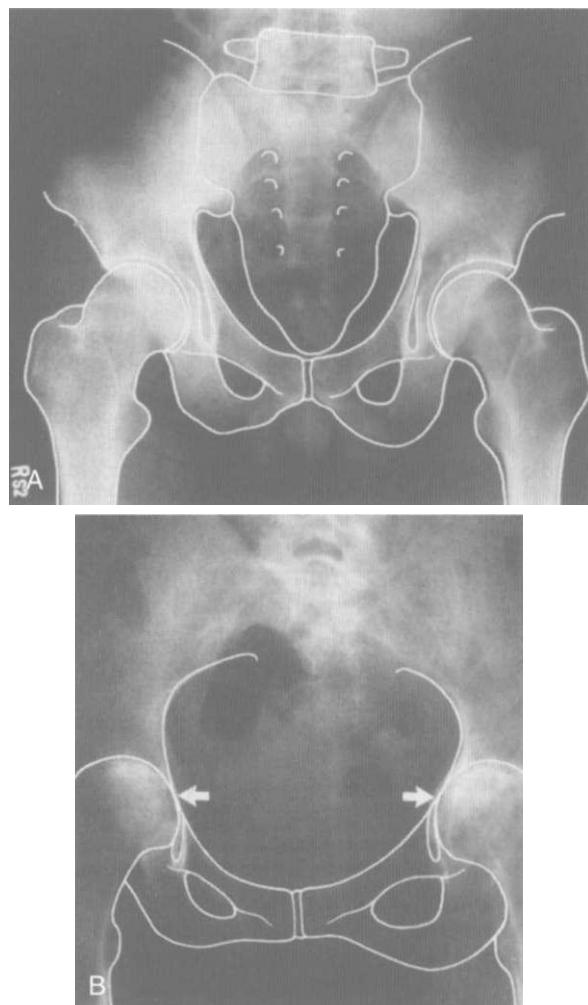


Figure 27-15

A, Radiograph of normal hips and pelvis. **B**, Radiograph of rheumatoid arthritis of the hips. Note the narrowed joint space (loss of articular cartilage) and periarthritis bone density changes. [From McKinnis LN: Fundamentals of radiology for physical therapists. In Richardson JK, Iglarsh ZA, eds: *Clinical orthopaedic physical therapy*, Philadelphia, 1994, Saunders, p 673.]

The presence of serum rheumatoid factor supports the diagnosis but can also be found in healthy persons. Synovial fluid analysis will reveal an elevated white blood cell count and protein content. Also, a decrease in synovial fluid volume and poor viscosity and an increased turbidity may be noted (see Table 27-1).

C-reactive protein, an acute-phase reactant, may be helpful when obtained in a series over time to predict those individuals who are at increased risk for joint deterioration and as a measure of response to treatment. Persistent elevation in C-reactive protein is a predictive factor for cervical spine subluxation.^{134,387}

Conventional radiography, ultrasonography, and MRI studies are allowing more accurate diagnosis of RA. The earliest joint changes (periarticular swelling and cortical thinning with erosion at the margins of the articular cartilage and joint space narrowing) are seen on plain radiographs (Fig. 27-15). Screening cervical spine radiographs should be considered for all individuals with RA but

Table 27-2 Criteria for the Classification of Acute Rheumatoid Arthritis*

Criterion	Definition
1. Morning stiffness	Morning stiffness in and around the joints, lasting at least 1 hr before maximal improvement
2. Arthritis of three or more joint areas	At least three joint areas simultaneously have had soft tissue swelling or fluid (not bony overgrowth alone) observed by a physician; the 14 possible areas are right or left PIP, MCP, wrist, elbow, knee, ankle, and MTP joints
3. Arthritis of hand joints	At least one area swollen (as defined above) in a wrist, MCP, or PIP joint
4. Symmetric arthritis	Simultaneous involvement of the same joint areas (as defined in criterion 2) on both sides of the body (bilateral involvement of PIPs, MCPs, or MTPs is acceptable without absolute symmetry)
5. Rheumatoid nodules	Subcutaneous nodules over bony prominences or extensor surfaces or in juxtaarticular regions, observed by a physician
6. Serum rheumatoid factor	Demonstration of abnormal amounts of serum rheumatoid factor by any method for which the result has been positive in <5% of normal control subjects
7. Radiographic changes	Radiographic changes typical of rheumatoid arthritis on posteroanterior hand and wrist radiographs, which must include erosions or unequivocal bony decalcification localized in or most marked adjacent to the involved joints (osteoarthritis changes alone do not qualify)

Data from Arnett FC: The American Rheumatism Association 1987 revised criteria for the classification of rheumatoid arthritis, *Arthritis Rheum* 31:315-324, 1988.

PIP, Proximal interphalangeal; MCP, metacarpophalangeal; MTP, metatarsophalangeal.

*For classification purposes, a person shall be said to have rheumatoid arthritis if he/she has satisfied at least four of these seven criteria. Criteria 1 through 4 must have been present for at least 6 wk.

especially for those with advanced peripheral joint disease.²⁴⁸

MRI is more sensitive than conventional radiography for detecting early RA and can show lesions of the synovium and cartilage, and joint effusions.¹¹⁰ MRI has the ability to visualize synovitis and detect bone edema, which is emerging as a predictor of future erosive bone changes.⁵²

Ultrasonography now allows the visualization of small superficial structures and can reveal synovial inflammation and tenosynovitis as well as effusions and bone erosions.

TREATMENT. Early treatment of RA is critical to improving long-term outcomes, since clinical evidence has clearly shown that joint destruction in RA begins early in the disease.⁴⁰⁸ The treatment goals for someone with RA are to reduce pain, maintain mobility, and minimize stiffness, edema, and joint destruction. Aggressive combination drug therapy (Box 27-10) in conjunction with other management techniques, including physical therapy, is the mainstay of treatment.

The management approach is individualized, especially in the presence of extraarticular manifestation. The physician has a challenging task trying to optimize the pharmacologic management when there is no way to identify which people will need aggressive therapy. The physician must balance the need for conservative care without being too conservative to avoid unchecked inflammation and joint damage, while at the same time avoiding being too aggressive, leading to exposure to potentially toxic medications when less expensive, safer drugs would have been as effective.⁵²

The chronic nature of the disease makes client education and continual adherence to the treatment program vital. Since the inflammatory process results in progres-

sive joint destruction, controlling inflammation is a primary goal. Medications, rest, ambulatory assistive devices, orthoses, and ice can be used during the acute phase.

Pharmacotherapy. Many medications are available now to help in the management of RA (see Box 27-10). Analgesics are used to help relieve pain. NSAIDs reduce pain, swelling, and inflammation. Corticosteroids help reduce inflammation and pain and can slow joint damage. Biotechnology has made new pharmacologic agents possible with genetically engineered products that can relieve symptoms and slow the progression of this disease. These new agents can "reset the inflammatory thermostat" and avoid joint damage.³⁸⁶

Disease-modifying antirheumatic drugs (DMARDs) and biologic response modifiers (BRMs) are two examples of newer types of drugs used in combination with analgesics, antiinflammatories, and steroids to alter the course and clinical presentation of RA. Some DMARDs block the activity of a protein (TNF) that triggers and prolongs the inflammatory process, leading to joint destruction. Others block IL-1, a protein present in excess in people with RA, thus inhibiting inflammation and cartilage damage.

DMARDs, often used in combination, are started as early as possible to reduce or prevent joint damage. These drugs take weeks to months to begin working and must be monitored carefully for adverse side effects. DMARDs are often given along with a steroid, which quiets inflammation and improves symptoms while the individual is waiting for the DMARD to take effect. Then the steroid is withdrawn slowly.

Immunosuppressants, such as methotrexate (MTX; Rheumatrex), azathioprine (Imuran), and cyclophosphamide (Cytoxan), may be used. MTX is currently the most widely used immunosuppressant for RA management

Box 27-10**PHARMACOTHERAPY FOR RHEUMATOID ARTHRITIS****Analgesics**

- Various over-the-counter and prescription drugs, including acetaminophen (Tylenol), tramadol (Ultram), and codeine

Nonsteroidal Antiinflammatory Drugs (NSAIDs)

- Over-the-counter and prescription formulas, including aspirin, ibuprofen (Advil, Motrin), naproxen (Aleve, Anaprox, Naprelan, Naprosyn), ketoprofen (Orudis, Oruvail), diclofenac (Voltaren), diflunisal (Dolobid), indomethacin (Indocin)

Corticosteroids

- Oral or injection formulas, including prednisone (Cortan, Deltasone, Meticorten), methylprednisolone (Medrol)

Disease-Modifying Antirheumatic Drugs (DMARDs)

- Antimalarials (hydroxychloroquine [Plaquenil])
- Antibiotics (sulfasalazine [Azulfidine], minocycline [Minocin, Dynacin])
- Injectable and oral gold (Ridaura)
- D-penicillamine (Depen, Cuprimine)
- Methotrexate (Amethopterin, Rheumatrex)
- Immunosuppressants (azathioprine [Imuran], cyclophosphamide [Cytoxan], cyclosporine [Neoral, Sandimmune], Leflunomide [Arava])

Biologic Response Modifiers (BRMs)**Cytokine Inhibitors ***

- Tumor necrosis factor (TNF) inhibitors† (etanercept [Enbrel], infliximab [Remicade], adalimumab [Humira])
- Interleukin-1 inhibitor (anakinra [Kineret])

Lymphocyte Inhibitors

- Rituximab (Rituxan) (antibody originally developed for the treatment of B-cell lymphoma)
- Abatacept (Orencia) (interrupts the activation of T cells, leading to T-cell anergy and apoptosis)

Investigational Drugs (in Clinical Trials)

- HuMax-CD20 (antibody that targets B cells)
- Belimumab (LymphoStat-B; inhibits B-cell growth and survival)
- Atacicept (inhibits B-cell growth and survival)
- Tocilizumab (anti-interleukin-6 receptor monoclonal antibody)
- Certolizumab (human monoclonal antibody to TNF- α)
- Golimumab (human monoclonal antibody to TNF- α)

Data from Yazici Y: Bright future for RA therapies, *J Musculoskelet Med Suppl*:32-35, November 2006.

*PEGylation is a new way to deliver anti-TNF- α agents that is site specific. Polyethylene glycol is added to enhance the pharmacokinetic properties of a molecule, decreasing its volume of distribution and clearance and increasing its half-life.

†May also be referred to as TNF- α antagonists.

because of its long-term efficacy. Although its exact mechanism remains unknown, it has been shown to alleviate pain and morning stiffness. Effects tend to plateau after 6 months, and side effects can be numerous; regular serum monitoring of liver and renal function is required.

NSAIDs are effective for pain and swelling associated with inflamed joints caused by RA, but these pharmaco-

logic agents do not affect disease progression. Corticosteroids may be prescribed in addition to DMARDs and NSAIDs to relieve pain and in clients with unremitting disease with extraarticular manifestations. Intraarticular injections can provide relief of acute inflammation. Administration of these drugs for periods of 3 to 6 months is often necessary for benefit to be noted.

The development of cytokine inhibitors has had an important role in treating individuals with RA. Targeting TNF- α , an important proinflammatory cytokine present in the rheumatoid synovium, has been a great help in treating this disease. IL-1 has several actions that overlap those of TNF- α ; TNF- α appears to be more important in early inflammation, while IL-1 may be more important in erosive arthritis. Cytokine targets such as IL-6, IL-12, and IL-18 remain under investigation.²⁵⁴

BRMs were developed to target the interaction sites in the pathologic pathway blocking the action of TNF- α , which initiates the inflammatory response, thereby suppressing inflammation more effectively. Etanercept (Enbrel) is a genetically engineered (recombinant) version of a receptor for TNF that helps bind and inactivate excess TNF, thereby reducing the inflammatory response (cytokine inhibitor). When used in combination with MTX, results are superior to those of MTX alone.^{355,356}

In fact, some people treated early with MTX and infliximab (Remicade) are experiencing drug-free remission. Two landmark studies have been reported: the first study (called PROMPT) and the second study (the BeSt study) found less radiographic evidence of joint damage, indicating an ability in the first group to prevent the disease from progressing to full-blown RA and in the second group, the ability to stop taking infliximab without relapsing and then taper off the use of MTX.¹⁴

Other drugs under investigation block protein signals that cause inflammation. Reducing the number of these proteins or blocking the receptors that receive their signals might help control RA. Researchers are investigating a number of targets, including IL-6. One investigational drug, tocilizumab, attaches to the receptors that accept signals from IL-6, so the messages cannot get through. Early studies show promise, but side effects such as elevated cholesterol levels and increased risk of infection have been observed.

Drugs that stop B cells from causing inflammation, such as rituximab, intercept B cells and stop them from completing their tasks. Several other approaches to stopping B cells are under investigation. One investigational drug, belimumab (LymphoStat-B), blocks signals that drive B cells. Early studies have had mixed results.³⁴⁸

The additional use of bone-active agents can reduce the rate of bone loss (e.g., OPG, a regulator of osteoclast formation, may prevent bone erosion without inhibiting the inflammatory process necessary in other parts of the body).¹⁵⁷ Minocycline, an antibiotic, is another effective DMARD that can be used in people with early seropositive RA, making it possible to reduce the amount of corticosteroids used.³⁷⁸

Surgery. Surgery may be indicated if conservative care is insufficient in achieving acceptable pain control and level of function. Synovectomy to reduce pain and joint damage is the primary operation for the wrist. Total joint

replacement procedures are performed at the shoulder, hip, knee, wrist, and fingers. The most common soft tissue procedure is tenosynovectomy of the hand. Studies support prophylactic stabilization of the rheumatoid cervical spine to prevent paralysis in high-risk individuals.^{84,360}

Complementary and Alternative Medicine/Complementary and Integrative Medicine (CAM/CIM). Some complementary approaches have been advocated by some in the treatment of RA, including fish and plant oils; vitamin, mineral, and other supplements (e.g., S-adenosylmethionine, or SAMe). Safety and effectiveness are not proven yet in long-term studies, but complementary approaches show promise.

PROGNOSIS. There is no known cure for RA at this time, and joint changes are usually irreversible. Restrictions in the ability to perform specific actions and difficulty in performing ADLs can result in functional limitations and disability. It is now established that the longer a person has RA, the greater the likelihood of having cervical spine disease.³⁷⁷

More specific predictors of neurologic recovery from brainstem or spinal cord compression include location of the disease (basilar invagination has a poorer prognosis than isolated atlantoaxial or subaxial instability), degree of preoperative neurologic deficit, and spinal canal diameter.⁴²⁰

Knowledge of the natural history of RA affecting the cervical spine is limited. Studies are small in size and limited in scope. Transition from reducible subluxation to irreducible subluxation often accompanies atlantoaxial impaction an average of 6 years after atlantoaxial subluxation. Up to 80% of individuals with rheumatoid subluxations demonstrate radiologic progression but may not experience corresponding clinical symptoms.²⁴⁸

There is a high rate of sudden death linked with untreated myelopathy. The presence of myelopathy increases the risk of mortality dramatically; without surgery, most people die within 1 year. Even with surgery to stabilize the spine, death from cord compression does occur.³⁶⁸

Quality-of-life issues are central to this disease when people who expected to be active and productive are severely incapacitated in early adulthood. The natural history of RA varies considerably, but people who present at an early stage and receive early intervention continue to do well years later, with reduced joint pain and inflammation and preservation of function.⁴⁸³

Mortality in adults with extraarticular manifestations of RA is significantly greater than in those whose disease is limited to the joints; in many people, the extraarticular manifestations are more debilitating than the arthritis itself.¹³⁵ Death from complications associated with RA and its treatment can occur. These complications include subluxation of the upper cervical spine; infections; gastrointestinal hemorrhage and perforation; and renal, heart, and lung disease. The same factors that contribute to joint inflammation also accelerate atherosclerosis and heart disease; early death resulting from coronary artery disease will be the focus of future treatment efforts.⁹⁸

As the complex pathogenesis is better understood, new drugs that can interrupt tissue and joint destruction without interfering with host defense mechanisms or causing other adverse effects may be developed to stop the progression of this disease.

SPECIAL IMPLICATIONS FOR THE THERAPIST 27-9

Rheumatoid Arthritis

PREFERRED PRACTICE PATTERNS

4A: Primary Prevention/Risk Reduction for Skeletal Demineralization (osteopenia, osteoporosis)

4B: Impaired Posture (cervical involvement)

4C: Impaired Muscle Performance

4D: Impaired Joint Mobility, Motor Function, Muscle Performance, and Range of Motion Associated with Connective Tissue Dysfunction

4H: Impaired Joint Mobility, Motor Function, Muscle Performance, and Range of Motion Associated with Joint Arthroplasty

4I: Impaired Joint Mobility, Motor Function, Muscle Performance, and Range of Motion Associated with Bony or Soft Tissue Surgery (tenosynovectomy, tendon reconstruction)

5H: Impaired Motor Function, Peripheral Nerve Integrity, and Sensory Integrity Associated with Nonprogressive Disorders of the Spinal Cord (cervical spine)

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

Medical Screening of Joint Pain

Therapists see many people with joint pain. Most of these people have joint pain secondary to degenerative OA as opposed to rheumatic joint disease, but the therapist must be aware of the symptoms and signs associated with RA. The therapist can ask a series of questions to help identify the cause of joint pain.¹⁵⁶ An abbreviated version of this list can include the following:

- Are you stiff in the morning? If yes, how long does this last?
- Does your stiffness increase after sitting?
- Where do you think the pain is coming from?
- If joints, which ones are involved?
- Do you ever notice any joint swelling or other changes?
- Does anyone else in your family have RA or other kinds of arthritis?
- Does aspirin or ibuprofen help you feel better?

Being aware of the clinical signs and symptoms of RA will help the therapist make an early referral. The distribution of joint involvement is an important clue. RA usually affects the small joints of the feet and hands symmetrically; generalized pain ("I hurt all over") is not characteristic of RA.

Quick, aggressive medical treatment is necessary to minimize joint destruction. Unexplained joint pain for 1 month or more, especially accompanied by systemic complaints, skin rash, or extensor nodules, no

matter how mild, should raise concern on the therapist's part. Cervical pain with reports of urinary retention or incontinence warrants immediate medical evaluation. Also, insidious onset of polyarthritis or joint pain within 6 weeks of taking a medication should raise suspicion regarding the nature of the pain complaints. Any of these red flags suggests the need for a medical referral if a physician has not recently evaluated the affected individual.

Anyone with known RA should be instructed in recognizing the signs and symptoms of progression of disease (e.g., increased duration of morning stiffness, increased number of tender and painful joints, increased intensity of joint pain, increased fatigue, increased gait disturbances, worsening of visible deformities) so that the intervention plan can be modified to meet the individual's needs.

When distinguishing articular pain from periarticular involvement, remember that true arthritis produces pain and limitation during both active and passive range of motion, while limitation from tendinitis is much worse during active than during passive motion. Inflammatory joint involvement typically produces warmth, erythema, and tenderness. Frequently, there is bogginess related to underlying synovitis or effusion. These indicators are not present with joint pain of a mechanical cause.

Patient/Client Education

Helping individuals affected by RA understand the disease, disease process, treatment, possible outcomes, and role of exercise and self-care is a major part of the therapist's task. Self-management includes learning pacing, joint protection, and energy conservation; monitoring symptoms; and maintaining or progressing an exercise program.

