CHAPTER 33

Arthritis

Joints

GENERAL CONSIDERATIONS

A joint is a connection between bones of the skeleton. Joints can be classified as fibrous, cartilaginous, or synovial. Fibrous joints are represented by the sutures of the skull, while an example of a cartilaginous joint is the symphysis pubis; neither of these joints types allows gross motion. Synovial joints, also termed diarthrodial, are the movement units of the skeleton and the main consideration of this chapter.

Synovial joints are composed of the ends of bones, which are covered with hyaline cartilage and encased in a fibrous and ligamentous capsule that is lined with synovium. Hyaline cartilage both functions as a shock absorber and provides a smooth gliding surface for motion. The synovium begins at the margins of the articular cartilage but normally does not overlie the cartilage. The ligaments and capsule, along with the muscles and tendons of the area, provide stability for the joint. The synovium secretes synovial fluid, which lubricates and nourishes the articular cartilage.

Every joint contains a small amount of synovial fluid, which is a combination of a dialysate of plasma and hyaluronic acid that is secreted by the synoviocytes. The lubricating qualities of the fluid come from the mixture of viscid hyaluronic acid and water. Coagulation proteins are not present in normal synovial fluid, and consequently it does not clot.[37] The combination of synovial fluid over articular cartilage produces a remarkably friction-free gliding surface. This is especially important because the articular surfaces are not a perfect fit, and the contact areas change in dimension as motion occurs. In adults, all of the cartilage nutrition is derived from synovial fluid, while in children there is a smaller contribution from the underlying bone.[22,132,187]

Symptoms arising from a joint are ordinarily associated with motion and with the stresses of standing and walking. Pain is an outstanding feature, as joints have numerous nerve endings in the synovial membrane and capsule. Oversecretion of synovial fluid produces distention of the joint capsule. Excess synovial fluid can be easily seen and palpated in superficial joints. In later stages of inflammation, proliferation and general thickening of the synovium occur and can be detected by careful palpation. With joint inflammation, active and passive motion of the joint are limited.

Muscle spasm, a visceromotor reflex response to painful stimuli, usually accompanies joint inflammation. Spasm is more predominant in the flexor muscle groups, producing a flexion deformity. Atrophy of muscles that are antagonists to those in spasm occurs early and lasts for the duration of the joint disease, often persisting after the spasm has resolved. If a weightbearing joint is affected, the child will walk with an antalgic limp.

Ultrasonography depicts fluid in the joint and distention of the joint capsule.[218] Radiographs show distention of the capsule, and magnetic resonance imaging (MRI) can delineate the synovial hypertrophy or other soft tissue pathology. Later changes include narrowing of the articular space from erosion of articular cartilage. Subjacent bone responds with sclerosis and osteophyte new bone formation, while from the cartilage loose bodies may form. The final stage is exposure of cancellous bone with "bone on bone" and fibrous ankylosis of the joint.

JOINT FLUID ANALYSIS

Examination of synovial fluid is an important tool in diagnosing joint disease. Aspiration of a joint should be performed under rigidly aseptic conditions. The area should be surgically prepared and draped to ensure sterility. The examiner wears a mask and gloves, and assistants should be available to control the apprehensive child.

The anatomic approach to aspiration of various joints is illustrated in Figure 33–1. It is best to use an 18-gauge lumbar puncture needle with a stylet inside. A local anesthetic, such 1% lidocaine (Xylocaine) or procaine, is used.

Gross Appearance. The gross appearance of the joint fluid often yields important information. Normal synovial fluid is clear and colorless or straw-colored. In the course of aspiration, blood vessels may be punctured, and sanguineous streaks may be found in the joint fluid. This uneven distribution of blood in the syringe is distinguishable from the appearance of the fluid aspirated in acute traumatic hemarthrosis, which is entirely sanguineous. In chronic hemarthrosis the fluid may be xanthochromatic. With inflammation the joint fluid becomes turbid. The greater the degree of inflammation, the more turbid is the synovial effusion. The fluid from a pyogenic joint has the creamy or grayish

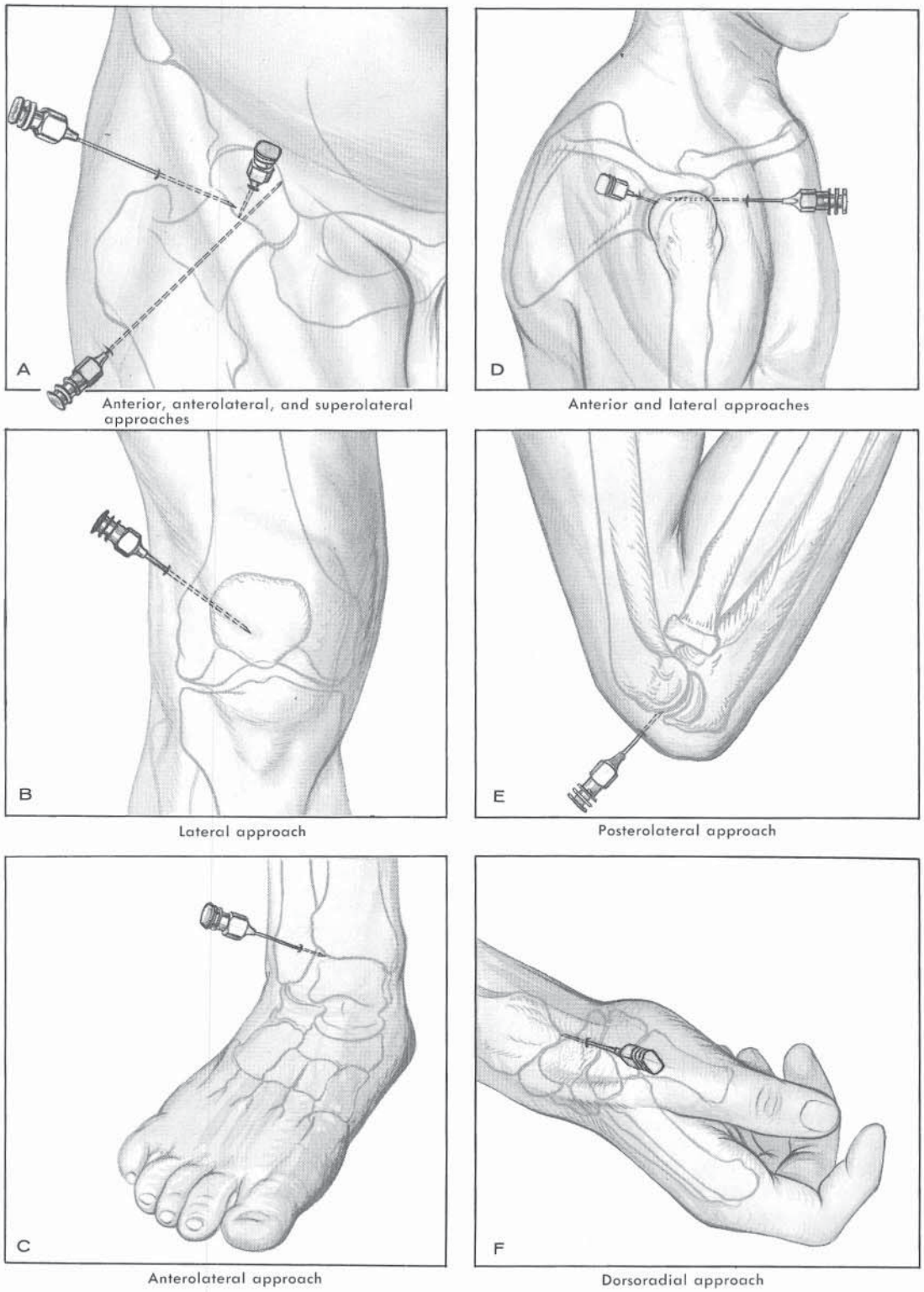


FIGURE 33–1 Routes of aspiration of joints. **A,** Anterior, anterolateral, and superolateral approaches. **B,** Lateral approach. **C,** Anterolateral approach. **D,** Anterior and lateral approaches. **E,** Posterolateral approaches. **F,** Dorsoradial approach.

appearance of frank pus. In rheumatoid arthritis the fluid may be clear in the early stages, but as inflammation increases, it becomes turbid in appearance. The fluid in acute gout is milky white because of its urate content. In degenerative arthritis, the joint fluid is almost normal in appearance.

Viscosity and Mucin Clot. The concentration and quality of hyaluronate are altered in inflammatory states, with resultant changes in the physical characteristics of the synovial fluid. The fluid should be examined at the time of aspiration, with the examiner noting its viscosity by "pulling" it between the gloved fingers and by letting it drop from a syringe. Normal fluid should form a string at least 1 inch in length. Mucin quality can be tested by adding the fluid to either distilled water or 5% acetic acid. The clot that forms is graded as normal; fair, characterized by some loss of clot continuity; poor, in which there are small friable masses of clot in a cloudy solution; and very poor, characterized by a few flecks in cloudy solution.

Microscopic Examination. Synovial fluid may be examined for cellular elements, intracellular inclusions, and crystals. Glucose and protein levels are also determined. Normal synovial fluid has less than 300 white blood cells (WBCs)/mm^3, and the cell types may be determined. Normal fluid has less than 25 percent polymorphonuclear leukocytes. Specialized microscopy may reveal cytoplasmic inclusions of immunoglobulin, rheumatoid factor, and complement components. Crystals may be seen with polarized light microscopy.

Other Examinations. It is important to determine glucose levels because the difference between serum and synovial fluid glucose levels increases with more severe inflammation. Septic arthritis lowers the joint fluid glucose level more than other conditions, and this finding is of diagnostic importance. The normal protein content of synovial fluid is about 30 percent that of serum. The total protein level is usually 1.8 g/dL, 70 percent albumin and 7 percent α_2-globulin. Normal values and common abnormalities are listed in Table 33–1. With inflammation, the permeability of the synovium to plasma increases and the protein content of the joint approaches that of serum. In addition, clotting factors enter the joint, and the inflammatory fluid forms clots (Table 33–1).

Juvenile Rheumatoid Arthritis

Juvenile rheumatoid arthritis (JRA) is the name applied to a group of disorders characterized by chronic arthritis of one or more joints with a duration of at least 6 weeks. The majority of cases are pauciarticular, with several joints involved, and are often accompanied by uveitis. The polyarticular form is sometimes associated with involvement of other systems with such manifestations as lymphadenopathy, splenomegaly, and fever. Systemic onset JRA is a severe multisystem disease with arthritis as an accompanying manifestation. In the past, the term Still's disease was used to identify these disorders, after G. F. Still, who published a description of 22 cases in 1897.[192] An earlier description by Cornil in 1864 predated Still's paper but lacked its completeness.[43]

DEFINITION AND CLASSIFICATION

The American College of Rheumatology has established five criteria for the diagnosis of JRA (or JA): (1) age at onset less than 16 years, (2) arthritis of one or more joints, (3) symptom duration of at least 6 weeks, (4) an onset type after 6 months' observation of polyarthritic form (five or more affected joints), oligoarthritic form (fewer than five joints affected), or systemic form with arthritis and characteristic fever, and (5) exclusion of other forms of arthritis. In 1977 the European League Against Rheumatism (EULAR) proposed the term juvenile chronic arthritis (JCA) for the same disorder. Their criteria include (1) onset before age 16 years, (2) arthritis in one or more joints, (3) disease duration of at least 3 months, (4) a pattern of pauciarticular (fewer than five joints affected), polyarticular (more than four joints affected), and rheumatoid factor (RF)-negative, or systemic arthritis with characteristic fever. They also include juvenile rheumatoid arthritis (more than four joints affected and RF-positive), juvenile ankylosing spondylitis, and juvenile psoriatic arthritis in the classification.