Each person must be taught ways to minimize trauma to inflamed joints by unloading joints and reducing mechanical joint stresses. This can be done through modification of activities such as using assistive devices to open doors and jars, and avoiding excessive weight bearing on inflamed joints by reducing movements such as bending and stooping. Energy conservation (see Box 9-8) should become a way of life for anyone with acute or subacute disease. The systemic nature of this disease produces global fatigue; the demand for energy to move joints may increase if biomechanics are altered.²²³

The need for frequent rest breaks, change in level of activity, and change in positions throughout the day should be taught and their use encouraged, but they should be balanced by the need to avoid muscle wasting and weakness from immobilization. Range of motion, stretching, and isometric exercises must be taught, monitored, and reinforced for as long as the therapist follows the individual, always teaching the client how to modify the program during periods of active inflammation.

Cervical Spine Involvement

Cervical collars may be used for comfort but do not protect against progressive subluxation or neurologic

Box 27-11

ACR CRITERIA FOR CLASSIFICATION OF FUNCTIONAL STATUS IN RHEUMATOID ARTHRITIS

Class 1:	Completely able to perform usual activities of daily living (self-care, vocational, avocational)
Class 2:	Able to perform usual self-care and vocational activities, but limited in avocational activities
Class 3:	Able to perform usual self-care activities, but limited in vocational and avocational activities
Class 4:	Limited in ability to perform usual self-care, vocational, and avocational activities

ACR, American College of Rheumatology.

compromise. Rigid cervical collars can partially limit anterior atlantoaxial subluxation, but they also prevent reduction of the deformity in extension.^{238,239} Rigid orthoses are also poorly tolerated in these individuals because of skin sensitivity and temporomandibular joint involvement. Anyone with cervical spine involvement should be taught to avoid cervical flexion. The therapist can focus on isometric strengthening of neck muscles and overall postural training.

In the case of conservative intervention with non-surgical management, the therapist must observe for (and teach the client to observe for) gradual deterioration in function that may indicate the development of subtle myelopathy (see Clinical Manifestations in this section). Radiographic evaluation is important in observing for impending neurologic compromise.⁴²⁰

Rehabilitation

RA is a chronic, progressive disease requiring an interdisciplinary team approach that is individual to the client and comprehensive, with long-range planning that extends beyond the initial acute phase. The role of the physical therapist as an integral part of the management of RA has been well established,^{59,287,370} with a renewed focus on outcome-based intervention.

The American College of Rheumatology has developed criteria for classification of functional status for individuals with RA that may help guide the therapist in designing and monitoring the results of an appropriate plan of care (Box 27-11).

The Ottawa Panel has identified nine goals in the rehabilitation of individuals with RA, including decreasing pain, effusion (swelling), and stiffness; correcting or preventing joint deformity; increasing motion and muscle force (decreasing weakness); improving mobility and walking; increasing physical fitness; reducing fatigue; and increasing functional status.³⁸³

Before initiating any rehabilitation program for this group of individuals, a thorough limb and joint examination must be done to provide an objective way to assess and document disease activity and progression or, conversely, remission and improved function. Numerous resources are available to assist the clinician in carrying out a thorough clinical assessment of the individual with RA, including a helpful joint-by-joint

Continued.

guide (describing where and how to palpate).^{347,431,459,460} Physical therapy examination should also include observation of functional performance, limitations, impairments, and a systems review as outlined in the *Guide to Physical Therapist Practice*.¹⁷

Complete bed rest is rarely indicated and is saved for those with severe, uncontrolled inflammation. For many people, a rest period of up to 2 hours during the day is important for dealing with general body fatigue and protection of involved joints. Splints can be applied to rest involved joints, prevent excessive movement, and reduce mechanical stresses. Crutches, canes, or walkers can be used to reduce weight-bearing stresses and enhance balance.

Adaptations may be necessary (e.g., platform crutches) because of upper extremity involvement. A home program of self-management will include instruction in proper body mechanics, positioning, joint protection, and energy conservation (see Box 9-8). Adaptive equipment designed to make tasks easier may include large key handle attachments allowing the person to use the whole palm to turn a key, spring-open scissors with big loops for anyone with hand involvement, jar openers and electric can openers, clip-on bottle openers, and ergonomic kitchen utensils with large handles and ergonomically angled handles.³¹⁸

Instability at any joint, particularly at the atlantoaxial segment, requires caution on the therapist's part. Such a joint may present with a marked reduction in range of motion, such as the shoulder or neck feeling stuck or caught with a certain movement. A history of periods of significant loss of range of motion alternating with full range of motion suggests joint hypermobility. Restoration of mobility is an important goal, but choosing techniques that are gentle or applying traction while stretching is necessary.

The extraarticular problems may affect the rehabilitation program. For example, if fatigue is present, the therapist may have to allow periods of rest during the treatment session. During periods of symptom exacerbation there is a fine line between overextending the client and maximizing activity. There are times when active exercise may have to be curtailed, but passive stretching remains important to prevent contractures.

Splenomegaly may account for tenderness on palpation and fullness or increased resistance of the left upper abdominal quadrant. Deep soft tissue techniques are contraindicated in this area. Percussion techniques may help the therapist delineate the caudal boundaries of the spleen.

Remission

Whether or not remission from RA was possible was a point of discussion and debate in 1981 when preliminary criteria for remission were first proposed.⁴⁰³ These criteria have been modified over the years to reflect current treatment, trends, and outcomes. For example, the complete absence of tender or swollen joints is the most important sign of remission.³⁸⁰

Changes must last for more than 2 months, but a definitive time line has not been established. Today

even some people with long-standing disease may achieve remission, although this is not possible for everyone. After remission, researchers hope to be able to effect a curative approach.

Postoperative Care

Surgical treatment of RA is often complicated by the client's generalized debilitated condition. People with RA tend to have poor skin condition, poorly healing wounds, and osteopenic bone. Generalized bone loss occurs early in the course of RA and correlates with disease activity. This condition is further affected by the use of corticosteroids.

Poor nutritional status has been associated with higher complication rates following surgery, including infection. Following Standard Precautions with adequate handwashing is important. Likewise, promoting respiration with good breathing techniques is an important component of the therapist's postoperative intervention.

Anyone with RA should be taught early on that, if surgery is ever indicated, a program of isometric exercises and range of motion before surgery is advised. Review dislocation precautions and restrictions prior to the surgical procedure. After arthroplasty, correction of deformity and relief of pain are typical, but recurrence of deformity can occur even with appropriate rehabilitation. Many clients are still very satisfied with the improved cosmesis, reduced pain, and improved function. Maximum benefit from arthroplasty may not occur for up to 1 year after surgery.⁷⁷

The postoperative rehabilitation regimen must be tailored to the specific needs of each individual. The surgeon's intraoperative assessment of the quality of tissues, component stability, and any associated repairs is critical to the rehabilitation protocol selected. Specific motion limitations vary depending on intraoperative repairs made, complications, and adverse events (e.g., infection, wound dehiscence, dislocation, implant fracture or failure, nerve damage).

Exercise and Rheumatoid Arthritis

A group of experts from the University of Ottawa reviewed comparative controlled trials and compiled evidence to suggest and support the conclusion that therapeutic exercises, including specific functional strengthening and whole-body functional strengthening, are a beneficial intervention for individuals with RA. The benefit may vary depending on the stage of disease (acute, subacute, inactive) but includes reduced pain, improved overall function, and decreased number of sick leaves.³⁸⁴

There is a need for more research to investigate the impact and effects of exercise on individuals with RA. Whereas rest has often been the treatment of choice, a balance must be attained between rest required during acute flare-ups and activity necessary to prevent the deconditioning effects of prolonged rest, immobilization, and inactivity. Education as to the efficacy of exercise and its proper use in self-management of RA has been shown effective in reducing stiffness and improving function in as little as 4 hours of a

community-based physical therapy intervention delivered over a period of 6 weeks.³⁹

To date, there is no evidence that active exercise beneficially affects the inflammatory processes associated with adult RA, but it has been shown that a short-term intensive exercise program in active RA is more effective in improving muscle strength than a conservative exercise program and does not have deleterious effects on disease activity.⁵¹⁶

Individual studies of long-term intensive exercise have not shown an increase in joint swelling or pain with such a regimen. In fact, those who exercise rigorously at least twice a week for an hour show more improvement in physical abilities such as stair climbing and reduced psychologic distress compared to those who received standard care.¹⁰⁰

The beneficial effect of exercise in lowering cytokine levels and increasing antiinflammatory compounds in plasma has been demonstrated in children with juvenile RA.⁵²² Other effects of exercise on the immune system are discussed in the section on Exercise Immunology in Chapter 7.

GENERAL CONCEPTS

Exercises to prevent contractures, improve strength and flexibility, and enhance cardiorespiratory or aerobic conditioning are important components of the rehabilitation program.^{176,255,370} Joint pain leads to a reflex inhibition of muscle surrounding the joints, causing disuse atrophy of these muscles. Use of corticosteroids may lead to an additional decrease in strength and function.³⁷⁰

The feet are often overlooked in people with RA, but foot involvement occurs frequently, can impair gait, and can prevent safe participation in an exercise program. Foot involvement resembles that of the hand, with one important difference being that alterations in biomechanics may cause excessive stress to proximal lower extremity joints and to the spine areas forced to compensate for the altered gait.

Careful assessment of the feet may reveal uneven or pathologic weight-bearing patterns. Gait analysis and assessment of shoe wear can provide additional significant information regarding altered biomechanics. Providing assistive devices or orthotics before initiating an exercise program may be essential.^{431,459,460}

Avoid overloading and overtraining. For the individual with active (acute) disease, adequate sleep is essential. Encourage 8 to 10 hours of rest each night. Range-of-motion exercises should begin with low repetitions several times throughout the day. Isometric exercises with short holds (4 to 6 seconds) have been suggested, once again using low repetitions (start with one or two and build up gradually to four to six).²²³

Range-of-motion exercises can be increased up to 8 to 10 repetitions in subacute cases, with the addition of dynamic strengthening exercises. Stable, quiescent, or inactive disease makes it possible to add an aerobic component such as walking, aquatics, or biking for at least 15 minutes each day three times a week. Range of motion and strengthening can be continued and monitored.²²³

EXERCISE PRESCRIPTION

A helpful guide in establishing the level of acceptable exercise intensity is as follows: acute pain during exercise indicates a need to modify the program; if joint pain persists for more than 1 hour after exercise is completed, the exercise was probably excessive.¹⁴⁴ Recent research indicates that it is not necessary to work out as vigorously as once thought to derive benefits from exercise.

Engaging in moderate-level exercise for 30 min/day 4 or 5 days/wk appears to substantially increase physical fitness, even for older adults.⁴⁸⁴ Exercise spaced out over the course of the day can help loosen up stiff, achy joints while still providing cardiovascular benefits.^{243,390}

The Arthritis Foundation provides a 24-page booklet (*Exercise and Your Arthritis*) describing the benefits and types of exercises, how to start, and how to keep going. This publication educates people about the possible effects of not exercising (e.g., increased joint stiffness, muscle weakness, muscle atrophy, increased risk of fracture and deformity). The foundation also provides valuable exercise tips for before, during, and after exercise.²³

Currently the National Arthritis Foundation offers the largest standardized exercise program to individuals with arthritis through two community-based programs: an on-land exercise program called People with Arthritis Can Exercise (PACE) and an aquatic exercise program called the Arthritis Foundation Aquatic Program (AFAP).

The PACE program involves 72 exercises to improve motion, strength, endurance, balance, coordination, posture, and body mechanics. Other recommendations and guidelines for including conditioning exercise in a comprehensive management program for RA are available.^{15,26,345}

STRENGTH TRAINING

Dynamic strengthening (gradually increasing resistance) through the full available range of motion helps stabilize joints, reducing erosive wear and tear on the structures. Regular, dynamic strength training combined with endurance-type physical activities improves muscle strength and physical function but not bone mineral density (BMD) in adults with early and long-standing RA, without causing detrimental effects on disease activity.^{168,169}

Low-load resistive muscle training has been shown to increase functional capacity and is a clinically safe form of exercise in mild to moderate RA.²⁵⁵ Other studies report that moderate- or high-intensity strength training programs have better training effects on muscle strength in RA. It is essential to maintain the training routine to obtain long-term benefits.¹⁶⁷

Strengthening in some cases of RA can be difficult, because exercise may lead to increased joint swelling and subsequent joint destruction. Before initiating a strengthening program, the physical therapist must be sure that pain is controlled, range of motion is optimized, and contractures are minimized.³⁷⁰ Paying

Continued.

attention to biomechanics, deformities, and muscle imbalances is important. Exercising misaligned joints without properly distributing the load can place too much pressure on vulnerable joints.⁴⁵²

AEROBIC EXERCISE

Aerobic exercise in this population is necessary to help reduce weight and improve cardiovascular fitness without increasing pain.³³⁰ Aerobic exercises are safe to perform during the subacute and inactive stages of RA. Aerobic capacity can be estimated using a single-stage submaximal treadmill test.³⁴⁴ Training programs begin at 50% (and work towards 80%) of maximal oxygen uptake based on the baseline test results. Without baseline testing, the therapist can rely upon (and teach the client to use) the Borg scale for rate of perceived exertion (RPE; see Table 12-13). Heart rate monitors are also helpful in enabling clients to track their cardiovascular responses.²²³

Screening for unknown coronary artery disease is recommended before initiating resistance or aerobic exercise. RA can also affect the bony structures of the rib cage and cause a decrease in chest expansion. The usual precautions for cardiopulmonary screening still apply based on the individual's age, risk factors, and health history (especially heart health history).

People with RA may have normal pulmonary function tests but reduced respiratory muscle strength and endurance with reduced aerobic capacity compared to adults without RA.⁸² Assessment of respiration and intervention to improve breathing patterns are important components of the rehabilitation program.

AQUATIC THERAPY

Aquatic therapy may be beneficial for conditioning, strengthening, and flexibility while reducing mechanical stress on the joints. Water exercise provides the means by which people with RA can reach needed training levels in a comfortable environment. The AFAP program consists of 72 exercises similar to the PACE program that can be done in water with decreased joint loading, a reduced effect of gravity, increased buoyancy, and increased circulation.

Modalities and Rheumatoid Arthritis

Various modalities provide temporary pain relief and may be used in effectively and safely controlling symptoms of the acute inflammatory phase of RA. Information on the rationale for use and effectiveness of the various physical modalities is available.^{64,335,346}

Although cold may be more suitable in acute inflammation, people with RA usually prefer heat. Superficial heat (e.g., paraffin baths, moist hot packs, hydrotherapy or aquatic therapy) is recommended, whereas prolonged or deep heat is contraindicated, since it may increase intraarticular temperature, leading to increased collagenase activity, possibly contributing to joint destruction.¹⁹⁴

Electrotherapeutic modalities and thermotherapy physical agents are often used as part of a rehabilitation program mainly for pain relief, to control inflammation, and to reduce joint stiffness.

The Ottawa Panel recommends the use of low-level laser therapy, therapeutic ultrasound, thermotherapy, electrical stimulation, and transcutaneous electrical nerve stimulation for the management of RA. This recommendation is based on the analysis of systematic and literature reviews. Specifics of studies reviewed and a summary of the findings have been published in our own journal (*Physical Therapy*).³⁶

The Ottawa Panel reported on available systematic reviews on the efficacy of ultrasound in the management of RA and noted that these are limited and do not offer conclusive evidence-based support for the use of ultrasound alone or combined with exercise.

Medications

Because of the long-term nature of RA, intervention is usually an ongoing process. Aspirin and NSAIDs are potentially ulcerogenic, and prolonged use of corticosteroids can lead to osteoporosis.

The analgesics and slow-acting antirheumatic drugs such as gold and penicillamine can impair renal function. Periodic screening of each of the body systems is imperative when working with this population; anyone taking DMARDs but still having joint pain and swelling should be reevaluated by the rheumatologist.

Numerous other side effects can occur with any of the pharmacologic agents used in the management of RA. The therapist should be aware of the potential side effects with any of the medications each client is taking.

For the individual with Felty's syndrome (RA in combination with low white blood cell count or leukopenia and an enlarged spleen), there is an increased risk of infection, even when treatment with DMARDs raises the white blood count. Careful handwashing and Standard Precautions are always warranted, but especially in the case of this syndrome.

Juvenile Idiopathic Arthritis

Overview and Incidence. Although the term *juvenile idiopathic arthritis* (JIA; formerly juvenile rheumatoid arthritis [JRA]) brings to mind a single disease similar to adult RA, it is actually an umbrella term for a heterogeneous group of arthritides (Box 27-12) of unknown cause that begin before 16 years of age and occur in all races. Each subtype has a different presentation, genetic background, and prognosis.^{48,415}

Many other forms of arthritis (e.g., SLE, dermatomyositis, scleroderma) that affect adults occur less frequently in children. Approximately 30,000 to 50,000 children in the United States are affected by one of the subtypes discussed here (Fig. 27-16). The general classification of JIA is based on the number of involved joints and the presence of systemic signs and symptoms.

Pauciarticular JIA, (PaJIA; also known as oligoarthritis), meaning "few joints," generally affects four or fewer joints, usually in an asymmetric pattern, and most commonly involves the knees, elbows, wrists, and ankles.

Box 27-12**SUBCATEGORIES OF JUVENILE IDIOPATHIC ARTHRITIS (JIA)**

- Pauciarticular JIA (PaJIA; oligoarthritis)
- Polyarthritis JIA (PoJIA RF+)
- Polyarthritis JIA (PoJIA RF-)
- Systemic-onset JIA (SoJIA)
- Psoriatic JIA
- Enthesitis-related arthritis
- Other (undefined)

Data from Petty RE: ILAR classification of JIA: second revision, Edmonton 2001, *J Rheumatol* 31(2):390, February 2004.

RF+, Rheumatoid factor positive; RF-, rheumatoid factor negative.

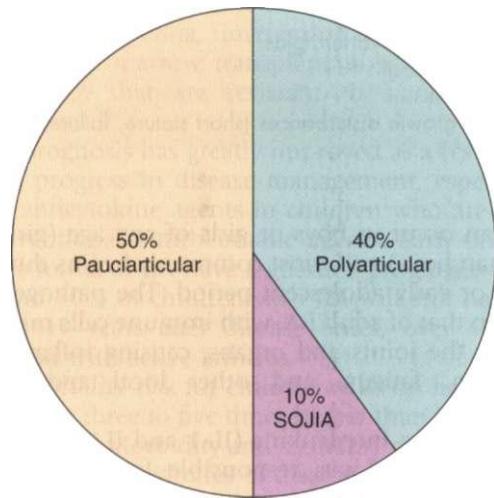


Figure 27-16

Breakdown in types of juvenile idiopathic arthritis (JIA).