Debate continues regarding the proper terminology for these various disorders. The term rheumatoid is considered inappropriate by many authors because so few children carry RF.[62,156]

INCIDENCE AND PREVALENCE

The reported incidence of JRA ranges from 3 to 13.9 cases per 100,000 per year.[68,208] The prevalence of the disorder is in the range of 113 per 100,000 children (95 percent confidence limits: 69, 196).[200]

DEMOGRAPHICS

The most common age at onset is between 1 and 3 years, and in this age group girls predominate and most often have pauciarticular disease.[44,52,53,76,126] A second peak of onset occurs around age 9, and at this age the proportion of boys affected approaches that of girls. Overall, JRA is twice as common in girls as boys. With pauciarticular disease the ratio is 3:1, and with uveitis and arthritis girls outnumber boys by 5 or 6 to 1.[1,38,195,199,201] It may be that black children are less often affected than white children, but this is uncertain.[84]

ETIOLOGY

The etiology of JRA remains unknown, but a number of factors relating to etiology have been reported. The predominant common factors involve the immune system. Children with JRA have altered immune systems, as shown in several studies.[117,122] Specific immunodeficiencies are associated with JRA, and there is much evidence that immune reactions are involved in joint inflammation. T-lymphocyte abnormalities have been reported frequently, but their exact role in pathogenesis has yet to be determined.[12,122,147,173,214] HLA product, T-cell receptor, and an antigen, together called a trimolecular complex, play a critical role in JRA pathogenesis.[77]

Heredity may also play a role in the etiology of JRA. The reported familial incidence of the disorder ranges from 23 percent to 41 percent,[52,114,172] and twin concordance has been reported.[13,106]

TABLE 33–1 **Synovial Findings in Joint Affections**

Parameter Measured	Group I: Noninflammatory			Group II: Noninfectious Inflammatory				Group II: Severe Noninfectious Inflammatory	Group III: Infectious Inflammatory	
	Normal	*Traumatic Arthritis*	*Degenerative Joint Disease*	*Systemic Lupus Erythematosus*	*Pigmented Villonodular Synovitis*	*Rheumatic Fever*	*Gout*	*Rheumatoid Arthritis*	*Pyogenic Arthritis*	*Tuberculous Arthritis*
Appearance	Straw or clear yellow	Clear yellow, bloody, or xanthochromatic	Clear yellow	Straw	Xanthochromatic	Yellow	Yellow to turbid milky	Yellow to greenish	Grayish or bloody	Yellow
Clarity	Transparent	Transparent or turbid	Transparent	Slightly cloudy	Turbid	Slightly cloudy	Cloudy	Cloudy	Turbid purulent	Cloudy
Viscosity	Normal	Normal	Normal	Normal or decreased	Normal	Decreased	Decreased	Decreased to poor	Decreased to poor	Decreased to poor
Mucin clot	Good	Good	Good	Good or fair	Good	Good	Poor	Poor	Poor	Poor
Total WBC	≤200	≤2,000 (few to many RBCs)	≤1,000	5,000 (10% DNA particles)	≤3,000 (some RBCs)	10,000	10,000–14,000	15,000 (1,000–60,000)	60,000	20,000
Polys (%)		<20	<20		<20	50	60–70	55	90	60
Crystals	Negative	Negative	Negative	Negative	Negative	Negative	Urate + (in pseudogout, calcium pyrophosphate)	Negative	Negative	Negative
"RA" or "LE" cells	Negative	Negative	Negative	"LE" cells	Negative	Negative	Negative	"RA" cells	Negative	Negative
Bacteria	Negative	Negative	Negative	Negative	Negative	Negative	Negative	Negative	Positive	Positive
Glucose—difference between levels in joint fluid and blood	20 mg/mL	20 mg/100 mL	20 mg/100 mL	20 to 30 mg/100 mL	20 mg/100 mL	20 mg/100 mL	20 mg/100 mL	≥30 mg/100 mL	30–50 mg/100 mL	30–50 mg/100 mL
Total proteins	1.8 g/100 mL	3.3 g/100 mL	3.0 g/100 mL	3.2 g/100 mL	3.0 g/100 mL	3.0 g/100 mL	5 g/100 mL	4.1 g/100 mL	4.2 g/100 mL	4.2 g/100 mL
Albumin (%)	60–70	60	60	60	57	60	70	42	45	45
γ-Globulin (%)	14	16	16	15	17	14	9	25	25	25
Immunoglobulin		Normal	Normal	Elevated	Normal	Normal or slightly elevated	Normal	Elevated	Normal	Normal
Complement—total and B_1-C		Normal	Normal	Decreased	Negative	Normal	Normal or elevated	Decreased	Normal	Normal
Latex fixation and sensitized sheep cell agglutination	Negative	Negative	Negative	Occasionally positive	Negative	Negative	Negative	Positive	Negative	Negative

Infection has long been proposed as a factor in the etiology of JRA, and many different hypotheses have been supported. Studies in the late 1960s implicated *Mycoplasma fermentans,* which was isolated from 31 of 79 samples of synovial fluid.[213] More recently, infection with rubella virus has been found in children with rheumatic diseases, with virus isolated from both serum and synovial fluid in 7 of 19 patients.[31] Perinatal infection with influenza virus with expression of the disease many years later has also been proposed as an etiology.[158] The infectious agent may supply the antigen that initiates the immune reaction.[159]

Physical and psychological trauma have been associated with the onset of JRA. However no clear causal relationship has been identified for either type of trauma, and they are considered aggravating factors at best. Barometric changes and weather patterns have anecdotally been associated with disease severity but most likely have no causal role.

PATHOLOGY

The histologic changes of the synovium in these disorders are those of chronic inflammation and are not specific to or diagnostic of rheumatoid disorders. The inflamed synovium is hypervascular and infiltrated with small lymphocytes and polymorphonuclear leukocytes in the acute phases (Fig 33–2). There is excessive synovial fluid, which is thin and watery. Later the synovium proliferates and forms granulation tissue, which may cover the articular cartilage and is

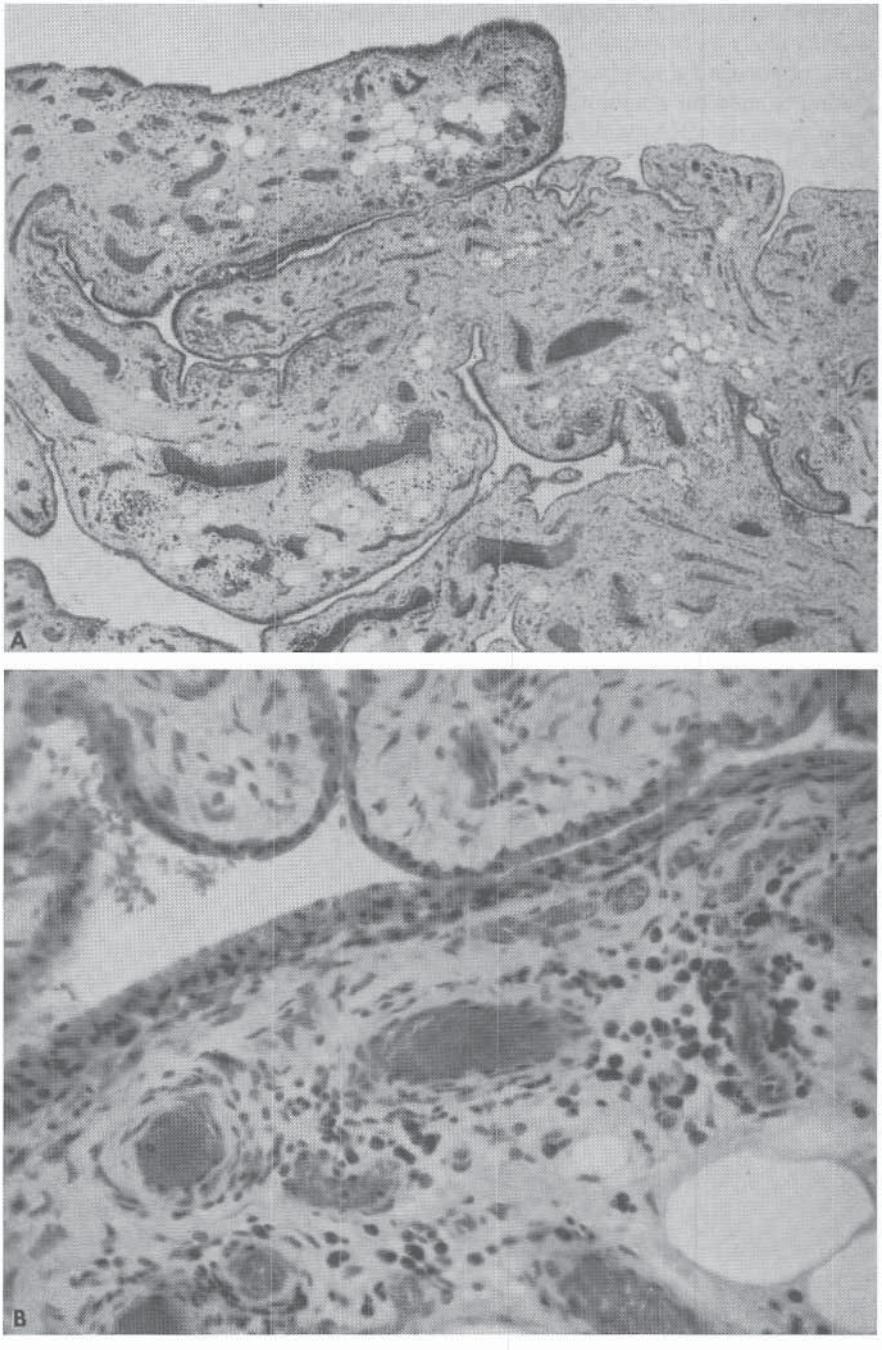


FIGURE 33–2 Histologic appearance of synovium in rheumatoid arthritis. **A,** ×100. **B,** ×250.

termed a pannus (Fig. 33–3). Precipitated fibrin may form small, solid pieces called rice bodies, which may float freely in the joint.

Reactions in the bone are secondary to the aggressive inflammation of the synovium. Erosion of bone at the sites of synovial attachments occurs, and subchondral bony resorption is common. Loss of cartilage beneath the pannus is followed by subchondral bony destruction, and this sequence may lead to ankylosis of the joint.

CLINICAL FEATURES

Pauciarticular JRA. Approximately half of cases of JRA in children are of the pauciarticular form, which by definition includes only cases with fewer than five joints involved. Girls affected by this variety of the illness outnumber affected boys by a ratio of 7:3. In other words, an affected child is twice as likely to be female than male. The peak period of onset is between 2 and 4 years, with half of affected children coming to medical attention before age 4.