Girls are affected more often than boys and usually between the ages of 1 and 5 years. This type of JIA is relatively mild with few extraarticular features. Parents may notice a swollen joint and limp or abnormal gait, usually early after the child wakes up in the morning. Leg length discrepancy is common. Pain is not a central feature at first, and the disease rarely manifests any constitutional symptoms.³⁵¹

PaJIA is the most common type of JIA, comprising one half of all JIA cases, and has three subtypes. The first is characterized by the presence of antinuclear antibodies (ANAs; see discussion in Chapter 40) and a high risk of uveitis, a potentially dangerous inflammation of the eye resulting in irreversible damage and blindness. The second subtype affects the spine as well as other joints, although spinal involvement may not occur until the child reaches late adolescence. Children with this subtype may test positive for the HLA-B27 gene (see Table 40-20), which is common in adults with AS (this subtype is sometimes referred to as juvenile AS). In the third subtype, joint involvement is the extent of the disease. Usually PaJIA runs a benign course; recurrences occur in up to 20% of children, most often during the first year but possibly delayed by as much as 5 years after the initial

diagnosis. There are some children who develop persistent joint disease, referred to as extended oligoarthritis.³⁵¹

Polyarticular JIA (PoJIA) affects five or more joints, most commonly including the large and small joints (wrists, cervical spine, temporomandibular joint, small joints of the hands and feet, as well as the knees, ankles, and hips). Joint involvement is usually symmetric and is most like that of adult RA, with the potential for severe, destructive arthropathy.

PoJIA comprises 40% of all cases of JIA and affects girls more than boys. There are two subtypes depending on whether children are rheumatoid factor positive or negative. The rheumatoid factor-positive subtype is characterized by the presence of rheumatoid factor (a type of autoantibody found in the blood of adults with RA) and the *DR4* genetic type, also common in adults with RA. Subcutaneous nodules, cervical spine fusion, chronic uveitis, and destructive hip disease can occur in this type of PoJIA.³⁵¹

The second subtype is characterized only by joint involvement, usually less severe. Children with this subtype do not test positive for rheumatoid factor.¹⁰⁹ Morning stiffness and fatigue with possible low-grade fever are common clinical manifestations of this type of JIA.

Systemic-onset JIA (SoJIA; also called Still's disease; sometimes also affecting adults, although rare) affects boys and girls equally with involvement of any number of joints. This subtype has the most severe extraarticular manifestations, affecting many body systems, and comprises 10% of all cases of JIA.

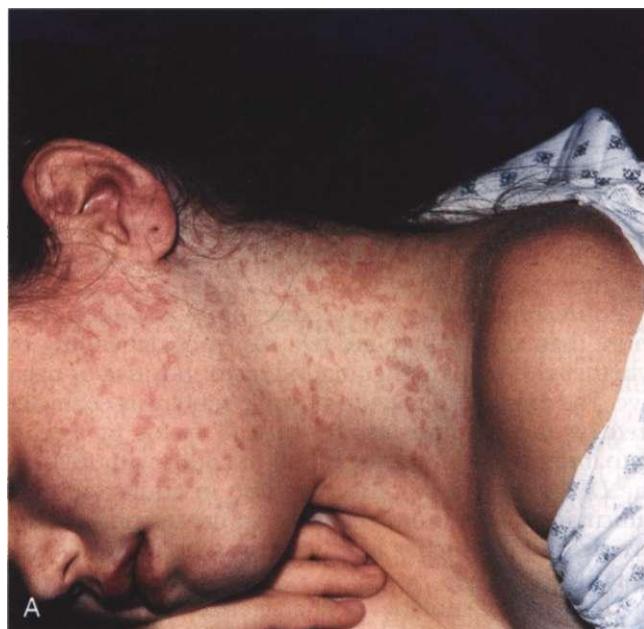
It often begins with a high-spiking fever and chills that appear intermittently for weeks and may be accompanied by a rash on the thighs and chest that often goes away within a few hours (Fig. 27-17). The fever pattern is marked by spikes exceeding 102° F (39° C) and periods between the spikes during which the child feels much better.

Inflammatory arthritis typically develops at some point, and 95% of the children have joint complaints within 1 year of the initial presenting symptoms. Approximately half of the children who have SoJIA recover almost entirely; unfortunately, one third of the children remain ill, with persistent inflammation manifesting as fever, rash, and chronic destructive arthritis.³⁵¹

In addition to inflamed joints, the child may experience enlargement of the spleen (hepatosplenomegaly) and lymph nodes (lymphadenopathy); inflammation of the liver, heart, and surrounding tissues; and anemia.⁴⁴ Box 27-13 lists clinical manifestations associated with Still's disease.

Psoriatic JIA presents with psoriasis, arthritis, and at least two of the following: dactylitis, nail abnormalities, and a family history of psoriasis. Treatment with aggressive immunosuppressives may be required; uveitis is a feature in some cases.

Enthesitis-related arthritis presents as inflammation of the tendon attachments to the bone, especially along the spine and Achilles tendon, along with arthritis and any two of the following: sacroiliac joint tenderness, inflammatory spinal pain, the presence of HLA-B27, positive



A



B

Figure 27-17

A and **B**, Skin rash associated with juvenile idiopathic arthritis. (A, from Paller AS, Mancini AJ: *Hurwitz clinical pediatric dermatology: a textbook of skin disorders of childhood adolescence*, ed 3, Philadelphia, 2006, Saunders. B, from James WD, Berger T, Elston D: *Andrews' diseases of the skin: clinical dermatology*, ed 10, Philadelphia, 2006, Saunders.)

family history, acute uveitis, and pauciarticular or polyarticular arthritis in boys older than 8 years.

Etiologic and Risk Factors and Pathogenesis. The cause is still poorly understood, but JIA may be triggered by environmental factors and infection (viral or bacterial) in children with a genetic predisposition. Genomic studies hope to identify genetic traits that will predict disease risk and other characteristics such as disease course, age of onset, and disease severity. Eventually, researchers may be able to identify molecular biomarkers to help diagnose and treat this group of arthritides.⁴⁰¹

Box 27-13

CLINICAL MANIFESTATIONS ASSOCIATED WITH STILL'S DISEASE

Systemic

- Fever
- Rash
- Lymphadenopathy
- Polyarthritis
- Pericarditis
- Pleuritis
- Peptic ulcer disease
- Hepatitis
- Anemia
- Anorexia
- Weight loss

Musculoskeletal

- Polyarthritis, polyarthralgias
- Myalgia, myositis
- Tenosynovitis
- Skeletal growth disturbances (short stature, failure to thrive)

JIA can occur in boys or girls of any age (girls more often than boys) and most commonly begins during the toddler or early adolescent period. The pathogenesis is similar to that of adult RA, with immune cells mistakenly attacking the joints and organs, causing inflammation, destruction, fatigue, and other local and systemic effects.

TNF and the interleukins (IL-1 and IL-6) seem to be the primary cytokines responsible for many systemic features. These cytokines increase collagenase activity, osteoclast activation, body temperature, and muscle and fat breakdown, as well as acute-phase reactants.

MEDICAL MANAGEMENT

DIAGNOSIS. Early disease recognition is needed to help improve the clinical outcome, but symptoms of rheumatic disease are often mistaken for "growing pains," delaying diagnosis by many months. Diagnosis involves a medical history, physical examination, and laboratory tests, including serum evaluation to measure inflammation and to detect ANAs, rheumatoid factor, or sometimes HLA-B27.

For a diagnosis of JIA, objective arthritis must be seen in one or more joints for at least 6 weeks in children younger than 16 years; it may take up to 6 months to determine which subtype is present. Pain is often dull and aching and less severe but presents in the morning and early during the day rather than the more common presentation of growing pains at night. The systemic features in SoJIA are more readily diagnosed.

TREATMENT AND PROGNOSIS. Some of the immunomodulatory medications used in adult RA can be used in cases of JIA, but none of the current medications used has a curative potential. The goal of treatment is to control pain, preserve joint motion and function, minimize systemic complications, and assist in normal growth and development.

Early aggressive combination medications are replacing the previous gradual add-on approach to treatment (i.e., start with one drug and slowly add another and another to gain the desired effects without too many side effects).⁵³² Medications are administered to control the systemic and articular complaints and, in some cases, halt the progression of the disease.

These agents may include immunosuppressives, DMARDs (e.g., MTX), and biologic agents such as TNF inhibitors (etanercept [Enbrel], infliximab [Remicade]).¹⁶⁶

Adverse effects from taking anti-TNF agents (e.g., neurologic disorders, weight gain, severe infection, hemorrhagic diarrhea) have been reported and should be monitored for carefully. In systemic JIA, approximately 50% respond to anti-TNF agents, but in many children, the response is not sustained.²⁹³ Corticosteroids are indicated if severe anemia, unrelenting fever, or vasculitis is present. Bone marrow transplantation may be used in cases of JIA that are resistant to standard medical management.⁴⁸²

The prognosis has greatly improved as a result of substantial progress in disease management, especially the use of anticytokine agents in children who are resistant to conventional antirheumatic agents. Early-onset, progressive forms of JIA have a guarded prognosis. Between 25% and 70% of children with JIA will still have active arthritis 10 years after disease onset; over 40% enter adulthood with active arthritis.²⁹³

The mortality risk for children with JIA has been estimated to be three to five times higher than in the general population.⁴⁹⁸ Morbidity and mortality may be improved (including increased rates of disease remission) with the changes in treatment approaches, but this has not been documented as yet.²⁹³

Autologous stem cell transplantation (ASCT) is used for some individuals whose disease is refractory to MTX and other DMARDs. Complete remission is possible for up to half of the individuals receiving ASCT, with improvement reported in those individuals who are not resistant. Infection is a common morbidity associated with this treatment, observed in up to 71% of cases, in addition to an associated death rate of 15%.⁹⁷

As the immune mechanisms and inflammatory processes are better understood, new drugs able to inhibit single molecules or pathways will be developed.⁴¹⁵ Investigations continue to study biologic therapies that block IL-1 and IL-6 in systemic JIA.²⁹³

SPECIAL IMPLICATIONS FOR THE THERAPIST 27-10

Juvenile Idiopathic Arthritis

PREFERRED PRACTICE PATTERNS

See also appropriate practice patterns outlined for adult RA.

4A: Primary Prevention/Risk Reduction for Skeletal Demineralization (low bone mineral density)

4E: Impaired Joint Mobility, Motor Function, Muscle Performance, and Range of Motion Associated with Localized Inflammation

5B: Impaired Neuromotor Development

Children with JIA may have no disability, especially if they have the oligoarticular form of the disease. Severe disability is seen most often in cases of rheumatoid factor-positive polyarticular and systemic disease, followed by rheumatoid factor-negative polyarthritis, enthesitis-related arthritis, and psoriatic arthritis. Physical therapy is an important adjunctive therapy for JIA.⁴⁸

Efforts have been made to identify early predictive clinical, laboratory, demographic, genetic, or treatment-related factors for functional disability. It appears that there are complex interactions between disease subtypes but with specific predictors of outcome for each disease subtype. More study is needed to completely define predictive factors.

Physical therapy and occupational therapy are utilized for pain control, facilitation of mobility, and function. Equally important is the role of exercise in improving strength, endurance, and aerobic capacity. In children with JIA, resistive exercise produces a change in the immune response, with significantly lower levels of cytokines and higher levels of antiinflammatory compounds compared with those who did not exercise.⁵²²

With respect to joint impairment, loss of joint motion is the strongest indicator of functional disability in children with systemic JIA, and loss of joint motion has a greater effect on lower limb function than on upper limb function.³⁸ By the time children become adults, only 20% of them have moderate to severe limitations.

The role of the therapist in preventing joint loss should not be underestimated. Cervical spine involvement can occur in PoJIA and systemic JIA, with cervical stiffness as the most common finding. Neck pain is uncommon, and neurologic complications are less likely to develop in JIA than in adult RA.

The therapist should keep in mind the significant impact JIA has on the children and their families. The cost of this disease can be staggering, with pharmacotherapy, hospitalizations, medical visits, and other professional services, including physical and occupational therapy.

Psychosocial-spiritual and quality-of-life issues should also be addressed. Appearance and body image are affected directly by JIA (e.g., generalized growth failure, local growth anomalies such as micrognathia), side effects of drug therapy, surgical scars, and severity of pain and fatigue. The therapist may be the first one to recognize overall problems with adjustment reflected by anxiety, depression, and/or social withdrawal.⁴⁸

For those children with JIA who reach adulthood, significant problems can result from arthritis and uveitis, medication morbidity, and lifelong disability.³⁴¹ Young women may face the issue of pregnancy and childbirth with fears of transmitting JIA to the offspring, and possible reduced ability to conceive along with increased rates of miscarriages have been reported by some.³⁸¹

If the therapist is seeing a young child with joint pain who has not been diagnosed, sensitivity to soft

Continued.

tissue manipulation despite improved or restored joint range of motion warrants further medical investigation. For children with a known diagnosis, the use of physical modalities may be appropriate but must be used with caution as children are not always able to perceive and/or report discomfort.

Exercise and Juvenile Idiopathic Arthritis

Very few studies have been done to determine the safety and efficacy of exercise for children with JIA. Studies that have been done have a small sample size, and results cannot be generalized to all children with this condition. For example, the results of at least one study of nine children suggest that bicycle and treadmill exercise is safe and feasible for children who do not have severe hip disease.^{46,3}

Aquatic physical therapy is an excellent way to complete exercises while providing joint protection and engaging the child in a fun activity. One study engaged 54 children with JIA ages 5 to 13 in a supervised aquatic training program 1 hr/wk for 20 weeks. Measures of functional ability, health-related quality of life, joint status, and physical fitness showed no improvement, but there were also no signs of worse health status, suggesting that swimming is a safe exercise program.^{48,7} More studies are needed to determine at what frequency, level, intensity, and duration a swimming training program would make a significant difference (improvement) in the areas tested and measured.

Strengthening programs for children with rheumatic disease can be part of the exercise program, even for children under 6 years of age. Twice-daily sessions of 15 to 20 minutes are advised.^{42,1} Research at the University of Buffalo showed significant improvement in function and better strength, endurance, and aerobic capacity. Equally important, pain, disability, and use of medications decreased significantly. The researchers found that the children who participated in the exercise program had significantly lower levels of cytokines (proinflammatory proteins) and higher levels of anti-inflammatory compounds in their plasma than those who did not exercise.^{12,4}

Active exercise is not advised during flare-ups when joints are inflamed, but most children usually self-limit their physical activity according to symptoms. Passive stretching and modified aquatic physical therapy are better choices during exacerbations. Parents and children should be educated on the importance of avoiding forced or deep flexion of inflamed joints. Some activities may need to be modified or avoided.

Children with quiescent JIA can participate in some sports activities. With improved medical therapies, children with JIA are able to lead a more active lifestyle compared with similar children even 10 years ago. The therapist can be helpful in assessing each child for abnormal biomechanics that can place him or her at increased risk for injury or future articular damage. Neuromuscular training may be needed to improve neuromuscular function and biomechanics. Proper technical performance during athletics may allow children with JIA to use joint-loading techniques (e.g.,

during jumping and landing) in a safe and controlled manner.^{35,9}

Low BMD is a common secondary condition associated with JIA. Weight-bearing exercise programs to reduce the risk of low BMD are safe and effective for children with JIA who are healthy and should be included in the plan of care. More research is needed to determine the amount, duration, and frequency of weight-bearing activity needed to reduce the risk for low BMD in this population.^{13,6}

The role of physical activity and an active lifestyle in cardiorespiratory fitness has been documented, but long-term follow-up is still needed to show if such a program protects from loss of aerobic fitness in this population group.^{48,8} The 6-minute walk test has been shown to be a good test for measuring functional exercise capacity in a small study of children (boys and girls) with JIA ages 7 to 17 years.^{38,5} This might be a good place to begin baseline studies and evaluate response to the aerobic component of the plan of care for individuals with JIA.

Spondyloarthropathies

Spondyloarthropathies (SpAs), a group of disorders formerly considered variants of RA, are in fact distinct entities with similar features affecting the spine (Box 27-14). SpAs are characterized by inflammation of the joints of the spine and include several distinct entities: AS, Sjogren's syndrome, psoriatic arthritis, and reactive arthritis, including arthritides that accompany inflammatory bowel disease (IBD; known as enteric arthritides), and Reiter syndrome. Inflammatory eye disease (e.g., uveitis, conjunctivitis, iritis) occurs in approximately 25% of clients.

Enteropathic arthritis (arthritis associated with IBD) occurs in about 20% of clients who have IBD (e.g., Crohn's disease, ulcerative colitis) and is discussed further in Chapter 16. Arthritic symptoms flare with IBD and usually affect the lower extremities in an asymmetric pattern. Vasculitis, clubbing of the fingers, and skin changes may be present.

New biomarkers for SpAs have been reported recently. Changes in synovial tissue may be good biomarkers to assess the effectiveness of treatment. Specific changes in the cells of the synovial tissue correspond to treatment with TNF blockers. Biopsy specimens of synovial tissue taken after treatment showing changes in synovial macrophages, polymorphonuclear leukocyte (PMN) levels, and expression of metalloproteinases (MMP-3) thought to play a role in the degradation of collagen may lead scientists to use these same biomarkers for early identification and treatment of RA.^{26,3}

Ankylosing Spondylitis

Overview and Incidence. AS, sometimes referred to as Marie-Strumpell disease, is an inflammatory arthropathy of the axial skeleton, including the sacroiliac joints, apophyseal joints, costovertebral joints, and intervertebral disk articulations.

Box 27-14**COMMON FEATURES OF THE SPONDYLOARTHROPATHIES**

- Chronic inflammation of the axial skeleton and sacroiliac joints
- Asymmetric involvement of a small number of peripheral joints
- Young males (late teens, early adulthood) most commonly affected
- Familial predisposition
- Inflammation at sites of ligament, tendon, and fascial insertion into bone
- Seronegativity for rheumatoid factor, but an association with histocompatibility antigens, including HLA-B27
- Extraarticular involvement of eyes, skin, genitourinary tract, cardiac system

Approximately one third of those with AS have asymmetric involvement of the large peripheral joints, including the hip, knee, and shoulder. Fig. 27-18 shows the most commonly involved joints. The disorder can ultimately lead to fibrosis, calcification, and ossification with fusion of the involved joints. The pain, resultant postural deformities, and complications associated with this disease can be disabling.

Prevalence of AS is 0.1% to 0.2% in the U.S. general population. Nearly 2 million people in the United States have this condition, making it almost as common as RA. It is higher in Caucasians and some Native Americans than in African Americans, Asians, or other nonwhite groups.³

AS typically affects young people, beginning between the ages of 15 and 30 years (rarely after age 40 years). This differs from back pain of mechanical origin, which is much more likely to develop between 30 and 65 years of age. Men are affected two to three times more often than women, although this disorder may be just as prevalent in women but diagnosed less often because of a milder disease course with fewer spinal problems and more involvement of joints such as the knees and ankles.⁵¹⁹⁻⁵⁴⁵ Overall sibling risk is about 5.9%.²⁸²

Etiologic and Risk Factors. Although the molecular basis of AS remains unclear, evidence points to a genetic or environmental link. Approximately 90% of those with AS are HLA-B27 positive, but of all of the people with this antigen only 2% develop AS. Proof that this disorder is an autoimmune disease attributable to cross-reactivity between bacteria and HLA-B27 is still lacking.²²⁰ However, approximately 20% of people who develop AS have a first-degree relative who is HLA-B27 positive.