Pauciarticular JRA manifests as a low-grade inflammation of one or several joints in an otherwise well child. In about half of patients only one joint is involved. The knee is most often affected, with the ankle-subtalar and elbow joints next in frequency (Fig. 33–4). Hip involvement is quite unusual and, when present, may raise other diagnostic considerations. The small joints of the hands and feet are usually spared. Cervical spine involvement is extremely rare.[88] On presentation, one or several joints may be involved. Over several months other joints may become inflamed, but in half of the pauciarticular cases only one joint is involved.[30,178]

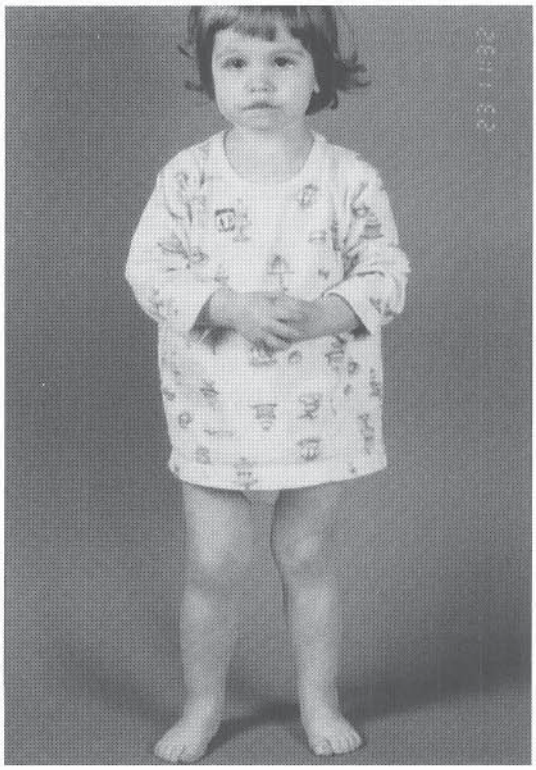


FIGURE 33–4 Typical appearance of a young girl with a swollen knee of pauciarticular JRA.

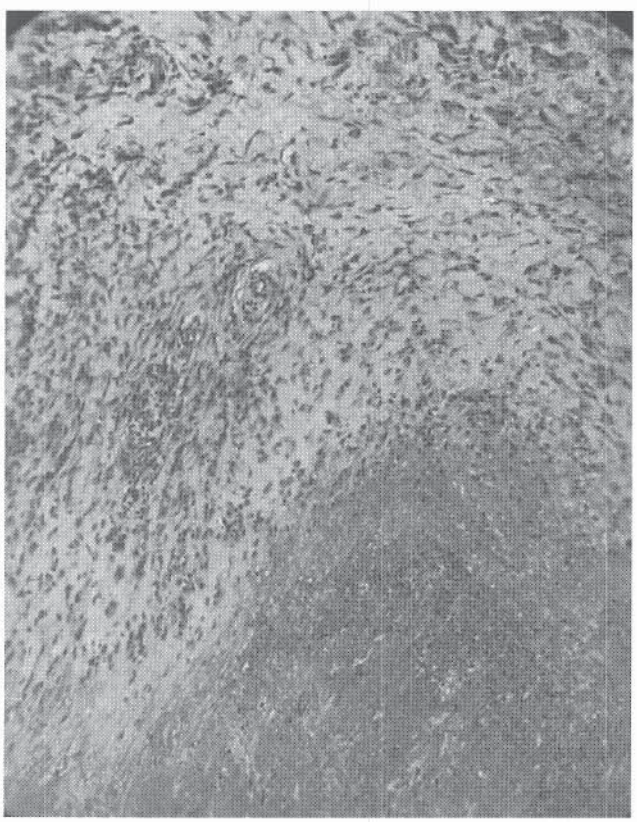


FIGURE 33–3 Microscopic appearance of rheumatoid nodule. Note the focus of fibrinoid degeneration surrounded by fibroblasts arranged in palisade formation.

A history of insidious onset without precipitating trauma is common, although occasionally some traumatic event calls attention to the joint. Morning stiffness is a frequent complaint, with symptoms decreasing during the day as the joint is used. The swelling is persistent; it may gradually increase but usually does not change dramatically from day to day. By convention a duration of 6 weeks of arthritis is necessary for the diagnosis of JRA to be considered.

The involved joints are usually mildly tender and swollen. The swelling is a combination of synovial thickening and joint effusion. The degree of swelling is often out of proportion to the degree of tenderness or the amount of pain. The joint is warm but usually not erythematous, and there is some loss of range of motion and some pain when the joint is moved. By comparison, a septic joint is exquisitely tender and the limitation of motion is much greater than in the inflamed joint in JRA.

Uveitis is a serious associated problem and may ultimately affect the child's vision. It may be present at onset, and over the course of the disease 20 percent of children will develop it. An early diagnosis can be made on finding increased protein levels and inflammatory cells in the anterior chamber of the eye on slit lamp examination. Later, posterior synechiae form and tether the iris to the lens, resulting in an irregular and poorly reactive pupil. Band keratopathy and cataracts occur late but eventually may involve 42 percent to 58 percent of patients with uveitis.[105,154] Most cases are asymptomatic, and ophthalmologic examination is essential to allow early treatment.

The course of the disease is relatively benign. The arthritis

TABLE 33–2 **Pauciarticular Pearls**

Often presents to an orthopaedist
One or two joints, often knee, subtalar
Morning stiffness
Joints swollen, minimally tender
Erythrocyte sedimentation rate and C-reactive protein level mildly elevated or normal
Uveitis present

waxes and wanes and is usually responsive to medical control. Over a period ranging from 3 to 11 years the disease usually resolves. The average duration of disease is 2 years 9 months, and in half of cases it is less than 2 years.[75] In approximately one-third of cases there is progressive involvement of more joints so that the disorder resembles typical polyarthritis with somewhat fewer joints involved (Table 33–2).

Polyarticular Onset JRA. When five or more joints are involved, the syndrome is by definition polyarticular JRA. Two peaks of onset exist, the first between 1 and 3 years and the second between 8 and 10 years. Females predominate in the later age group. Polyarticular JRA has many characteristics in common with the pauciarticular form. The onset is insidious, the large joints of the lower extremity are often involved, the inflammation is chronic, and pain and swelling are moderate. The small joints of the hands and feet are commonly involved, as are the joints of the cervical spine and the temporomandibular joints (Figs. 33–5 and 33–6). The affected joints are warm, tender, painful on motion, and swollen, with synovial thickening and effusion. Limitation of joint range of motion is almost always present; this is initially caused by protective muscle spasm and later by destruction of articular cartilage and fibrosis. Affected children typically appear apprehensive and guard their painful limbs against movement. Symptoms arising in the temporomandibular joint are often described as "earache," and symptoms arising from the sternoclavicular and costochondral joints are described as "chest pain." On occasion, hoarseness and laryngeal stridor may result from inflammation of the cricoarytenoid joints. Cervical spine involvement with fusion of the apophyseal joints results in limitation of neck motion. Involvement of the temporomandibular joint will cause failure of development of the lower jaw and result in a receding chin.

Some systemic manifestations may be present and include low-grade fever, hepatosplenomegaly, lymphadenopathy, and subclinical pleural and pericardial inflammation.

A major distinction is made between children with RF-positive polyarticular disease and those with RF-negative disease. The RF-positive disease in children is in many ways similar to the adult form of rheumatoid arthritis. The children have rheumatoid nodules, erosion of joint surfaces, and a disease course that extends well into adulthood (Fig. 33–7). Children with RF-negative disease have less involvement of the small joints of the hands and feet and do not form nodules.

Systemic Onset JRA. The systemic form of JRA is a serious disease in which arthritis is only one manifestation of a generalized disorder. Many organs and systems may be involved, including the liver, spleen, pleura, pericardium, and skin. Uveitis is rare. A febrile course with one or two daily spikes from normal to 39°C or 40°C is typical. The temperature spikes most often occur late in the afternoon, and the temperature rapidly returns to baseline. During the febrile periods the children are listless and appear ill but may seem well once they defervesce. The fever usually does not respond to salicylates or nonsteroidal agents.

Affected children usually have a characteristic skin rash with discrete, erythematous maculas 2 to 5 mm in diameter (Fig. 33–8). The skin rash is classically a salmon color but may be more reddish in the early stage. It is located on the trunk, face, palms, soles, and proximal extremities, and tends to migrate fairly rapidly. There is often a clear halo around the maculas, and the larger ones may be clear in the center.

Hepatosplenomegaly and generalized lymphadenopathy are often present. Enlarged, inflamed mesenteric nodes may cause abdominal pain and distention, suggesting an acute

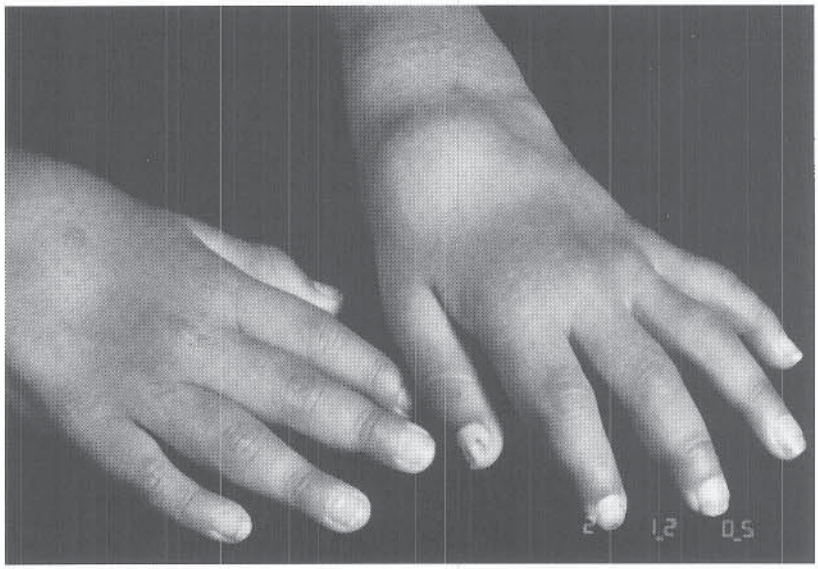


FIGURE 33–5 Swelling of the wrist, metatarsophalangeal, and proximal interphalangeal joints of the hand in polyarticular JRA.