Gender, age, race, and family history are all important factors related to the risk of developing AS. Although it is more prevalent in males, there is significantly less disparity in incidence between the genders than was once thought. The belief is that the disorder has been grossly underdiagnosed in women because the disease tends to be milder and peripheral joint involvement is more common, confusing the clinical picture.

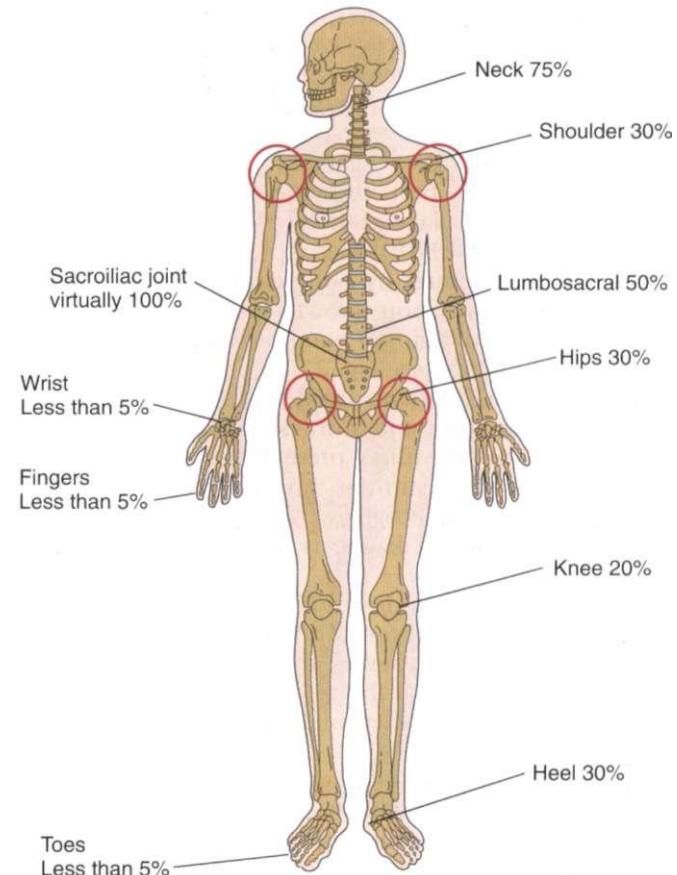


Figure 27-18

Joints most commonly involved in ankylosing spondylitis and incidence of involvement. Not shown: jaw 15%, eye 20%, ribs 20%, costovertebral junction 70%. (From Ramanujan T, Schumacher HR: Ankylosing spondylitis: early recognition and management, *J Musculoskelet Med* 1[1]:75-91, 1992.)

No direct linkage of the X chromosome with susceptibility to AS has been found, but the influence of female gender is greater than that of male gender in determining increased susceptibility to AS in children. The striking maternal effect is greatest for women with young age at onset, which is not seen in men.⁶³ It has been hypothesized that women tend to have a milder form of AS because they have the susceptibility genes, which they pass on to their children, whereas men have the severity genes.⁵⁶

Studies under way using the results of the Human Genome Project are searching for other genes that in combination with HLA-B27 constitute risk for AS. The results of the first genome scan show areas where there appear to be some other genes that have been implicated in Crohn's disease and psoriasis, suggesting a shared genetic basis in these conditions. Other researchers have demonstrated that environmental factors, such as infectious microbes, are essential for the development of AS, in particular normal bacteria in the bowel.^{22,217,282}

Pathogenesis. The pathogenesis of AS is poorly understood, but the fundamental lesion appears to be chronic inflammation at sites of attachment of cartilage, tendons,

ligaments, and synovium to the bone.⁴¹⁰ AS is marked by a chronic nongranulomatous inflammation at the area where the ligaments attach to the vertebrae (an area called the enthesis), initially in the lumbar spine and then in the sacroiliac joint.

Disruption of this ligamentous-osseous junction results, and reactive bone formation occurs as part of the repair process. Cartilage of the sacroiliac joints may also be involved (Fig. 27-19). The replacement of inflamed cartilaginous structures by bone contributes to progressive ossification with bony growth between the vertebrae, leading to a fused, rigid, or bamboo spine, characteristic of end-stage disease (see Fig. 27-20).

Clinical Manifestations. Insidious onset of low back, buttock, or hip pain and stiffness lasting for at least 3 months are often the initial presenting complaints. Early onset of back pain, stiffness, and fatigue during childhood often goes unrecognized for what it is. Onset of symptoms leading to a diagnosis occurs most often during early adulthood.⁴¹⁰

At first the pain is described as a dull ache that is poorly localized, but it can be intermittently sharp. Over time, pain can become severe and constant, increased by prolonged rest or immobility and decreased by active movement. Coughing, sneezing, and twisting may worsen the pain.²⁸²

Pain may radiate to the thighs but does not usually go below the knee.³ Buttock pain is often unilateral but may alternate from side to side. Significant morning stiffness, lasting more than 1 hour, is often present. There may be tenderness over the spinous processes and sacroiliac areas with associated paraspinal spasms.

Enthesitis (inflammation of the tendons, ligaments, and capsular attachments to bone) may produce tenderness, pain and/or stiffness, and restricted mobility in the costosternal, costovertebral, and manubriosternal joints, iliac crest, ischial tuberosities, greater trochanters, spinous processes, or ligamentous attachments at the calcaneus.

Other clinical features include early loss of normal lumbar lordosis with accompanying increased kyphosis of the thoracic spine, painful limitation of cervical joint motion, and loss of spine mobility (flexibility) in all planes of motion.

In some cases the initial complaints may occur in the extremities (e.g., hips or knees) with back symptoms appearing on average 3 years after onset of the peripheral involvement. Shoulder symptoms and loss of shoulder mobility are common but are rarely disabling. Involvement of the shoulder joint in AS correlates with involvement of other peripheral joints as well as the extent of radiographic change on shoulder films.⁵⁴²

Hip flexion contractures are often present bilaterally and can be assessed using the Thomas test (in the supine position, ask the individual to maximally flex the contralateral hip joint; observe for loss of lumbar lordosis and flexion of the opposite leg, signaling a positive Thomas test result for the presence of a hip flexion contracture; repeat on the other side). Loss of hip mobility can result in reduced functional ability.

Loss of chest wall excursion is an indicator of decreased axial skeleton mobility because of involvement of the thoracic spine and the costovertebral and costosternal

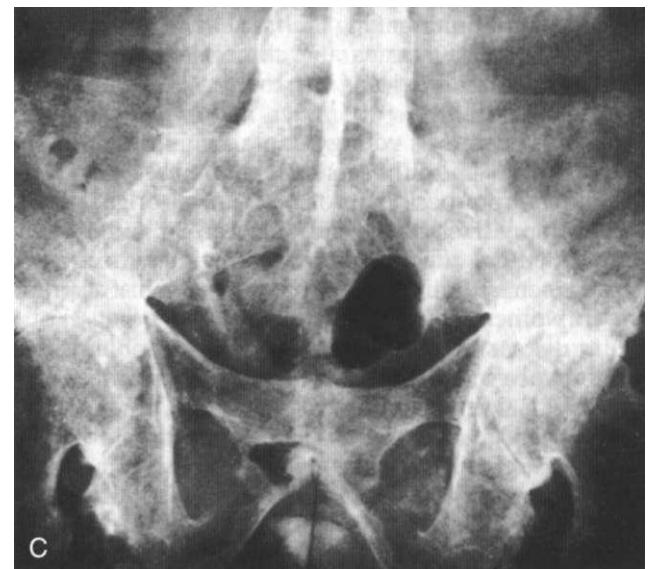
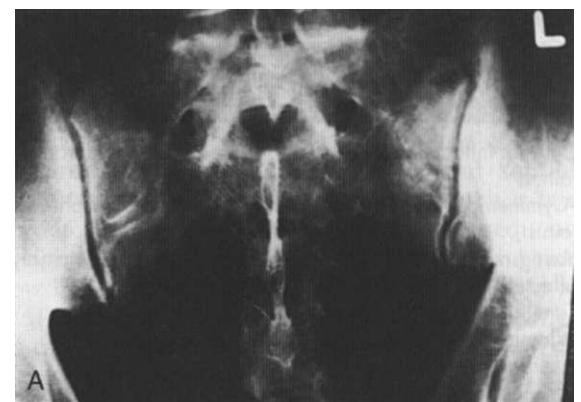


Figure 27-19

Progression of ankylosing spondylitis of the sacroiliac joints. **A**, Normal sacroiliac joints. **B**, Fusion of sacroiliac joint spaces; the sclerosis has resorbed, and there is slight narrowing of the left hip joint. **C**, Advanced ankylosing spondylitis with generalized osteoporosis and fusion of the spinous process, intervertebral disks, sacroiliac joints, and symphysis pubis. The entire skeletal unit has been transformed into one continuous osseous mass. (**A**, from Magee D: *Orthopaedic physical assessment*, ed 2, Philadelphia, 1992, Saunders, p 329. **B**, from Rothman RH, Simeone FA: *The spine*, Philadelphia, 1982, Saunders, p 921. **C**, from Bullough PG: *Orthopaedic pathology*, ed 3, London, 1997, Mosby-Wolfe, p 300.)

joints. If the ligaments that attach the ribs to the spine (costovertebral junction) become ossified, the chest may be unable to expand, compromising breathing. Circumferential measurements taken at the fourth intercostal space (or just below the breasts in women) before and at the end of inspiration should reveal an excursion of 4 to 5 cm. Excursion less than 4 cm, and especially anything less than 2.5 cm, is a suspicious finding.

COMPLICATIONS. Long-standing disease is associated with multiple complications. Skeletal complications include osteoporosis, fracture, atlantoaxial subluxation, and spinal stenosis. In the most severe cases, the spine becomes so completely fused that the person may become locked in a rigid upright or stooped position, unable to move the neck or back in any direction. Flexion contractures, rigid gait, and flexing at the knees in order to maintain an upright position are not uncommon findings.

The stiff and osteoporotic spinal column is prone to fracture from even a minor insult; a significant proportion of individuals with AS experience vertebral fracture during the course of the disease. Fracture sites range from T7 to S1.^{117,197} In fact, the incidence of thoracolumbar fractures in AS is four times higher than in the general population.^{89,197}

Fractures can occur in the lower cervical spine. The atlantoaxial segment is among the last areas of the axial skeleton to fuse. Because of the inherent mobility of C1 and C2 and the immobility of the remainder of the cervical spine secondary to the disease, attempts to move the head could result in subluxation. Spinal stenosis can result from the proliferation of bony tissue from the spinal ligaments and facet joints. Stenosis may cause neurogenic claudication and cauda equina syndrome.

AS is a systemic disease with widespread effects. In addition to arthritis in the spine, arthritis in other joints may be accompanied by inflammatory bowel syndrome with fever, fatigue, loss of appetite, weight loss, and other extraarticular complications. These clinical features distinguish AS from mechanical pain.

The most common extraarticular manifestation is uveitis, occurring in 20% to 30% of affected individuals. Cardiomegaly, pericarditis, aortic regurgitation or insufficiency, amyloidosis (rare), and pulmonary complications may also occur. Pulmonary problems include upper lobe fibrosis and decreased total lung capacity and vital capacity (late stages of AS).³¹⁰

MEDICAL MANAGEMENT

DIAGNOSIS. AS is not easily detectable in the early stages without an MRI and only then with specialized MRIs, which are not routinely ordered. Early, accurate diagnosis allows treatment to start before the onset of permanent rigidity and deformity.

Intraarticular inflammation, early cartilage changes, and underlying bone marrow edema and osteitis can be seen with a specific MRI technique called short tau inversion recovery (STIR) sequences.³¹⁰ Diagnosis is usually based on identification of the clinical manifestations and radiographic findings.³ History, physical examination, radiography, and laboratory tests are all used in the diagnosis.

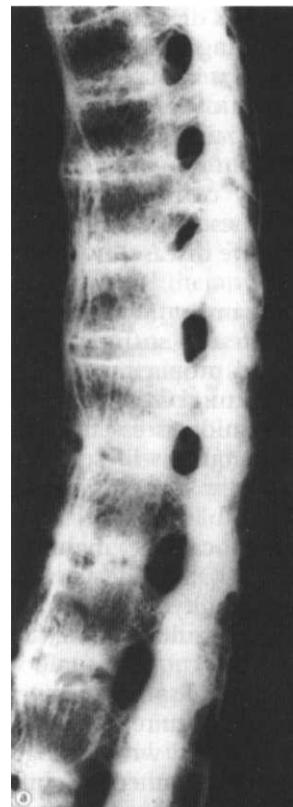


Figure 27-20

Radiograph of a sagittal vertebral column in a person with ankylosing spondylitis. There is complete fusion of the spine, accentuated kyphosis, and loss of lumbar and cervical lordosis. There is also complete fusion of the intervertebral disk spaces. (From Bullough PG: *Orthopaedic pathology*, ed 3, London, 1997, Mosby-Wolfe, p 301.)

In the physical examination, range-of-motion tests provide important information. The Schober test, chest expansion (measured at the fourth intercostal space or just below the breasts in women on inspiration after forced maximal expiration), and military stand against the wall are clinical tests used most often. A distance of less than 15 cm for the Schober test and less than 2.5 cm for chest expansion on inspiration are suspicious findings.

Clinical manifestations alone are not diagnostic, but when taken along with the history and radiographic results, they become significant for the diagnosis. Radiographs may be negative in the early stages of the disease; MRI enables the examiner to identify sacroiliitis earlier than plain radiography.

Radiographic findings of symmetric, bilateral sacroiliitis include blurring of joint margins, juxtaarticular sclerosis, erosions, and joint space narrowing. The replacement of ligamentous tissue by bone at the site where the annulus fibrosus of the intervertebral disk inserts into the vertebral body results in a characteristic square-shaped vertebral body. In addition, as bony tissue bridges the vertebral bodies and posterior arches, the thoracic and lumbar spine takes on the appearance of a bamboo spine on radiographs (Fig. 27-20).

No laboratory test is diagnostic of AS; laboratory tests assist primarily by ruling out other diseases. The presence of the HLA-B27 antigen is a useful adjunct to the diagnosis but is not diagnostic itself, since so many people with other causes of back pain also are HLA-B27 positive. ESR and C-reactive protein are elevated in 75% of affected individuals and may correlate with disease activity in some people, but these levels can be normal even in individuals with active disease.^{410,461}

TREATMENT. The primary focus of intervention is to reduce inflammation and stiffness in the joints, maintaining mobility and proper postural alignment of the spine to prevent structural damage, while providing pain relief. Effective education is essential, because much of the management requires lifestyle adjustments and cooperation, especially compliance with the exercise program.²⁸²

Joint involvement can be managed with NSAIDs (including the new COX-2-specific inhibitors), but in some cases disease-modifying drugs (DMARDs) such as methotrexate (MTX) or sulfasalazine (SSZ) may be used for peripheral disease.

Persistent symptoms of spinal involvement, peripheral arthritis, or enthesitis unresponsive to NSAIDs or DMARDs can be managed with biologic agents such as TNF- α antagonists (e.g., etanercept, infliximab, adalimumab). These agents have been shown effective in preventing the progression of disease by reducing disease activity, decreasing inflammation, and improving spinal mobility.^{3,51}

Other targeted therapies may be needed to treat specific organ involvement, such as eye inflammation. To avoid long-term complications associated with severe postural deformities, a lifetime commitment to exercise is important.

Surgery has a limited role in the management of AS and is most appropriate for individuals with severe deformity that impedes vision, walking, eating, abdominal expansion, or respiratory function. Spondylodiskitis or spinal fracture may require surgical intervention.⁴⁹ Spinal fusion may be needed but is not routinely recommended. The most valuable surgical intervention is total joint arthroplasty, especially a total hip replacement.²⁸² It is expected that with the new biologic therapies, the need for surgical intervention will become a rare event.⁴⁹

PROGNOSIS. The extent of disability in persons with AS varies considerably, but fewer than 1% experience complete remission and more than 80% who are ill for longer than 20 years still have daily pain.^{24,545} Periods of exacerbation and remission are common during the course of the disease.

The severity of symptoms during the first decade indicates the long-term severity and disabling nature of the disorder. Severe disease is usually marked by peripheral joint and extraarticular manifestations. The onset of hip disease in anyone with AS at any stage of the disease is a major prognostic marker for long-term severe disease and is more common in people with onset at a young age.⁵⁶

Individuals with AS have an increased mortality rate. The impact of this disease can be seen in various aspects

of workforce participation, such as needing more assistance, withdrawal from the workforce, and reduced quality of life. Early diagnosis and management will likely help prevent functional disability and improve outcomes.⁴⁷

SPECIAL IMPLICATIONS FOR THE THERAPIST 27-11

Ankylosing Spondylitis

PREFERRED PRACTICE PATTERNS

4A: Primary Prevention/Risk Reduction for Skeletal Demineralization (osteopenia, osteoporosis)

4B: Impaired Posture

4E: Impaired Joint Mobility, Motor Function, Muscle Performance, and Range of Motion Associated with Localized Inflammation

4F: Impaired Joint Mobility, Motor Function, Muscle Performance, Range of Motion, and Reflex Integrity Associated with Spinal Disorders (spinal stenosis)

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning (other cardiovascular/pulmonary patterns may apply depending on clinical presentation)

Whenever someone presents with new onset of back, sacroiliac, or hip pain in the absence of trauma or overuse that is accompanied by associated signs and symptoms of systemic disease (e.g., fever, fatigue, respiratory compromise) the therapist must recognize that these are not symptoms typically associated with mechanical causes of back pain. If present, these features are considered red flags that should raise concern regarding the underlying cause of the complaints.

The therapist can assess for limitation of spinal mobility, most evident as a decrease in forward flexion of the spine, using the Schober test. In the standing position, place a mark at the lumbosacral junction, which is represented by the intersection of a line joining the dimples at the posterosuperior iliac spines (PSISs). Place a second mark 5 cm below and 10 cm above the lumbosacral junction. The client is then asked to bend forward and attempt to touch the toes.

Now remeasure the distance between the upper and lower marks. The distraction between these two marks has been found to correlate very closely with anterior flexion measured radiologically. The distance increases more than 5 cm in the absence of AS and less than 5 cm when AS is present.

Alternatively, measure the horizontal distance between the posterior inferior iliac spine in both the prone and sitting positions, observing for a change in the distance with a change in position. Expect to see the posterior inferior iliac spine 1/2 inch closer in the prone position compared to sitting in the normal young adult and suspect AS if there is no change.

Treating a client with a diagnosis of AS requires that the therapist entertain different considerations. If the client complains of sharp pain associated with a fall, sneeze, or lifting a moderately heavy object, one must consider the possibility of a fracture. With osteoporosis being a potential complication, technique modification is warranted. See Special Implications for the

Therapist box in the section on Osteoporosis in Chapter 24 for further discussion.

Management should follow guidelines similar to those outlined for fibromyalgia (see Chapter 7). With the fragility of the spine in AS and the risk of fractures from even a minor injury or fall, falls prevention is also important. This is an area for careful attention, because a number of people with AS suffer neurologic deficit after injury, especially spinal fracture.¹⁹⁷

Trunk range-of-motion and strengthening exercises to minimize thoracic kyphosis are essential. The more severe the kyphosis, the more hindered pulmonary function will be and the more pronounced the compensatory forward head posture. Postural deformities contribute to cervical pain and headaches and may also affect balance. Avoiding obesity is recommended to reduce stress on weight-bearing joints and the cardiopulmonary system.

Finally, smoking should be discouraged because of its adverse effects on the cardiopulmonary system. The therapist can also monitor clients taking NSAIDs for early signs of gastrointestinal bleeding or other adverse side effects (see Chapter 6).

Exercise and Ankylosing Spondylitis

Therapists play a potentially important role in the rehabilitation of this population. Although more studies are available now than ever before in this area, there still is not sufficient evidence to base recommendations for or against specific physical therapy interventions. Continued research is needed to address those modalities and applications commonly used in practice.⁹⁴ A summary of the available scientific evidence on the effectiveness of physical therapy interventions in the management of AS is presented here and elsewhere.⁵¹⁹

GENERAL CONCEPTS

Although exercise is a commonly recommended intervention for AS, little is known about the effectiveness of unsupervised recreational and back exercises. Findings from one study⁵¹¹ suggest that exercise improves pain, stiffness, and function when performed at least 30 min/day and back exercises at least 5 days/week, although these effects vary with the duration of the AS. There may be an optimal duration for exercise performed independently over a weekly period, but consistency rather than quantity appears to be a much more significant variable. Although there is no difference in disease activity with a regular program of exercise, improved function has been demonstrated.⁴³⁷

Interpreting the results of some studies can be difficult. For example, one study showed that younger individuals with AS who had a shorter duration of disease and were less disabled also exercised less compared to older, more disabled adults who exercised more often. Without careful analysis and understanding of the results, the conclusion could be made that those people who exercised more frequently were more disabled, when in fact, affected individuals did not get serious about exercising consistently until the disease had progressed.¹¹⁵

EXERCISE PRESCRIPTION

The question of what type of exercise is best for this condition has not been adequately answered. Exercising has been shown to benefit people with AS, but again, consistency, not quantity, seems to be the key factor in influencing posture, strength, motion, mobility, fitness, and overall health.⁴³⁶ Home programs and group programs have both been shown to be beneficial for individuals with AS.^{93,122}

A multimodal physical therapy program including aerobic, stretching, and pulmonary exercises along with routine medical management has been shown to yield greater improvements in spinal mobility, work capacity, and chest expansion compared with medical care alone. Individuals in a 50-minute, three-times-a-week multimodal exercise program were significantly improved after 3 months in chest expansion, chin to chest distance, occiput to wall distance, and the modified Schober flexion test.²¹⁹

Functional and breathing capacity as well as balance should be assessed and developed. Stretching of the shortened muscles and chest expansion exercises should be encouraged. Improving and/or maintaining cardiovascular fitness is important.

Strengthening of the trunk extensors is equally important, so that if and when spinal fusion occurs, the spine is aligned in the most functional position possible. This requires a coordinated effort among all team members, including the affected individual and family.

High-impact and flexion exercises should be avoided, whereas low-impact, aerobic exercise with extension and rotational components can be emphasized. Each individual will need to identify personal limitations and safe levels of participation.

In general, contact sports and high-risk activities such as downhill skiing, horseback riding, boxing, football, soccer, and water skiing should be avoided; aquatic therapy is an excellent option for most people provided extension principles are emphasized. In the presence of spinal fusion and osteoporosis, other activities requiring high levels of balance, agility, and coordination (e.g., bicycling, ice skating, rollerblading) can result in falls and fractures.

Overexercising can be potentially harmful and can exacerbate the inflammatory process; principles of relaxation, proper body mechanics, and energy conservation should be a part of the education program offered (see Box 9-8), including assessment of ADLs and providing necessary aids or devices such as long-handled reaching tools, adaptations for the car, special garden tools, or elastic shoelaces. Learning and using proper breathing techniques throughout all activities will help maintain chest expansion, improve oxygenation, and minimize muscle fatigue.

At present, aggressive but careful stretching to address the areas of hypomobility and the muscle imbalances is purported to help maintain an optimal posture as possible. An exercise program focusing on specific strengthening and flexibility exercises of the shortened muscle chains offers promising short-

Continued.

and long-term results in the management of this condition.¹²¹

OUTCOMES MEASUREMENTS

Mobility tests to establish a baseline and measure the effectiveness of short-term intensive physical therapy and exercise on spinal, hip, and shoulder motion have been evaluated.

Finger to floor distance, thoracolumbar rotation, and thoracolumbar lateral flexion are the most sensitive tests to detect improvements (or progression) in short-term clinical trials, whereas the Schober test, thoracolumbar flexion, and occiput-wall distance are insensitive measures of improvement. Hip internal rotation, shoulder flexion, and abduction measurements are also sensitive, although more suitable for individuals with articular symptoms. Thoracolumbar rotation and hip rotation are the only measurements that correlate with disease duration but not with age.^{74,186}

The Bath Ankylosing Spondylitis Functional Index (BASFI), along with the Dougados Functional Index score, Bath Ankylosing Spondylitis Metrology Index, and Bath Ankylosing Spondylitis Disease Activity Index are self-assessment tools that can be used both by the therapist and by the client to establish a baseline and document improvement or decline.^{33,42,361}

The therapist must be vigilant for an exacerbation of the inflammatory process. Assess (and periodically reassess) the spine and peripheral joints for mobility, range of motion, and strength. Modalities such as ultrasound, heat and cold, and electrotherapy may be effective during the acute phase if used judiciously.

Aggressive stretching must be avoided during these phases; teaching the individual a progressive relaxation program may be helpful. Communication with the physician is necessary for the development of a proper medication regimen.

POSITIONING

The affected individual may need help modifying home or work situations. Appropriate footwear advice should be provided; some people benefit from functional foot orthoses. Resting in the prone position is advised to help avoid hip and spine flexion contractures. The Spondylitis Association of America⁴⁷² recommends a firm, supportive sleeping surface to maintain good spinal alignment. Soft mattresses or waterbeds can contribute to excessive flexion and stooped postures.

The therapist can provide additional recommendations for the use of pillows or towel rolls for proper alignment in the various positions. The person may not stay positioned all night, but with time and training, changing positions and realigning props can be incorporated into the sleep cycle at least part of the time.

Proper lifting techniques can be demonstrated, with return demonstration provided by the client. The therapist should assess the work area or instruct the individual in appropriate ergonomics given the diagnosis of AS.

Sjogren's Syndrome

Overview. Sjogren's syndrome is a chronic arthritis-related disease that can affect several organs, most commonly the moisture-producing glands (e.g., mouth, eyes) but also joints, lungs, kidneys, or liver. Sjogren's syndrome is an autoimmune disease, sometimes also considered as a connective tissue disease, that is characterized by the body's inability to distinguish healthy cells from foreign substances.

In Sjogren's, the immune system mistakenly attacks its own moisture-producing glands and other organs. It may be a primary condition occurring alone or secondary to other autoimmune diseases, such as RA or lupus.⁸³

Incidence and Risk Factors. Sjogren's syndrome is the second most common autoimmune rheumatic disease, affecting an estimated 2 to 4 million Americans, developing most often in postmenopausal women. It can occur in children and men or women of any age; women are affected nine times more often than men.¹³⁰

Information from rheumatology clinics suggests that primary Sjogren's syndrome is as common as SLE and that approximately 30% of people with RA or systemic sclerosis have histologic evidence of Sjogren's. Other risk factors include having another autoimmune disease or having a family member with Sjogren's.

Etiologic Factors and Pathogenesis. The primary symptoms of Sjogren's syndrome are the result of exocrine gland (mainly salivary and lacrimal gland) destruction by focal T-lymphocytic infiltrates. The infiltrating T and B cells interfere with glandular function at several points.¹³⁸ Additional potential contributing factors are B-cell hyperreactivity (these locally produce immunoglobulins having autoantibody reactivity) and long-term immune system stimulation.⁴⁶⁴

Evidence supports a genetic component in its etiology, but there is no strong evidence for a specific candidate gene.²⁰ Neurogenic regulation of the salivary gland is impaired, with structural abnormalities of the secretory acinar apparatus. The acinar basement membrane is abnormal, as it lacks the laminin α1 chain; this loss may impair its ability to induce stem cells to differentiate into acinar cells.²⁵⁷

Organ-specific autoantibodies are present, but the role of the autoantibodies in the disease process is not clear, and it is unknown whether they contribute to tissue dysfunction before tissue inflammation is observed.⁴²³ Researchers suspect that a common immunologic mechanism (e.g., infiltration by activated T cells and expression of HLA-class molecules on epithelial cells) is involved in the development of autoimmune disorders, especially autoimmune thyroid diseases and Sjogren's syndrome, but the details remain unknown.

The interactions between the neuroendocrine and immune systems as these relate to autoimmune diseases such as Sjogren's syndrome are the topic of numerous research studies.²³⁰ Significantly lower basal adrenocorticotropic hormone (ACTH) and Cortisol levels have been found in individuals with Sjogren's, associated with a blunted pituitary and adrenal response to ovine corticotropin-releasing factor (oCRH) compared to normal controls. Research findings suggest both adrenal

axis hypoactivity as well as adrenal and thyroid axes dysfunction.²²⁹

Clinical Manifestations. Clinical manifestations vary according to the systemic problems present from integumentary, respiratory, renal, hepatic, neurologic, and vascular involvement. Associated symptoms may include extremely dry throat, esophagitis, gastritis, and dental cavities from a lack of saliva; vaginal dryness with painful sexual intercourse; fatigue; joint and muscle pain; joint and muscle stiffness; swelling; rashes (vasculitis); numbness (peripheral neuropathy as a consequence of small-vessel vasculitis); Raynaud's phenomenon; B-cell lymphoma; and inflammation of the lungs, kidneys, or liver.

The hallmark symptoms of Sjogren's syndrome are dry eyes and dry mouth. This syndrome may also cause dryness in other areas, such as the kidneys, gastrointestinal tract, blood vessels, sinuses, respiratory tract, liver, pancreas, and central nervous system.

Some of the problems (e.g., recurrent bronchitis or sinusitis) arise from exocrine dysfunction in other organs, while other problems (e.g., interstitial lung disease, interstitial nephritis) occur as a result of extraglandular spread of lymphocytic infiltration discussed in the pathogenesis of this disease.⁵²⁹ Primary Sjogren's causes salivary gland swelling and tenderness. The dry eyes (*keratoconjunctivitis sicca*) are described as the feeling of sand or a burning sensation in the eyes with decreased secretion of tears. Dry mouth (*xerostomia*) and dry cough make it difficult for affected individuals to chew and swallow food or speak continuously.

Depression, anxiety, thyroiditis, and fibromyalgia are frequent comorbid illnesses requiring a comprehensive management approach to this condition. Quality of life is decreased by complications such as sleep loss, loss of teeth and poorly fitting dentures, loss of vision, profound fatigue, musculoskeletal pain, morning stiffness, and so on.

MEDICAL MANAGEMENT

DIAGNOSIS. Many conditions present similarly to Sjogren's syndrome with dry eyes and dry mouth, such as lupus, vasculitis, thyroid disease, and scleroderma; side effects of some medications (e.g., tricyclic antidepressants, antihistamines, radiation treatments of the head and neck) can mimic Sjogren's. Sjogren's is a systemic disease with the potential to affect almost every organ system in the body, so the proper diagnosis is important.

Diagnosis is based on a complete physical examination; medical history; and specific tests such as a slit-lamp test to detect damage to the surface of the eye by using a dye that exposes eroded areas of the conjunctiva (the membrane that covers the eye and lines the inside of the eyelids), Schirmer's test to assess degree of dryness in the eyes, lip biopsy to show inflammation of the salivary glands, and blood tests to detect antibodies (e.g., rheumatoid factor, ANA, anti-SSA, anti-SSB) that are associated with primary Sjogren's.

Many serum and salivary biomarkers for Sjogren's have been proposed, but none has been specific enough for diagnostic purposes or correlated with disease activity

measures. Modern genomic investigation is looking for candidate biomarkers and possible etiopathologic mechanisms underlying this disorder.¹³⁰

TREATMENT. There is no cure for Sjogren's, but it can be managed effectively. Ocular involvement is managed with local and systemic stimulators of tear secretion. Treatment of oral manifestations includes intense oral hygiene and prevention and treatment of oral infections. The use of saliva stimulants and mouth lubricants can help with the dryness.³¹⁶ Avoiding situations and activities that contribute to dryness and moisturizing other areas of dryness such as the skin and vagina (women) are advised.

Intervention typically involves medications (e.g., corticosteroids such as prednisone, NSAIDs, or hydroxychloroquine [Plaquenil]) to help reduce joint pain and stiffness and ease fatigue and muscle pain as well as other palliative measures for symptomatic relief. Exercise and proper nutrition may help with the fatigue and joint symptoms.

Mild cases of peripheral neuropathy can remit spontaneously, but usually symptomatic treatment (e.g., gabapentin) is needed. More severe involvement affecting ambulation may require the use of steroids, azathioprine, or intravenous gamma globulin or cyclophosphamide.⁵²⁹ Anti-B-cell therapy is a new potential therapy for glandular and extraglandular manifestations such as glomerulonephritis or vasculitis. Gene transfer has been attempted in animal models with promising results.³¹⁶ The use of green tea polyphenols (GTPs), which have both antiinflammatory and antiapoptotic properties, is also under investigation based on the knowledge that the incidence of Sjogren's is much lower in China and Japan, two leading green tea-consuming countries. Animal studies show that GTPs could provide protective effects against autoimmune reactions in skin and salivary glands.²⁰⁶

PROGNOSIS. Sjogren's syndrome progresses slowly, with the interval between first symptoms and diagnosis ranging from 2 to 8 years. Left untreated, dryness of the eyes can lead to eye infections and may result in damage to the cornea and visual loss.

Sjogren's is a benign disease that affects quality of life. When extraoral and extraocular exocrine gland dysfunction or lymphocyte-mediated tissue destruction involves other organs, significant morbidity and mortality can occur. There is a high risk of malignant transformation that requires close follow-up.

SPECIAL IMPLICATIONS FOR THE THERAPIST 27-12

Sjogren's Syndrome

Special implications and preferred practice patterns are determined by the presenting clinical features but follow the general guidelines for RA. See Special Implications for the Therapist box in the section on Rheumatoid Arthritis.

Physical capacity is reduced in Sjogren's, and fatigue is a dominating and disabling symptom.

Continued.

Evidence-based studies on the effect of exercise in Sjogren's are limited, with small sample sizes. The available studies indicate that clients with Sjogren's can benefit from moderate- to high-intensity levels of exercise with positive effects on aerobic capacity, fatigue, physical function, and depression (mood).

Further research is needed to evaluate the effect of exercise on groups with varying degree of disease severity and to document the long-term impact on the disease.⁴⁷⁹

Psoriatic Arthritis

Overview and Incidence. Psoriatic arthritis is a seronegative inflammatory joint disease afflicting a small percentage of people who have psoriasis. This joint disorder is associated with radiographic evidence of periarticular bone erosions and occasional significant joint destruction. Psoriatic arthritis tends to progress slowly, and for most of those affected it is more a nuisance than a disabling condition.

Approximately 1% of the population of the United States has psoriasis. Psoriatic arthritis occurs in about 20% of persons with psoriasis and more often in those with severe psoriasis. Uncomplicated psoriasis typically presents during the second and third decades of life, with the onset of the arthritis occurring up to 20 years later. The disease can occur in children, with onset typically between the ages of 9 and 12 years. Psoriatic arthritis does not appear to have a strong predilection for one gender.

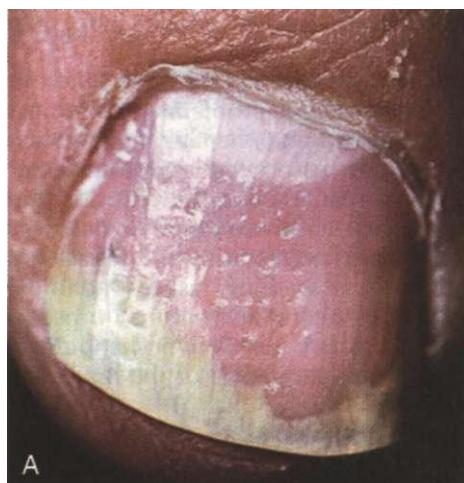
Etiologic and Risk Factors. A strong familial association has been noted with this disease. Although specific marker genes have not been discovered, there is general agreement that a genetic predisposition exists for psoriatic arthritis. There is approximately an 80% to 90% chance of contracting psoriatic arthritis if one has a first-degree relative with the disorder.

Pathogenesis. An inflammatory synovitis results in the joint changes associated with psoriatic arthritis. Lymphocyte infiltration into the synovium occurs. Initially the synovium is pale, with edematous granulation tissue extending along the contiguous bone. The synovium later becomes thickened with villous hypertrophy. Eroded articular margins begin to appear at this time. In severe cases the joint space tends to be filled in with dense fibrous tissue.

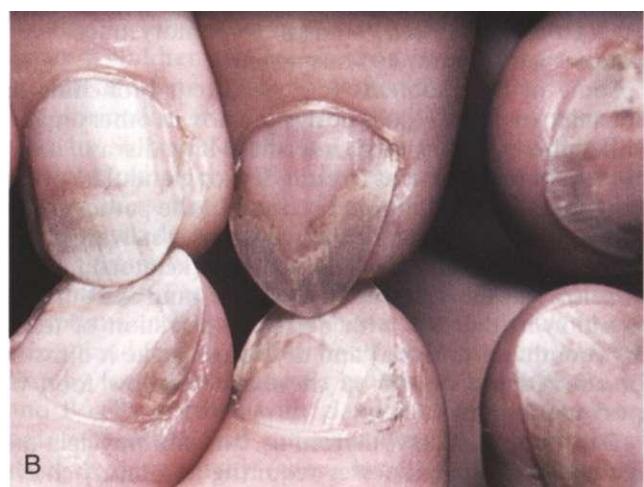
Clinical Manifestations. The arthritis can be oligoarticular or polyarticular. There is a predilection for the distal interphalangeal joints of the hands. Other joints of the digits may be involved. The joint changes may lead to significant hand deformities, including claw deformity. The digital joint changes and associated flexor tenosynovitis can result in an edematous, thickened digit.

Joints of the axial skeleton can also be affected but typically become involved several years after the onset of the peripheral joint disease. The sacroiliitis is usually unilateral, unlike that in AS. Sacroiliitis can occur in 20% to 40% of clients.

Although not as common as in RA or Reiter syndrome, extraarticular manifestations can occur with psoriatic arthritis. Inflammatory eye disease, including conjuncti-



A



B

Figure 27-21

Nail changes associated with various forms of arthritis. **A**, Pitting of the nail beds associated with psoriasis. **B**, Onycholysis associated with reactive arthritis, a separation of the nail plate from the nail bed beginning at the free margin and progressing inward. (A, from James WD, Berger T, Elston D: *Andrews' diseases of the skin: clinical dermatology*, ed 10, Philadelphia, 2006, Saunders. B, from Arndt KA: *Primary care dermatology*, Philadelphia, 1997, Saunders.)

vitis and iritis; renal disease; mitral valve prolapse; and aortic regurgitation have been associated with this disorder. Pitting of the nails and onycholysis (Fig. 27-21) are also commonly associated with psoriatic arthritis.