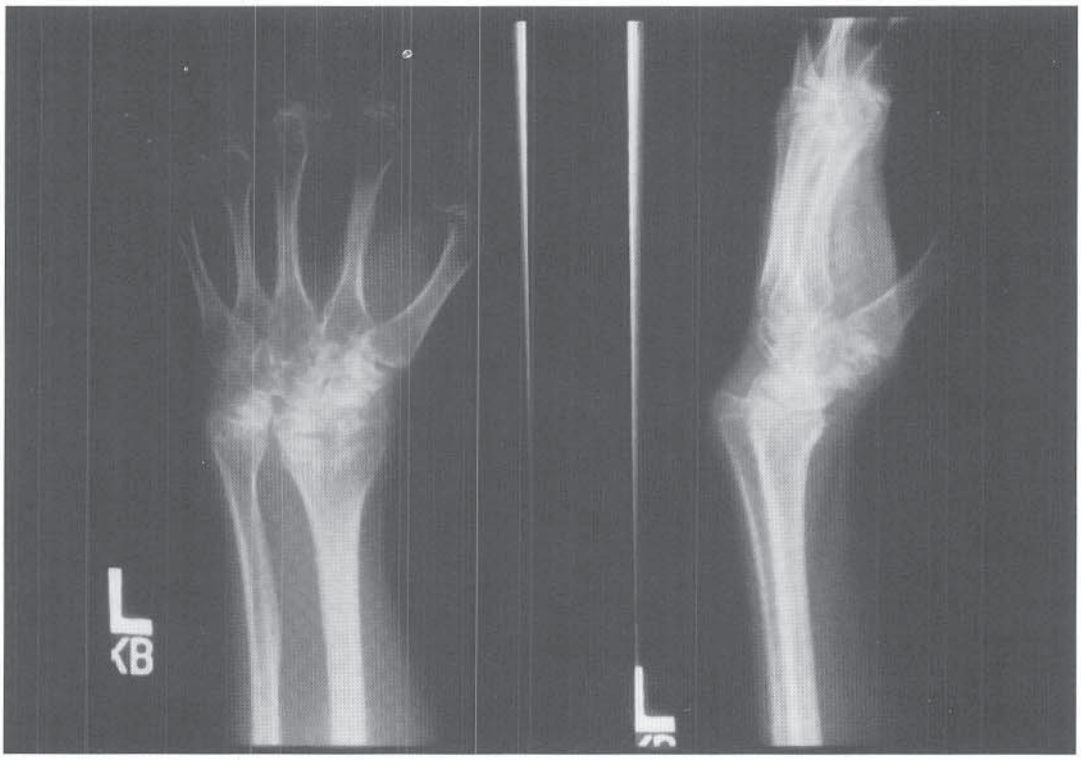


FIGURE 33–6 Radiographic changes of JRA of the wrist. Carpal destruction and volar subluxation are common findings.

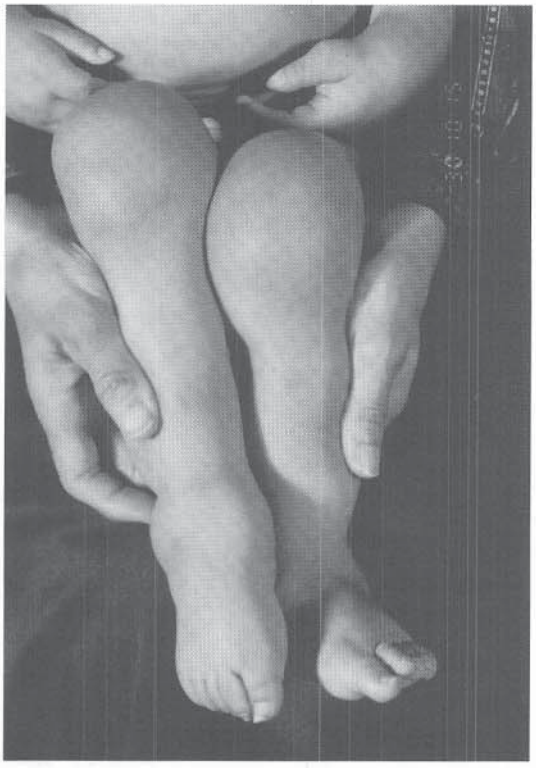


FIGURE 33–7 Arthritis of the knees and ankles in a child with seropositive polyarticular JRA.

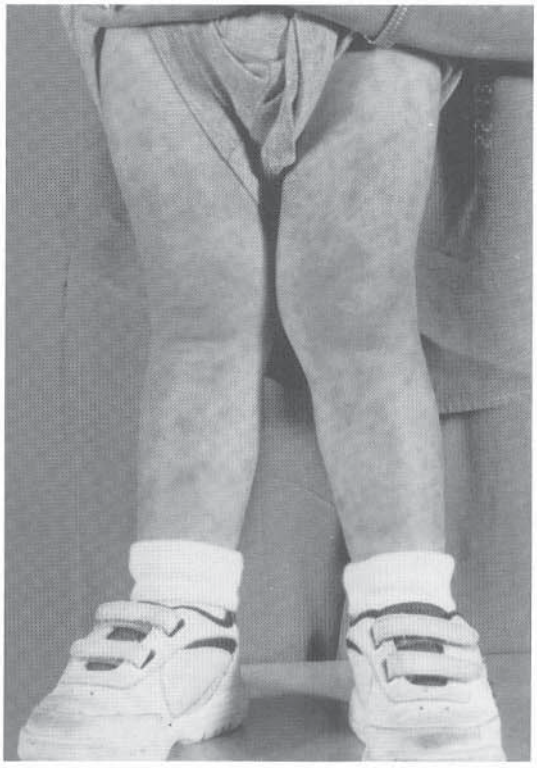


FIGURE 33–8 Typical rash of systemic JRA.

surgical abdomen. The enlargement of abdominal organs usually resolves over a few months. Pericarditis and pleural effusions occur in about 10 percent of those with systemic disease and may manifest with nonspecific chest pain.[72] Electrocardiographic changes are present and the cardiac silhouette is enlarged on the chest radiograph. The cardiac manifestations are usually transient and rarely result in congestive heart failure.[48,119] The presence of pericarditis is not related to the severity of the disease in general or to the joint manifestations.[21]

Amyloidosis is a grave complication that is rare in North America, but in Great Britain it occurs in about 7.5 percent of cases.[7] It presents with proteinuria and hypertension, and IgG and C-reactive protein (CRP) levels are elevated in those who develop amyloidosis. Control of the activity of the inflammatory disease is the mainstay of prevention of amyloidosis.[7]

LABORATORY EVALUATION

There is no single or definitive test for rheumatoid disease; rather, the diagnosis is made from clinical findings coupled with suggestive laboratory findings. Anemia, leukocytosis, and inflammatory indices generally correspond to the severity of the disease. WBC counts of 30,000 to 50,000/mm^3 may occur in children with systemic disease. Elevation of the platelet count also may accompany severe disease. The erythrocyte sedimentation rate (ESR) and CRP counts are again elevated and are related to the severity of systemic disease.

Synovial biopsies show villous hypertrophy, vascular endothelial hyperplasia, and infiltration by lymphocytes and plasma cells. These changes are typical of chronic inflammation. Over time, the inflamed synovium forms a pannus of tissue that covers and destroys articular cartilage. Rheumatoid nodules are not seen in children with JRA except for those with the seropositive polyarticular form.[30]

RADIOGRAPHIC EVALUATION

Although plain radiography remains the mainstay of radiographic evaluation, ultrasound and MRI are useful in the early stages of disease to identify joint effusion and synovial hypertrophy.[164] The earliest changes seen on plain films include periarticular soft tissue swelling, osteopenia, especially around the joint, and widening of the joint space.

As the disease progresses the radiographic joint space narrows, owing to destruction of articular cartilage (Fig. 33–9). Adjacent osteopenia causes loss of the subchondral bony plate. In late disease, erosive changes produce notching of the bone, especially in the carpals (see Fig. 33–6). Epiphyseal overgrowth may occur secondary to hyperemia, or disuse may retard growth.

Both large and small joints may become subluxated. Most commonly this occurs with volar subluxation of the wrist, posterolateral subluxation of the hip, and ulnar subluxation of the metacarpophalangeal joints. In the final stages fibrous or bony ankylosis occurs.

Some of the most specific radiographic changes occur in the cervical spine.[60,133] Erosion of the odontoid in a "napkin ring" pattern may be associated with atlantoaxial instability. An atlanto-dens interval greater than 4.5 mm may be seen in 20 percent of patients but is rarely related to neurologic dysfunction.[58] Fusion between cervical segments is common and most often occurs at C2–3. Cervical spine involvement is rarely present in patients with pauciarticular disease.[88]

Involvement of the temporomandibular joint is also common and may result in mandibular undergrowth. This produces a micrognathia that is characteristically seen in children with long-standing disease.[15]

TREATMENT

As with other complex disorders, juvenile arthritis is best managed by a specialized team that includes the rheumatologist, orthopaedists, ophthalmologists, physical and occupa-

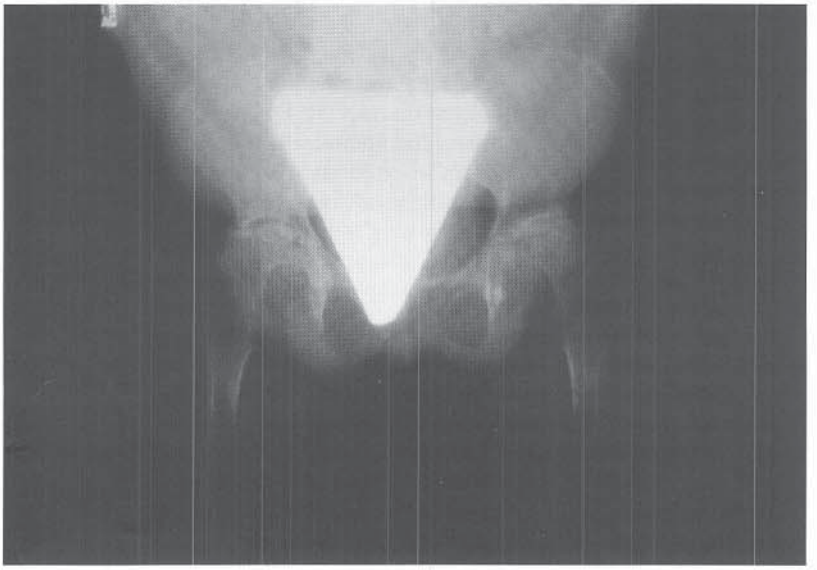


FIGURE 33–9 Bilateral hip involvement with systemic onset JRA. There is almost total loss of joint space on the right. Total hip replacement is usually quite successful in this situation.

tional therapists, nurses, and social workers. The nonsteroidal anti-inflammatory drugs (NSAIDs) are the mainstay of treatment of most patients. Although aspirin was the initial agent of choice, modern agents supply more potency with fewer side effects. Aspirin is given in a dosage of 75 to 90 mg/kg/day, usually in four divided doses with food, seeking a therapeutic salicylate level of 20 to 25 mg/dL. Ibuprofen is given in a dosage of 35 mg/kg/day, again in four doses with food. Naproxen is used at 15 to 20 mg/kg/day twice a day. Tolmetin sodium is given in a dosage of 25 to 30 mg/kg/day in three doses. At this time only these three NSAIDs are approved by the Food and Drug Administration (FDA) for use in children.

In more severe disease not responsive to NSAIDs, other, more toxic drugs may be efficacious. Low-dose methotrexate and other cytotoxic drugs may have a rapid effect on resistant disease. Hematologic, hepatic, and pulmonary monitoring is mandatory when these agents are used. A group of agents, the slow-acting remittitive drugs, or SARDs, includes antimalarial agents (hydroxychloroquine), parenterally administered gold compounds, and penicillamine. For patients who fail to respond to first-line drugs these drugs may be effective, but they are slow-acting, requiring 3 to 6 months for full effect.

Intra-articular glucocorticoids are indicated and may be quite effective treatment for recalcitrant joint inflammation. Systemic steroids are indicated for life-threatening systemic disease but are not indicated long term because of the side effects of iatrogenic Cushing's syndrome. Ophthalmic glucocorticoids are used to treat chronic uveitis.

Physical and Occupational Therapy. Physical and occupational therapists should be involved in the clinic team managing children with chronic arthritis. The goals of such therapy are to relieve pain, increase range of motion, improve muscular coordination, and help the patient relearn physiologic functional patterns. Therapists also help children learn about joint protection and self-care. Splinting on a selective basis is useful, and adapted footwear and walking aids may be used.[81] Splinting of the wrists and hands may reduce the tendency toward joint contracture and subluxation. Occasionally, splinting of the knee or ankle at night is indicated to maintain range of motion.

Physical conditioning in children with arthritis is often poor, with decreased aerobic capacity and exercise tolerance in proportion to the severity of the disease.[70,110] Disuse atrophy of muscles, joint contractures, and anemia all contribute to deconditioning. Rehabilitation of children with arthritis should include conditioning training in addition to standard physical therapy activities. Conditioning requires that muscles be challenged with repetitive, progressive stress with exercises aimed at specific muscle groups. When joints are acutely inflamed, isometric exercises are recommended. Dynamic exercise can be begun when the arthritis is in subacute or chronic stages. A general guideline is to have the child lift the maximum weight he or she can lift for 10 repetitions. That weight is then used for two to three sets of 2 to 10 repetitions for each muscle to be exercised. That weight is then gradually increased. Low-impact sports such as walking, swimming, cycling, or low-impact aerobic dance are more appropriate than highly competitive sports.[111] Although excessive exercise may aggravate an inflamed joint, specific restrictions should be applied only when the management team is relatively certain of a deleterious effect.

Controlled studies of physical therapy interventions with standardized measurement techniques have not shown a positive long-term effect on the arthritic disease,[83,86,191] nor have they shown a negative effect.[80,192] One study reported that children with juvenile arthritis who receive massage therapy from their parents for 15 minutes a day for 30 days showed significant reduction in pain and anxiety and improved activity level compared to a control group of children who engaged in relaxation therapy.[61]

Orthopaedic Treatment. Chronic joint inflammation results in a cycle beginning with muscle spasm to protect the painful joint from motion. If continued, the cycle results in contracture of the muscle and joint capsule and disuse of the extremity, with resultant osteopenia. The orthopaedic management of JRA is concerned with interrupting this cycle. Thus, management emphasizes maintaining of joint range of motion and extremity alignment and length, reducing synovial proliferation, observation of cervical spine stability, and ultimately joint replacement as needed. Synovectomy remains a controversial modality. Releases about the hip and knee are sometimes needed; rarely, cervical spine instability requires treatment; and hand and foot deformities are sometimes correctable. All operative procedures in patients with JRA require careful preanesthetic evaluation. Cervical spine stiffness and instability, reduced mobility of the jaw with hypoplasia of the mandible, and coexisting medical conditions may require specialized approaches for intubation and recovery.

Synovectomy may be helpful for severely affected, recalcitrant joints but has not been shown to alter the long-term outcome of joint disease.* Although some studies have shown that following removal of inflamed synovial tissue a new, relatively normal synovial lining will regenerate,[65,97] others have reported frequent recurrence of disease.[73,150,161] The results of synovectomy are best in large joints, and the knee is the commonest joint so treated. Likewise, the best results with synovectomy are obtained if the procedure is performed early, before significant joint destruction has occurred.[104] Successful synovectomy results in a reduction in swelling and pain. Range of motion is not improved after synovectomy, and care is necessary to avoid losing motion. Arthroscopic synovectomy is associated with less postoperative stiffness and morbidity, and postoperative continuous passive motion may be helpful. Synovectomy is indicated when a trial of medical management for more than 6 months (including intra-articular steroids) has failed.

The development of flexion contractures of the hip and knee results in loss of walking efficiency, with both increased loading on the knee and increased pain.[196] When contractures of the knee exceed 15 to 20 degrees, significant loss of walking ability occurs.[152] Surgical releases of the hip and knee may result in long-term improvement in range of motion and function. Witt and McCullough reported a reduction in flexion deformities of the hip from an average of 35 degrees to 9.5 degrees, with loss of correction to 18 degrees at 3 years. This improvement was maintained in patients followed as long as 12 years.[220] In another study of

*See references 8, 56, 57, 63, 65, 96, 99, 119, 133, 149, 168, 171.

soft tissue releases of the hip or knee (or both), 10 of 27 patients were able to walk preoperatively and 22 could walk after release(s).[142] After 3 years there was some loss of correction. Other authors have reported similar reductions in contracture, with acceptable recurrence rates.[170]

Release of knee flexion contracture is best done with the patient prone. Usually the hamstrings, lateral intermuscular septum, and iliotibial band are released. If necessary, one or both tendinous portions of the gastrocnemius muscles are sectioned and the posterior capsule of the knee is also released. Occasionally the anterior cruciate must be cut to correct posterior tibial subluxation.[39,170] The postoperative cast must be molded to displace the tibia anteriorly as the knee is extended, to avoid posterior subluxation. If full correction cannot be obtained without neurovascular compromise, subsequent cast changes under anesthesia may be required.[39] Postoperative night splinting for up to 6 months is recommended to prevent recurrence.

Flexion contractures of the hip also respond to soft tissue releases. These are indicated when a significant contracture that interferes with ambulation persists after 6 months or more of aggressive medical therapy. Swann and Ansell reported a reduction in flexion contractures after psoas and adductor tenotomy, with improvement still evident 3 years postoperatively.[197]

Growth disturbances are most often seen at the knee. When a valgus deformity is present, either an epiphyseal stapling or percutaneous partial epiphysiodesis will correct the deformity without major surgical trauma. The epiphysiodesis approach is preferred because of the minimal incision required. It should be done when growth prediction based on bone age shows 2 to 3 cm of growth remaining at the distal femoral epiphysiodesis. Rydholm and colleagues reported that stapling of the distal femoral epiphysis for valgus was effective in correcting the deformity in 15 of 17 patients so treated. Stapling was also effective in correcting leg length inequality.[169]

Scoliosis occasionally occurs in patients with JRA and may be managed by conventional means.[143]

Total Joint Arthroplasty. Total joint arthroplasty is an appropriate and effective therapy for adolescents with polyarticular disease and painful, stiff, destroyed joints. Hip and knee replacements have a well-established role in improving the function and well-being of the patient. Wrist, elbow, and ankle replacements may be useful, but there is less clinical experience behind them.

Total hip or knee arthroplasty is indicated in the adolescent with marked functional impairment or severe disabling pain from advanced structural hip or knee joint involvement (see Fig. 33–9).[182] Careful planning with a team approach is essential and should include consideration of high school and college education, use of crutches, medications, and emotional status. Preoperative planning includes procuring miniature or custom-made hip prostheses in up to half of patients. When both hip and knee replacements are necessary, it is best to approach the hip first. If the knee is approached first, it is more difficult to rehabilitate the knee in the presence of a painful, contracted ipsilateral hip. In addition, it is useful at times to manipulate and cast the knee to gain extension at the time of the total hip arthroplasty.[182]

Total hip replacement may be performed in a child with growth remaining. Knee replacement in the setting of open epiphyses is indicated if minimal growth remains. One series of knee replacements with open physes reported no growth disturbances, but all epiphyses had closed within 2 years of replacement.[175]

Total joint replacements are difficult to perform in these patients owing to osteopenia, contractures, and coexisting medical conditions. Cementless arthroplasties are gradually replacing cemented prostheses because of late loosening.

The results of hip and knee replacement are quite remarkable. Relief of pain is reported in almost all patients after hip replacement and in a high percentage after knee replacement. Improvement in range of motion is excellent at the hip and quite good at the knee. Most important, functional status improves in a high percentage of patients, often to a remarkable degree. Rates of loosening of hip components range from 12 percent at 4.5 years to 43 percent at over 5 years postoperatively. Prosthesis survival rates (still functioning) are up to 92 percent at 10 years and 83 percent at 15 years.*

There are fewer reports available evaluating replacement of other joints. Connor and Morrey evaluated 19 patients following total elbow replacement and found that 96 percent had pain relief. Although improvement in motion was less predictable, most patients gained a functionally significant range, including those with ankylosed joints preoperatively.[41] Total ankle replacements are rarely performed in children, and reports in adults are mixed, with a significant number of failures reported.[28,35,100,205,221]

Spondyloarthropathies

The spondyloarthropathies, often termed seronegative because of the absence of RF, are a group of disorders that include ankylosing spondylitis, Reiter's syndrome, arthritides associated with inflammatory bowel disease, and psoriasis. Enthesitis, or inflammation of ligament, tendon, and fascial insertions, is a common manifestation of these disorders and is not typical of rheumatoid arthritis. Joints of the axial skeleton are often involved as well as peripheral joints. Iritis, often acute, may occur in any of these related disorders. These entities are uncommon in children. Males are affected more often than females. A family history is more likely to be present in these disorders than in rheumatoid arthritis. The familial occurrence is related to the common finding of the HLA-B27 histocompatibility antigen.

JUVENILE ANKYLOSING SPONDYLITIS

Juvenile ankylosing spondylitis is characterized by arthritis of the sacroiliac joint and spine, along with involvement of one or several peripheral joints of the lower extremity. Large joints are more often affected than small. In most series males predominate, but the apparent male predominance may be misleading, because females may have less symptomatic disease and more peripheral joint involvement.[134,165] Current reports of large series of juvenile cases indicate a male-female ratio of 2.7:1, with 73 percent of patients being male.[74] Ankylosing spondylitis is much less common

*See references 19, 25, 29, 36, 78, 79, 113, 115, 118, 131, 182, 215.

in children than JRA and usually occurs in adolescence rather than childhood. The HLA-B27 assay is positive in 90 percent of Caucasians with the disorder.[177,179]

There is often a striking familial occurrence of ankylosing spondylitis.[155] The risk of a parent with the HLA-B27 antigen having a child with the disease is 5 to 10 percent.[206]

Involvement of the spine and sacroiliac joint is the diagnostic hallmark of ankylosing spondylitis (Fig. 33–10); however, many patients present with other symptoms and develop axial involvement later. Burgos-Vargas and Vazquez-Mellado have shown that enthesopathy, tarsal disease, pauciarthritis, and knee involvement are frequent presenting symptoms and can differentiate this disorder from JRA. In their series, definite spinal and sacroiliac involvement occurred after a mean of 7.3 years of other symptoms.[24]

Patients often are misdiagnosed, and physicians should consider juvenile ankylosing spondylitis when evaluating a patient with chronic pain or joint symptoms. Pain in the buttocks, groin, thigh, heel, or shoulder may be vaguely localized and evanescent. Only 25 percent of children will have definite findings about the spine or sacroiliac joint at presentation. Peripheral joint symptoms predominate and resemble those of JRA. The joints most frequently affected are the knees, the metatarsophalangeal joint of the first toe, and the ankle. Enthesitis is the most distinguishing feature, with tenderness about the patella, tibial tuberosity, calcaneal apophysis, plantar fascia, and metatarsal head or base being most frequent. Occasional tender sites include the greater trochanters, anterior superior iliac spines, pubic symphysis, and ischial tuberosities.

Sacroiliac joint inflammation is noted by tenderness over the joint, pain on compression of the pelvis, and pain on distraction of the sacroiliac joint, known as Patrick's test. Spinal involvement may manifest as loss of lordosis, increased thoracic kyphosis, and a list to the side.