There are some differences in the manifestations of this disease between children and adults. A slight predilection for females is noted in children. In addition, the arthritis may appear before the skin manifestations in a number of children. Compared with adults, the onset of the arthritis tends to be more acute in children, with the involvement of multiple asymmetric joints. The hip joint is much more commonly involved in children.

MEDICAL MANAGEMENT

DIAGNOSIS. The diagnosis of psoriatic arthritis is usually easily made because of the onset of inflammatory arthritis in the presence of obvious psoriasis. Differential diagnosis can be difficult, however, if the psoriasis is absent or equivocal. Laboratory tests do not help except to rule

Box 27-15**RADIOGRAPHIC FEATURES OF PSORIATIC ARTHRITIS**

- Asymmetric oligoarticular distribution of disease
- Relative absence of osteopenia
- Involvement of distal interphalangeal joints
- Involvement of sacroiliac joint (unilateral)

out RA. Box 27-15 lists common radiographic findings in psoriatic arthritis.

TREATMENT AND PROGNOSIS. There is currently no cure for psoriasis or psoriatic arthritis. People with mild arthritis are treated symptomatically with NSAIDs. If there is an acute flare of only one or two joints, local corticosteroid injections may help.

Anyone with more aggressive disease may benefit from DMARD therapy with MTX, SSZ, and the TNF- α antagonists.

Because of the association between severe skin involvement and severe arthritis, treatment of the psoriasis is emphasized with the hope of reducing the arthritis. Multiple medications have been used in an attempt to control progressive psoriatic arthritis but with equivocal results. As noted earlier, for most persons with psoriatic arthritis, the disease is mild, not destructive.

SPECIAL IMPLICATIONS FOR THE THERAPIST 27-13***Psoriatic Arthritis*****PREFERRED PRACTICE PATTERNS**

See *Special Implications for the Therapist* box in the section on *Rheumatoid Arthritis*.

If a flare-up of the skin condition is noted, encourage the client to see her or his physician. If the joint inflammation worsens, prompt communication with the physician should occur so the client can be placed on an appropriate medication regimen.

Reactive Arthritis

Overview. This section is confined to the discussion of reactive arthritis, which differs from bacterial arthritis (discussed in Chapter 25) in several ways. Reactive arthritis is defined as the occurrence of an acute, *aseptic*, inflammatory arthropathy arising after an infectious process but at a site remote from the primary infection, whereas bacterial arthritis may be a local response with joint destruction and sepsis.

The borderline between reactive arthritis and true septic arthritis may be obscure, since several organisms can cause both, with overlapping symptoms and laboratory features. Other infectious causes of arthritis are discussed in other chapters (e.g., HIV in Chapter 7; Lyme disease and Epstein-Barr virus in Chapter 8; rheumatic fever in Chapter 12).

Etiologic and Risk Factors and Pathogenesis. Reactive arthritis is a recognized sequela of infection with a

number of enteric pathogens, such as *Campylobacter jejuni* (gastrointestinal tract), *Salmonella typhimurium*, *Shigella* (dysentery), *Chlamydia trachomatis* (genitourinary tract), *Chlamydia pneumoniae* (respiratory tract), *Yersinia*, *Mycoplasma fermentans*,²⁴ and *Clostridium difficile* (colitis associated with antibiotic therapy; see discussion in Chapter 16).

The overall prevalence of reactive arthritis has declined, although an increase has been seen in a small population group composed of intravenous drug users with acquired immunodeficiency syndrome. Reactive arthritis is most common in young, sexually active adults, especially men who have been infected with *C. trachomatis*. However, children and older adults of both genders are affected by the postenteric form. Reactive arthritis following urogenital infection is underdiagnosed in women. The tendency for chlamydial infection to be subclinical or asymptomatic and the relative infrequency of pelvic examinations are contributing factors.³⁵⁸

A particular MHC class I antigen, HLA-B27, is well recognized as a genetic marker of susceptibility to reactive arthritis (see Table 40-20). Bacteria in the joint may stimulate the immune system to produce antibodies and protein factors (cytokines), several of which produce local inflammation and tissue damage, leading to an arthritic joint.

Clinical Manifestations. The arthritis first manifests 1 to 4 weeks after the infectious insult and is usually asymmetric affecting more than one joint, typically the large and medium joints of the lower extremities. Sacroiliac joint involvement occurs in about 10% of acute cases and 30% of chronic cases. The clinical picture varies from mild arthralgia and arthritis to incapacitating illness that may result in bed rest for several weeks. Joint pain may be minimal with no signs of inflammation, but stiffness, pain, tenderness, and loss of motion are often present.³⁵⁸

Associated findings may include uveitis, enthesitis (inflammation involving the sites of bony insertion of tendons and ligaments), sacroiliitis, urethritis, and conjunctivitis. Reactive arthritis encompasses a subgroup that demonstrates the classic clinical triad of arthritis, urethritis, and conjunctivitis, which is called Reiter syndrome (see further discussion in the next section). Reactive arthritis is a broader category that includes some but not all of the more restrictive features associated with Reiter syndrome. The distinction between these two conditions is somewhat arbitrary.²⁴¹

Extraarticular manifestations of reactive arthritis may include onycholysis of the fingernails or toenails, dactylitis (sausagelike swelling of the toes and fingers because of joint and tenosynovium inflammation), painless mucosal ulcers in the mouth, discharge from the vagina or penis, urologic symptoms (urgency, frequency, difficulty starting or continuing a flow of urine), or various types of skin lesions. Rarely, neurologic or cardiac involvement occurs secondary to inflammatory and fibrotic lesions.

MEDICAL MANAGEMENT

DIAGNOSIS. There is considerable clinical overlap among the various types of inflammatory arthritides. Usually, a

careful clinical and family history and physical examination will lead to the diagnosis. Laboratory evaluation, synovial fluid aspiration, cultures for bacteria, antibody testing, measurement of serum immunoglobulin, and imaging studies contribute to the differential diagnosis.

TREATMENT AND PROGNOSIS. NSAIDs and disease-modifying drugs are the basis of medical management. A short course of corticosteroids may be necessary in some cases, and antirheumatic agents may be beneficial in chronic reactive arthritis. Antibiotics are recommended if the infection is identified.

The overall prognosis for reactive arthritis is good even in severe cases, but full recovery does not always occur. Many people will experience some form of persisting symptoms that can lead to chronic disability.

Recurrence is possible, and a chronic form of this condition can develop, characterized by recurring arthritis that is accompanied by tendinitis or tenosynovitis. Sacroiliitis and spondylitis may not resolve but may persist, with ongoing pain and stiffness of the neck and back.³⁵⁸

SPECIAL IMPLICATIONS FOR THE THERAPIST 27-14

Reactive Arthritis

PREFERRED PRACTICE PATTERNS

See *Special Implications for the Therapist* box in the section on *Rheumatoid Arthritis*.

The relationship of infections of the gastrointestinal or genitourinary system to the joint is well documented (see the section on Arthritis and Inflammatory Intestinal Diseases in Chapter 16), so that anyone with new onset of joint involvement must be medically evaluated for an underlying bacterial or infectious cause.

Past medical history may reveal a recent infectious process, use of antibiotics, presence of a sexually transmitted disease, or bowel disease to alert the physician. The presence of joint involvement accompanied by (or alternating with) gastrointestinal signs and symptoms such as diarrhea, abdominal pain or bloating, constitutional symptoms (e.g., fever, night sweats), or positive iliopsoas or obturator sign (see Figs. 16-14 and 16-15) must be reported to the physician.

Anyone taking NSAIDs for reactive arthritis must take them as prescribed and not just for analgesia or on occasion. The therapist can help educate affected individuals that a stoic attitude of enduring the pain and restricted mobility with a refusal to "take pills" will result in less optimal and delayed recovery.

Physical therapy intervention is very valuable during convalescence to regain full motion, strength, and function. Temporary splinting may be advised in the most painful cases, but muscle atrophy can be rapid, and therefore immobilization should be minimized.³⁵⁸

If new symptoms develop or the person does not respond to therapy, medical evaluation is advised; modification of medications may be needed.

Reiter Syndrome

Overview. Reiter syndrome is one of the most common examples of reactive arthritis. Reiter syndrome usually follows venereal disease or an episode of bacillary dysentery (enteric infection) and is associated with typical extraarticular manifestations.

The prevalence and incidence of Reiter syndrome are difficult to establish because of (1) the lack of consensus regarding diagnostic criteria, (2) the nomadic nature of the young target population, (3) the underreporting of venereal disease, and (4) the asymptomatic or milder course in affected women.

Etiologic and Risk Factors. The most common microbial pathogens are *Shigella*, *Salmonella*, *Yersinia*, *Campylobacter*, and *Chlamydia* species. Age, gender, and medical history are important risk factors associated with Reiter syndrome. The peak onset of this disorder occurs during the third decade of life, although children and older adults can also develop this disease.

Males are more commonly affected than females but not to the extent once thought. The incidence in women is potentially underestimated because their clinical manifestations are less severe than men's and women are more prone to occult genitourinary disease, leading to misdiagnosis.

A history of infection, especially venereal or dysenteric, is associated with increased risk of developing this condition. Men and women are equally affected by enteric infections. Reiter syndrome is the most common form of reactive arthritis observed in HIV-infected adults and appears to be more strongly associated with male homosexuality than with injection drug use or other risky behaviors.⁴⁴⁸

Pathogenesis. Reiter syndrome is primarily marked by inflammatory synovitis and inflammatory erosion at the insertion sites of ligaments and tendons (enthesis). Heterotopic bone formation can occur at these sites. Synovial findings include edema, cellular invasion (lymphocytes, neutrophils, plasma cells), and vascular changes. Extensive pannus formation is rare, unlike in RA.

Clinical Manifestations. The triad of symptoms classically associated with Reiter syndrome includes urethritis, conjunctivitis, and arthritis. The urethritis and conjunctivitis often occur early in the disease. Other ocular manifestations include uveitis and keratitis (fungal infection of the cornea).

Three musculoskeletal manifestations are acute inflammatory arthritis, inflammatory back pain, and enthesitis. Only about one third of individuals affected by Reiter syndrome have all three. As discussed in the previous section, the arthritis is usually asymmetric, is often acute, and typically involves joints of the lower extremity, including the knees, ankles, and first metatarsophalangeal joint. Isolated hand joints can be involved. Although most of the symptoms and signs disappear within days or weeks, the arthritis may last for months or years.

Extraarticular manifestations are as previously mentioned for reactive arthritis. The skin lesions may be indistinguishable from those of psoriasis. Low back pain is also a common complaint. The arthritis can progress and spread to the spine and even to the upper extremities.

MEDICAL MANAGEMENT

DIAGNOSIS. The diagnosis of Reiter syndrome may require months to establish, because the various manifestations can occur at different times. The combination of peripheral arthritis with urethritis lasting longer than 1 month is necessary before the diagnosis can be confirmed.

Laboratory tests typically reveal an aggressive inflammatory process. Elevated ESR and C-reactive protein are detected, and thrombocytosis and leukocytosis are common findings. Urine samples, genital swabs, and stool cultures are useful laboratory tests for identifying the triggering infection.

Up to 70% of those with established Reiter syndrome may have radiographic abnormalities, including (1) asymmetric involvement of the lower extremity diarthroses, amphiarthroses, symphyses, and entheses; (2) ill-defined bony erosions with adjacent bony proliferation; and (3) paravertebral ossification.

TREATMENT AND PROGNOSIS. Although Reiter syndrome is precipitated by an infection, there is no evidence that antibiotic therapy changes the course of the disorder. Treatment in general is largely symptomatic, with NSAIDs being the primary intervention.

If the arthritis persists, joint protection and maintenance of function become important. Immobilization and inactivity are usually discouraged, whereas range-of-motion and stretching exercises are emphasized. TNF- α antagonists may improve the outcome, but no controlled trials have been performed. Typically the arthritis resolves in 3 to 12 months but can recur. Chronic articular or spinal disease affects 30% of the population affected; severe disability occurs in less than 15% of those afflicted.³

SPECIAL IMPLICATIONS FOR THE THERAPIST 27-15

Reiter Syndrome

PREFERRED PRACTICE PATTERNS

See Special Implications for the Therapist box in the section on *Rheumatoid Arthritis*.

Questions related to the presence and treatment of infection, current and past, should be asked during the history taking. Also inquire into the person's general health, both current and before the onset of the presenting pain complaints. Typically, the onset of joint pain associated with concurrent systemic complaints would raise suspicion.

New-onset inflammatory joint disease with a history of recent enteric or venereal infection or new sexual contact strongly suggests a systemic origin of symptoms. Reiter syndrome is one condition in which past medical history and general health status may provide the most important information.

If the client is undiagnosed or has not yet been seen by a physician, medical evaluation is required. See also the previous section on Reactive Arthritis, including the Special Implications for the Therapist box.

Gout

Overview. Gout represents a heterogeneous group of metabolic disorders marked by an elevated level of serum uric acid and the deposition of urate crystals in the joints, soft tissues, and kidneys. Gout is the most common crystalopathy in the United States.

Hyperuricemia and gout are generally classified into one of three groups. *Primary hyperuricemia* is an inherited disorder of uric acid metabolism. *Secondary hyperuricemia* occurs as a result of some other metabolic problem, such as glucose-6-phosphatase dehydrogenase (G6PD) deficiency, reduced renal function (from any number of causes), certain medications that block uric acid excretion, or neoplasms. The third category, *idiopathic hyperuricemia*, encompasses conditions that do not fit into either of the other categories.

Although gout is a metabolic disorder and could be presented in Chapter 24 as such, it is so predominantly viewed as a form of arthritis because of its clinical presentation (gout can be manifested as a joint disorder characterized by acute or chronic arthritis) that it is included here instead. Crystals other than uric acid crystals can also form inside joints, such as occurs in a condition called *pseudogout* when calcium pyrophosphate dihydrate crystals are present.

The presence of calcium pyrophosphate dihydrate crystals in the synovial fluid can cause symptoms identical to those of acute gout. Unlike gout, however, calcium pyrophosphate dihydrate most often affects the knees of older women and may have polyarticular involvement. Pseudogout, also known as chondrocalcinosis, is associated with a number of metabolic disorders, such as hypothyroidism, hemochromatosis, hyperparathyroidism, and diabetes mellitus.

Incidence. Primary gout is predominantly associated with middle-aged men, with a peak incidence during the fifth decade of life. It is the most common inflammatory disease in men older than 30 years, affecting 2.1 million people in the United States, generally becoming symptomatic after a period of hyperuricemia lasting 10 to 20 years.³⁴⁸

Gout is rare in children, and less than 10% of the cases occur in women. Most women with gout are 15 years or more postmenopausal (later for women taking hormone replacement therapy; a few years of estrogen deficiency are necessary before gout becomes evident in this population).²¹⁰

Risk Factors and Etiologic Factors. A family history of gout increases the risk of developing the disorder. The prevalence of gout increases with increasing serum urate concentration and age; with the aging of the American population, decreased renal function is becoming more prevalent, accompanied by a rise in the number of cases of gout.

Secondary hyperuricemia (gout) can be a result of urate overproduction or decreased urinary excretion of uric acid. People at risk for urate overproduction are those with a history of leukemia, lymphoma, psoriasis, or hemolytic disorders and those receiving chemotherapy for cancer.

Heavy alcohol consumption, obesity, fasting, medications (e.g., thiazide diuretics, levodopa, salicylates), renal

insufficiency, hypertension, hypothyroidism, and hyperparathyroidism can all lead to decreased excretion of uric acid. Among the associated factors, age, duration of hyperuricemia, genetic predisposition, heavy alcohol consumption, obesity, thiazide drugs, and lead toxicity contribute the most to the conversion from asymptomatic hyperuricemia to acute gouty arthritis.⁴⁶⁹

A diet rich in purines (nitrogen-containing compounds found in foods such as shellfish, trout, sardines, anchovies, meat [especially organ meats], asparagus, beans, peas, spinach) can increase the risk of gout or make gout attacks more severe. Conversely, there is a lower prevalence of gout in vegetarians.²¹⁰

In many cases of primary gout the specific biochemical defect responsible for the hyperuricemia is unknown. A majority of cases probably result from an unexplained impairment in uric acid excretion by the kidneys. This impairment could result from decreased renal filtration, increased reabsorption, or decreased urate excretion by the renal tubules.

Pathogenesis. Uric acid is a substance that normally forms when the body breaks down cellular waste products called *purines*. In healthy people, uric acid dissolves in the blood, passes through the kidneys, and is then excreted through the urine. If the body produces more uric acid than the kidneys can process or if the kidneys are unable to handle normal levels of uric acid, then the acid level in the blood rises.

When the uric acid in the blood reaches high levels, it may precipitate out and accumulate in body tissues, forming supersaturated body fluids, including in the joints and kidneys. These crystals frequently collect on articular cartilage, epiphyseal bone, and periarticular structures. The crystal aggregates trigger an inflammatory response, resulting in local tissue necrosis and a proliferation of fibrous tissue secondary to an inflammatory foreign-body reaction.

Clinical Manifestations. The disease occurs in four stages: asymptomatic hyperuricemia (defined as serum urate of more than 7 mg/dl), acute gouty arthritis, intercritical gout, and chronic tophaceous gout.³⁵⁰ Many people with elevated uric acid levels for prolonged period of time never develop signs or symptoms.

The most common clinical presentation is the acute, monoarticular, inflammatory arthritis manifested by exquisite joint pain, occurring suddenly at night. Although the first metatarsophalangeal joint (i.e., the big toe) is a common site of pain, the ankle, instep, knee, wrist, elbow (olecranon bursa), and fingers can all be the site of the initial attack (Fig. 27-22). Besides local, intense pain of quick onset, erythema, warmth, and extreme tenderness and hypersensitivity are typically present. Chills, fever, and tachycardia may accompany the joint complaints.

After recovering from the initial episode the person enters an asymptomatic phase called the *intercritical period*. This period can last months or years despite persistent hyperuricemia and synovial fluid that contains monosodium urate crystals.³⁵⁰

The gouty attacks return suddenly with increasing frequency and severity and often in different joints. These attacks may be precipitated by trauma, surgery, alcohol consumption, or overindulgence in foods with high

purine content. The arthritis can enter the chronic phase up to a decade after the initial attack, characterized by joint damage, functional loss, and disability. Deposits of monosodium urate crystals in soft tissue (tophi) and bone abnormalities are the hallmarks of chronic disease (Fig. 27-23).⁴⁶⁹ Tophi can be located in tendons, ligaments, cartilage, subchondral bone, bursae, synovium, and subcutaneous tissue around the joints. Common sites of these hard, sometimes ulcerated masses that extrude chalky material include the helix of the ear, forearm, knee, and foot.³⁵⁰

MEDICAL MANAGEMENT

DIAGNOSIS. Often termed "the great imitator," gout may masquerade as septic arthritis, RA, or neoplasm. The diagnosis can be delayed for weeks or months. A definitive diagnosis of gout is made when monosodium urate crystals (tophi) are found in synovial fluid, connective tissue, or articular cartilage.