Involvement of the costosternal and costovertebral articulations may manifest as tenderness over those sites. Thoracic cage excursion during inspiration and expiration may be diminished, and a measured excursion of less than 5 cm suggests thoracic cage involvement.

The radiographic diagnosis is definitive when the sacroiliac joint shows widening, sclerosis, or fusion. The widening is termed pseudo-widening because it relates to erosions of the joints and disruption of subchondral borders due to inflammation.

The long-term prognosis is relatively good for juvenile onset ankylosing spondylitis.[176] Flato and co-workers found that 60 percent of patients reviewed had no disability after 9.7 years of disease, and 60 percent were in remission. Twenty-five percent had articular erosions and disability, and they tended to have started treatment later in the disease than the others.[64] Affected patients respond well in general to NSAIDs, but not so well to aspirin. Indomethicin (1 to 2 mg/kg/day in three divided doses) is often remarkably effective but may produce significant toxic effects. Sulfasalazine is also useful for treating this disorder.[30]

Orthopaedic management is similar to that for the other arthritides. Spinal extension exercises to prevent loss of lordosis or lumbar flexion may be helpful.

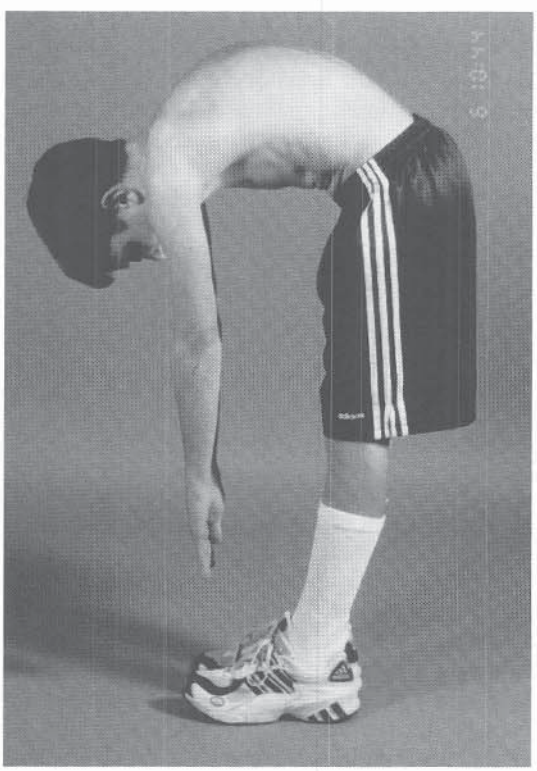


FIGURE 33–10 Limited forward bending with increased kyphosis in a boy with juvenile ankylosing spondylitis.

REITER'S SYNDROME

Classic Reiter's syndrome is diagnosed by the triad of arthritis, conjunctivitis, and urethritis. It is a postinfectious arthritis with a genetic predisposition, and those affected are usually HLA-B27 positive. In children it usually follows a diarrheal illness.

The arthritis usually affects a few joints. Unlike ankylosing spondylitis, upper extremity joints are often involved. Enthesitis is also frequent, but axial skeletal involvement is less common. Frank urethral discharge is present in 30 percent of children, and many more have pyuria on urinalysis. In the dysenteric form urethritis is often present as well.

Conjunctivitis is present in two-thirds of affected children, and occasionally more severe ocular disease occurs. Characteristic ulcerative oral lesions are seen, as well as keratoderma blennorrhagicum, which is a characteristic skin manifestation with papular eruptions on the soles of the feet. Like the other spondyloarthropathies, Reiter's syndrome is self-limiting and responds to standard treatment regimens.[45,125,145]

PSORIATIC ARTHRITIS

Psoriatic arthritis is characterized by arthritis of one or more joints for at least 6 weeks, accompanied by a typical psoriatic rash. The diagnosis can be made in the absence of the typical rash if three of four minor criteria are present: dactylitis, nail pitting or onycholysis, a psoriasis-like rash, or a family history of psoriasis. The family history is positive in 40 percent of patients. The disorder is slightly more common in girls than in boys.

Differences from the other entities include the occurrence of dactylitis, in which flexor tendon sheath inflammation

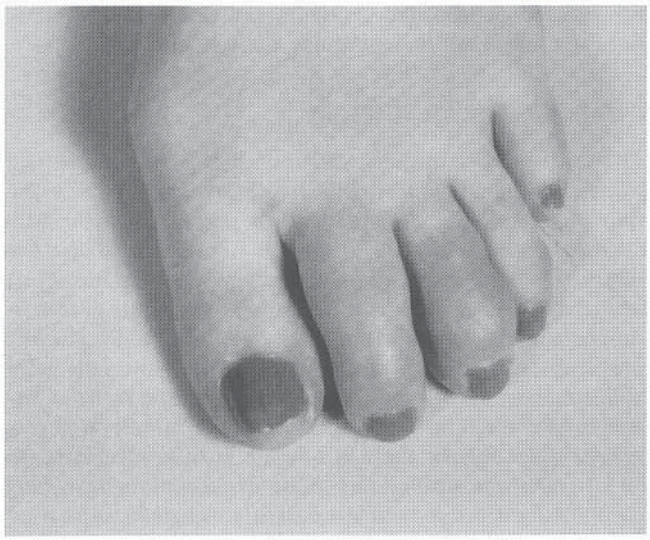


FIGURE 33–11 Sausage dactylitis of the second and third toes in a child with psoriatic arthritis.

produces a sausage-like toe or finger (Figs. 33–11 and 33–12). Large joint involvement and enthesitis are similar to what is found in ankylosing spondylitis, and these children also may have sacroiliac joint involvement. Psoriasis may precede or follow the onset of the other findings.[26,112,121,202] Uveitis occurs in psoriatic arthritis and may be resistant to treatment.

Treatment guidelines are similar to those for ankylosing spondylitis. The use of low-dose methotrexate was pioneered in psoriatic arthritis, and this drug may be very effective in those with multiple joints involved (Table 33–3).

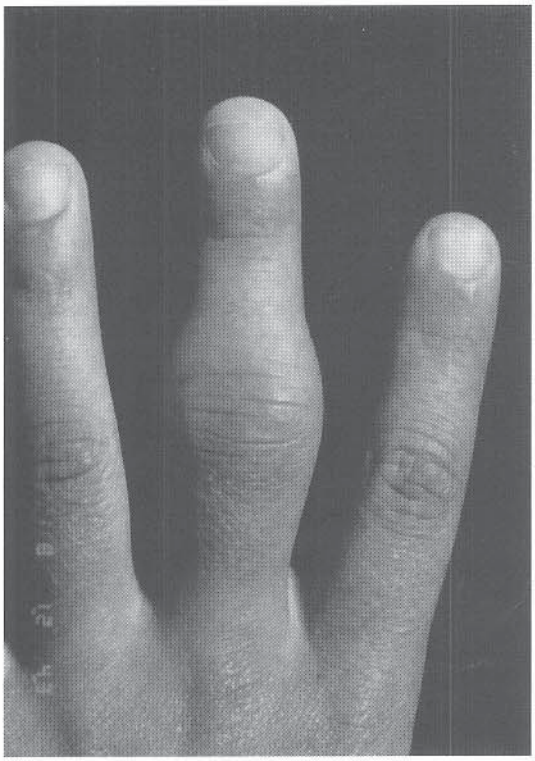


FIGURE 33–12 Sausage dactylitis of the proximal interphalangeal joint of the hand in psoriatic arthritis.

Acute Transient Synovitis of the Hip

Acute transient synovitis is the most common cause of hip pain in children. It is a self-limited inflammatory condition of the hip of undetermined etiology that occurs primarily in younger children. Although the disorder itself is benign, it must be distinguished from septic arthritis, which requires emergency treatment. Synovitis of the hip may also be the first symptom of Legg-Calvé-Perthes disease, early juvenile arthritis, or ankylosing spondylitis. Hip irritability also may accompany osteomyelitis in the femur or pelvis, or another bony lesion.

The incidence of transient synovitis in children between the ages of 1 and 13 years is estimated at 0.2 percent per year. During these childhood years a child has a 3 percent chance of sometime developing the disorder.[116] After a child has had an episode of synovitis, the annual risk of recurrence for that child is 4 percent.

The disorder has also been called toxic synovitis, irritable hip syndrome, observation hip, coxitis fugax, and acute transient epiphysitis. The first description was given in 1892 by Lovett and Morse, followed by many subsequent descriptions.[20,58,129,184]

TABLE 33–3 **Other Syndromes with Arthritis**

Systemic lupus erythematosus
Episodic disease
Inflammation of blood vessels and connective tissues
Antinuclear antibodies
Antibodies to native DNA
Malar (butterfly) rash
Multiple organ disease
Acute arthritis
Rapid response to steroids
Rheumatic fever (Jones' criteria)
Carditis
Polyarthritis (migratory)
Chorea
Erythema marginatum
Subcutaneous nodules
Minor fever (to 39°C), arthralgia
Elevated CSR, CRP
Evidence of prior streptococcal infection
Sarcoidosis
Boggy, nontender synovium
Pulmonary disease (rare in children)
Rash
Iritis
Hepatomegaly
Lyme disease
Rash
Acute or chronic arthritis
Carditis
Neurologic symptoms
Tick-borne spirochete
Juvenile dermatomyositis
Inflammation of muscle (weakness and pain)
Inflammation of skin (typical rash)
Calcinosis (late)
Vasculopathy

ETIOLOGY

The etiology of transient synovitis is unknown. The disorder occasionally follows an upper respiratory tract infection, but the nature of this relationship is unknown. A number of studies have sought evidence of a bacterial or viral etiology, without success.[85,116] In one study technetium bone scans showed a decrease in isotope uptake in the proximal femoral epiphysis in one-fourth of the hips with synovitis. In these hips, a rebound hyperemia was noted on follow-up scan 1 month later. Only one such patient later developed Legg-Calvé-Perthes disease.[87] The significance of this finding is uncertain. Another study found increased levels of proteoglycan antigen in children with both septic arthritis and transient synovitis.[127]

CLINICAL PICTURE

The usual presentation is a child with the fairly rapid onset of limping and subsequent refusal to walk or bear weight. This sometimes follows a recent upper respiratory tract illness, and parents may report a low-grade fever.

Boys are affected two to three times as frequently as girls.[207] Onset peaks between 4 and 10 years, with a mean age at onset of around 6 years.[116,207]

The examiner finds a child in mild distress who will not bear weight or walk, or who does so reluctantly and with an antalgic limp. The range of motion of the affected hip is moderately limited by pain and spasm, and the hip is held in flexion. Gentle short-arc motion may be tolerated, but an attempt to fully extend or internally rotate the hip will be resisted. The irritability of the hip is usually several grades less severe than in a child with septic arthritis. There may be low-grade fever.