Serum uric acid levels are elevated in approximately 10% of the affected population (more than 7 mg/dl); the presence of hyperuricemia alone does not equal a diagnosis of gout, nor does a normal serum level exclude its presence. The diagnosis is made most often on the basis of the triad of acute monoarticular arthritis, hyperuricemia, and prompt response to drug therapy.³⁵⁰

Bone abnormalities seen on imaging studies (e.g., calcification, overhanging edges of bone erosions with sclerotic margins but with normal bone density) may be present in a small number of affected individuals. These are usually late findings in the disease process, occurring most often in the chronic phase.

TREATMENT AND PROGNOSIS. The goals of intervention are twofold: (1) to end acute attacks and prevent recurrent attacks and (2) to correct the hyperuricemia. NSAIDs are effective in treating the pain and inflammation of an acute attack. Occasionally intraarticular injection of corticosteroids is used to manage acute attacks. Allopurinol can prevent or lessen future gout attacks by slowing the rate at which the body makes uric acid in cases of excess uric acid production.

Other medications can be used to lower uric acid levels in the blood by increasing the amount of uric acid passed in the urine. These pharmacologic agents must be taken on a continuous basis to maintain a lower concentration of uric acid in the blood. Colchicine is another medication given during the acute phase but is less commonly used now because of its narrow therapeutic range and numerous side effects. Involved joints should also be rested, elevated, and protected (e.g., crutches, foot cradle, assistive devices, orthotics, proper shoe wear).

Once the acute attack has been relieved, the hyperuricemia may be treated, especially in the case of recurrent attacks of acute gouty arthritis or chronic gout. This requires lifelong management, and compliance is absolutely necessary. Dietary changes, weight loss, and moderation of alcohol intake are all important. Controlling the hyperuricemia is the key to preventing this disease from becoming chronic and disabling.³⁴⁸



Figure 27-22

Tophaceous gout. **A** to **C**, Chronic gouty arthritis with tophaceous destruction of bone and joints. **D**, Tophaceous deposits in the digital pad of a 28-year-old man with systemic lupus erythematosus. **E**, Tophaceous enlargement of the great toe in a 44-year-old man with a 4-year history of recurrent gouty arthritis. (From Goldman L: Cecil textbook of medicine, ed 22, Philadelphia, 2004, Saunders.)

SPECIAL IMPLICATIONS FOR THE THERAPIST 27-16

Gout

PREFERRED PRACTICE PATTERN

- 4E:** Impaired Joint Mobility, Motor Function, Muscle Performance, and Range of Motion Associated with Localized Inflammation

An onset of severe joint pain with a swollen, hot joint should always concern the therapist. Gout, infection, and hemarthrosis are all conditions that could

account for this clinical scenario. Gout may be associated with fever and malaise, making it difficult to distinguish clinically from a septic joint.

A septic joint is an orthopedic emergency so anytime a red, hot painful joint is observed without prior medical diagnosis, immediate medical evaluation is necessary. Quick diagnosis and initiation of intervention are necessary to control or prevent damage to the joint structures.



Figure 27-23

Tophus, a chalky deposit of sodium urate present in the Achilles tendon and foot, occurs in cardiac transplant recipients who have an associated history of gout. These tophi form most often around the joints in cartilage, bone, bursae, and subcutaneous tissue, producing a chronic foreign-body inflammatory response. Tophi are not clinically significant for the therapist but indicate an underlying condition that requires medical attention. (From Howe S, Edwards NL: Controlling hyperuricemia and gout in cardiac transplant recipients, *Musculoskelet Med* 12:15-24, 1995.)

Neuroarthropathy

Neuroarthropathy, or neuropathic arthropathy, is an articular abnormality related to neurologic deficits, regardless of the nature of the primary disease. Other terms applied to this disorder are Charcot's joint, neurotropic or neuropathic joint disease, and neuropathic osteoarthritis. Many underlying diseases or conditions can cause neuropathy, such as syphilis, syringomyelia, meningomyelocoele, injury or trauma, multiple sclerosis, congenital vascular anomalies, diabetes mellitus, alcoholism, amyloidosis, infection (e.g., tuberculosis, leprosy), pernicious anemia, and intraarticular or systemic administration of corticosteroids.¹⁵³ See individual discussion of each condition.

Early joint changes as seen on imaging studies may look very similar to those of OA. Advanced neuropathy is more clearly defined, with enlarging and persistent effusion and minimal subluxation, fracture, or fragmentation. Microfractures can progress quickly into gross fragmentation, and the joint may appear to deteriorate quickly over a period of days to weeks.

Malalignment with angular deformity, subluxation, or dislocation leads to increased stress on the articular bone, contributing to sclerosis and fractures. Fracture lines can originate in the subchondral region and extend in an extraarticular direction. Management with arthrodesis or arthroplasty is often unsuccessful. More specific intervention approaches are discussed with each individual underlying condition.

BONE

Fracture

Overview

A fracture is any defect in the continuity of a bone, ranging from a small crack to a complex fracture with multiple segments. Fractures can be classified into four general categories: (1) fracture by sudden impact (traumatic), (2) stress or fatigue fracture, (3) insufficiency fracture, and (4) pathologic fracture.

A *stress* or *fatigue* fracture, sometimes referred to as a stress reaction or bone stress injury, is defined as a partial break (reaction) or complete break (fracture) in the bone caused by the bone's inability to withstand stress applied in a rhythmic, repeated, microtraumatic fashion. More simply stated, a fatigue fracture occurs if normal bone is exposed to repeated abnormal stress, and an insufficiency fracture occurs if normal stress is applied to abnormal bone.

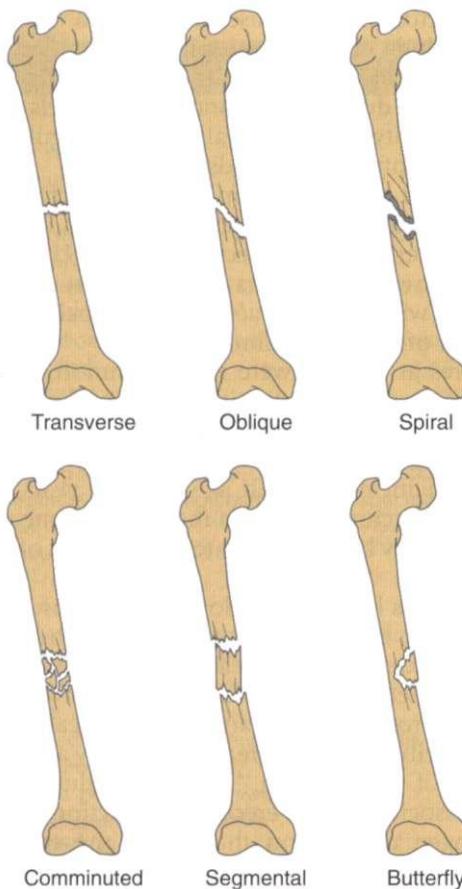
These types of overuse stress or fatigue fractures are most common in track and field athletes, distance runners, and soldiers in training. Most occur in the lower extremity and affect the tibial shaft and metatarsal bones, but they can also occur at the pubic ramus, femoral neck, or fibula; an increasing number of stress fractures have been reported in the knee (tibial plateau, proximal tibial shaft, femoral condyles).^{374,427}

The two kinds of stress fractures are compressive and distractive. Compressive stress fractures occur as a result of forceful heel strike during prolonged marching or running. Distractive stress reactions occur as a result of muscle pull and can become more serious if displacement occurs.

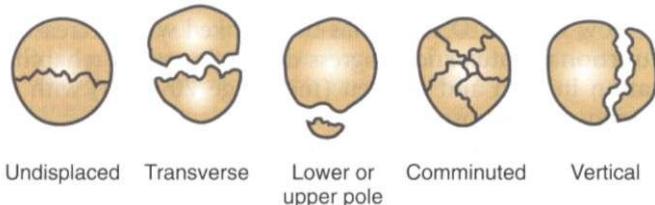
Insufficiency fractures (sometimes referred to as insufficiency stress fractures) result from a normal stress or force acting on bone that has deficient elastic resistance or has been weakened by decreased mineralization. Reduced bone integrity can result from many factors but occurs most commonly from the effects of radiation, postmenopausal or corticosteroid-induced osteoporosis, or other underlying metabolic bone disease (e.g., hyperparathyroidism, osteomalacia, rickets, osteodystrophy). Insufficiency fractures arise insidiously or as a result of minor trauma. It has been proposed that weight bearing alone can be enough "trauma" to transmit a traumatic force to the compromised spine.²⁷⁶

Pathologic fracture is a term used to describe a fracture that occurs in bone rendered abnormally fragile by neoplastic or other disease conditions. Insufficiency fractures can be thought of as a subset of pathologic fractures, occurring in bones with structural alterations due to osteopenia, osteoporosis, or disorders of calcium metabolism.

Fractures can also be further classified as *displaced* or *nondisplaced*, depending on whether the bone has moved on either side of the fracture, and as *open* (also called compound) or *closed*, depending on whether the skin is breached or not. Figs. 27-24 and 27-25 show the different types of fractures (Box 27-16). Displaced, open fractures are more likely to be unstable. Compressive or shear forces can cause stable fractures to shift, becoming

**Figure 27-24**

Classification of fractures. A complete fracture extends through the entire bone; a greenstick fracture does not. A greenstick fracture often has to be completed before effective healing occurs. Other incomplete fractures may be called torus (or buckle), crack, or hairline fractures. In a transverse fracture, the fracture line is at a right angle to the long axis of the bone; this fracture is usually produced by shearing force. An oblique or spiral fracture occurs following a twisting or torsional force; fragments displace easily in the oblique fracture, whereas nonunion rarely occurs in a spiral fracture because of the wide area of surface contact. A fracture is comminuted if the bone is broken into more than two fragments and segmental if a fragment of the free bone is present between the main fragments. The separation of a wedge-shaped piece of bone is called a butterfly fracture. See Box 27-16 for other types of fractures and their definitions.

**Figure 27-25**

Types of patella fracture. Patella fractures are classified as transverse, stellate, or vertical. These three categories can be further divided into displaced and undisplaced. The arterial blood supply to the patella is derived from two systems of vessels from branches of the geniculate arteries. These two systems supply the middle third and apex of the patella. In cases of displaced transverse fractures, the proximal blood supply may be compromised, leading to avascular necrosis of the proximal segment. [From Shankman G: *Fundamental orthopedic management for the physical therapist assistant*, St Louis, 1997, Mosby.]

Box 27-16**TYPES AND DEFINITIONS OF SOME FRACTURES**

- **Colles' fracture:** fracture of the distal radius and ulnar styloid in which the lower fragment is displaced posteriorly, usually from a fall on an outstretched hand
- **Galeazzi's fracture:** fracture of the middle third and distal third of the radius accompanied by dislocation of the distal radioulnar joint at the wrist
- **Jones fracture:** fracture of the base of the fifth metatarsal
- **Maisonneuve fracture:** tear of the anterior and interosseous tibiofibular ligaments and a fracture (usually oblique) of the fibula 3 or 4 inches above the ankle mortise
- **Monteggia's fracture:** fracture of the proximal third of the ulna with dislocation of the radial head
- **Nightstick fracture:** fracture of the ulna alone, usually midshaft
- **Piedmont fracture:** fracture of the radial shaft (rare)
- **Pott's fracture:** oblique fracture of the lateral malleolus and transverse fracture of the medial malleolus; the talus may be displaced posteriorly (avulsion)
- **Torus fracture:** sometimes referred to as a "buckle" fracture; a fracture in which there is localized cortical expansion but little or no displacement; most common in young children when a compression fracture may merely "buckle" the thin cortex surrounding the cancellous bone

unstable. Unstable fractures are more likely to require surgery to stabilize them.

An *epiphyseal* fracture occurs in the growth centers of children and adolescents, located in the long bones. Growth can be arrested or altered in this type of fracture, and immediate intervention is required. An *articular* fracture occurs on or near a joint and is described by the course of the fracture line (e.g., T or Y shaped, transcondylar, supracondylar, intercondylar).

Pelvic and *sacral* fractures were traditionally classified according to stability, but with improvements in orthopedic procedures, these types of fractures are more often classified based on causative force vectors; this system is more appropriate as they direct surgical fixation.⁵⁴⁹ The mechanisms of force vectors from the injury include anteroposterior compression, lateral compression, vertical shear, and combined/mixed mechanisms.

Pelvic and sacral fractures may include a single pubic or ischial ramus, ipsilateral pubic and ischial rami, pelvic wing of the ilium (Duverney's fracture), or fracture of the sacrum or coccyx. If the injury only results in a slight widening of the symphysis pubis or the anterior sacroiliac joint and the pelvic ligaments are intact, the fracture is considered stable. Unstable pelvic fractures can cause rotational instability, vertical instability, or both.

Vertically unstable pelvic fractures occur when a vertical force is exerted on the pelvis such as occurs when an individual falls from a height onto extended legs or is struck from above by a falling object. Disruption of the ligaments (posterior sacroiliac, sacrospinous, sacrotuberous) is usually complete, and the hemipelvis is displaced anteriorly and posteriorly through the symphysis pubis.

Sacral fractures occur from stress transmitted through the pelvic ring to the sacrum. Lateral compression fractures are seen most often in motor vehicle accidents.

Direct stress to the sacrum from a high fall onto the buttocks occurs less often and produces a transverse, rather than vertical fracture.²⁴⁹

Vertebral compression fracture (VCF) is one of the most common osteoporosis-related fragility fractures. VCFs often occur with only minor trauma. Only 20% to 25% of people who sustain a VCF develop symptoms severe enough to seek medical attention.²⁸⁶ VCFs are classified as wedge, crush, or biconcave according to their morphologic appearance.²²² The greater prevalence of wedge fractures may be related to DDD, a condition that causes normal intradiskal pressure to shift and concentrate load to the peripheral aspects of the vertebral body.²⁶⁵

Etiology

Bone mass is known to reach its maximum size and density (peak bone mass) by the time an adult reaches age 30. Women have a tendency to lose bone mass sooner than men, often beginning in their late thirties during the perimenopausal years. Bone loss is accelerated for women during and after menopause; men are more likely to experience bone loss in their mid to late sixties.

Cancellous bones have a greater percentage of trabecular bone (e.g., spine, ribs, jaw, wrist) and are more porous with a greater surface area and are therefore more susceptible to bone loss and fractures. But low bone mass does not always result in fractures. Scientists are actively studying the differences between bones that fracture and those that do not, especially among people of different ethnic backgrounds. It appears that differences in bone structure and repair capability are two major factors to explain differences in fracture rates.

Risk Factors and Incidence

By far the most common traumatic fractures are those associated with sudden impact, such as occurs with assault, abuse, traumatic falls, or motor vehicle accidents.

Motor vehicle accidents involve fractures of the skull, nasal bone, and mandible most often; high-velocity injuries including automobile or motorcycle accidents often result in open fractures of the lower extremity. In the general population, radius and/or ulna fractures comprise the largest proportion of upper extremity fractures. The most affected age group is children ages 5 to 14 years as a result of accidental falls at home.⁸⁰

Age is an important risk factor for fractures. The rate of hip fracture increases at age 50, doubling every 5 to 6 years. Increasing age and low BMD are the two most important independent risk factors for an initial vertebral or nonvertebral fracture.⁴⁶

Decreased BMD associated with osteoporosis accounts for the largest number of fractures among the older adult population (see the section on Osteoporosis in Chapter 24). In fact, a fracture may be the first sign of an underlying diagnosis of osteoporosis, and a serious fracture is a risk factor itself for future fractures in high-risk groups. There are an estimated 1.5 million osteoporosis-related fragility fractures in the United States each year.

VCFs are the most common osteoporosis-related fractures, accounting for approximately 700,000 injuries. The incidence increases with age and with decreasing bone

density. Factors that increase the risk of a first vertebral fracture include previous nonspine fracture, low BMD at all sites, low body mass index, current smoking, low milk consumption during pregnancy, low levels of daily physical activity, previous fall(s), and regular use of aluminum-containing antacids.³⁶⁹

A woman's risk of developing a hip fracture is equal to her combined risk of developing breast, uterine, and ovarian cancer.³⁶⁵ Data collected from the U.S. Medicare population over age 65 years revealed a pattern of rapidly rising rates with age for fractures of the pelvis, hip, and other parts of the femur among women.

Fractures at the hip were most common, accounting for 3.8% of the fractures identified. The proximal humerus, distal radius/ulna, and ankle also were common fracture sites. Fractures distal to the elbow or knee had only small increases in incidence with age over 65 years. Women have higher fracture rates than men of the same race, and whites generally have higher rates than blacks of the same gender.³²

Men are less likely to develop osteoporosis and subsequent fracture, but they are not immune to this condition and are frequently undertreated for osteoporosis even after a fracture.²⁴⁶ Epidemiologic studies have confirmed that osteoporosis in men is an increasing health problem, possibly attributable to increased longevity and increased awareness of the problem.¹⁴³ Bone loss due to hypogonadism associated with erectile dysfunction or induced by androgen deprivation therapy in the treatment of prostate cancer increases the risk of osteoporosis and thus fracture for some men.

Fracture risk has been consistently associated with a history of falls, including falls to the side, and attributes of bone geometry, such as tallness, hip axis, and femur length.³⁶⁴ The way a person falls, laterally landing directly on the trochanter versus falling backward, is an independent risk factor for hip fractures.^{159,389}

Other risk factors for fracture are listed in Box 27-17; see also Box 27-20. Some risk factors for fracture, such as age, low body mass index, and low levels of physical activity, probably affect fracture incidence through their effects on bone density and propensity to fall and inability to absorb impact.³⁶⁴ Vitamin D deficiency and its link with generalized muscle weakness leading to falls and fractures is likely more prevalent among older adults than previously thought.^{148,523}

Low vitamin D status has been linked with decreased functional status and progress during inpatient rehabilitation in men and women (mean age 70 years) with a variety of diagnoses.²⁴⁷

The long-term use of high-dose proton pump inhibitors (PPIs) such as Prilosec, Protonix, Prevacid, Aciphex, and Nexium used to reduce stomach acid has also been linked with hip fractures. The presumed mechanism is reduced bone density via interference with calcium absorption. A certain amount of acid is needed to absorb most forms of calcium. PPIs may also inhibit another proton pump important in bone remodeling. Until further information is known about this effect, individuals at risk for fractures who are also taking PPIs should talk with their physicians about fracture prevention.⁵⁴⁶

Box 27-17**RISK FACTORS FOR FRACTURES**

- Trauma
 - Motor vehicle accidents
 - Industrial or work-related accidents
 - Assault
 - History of falls; risk factors for falls (see also Boxes 27-19 and 27-20)
 - Overuse (marathon runners, military); sudden changes in training (duration, intensity)
 - Participation in sports, including dance (recreational or competitive)
- Advanced age
- Women: postmenopausal osteoporosis; military: stress fractures
- Men: hypogonadism (erectile dysfunction, prostate cancer)
- Any insufficiency* or fragility fractures, especially vertebral fractures
- Residence in a long-term care facility
- Poor self-rated health
- Low physical function
 - Slow gait speed; gait disorders or movement dysfunction; low levels of physical activity
 - Difficulty in turning while walking; inability to pivot
 - Use of a walking aid (cane, walker)
 - Decreased quadriceps strength (e.g., inability to rise from chair without using arms)
 - Increased postural (body) sway^{422,455†}
 - Impaired cognition, dementia
- Physical attributes
 - Low physical fitness
 - Decreased bone mineral density (BMD)
 - Bone geometry (see text description)
 - Leg length discrepancy
 - Height
 - Low body mass index (BMI); low muscle mass
 - Poor nutrition; eating disorder; vitamin D deficiency
- Alcohol and/or substance use
- Other diseases or conditions
 - Osteoporosis; failure to treat or undertreatment of osteoporosis
 - Osteogenesis imperfecta
 - Osteonecrosis
 - Neoplasm; skeletal metastases; surgical resection for tumor
- Radiation treatment
- High-dose, long-term use of proton pump inhibitors (PPIs)

*Fracture in bones with nontumorous disease (e.g., rheumatoid arthritis, osteoporosis, following radiation) at normal load.²⁵⁶

†Postural sway is a corrective mechanism associated with staying upright and can be used as a measure of balance. Postural sway increases with age (reflecting decreased balance) and with the use of benzodiazepines.⁴²²

Leg length discrepancy may also increase the risk of stress fracture, especially in female athletes. Decreased muscle mass and strength may play a role in the developing stress fractures by absorbing less of the force and distributing or exerting more load to the bone. Good muscle strength may decrease the strain on bone and delay muscle fatigue. Muscle fatigue may cause alterations in running mechanics that could increase ground reaction forces exerted on the bone.^{244,314}

Pathogenesis

The repair or regeneration of bone involves a complex sequence of cellular activities, beginning with acute hematoma formation and early inflammatory response and followed by granulation tissue infiltration, recruitment, proliferation, and differentiation of osteogenic and often chondrogenic cells; matrix formation and mineralization; and eventual remodeling.¹⁵⁴

The process is orchestrated and guided by a series of biologic and mechanical signals. Molecular signaling cascades and nutrition are key factors in the success of bone repair or regeneration. Bone response to injury and the phases of the reparative process are discussed in greater detail in Chapter 6 (see also Fig. 6-21).