DIAGNOSTIC STUDIES

The diagnosis of transient synovitis is one of exclusion. The laboratory evaluation may show mild elevations in the WBC count, ESR, and CRP level. Plain radiographs of the pelvis are usually normal or may show slight joint space widening. Capsular distention on plain films, long thought to indicate fluid within the joint, has been shown to be a radiographic artifact due to the positioning assumed by an irritable hip.[23] When there is joint space widening and a smaller femoral ossific nucleus on the involved side, one can make a presumptive diagnosis of early Legg-Calvé-Perthes disease.[116]

Ultrasonography of the hip is very useful in documenting the presence of an effusion in the hip joint. Ultrasonography is often performed prior to hip aspiration to be certain that the clinical findings are accompanied by an effusion. A number of studies have shown that ultrasonography reliably demonstrates fluid in the hip, and a negative study directs attention to other causes of hip pain.[91,136,140,198] One study noted a difference between the fluid present in transient synovitis and the synovial thickening of Legg-Calvé-Perthes disease, but others have not been able to make that distinction sonographically.[66] Another study reported that patients with Perthes disease had cartilage thickening in the femoral head that was detected sonographically.[166] Some have found that the effusion in septic arthritis is more echogenic than the effusion in transient synovitis, but this relative finding does not consistently distinguish the two entities.[140,185]

DIFFERENTIAL DIAGNOSIS

The most important diagnosis to exclude is septic arthritis. Classically there is a clear clinical difference in the presentation of the two, with septic arthritis presenting with more severe pain and marked limitation of motion of the hip due to pain. In actual practice, however, low-grade septic arthritis is not uncommon. Sometimes a less acute presentation occurs after the child has received antibiotics for another problem such as a respiratory illness. Others, especially older children, have low-grade septic arthritis caused by less virulent organisms. The WBC, ESR, and CRP are elevated to a greater degree when there is a septic arthritis. Del Beccaro and co-workers studied the value of laboratory screening combined with degree of temperature elevation and found the values to be useful.[49] Those with septic arthritis had a higher temperature (38.1°C versus 37.2°C), higher mean ESR (44 mm/hr versus 19 mm/hr), and higher mean WBC count (13,200/mm^3 versus 11,200 /mm^3). Because of a large degree of overlap in these values, however, a diagnosis could not be based on these findings alone.[49] They recommended aspiration of the hip for diagnosis when the ESR was above 20 mm/hr and the temperature was above 37.5°C.

Other septic processes should be excluded as well. Osteomyelitis of the upper femur or pelvis may produce similar manifestations, including joint effusion, moderate loss of motion, and mild pain on range-of-motion testing. A nearby Brodie's abscess may have quite similar laboratory characteristics, with minimal elevation in temperature, CSR, CRP, and WBC. A psoas abscess may manifest with a subacute course and specific limitation of hip internal rotation. Inflammatory nodes in the groin, avulsion injuries around the pelvis, and trauma may also be confused with transient synovitis.

Arthritis of the hip may be the presenting symptom of JRA or one of the seronegative spondyloarthropathies. In these conditions the arthritis will persist well beyond the 1- or 2-week period of transient synovitis. A careful general physical examination as well as an examination of all joints will clarify the diagnosis.

Legg-Calvé-Perthes disease may present with synovitis prior to definitive radiographic changes. Radiographic widening of the joint space is more characteristic of Perthes disease than of transient synovitis. Other findings in Legg-Calvé-Perthes disease include a smaller capital femoral ossific nucleus, subtle abnormalities of the contralateral hip, and subsequently an increased density of the femoral epiphysis. Bone scan or MRI can confirm the diagnosis before plain radiographic changes are seen.

CLINICAL COURSE

Transient synovitis by definition resolves spontaneously. Usually the child will present when unwilling to walk or when limping severely. The period of nonwalking generally lasts 1 or 2 days. The child then walks with a limp and has reduced range of motion of the hip for another few days to usually not more than 2 weeks before returning to normal.

An ultrasound study demonstrated that the effusion persisted longer than 1 week in 58 percent of patients.[197] Other authors found that the effusion was gone by 2 weeks in 73 percent of patients.[140] Significant deviations from this pattern should prompt further investigation to rule out other causes of synovitis. One must remember that transient synovitis is a diagnosis of exclusion, and septic arthritis can and does present at times with moderate signs and symptoms.

TREATMENT

Treatment begins almost spontaneously as the child refuses to walk or be moved, and thereby rests the hip. Hip joint aspiration is commonly necessary to rule out septic arthritis and may be beneficial. In one study the degree of capsular distention on ultrasonography was significantly less at 4 days and for the remainder of the follow-up period in children who had undergone joint aspiration than in those who had not.[108] The real purpose of aspiration is diagnostic, and the surgeon should have a low threshold for tapping the hip. Our practice is to aspirate the hip of any child who refuses to walk and has significant limitation of hip motion. When little or no fluid is obtained we perform arthrography to determine that the needle has entered the joint. Considerations that lower the threshold for aspiration even further are elevated infectious indices, significant fever, and leukocytosis.

The child is placed on bedrest until the symptoms and signs are improving. The toddler will pick his or her level of activity and should not be forced to lie down if able to stand. The older child should be allowed gradually increasing activity, as governed by severity of pain and muscle spasm. NSAIDs may be used and often result in rapid improvement. Antibiotics should not be used because the process is not infectious, and antibiotic therapy confuses the picture. In more severe cases, traction can be helpful for a few days.

Hospital admission is appropriate in cases in which septic arthritis remains a possibility or in which other diagnoses have not been eliminated. Close observation is essential and may require hospital admission, especially when the parents are not reliable. Rapid resolution of symptoms and return of range of motion are characteristic of transient synovitis. Worsening symptoms suggest sepsis, and a prolonged course suggests chronic inflammatory conditions such as rheumatoid arthritis and seronegative spondyloarthropathies.

NATURAL HISTORY

No negative long-term effects of transient synovitis have been demonstrated. Coxa magna of the involved hip has been noted in 32 percent of cases when defined as 2 mm or more enlargement of the involved hip compared to the contralateral hip.[102] Nachemson and Scheller also reported finding coxa magna at follow-up but failed to find any prearthritic abnormalities.[144]

It has been proposed that synovitis of the hip could cause ischemia of the femoral epiphysis by a tamponade effect from increased intra-articular pressure.[217,219] However, a number of studies have shown little or no evidence of a causative relationship.[69,103] The clinical confusion exists because at times synovitis of the hip is the presenting finding in early Legg-Calvé-Perthes disease.[20,129]

Neuropathic Arthropathies

Charcot in 1868 described a bizarre destruction of the knee joints with indolent swelling and instability in patients with tabes dorsalis, and proposed that the disease resulted from traumatization of a joint deprived of sensation.[33] Steindler subdivided the condition into the destructive atrophic and the hypertrophic proliferative forms.[190]

Charcot-like changes in joints are seen in patients who have absence or depression of pain and proprioceptive sensation and who engage in extended continuous physical activity. Consequently their joints sustain repeated trauma. In children, neurologic conditions causing neuropathic arthropathy are congenital insensitivity to pain, peripheral nerve injuries, and diabetic neuropathy, as well as a variety of chronic diseases of the spinal cord that lead to sensory disturbances of the limbs. In myelomeningocele, absence of pain sensation is associated with flaccid paralysis and marked limitation of physical activity; thus, owing to associated severe osteoporosis, the bone and joint changes produce a different picture.

The specific joints involved with neuropathic disease vary with the different etiologic conditions. In congenital insensitivity to pain and diabetic neuropathy, the destructive changes occur primarily in the tarsal and metatarsal joints, less commonly in the ankle, and rarely in the knee. In syringomyelia, the joints involved are those of the shoulder and elbow, whereas in tabes dorsalis, the knee, hip, ankle, and thoracolumbar spine are frequent sites of the disease.

CLINICOPATHOLOGIC FEATURES

When a limb with normal sensation is injured, the joint affected by a severe sprain or hemarthrosis is protected from further trauma by pain. In the absence of pain and proprioceptive sensation, however, the joint continues to be active and is repeatedly injured. Synovial effusion and hemarthrosis are aggravated and, together with the abnormal stresses on the joint, cause extreme stretching and weakening of the capsule and supportive ligaments. Local hyperemia causes bone atrophy and resorption. Cartilage destruction, bone erosion, and minute fractures soon follow. The reparative response results in the formation of callus and metaplastic changes in surrounding traumatized soft tissues. With repeated injury, the joint becomes totally disorganized, subluxation ensues, and severe degenerative changes take place.

Clinically, the affected joints are boggy, tense, swollen, nontender, and have an excessive range of motion, often in abnormal directions. The local triad of swelling, instability, and absence of pain is nearly always suggestive of a Charcot joint.

RADIOGRAPHIC FINDINGS

The neurotrophic joint will show varying degrees of destructive and hypertrophic changes (Figs. 33–13 and 33–14).

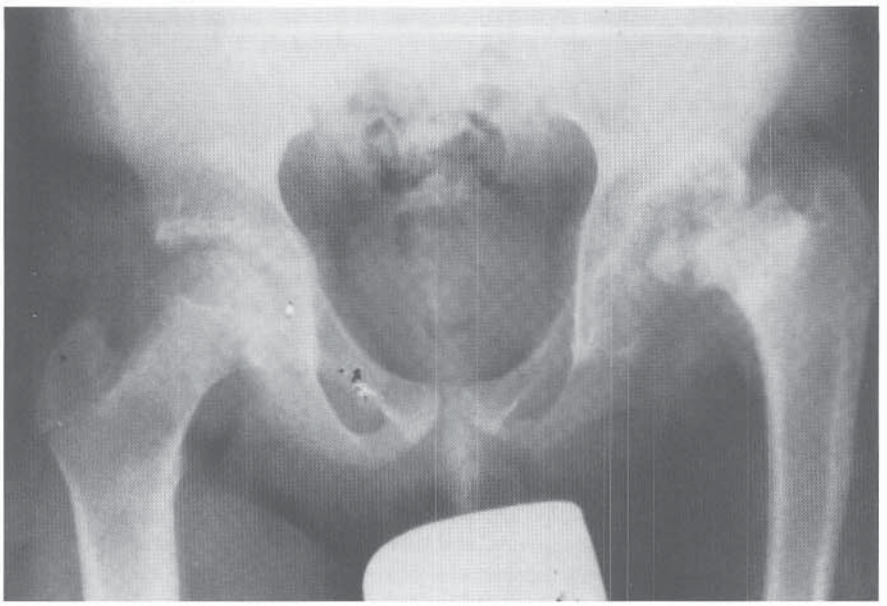


FIGURE 33–13 Pathologic fracture of the neck of the left femur with subluxation of hip in a 10-year-old boy with congenital insensitivity to pain.

Loss of articular cartilage, fragmentation and absorption of subchondral bone, and osseous proliferation of the joint margins also occur. The bone overgrowth may be enormous, bizarre in configuration, and so great as to surround the joint as a spongy mass. The periarticular soft tissues are thickened and contain scattered calcifications. Pathologic fractures involving the articular surface are common, as are irregular loose bodies within the joint.