When a bone is fractured, its normal blood supply is disrupted. Osteocytes (bone cells) will die from the trauma and the resulting ischemia. Bone macrophages will remove the dead bone cells and the damaged bone. A precursor fibrocartilaginous growth of tissue occurs before the laying down of primary bone, eventually followed by the laying down and remodeling of normal adult bone.

This complex process of fracture healing can be broken down into five stages: (1) hematoma formation, (2) cellular proliferation, (3) callous formation, (4) ossification, and (5) consolidation and remodeling. Some resources describe the phases of bone healing more succinctly as inflammatory, reparative, and remodeling.²⁰²

During the initial 48 to 72 hours after fracture, hematoma formation occurs as clotting factors from the blood initiate the formation of a fibrin meshwork. This meshwork is the framework for the ingrowth of fibroblasts and capillary buds around and between the bony ends.

During the cellular proliferation phase, osteogenic cells proliferate and eventually form a fibrocartilage collar around the fracture site. Eventually the collars and the ends of the bones unite. The cartilage is eventually replaced by bone as osteoblasts continue to move into the site (callous formation and ossification). Finally, the excessive bony callus is resorbed and the bone remodels in response to the mechanical stresses placed on it.

Clinical Manifestations

The primary manifestations of fracture are listed in Box 27-18. Point tenderness over the site of the fracture is usually present, but not all fractures are equally painful. Insufficiency fractures of the spine, pelvis, or sacrum often present with nonspecific low back, groin, or pelvic pain, mimicking other clinical conditions such as local tumor or metastatic disease or disk disease.

Stress Fractures. In the case of stress reactions and fractures, an abrupt increase in the intensity or duration of training (i.e., military trainees, athletes preparing for marathons) is often an additional risk factor.³⁷⁴ Female recruits are at increased risk for pelvic and sacral stress fractures. The generally increased risk of bone stress injuries among females has been explained by anatomic (wide pelvis, coxa vara, genu valgum), hormonal, and nutritional factors.³¹⁴

Box 27-18**CLINICAL MANIFESTATIONS OF FRACTURES**

- Pain and tenderness
- Increased pain on weight bearing
- Edema
- Ecchymosis
- Loss of general function
- Loss of mobility

With many fractures, attempts to move the injured limb will provoke severe pain, but in the presence of a fatigue fracture (stress reaction) active movement is typically painless. Resistive motions or repetitive weight bearing will cause pain, and the area will be exquisitely tender to local palpation. There may be edema observed in the area of the fracture. Clinical manifestations are most severe when the fracture is unstable.

In the presence of a compression fracture of a thoracic vertebra, the initial pain may be sharp and severe, but after a few days it may become dull and achy. The pain may be reproducible on examination with pressure over the spinous process of the involved level. Pain associated with VCFs tends to be postural (i.e., worse with spinal extension or even standing up straight); it can be debilitating enough to confine some older adults to a wheelchair or bed.

Complications. The deformity associated with an extremity fracture is often obvious, but the deformity of a spinal fracture is not always so. For example, a compression fracture of a thoracic vertebral body may result in an anterior wedging of the body but only a mildly accentuated thoracic kyphosis. When thoracic kyphosis does occur, decreased trunk strength and decreased pulmonary function are possible.²⁷⁵

Older adults with VCF are two to three times more likely to die secondary to pulmonary causes (e.g., congestive heart failure, pneumonia) and have an increased risk for hospitalization and mortality.¹¹¹ Urinary retention and gastrointestinal symptoms are also common manifestations in people with VCFs. Neurologic deficits can also occur, but these symptoms usually resolve; less than 5% of affected individuals need surgical decompression.⁴⁵³

Occasionally, in an adolescent or young adult who has not achieved mature bone growth, a persistent but painless prominence may occur 1 to 3 months after a minimally displaced fracture. It is located on the compression side of the fracture within the newly formed subperiosteal bone (intracortical) as a result of encapsulation or calcification of a hematoma. This transient postfracture cyst is benign but must be medically diagnosed as such, since it cannot be distinguished clinically from infection or tumors.⁴⁸⁹

The healing of a fracture can be abnormal in one of several ways. The fracture may heal in the expected amount of time but in an unsatisfactory position with residual bony deformity called *malunion*. The fracture may heal, but this may take considerably longer than the expected time (*delayed union*); or the fracture may fail to

heal (*nonunion*) with resultant formation of either a fibrous union or a false joint (*pseudoarthrosis*).

Loss of blood supply to the fracture fragments may impede healing by preventing adequate revascularization. Motion at the fracture site or an excessively wide gap can also contribute to nonunion. Individuals with nonunion often have pain, heat, and tenderness at the fracture site.

Other complications may include associated soft tissue injury, complications secondary to treatment, infection, skin ulceration, growth disturbances, posttraumatic degenerative arthritis, soft tissue or connective tissue adhesions, arthrodesis, myositis ossificans, osteomyelitis, refracture, nerve injury and neurologic complications, and vascular compromise.⁴³⁰

MEDICAL MANAGEMENT

PREVENTION. Therapists have a key role in the prevention of falls. Education and risk evaluation are two important variables in preventing fractures from occurring (Box 27-19). Combining BMD with fracture assessment (e.g., use of dual x-ray absorptiometry [DXA] to assess vertebral fractures) has a positive impact on lowering repeat fractures.¹⁴²

Studies are under way to determine the most cost-effective strategy for fracture prevention. In the case of hormone replacement therapy, treating those people with low BMD levels (secondary prevention) seems to be more cost effective than general treatment (primary prevention).

High-risk groups can be identified (e.g., long-term care residents) and treated with low-cost interventions (e.g., calcium plus vitamin D or external padded hip protectors). Use of hip protectors (padded, convex plastic shields worn inside specially designed undergarments) to prevent hip fracture for those people at risk has met with mixed results.^{43,216,388,442} Problems such as insufficient supply, discomfort while sleeping, functional incontinence, correct positioning of the shield, and ease of application for anyone who is overweight or obese and/or has arthritis has made the use of hip protectors less than optimal. Instead of relying on hip protectors, older adults should be encouraged to increase bone mass through nutrition and physical activity and take extra care with medications that cause dizziness. The use of hip protectors has been advocated for institutionalized individuals. Cognitive impairment is actually helpful in terms of compliance and positive results.^{379,441}

Fall prevention is important in adults over 60 years of age (see Box 27-19). Further studies comparing different preventive regimens are needed.⁵²⁴ Fracture prevention in the athlete begins with assessment of the athlete's past history, training variables, biomechanical factors, and shoe wear. In the military population, most bone stress injuries occur during the 8-week basic training period; injury-prevention programs to target this group are advised.³¹⁴ The reader is referred to other sources for more specific assessment techniques.^{15,53,54}

DIAGNOSIS. Fractures are often diagnosed by visual inspection and confirmed by plain radiographs. Many VCFs are detected incidentally on chest radiographs.

Box 27-19**PREVENTION OF FALLS**

- Wear low-heeled, closed footwear with rubber soles or good gripping ability; avoid smooth-bottomed shoes or boots. This applies to slippers; wear slippers or shoes when getting out of bed at night.
- Provide adequate lighting for hallways, stairways, bathrooms; use a flashlight outdoors. Wear glasses at night when getting out of bed for any reason.
- Conduct a home safety evaluation. Remove loose cords, slippery throw rugs; repair uneven stairs, steps, sidewalks.
- Avoid oversedation; carefully monitor medications (especially sleep medications, antidepressants) and drink alcohol in moderation (never drink alcohol if taking medications without your physician's approval).
- Provide sturdy handrails on both sides of stairways.
- Provide grab bars on bathroom walls and nonskid strips on mats in tub or shower and beside tub or shower.
- Avoid going outdoors when it is wet, icy, or slippery; wear footwear with good traction or clip-on ice grippers; avoid walking on wet leaves or garden or yard clippings or debris.
- Carry items close to the body and leave one hand free to grasp railings or for balance.
- Know the location of pets before walking through a room or area of the house or apartment; maintain floors free of clutter and small objects.
- Put aside pride and use an appropriate assistive device as recommended by the therapist (e.g., cane, walking stick, walker); walkers equipped with a seat work well for people with limited endurance.
- Encourage a program of physical activity and exercise that is attainable.
- Avoid changing position quickly, such as when getting out of a chair or bed. Stand for a moment to see if you are dizzy so that you can sit down again if necessary. See discussion of postural hypotension for prevention strategies (Chapter 12).
- Keep items on shelves in the kitchen and elsewhere within reach. Do not stand on a chair or stepladder to reach items. Consider the consequences of a fall and broken hip if you are tempted and if you are thinking, "Nothing will happen, I will be fine."

ing pathophysiologic changes associated with stress injuries but is more expensive and is reserved for cases in which other imaging findings are indeterminate.¹⁵⁸

TREATMENT. The medical approach to management of fractures is based on the location of the fracture, assessment of fracture type, need for reduction, presence of instability after reduction, and functional requirements of the affected individual. For example, stress fractures are usually uncomplicated and can be managed by rest and restriction from activity,³⁰⁸ whereas an unstable fracture of any bone may require immediate surgical intervention.

Individual factors such as age, activity level, the person's general health and overall condition, and the presence of any other injuries must also be taken into consideration. The goal of treatment is to promote hemostasis, hemodynamic stability, comfort, and early mobilization to prevent potential complications from immobility (e.g., constipation, deep vein thrombosis, pulmonary embolism, pneumonia). In the case of stress fractures, the initial period of rest is followed by a gradual return to activity. The progression of return to sports is based on symptomatic response to increasing activity.

The presence of osteoporosis complicates the need for immobilization or spinal fusion. Nonoperative treatment for VCFs includes activity modification, bracing, assistive devices, pharmacology (e.g., narcotic analgesics, calcitonin), and physical therapy. Hospital admission and bed rest is required for up to 20% of the population for whom conservative care is not possible or adequate.

The debilitating effects of immobilization and keeping older adults bed bound is well recognized, with increased risks for developing pulmonary complications, pressure ulcers, deep vein thrombosis, and urinary tract infections. And BMD is further reduced by immobility and bed rest, thereby increasing the risk of additional VCFs and other fragility fractures.⁴⁵³

Surgery. Surgical intervention may be required for VCFs, including bone grafts or bone graft substitutes, internal fixation (e.g., metal plating, wiring, screws), traction, or reduction and casting or immobilization. VCFs also may be treated by surgical decompression and fusion, vertebroplasty, and kyphoplasty. Analgesic therapy is effective for most people with VCFs from bone metastases.⁴⁰⁴

Minimally invasive procedures for the management of acute vertebral fracture have been developed. Injection of FDA-approved polymethylmethacrylate bone cement into the fractured vertebra is being used around the United States in procedures known as *vertebroplasty* or *kyphoplasty*.

In kyphoplasty, using a fluoroscope the surgeon locates the spinal fracture, inserts a needle into that vertebra, and inflates a tiny balloon at the tip of the needle, pushing the vertebral body as close to its normal position as possible and leaving a defined cavity that can be filled. Once the collapsed portion of the vertebra has been raised, the balloon is deflated and removed and bone cement is injected through the same needle into the vertebral body. The cement hardens, quickly sealing the fracture. No postsurgery bracing is required.

Fractures can often involve surrounding soft tissue, vascular, and neurologic structures, requiring careful assessment at the time of injury.

In the case of stress reactions (stress fractures) conventional radiographic studies (x-rays) are usually inadequate; often the lag time between manifestation of symptoms and detection of positive radiographic findings ranges from 1 week to several months. Up to 35% of sacral fractures are undetected on plain radiographs; cross-sectional imaging such as CT or MRI may be needed to identify and confirm sacral fractures. MRI is as sensitive as scintigraphy for identifying bone stress injuries of the lower extremities, especially during the early stages of developing injury. CT is the imaging technique of choice to identify pathologic fractures.^{314,540}

Radionuclide bone scanning (scintigraphy) has become a useful imaging study because it can demonstrate subtle changes in bone metabolism long before conventional radiography. MRI is also sensitive for detect-

Reports of acute pain relief have been documented, but the long-term effect of one or more reinforced rigid vertebrae on the risk of fracture of adjacent vertebrae remains unknown at this time.³⁶⁴ Another new technique under investigation is the use of a titanium implant for vertebral replacement. Data are very limited on this intervention modality at this time.²⁵³

A similar technique is being developed for the treatment of a fractured distal radius with calcium phosphate bone cement injected into the trabecular defect of the fracture site. Using a gene transfer vector, this remodelable bone cement allows for earlier removal (at 2 weeks instead of 6 weeks or more) of the cast or splint and early mobilization.

Results have been very encouraging, with better clinical and radiologic results than with conventional treatment.⁴³⁵ In the United States, other researchers are experimenting with the use of this cement on other bones, such as the calcaneus, and for use in cranial reconstruction.^{92,252} Whereas this product is approved only for limited use in the United States, it is currently used on fractures of the lower extremities and hips in Europe.

Rehabilitation and Fractures. With or without surgical intervention, following bone fracture there is usually a period of immobilization (casting or splinting, fracture brace) to remove longitudinal stress. This period allows for the phagocytic removal of necrotic bone tissue and the initial deposition of the fibrocartilaginous callus.

For any type of fracture, management during the peri-fracture period is directed toward blood clot prevention (mechanical and/or pharmacologic), the avoidance of substances that inhibit fracture repair (e.g., nicotine, corticosteroids), and the possible need for supplemental caloric intake. Treatment should be initiated for anyone with osteoporosis, including calcium and vitamin D supplements, oral bisphosphonates, selective estrogen receptor modulators, calcitonin, and teriparatide (see discussion of treatment for osteoporosis in Chapter 24).

Gradually progressive stress will be applied to stimulate fracture callus formation and healing. In the case of pelvic or lower extremity fractures, the timing and extent of mobilization depend on the type of fixation used. For example, if an external fixation is applied for fracture stabilization, mobilization can occur within tolerance of the person's symptoms almost immediately.

Modalities and Fractures. Many studies carried out on the effect of ultrasound waves on fracture healing show that bone heals faster when it responds to applied pressure. Low-intensity (0.1 W/cm²), pulsed ultrasound (2-msec bursts of sine waves of 1.0 MHz [frequency]; duration of 20 minutes daily) is an established therapy for fracture repair.⁵³⁵

In both animal and human trials, such ultrasound has been shown to facilitate fresh fracture repair and initiate healing in fractures with repair defects. However, the mechanism by which ultrasound achieves these outcomes is not clear. One possible mechanism is the direct stimulation of bone formation. Ultrasound has a direct effect on blood flow distribution around a fracture site, resulting in greater callous formation. This increased circulation serves as a principal factor facilitating the acceleration of fracture healing by ultrasound.^{27,486,535}

Ultrasound and electromagnetic stimulation are used most often for fracture healing where physicians anticipate healing problems or where nonunion has already occurred. A relatively new fracture management tool that incorporates the application of a specifically modified diagnostic ultrasound unit to heal fractures with the intention of accelerating repair is available.⁵³⁴ Therapists in some parts of the United States are involved in the use and study of this modality.

Bone Grafting. Bone grafting to enhance bone repair can be applied during the repair stage of bone formation. Autogenous bone grafting takes bone from another part of the body and implants it in the bony defect that requires healing. The graft is most often taken from the iliac crest or fibula and contains all the components needed for bone healing. Donor site pain is a common complaint and the primary reason why some people prefer to use allogenic bone graft material from a donor (bone bank).

Tissue engineering of bone has emerged as a new treatment alternative in bone repair and regeneration. The use of biodegradable plastics has been developed to provide scaffolding for the regrowth of tissue with the potential for healing fractures and repairing bone lost to tumors, osteoporosis, trauma, and other disorders.

Commercially available demineralized bone matrix can be used to enhance bone healing, especially in people with nonunions or after the removal of bone cysts or fibrous lesions. Demineralized bone matrix still retains some of the original trabecular structure, which can function as a scaffold for osteoconduction.¹⁷²

The addition to this scaffolding of growth hormones or other bioactive molecules that enhance bone repair to create a "smart matrix" has the potential of speeding up the healing of fractures and repair of more serious crush injuries or nonunion of bone. Further development of the concept includes gene transfer as a cellular vehicle for delivery of BMP to promote bone formation.^{13,269}

Gene Therapy. Gene therapy involves the introduction of DNA into cells (exogenous or endogenous) in an effort to direct them to overexpress a selected biofactor and thus promote bone repair. Gene transfer may be accomplished in one of several different ways. Cells may be grown in culture and reimplanted into the wound. DNA may be mixed into bone marrow during surgery and then implanted. Or DNA may be injected directly into the wound site. Gene-based strategies are still in the laboratory phase. Introduction into the clinic is expected in the next decade.¹⁵⁴

PROGNOSIS. In general, fractures in children heal in 4 to 6 weeks; in adolescents in 6 to 8 weeks; and in adults in 10 to 18 weeks. This process from fracture to full restoration of the bone will take weeks to months, depending on the type of fracture, location, vascular supply, health, and age of the individual. Nonunion or delayed union is more likely to occur in adults and occurs in up to 10% of all fractures (affecting nearly 500,000 people each year in the United States).⁴⁰⁹

Older adults who have suffered a hip fracture have the highest rate of nonunion complications (15% to 30%). These individuals are almost four times more likely to die