TREATMENT

The goal of treatment of a neuropathic joint is to reduce the stresses on it to allow healing of the traumatized synovium, ligaments, and cartilage. Immobilization of the joint in a cast or a brace is usually the first line of treatment. When the skin is insensitive to pressure, any form of immobilization carries a risk of severe pressure sores. For the ankle and foot, a well-molded ankle-foot orthosis or cast is appropriate. The knee may be supported with a knee-ankle-foot orthosis, and upper extremity orthoses may also be used. Schon and co-workers have shown that displaced ankle fractures are best managed with open reduction and internal fixation. Forefoot malalignment due to midfoot neuropathy often required operative correction in their series as well to prevent pressure ulceration.[180]

When joints are more severely involved, gradual destruction of cartilage and underlying bone produces irreversible damage. In these cases arthrodesis is often the only option. Total joint replacement is an alternative but has a high likelihood of loosening of components and ultimate failure. Arthrodesis is often difficult to achieve and results in increased stress on adjacent joints.[222]

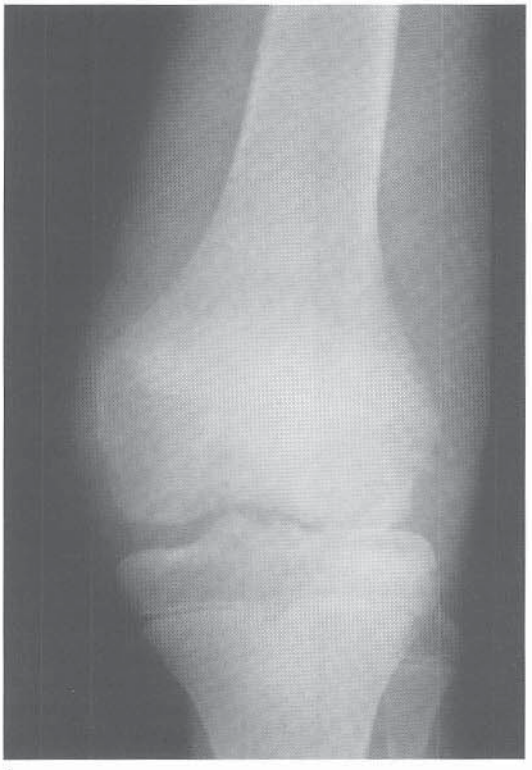
Tuberculous Arthritis

Tuberculosis of bones and joints is a granulomatosis inflammation caused by *Mycobacterium tuberculosis*. It is a localized and destructive disease that is usually blood-borne from a primary focus such as infected peribronchial or mesenteric lymph nodes. The infection may be of the human or the bovine type. In countries where raw milk is used extensively, bovine transmission is common, whereas in areas where milk is pasteurized and there is rigid control of dairy herds, the bovine type is extremely rare and the human type is more common.

After a period of marked decline of tuberculosis, especially in North America, a gradual increase in the incidence of the disease has been noted in the past two decades.[51,130,139,167,189] This increase has been closely associated with the AIDS epidemic. The incidence of tuberculosis has been projected to increase by 41 percent between 1998 and 2020 if better prevention is not practiced.[51] Especially alarming is the appearance of drug-resistant strains in various parts of the world.[47,138,153,186,223] Not too long ago, tuberculosis was the most common disease affecting the skeleton; this is still true in certain areas of the world. Even in economically well-developed countries it is still prevalent. Tuberculosis of bones and joints is more common in children, although it may occur at any age.

The invasion of a joint by the tubercle bacillus may occur by direct hematogenous infection of the synovial membrane (synovial tuberculosis) or by indirect spread from a focus in an adjacent bone—for example, in the metaphysis or epiphysis. Tuberculous osteomyelitis is characterized by destruction of bone, with little or no tendency for new bone formation. The tuberculous bone focus spreads centrifugally with increasing destruction of surrounding bone, until finally the joint is broken into. The synovial membrane reacts first by secreting excessive fluid and later by proliferation, thickening, studding of its inner surface with tubercles, and fibrosis of its outer surface (Fig. 33–15).

The tuberculous granulation tissue soon covers the hyaline articular cartilage as a pannus that eventually destroys the underlying articular cartilage and subchondral bone. The destruction of articular surfaces is most extensive around the periphery in areas where tuberculous granulations involve the synovial membrane.



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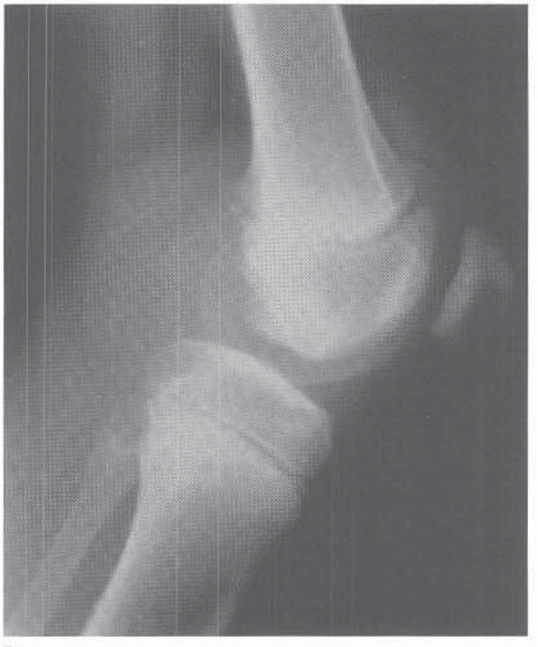

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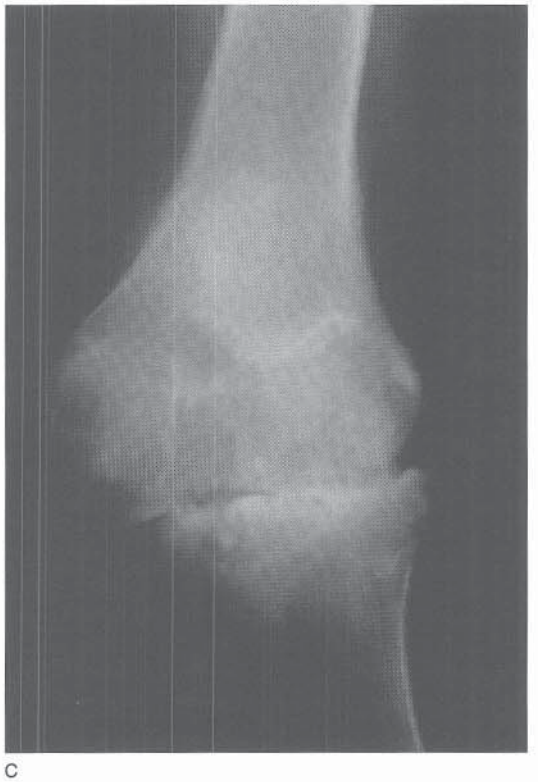

C

FIGURE 33–14 Progressive Charcot neuropathy in a boy with a demyelinating disease. **A,** AP radiograph of the knee showing early changes. There is lateral subluxation of the tibia and destructive, erosive changes in the lateral femoral condyle. **B,** Lateral radiograph of the knee showing destruction of the posterior portion of the lateral femoral condyle. **C,** AP radiograph of the knee obtained 2 years later. There has been total destruction of the lateral condyle of the femur and tibial plateau with severe subluxation of the knee.

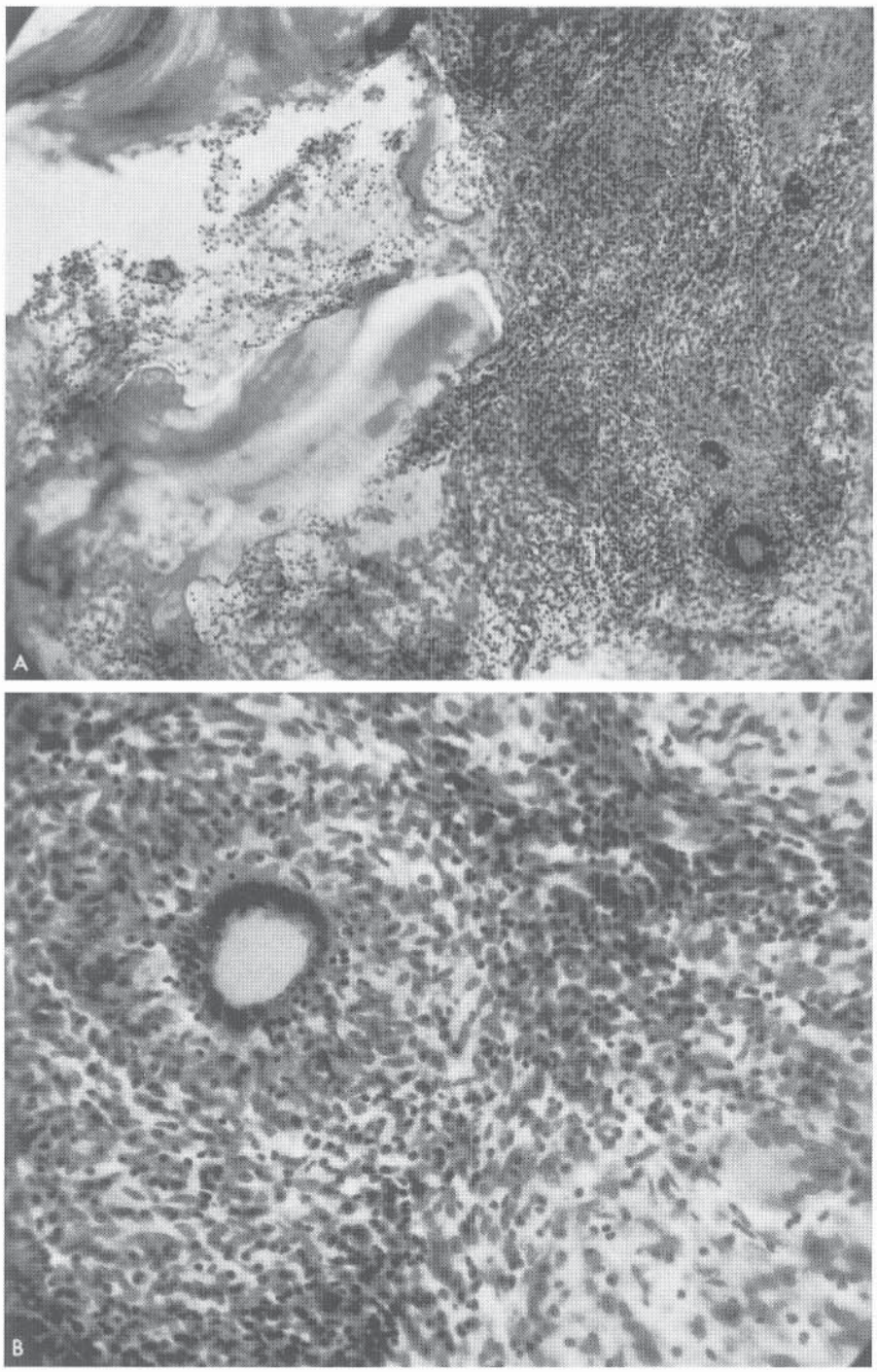

FIGURE 33–15 Microscopic picture of tuberculous arthritis (hematoxylin-eosin stain). **A,** ×100. **B,** ×250. Note the granulomatous inflammation and the Langhans giant cells.

With progression of the disease, increasing amounts of caseous necrotic material and tuberculous exudate reproduce. Soon, with increasing intraosseous or intra-articular pressure, the bony cortex or joint capsule becomes perforated and the so-called cold abscess forms. These tuberculous abscesses are so named because of the absence of acute inflammation. They spread by dissecting along tissue planes between muscles or between muscle sheaths, being limited by the deep fascia. With increasing tension the deep fascia is perforated and the wall lining the tuberculous abscess becomes subcutaneous. A thick fibrous wall lines the tuberculous abscess, which contains serum along with caseous necrotic tissue, tubercle bacilli, and degenerating leukocytes (Fig. 33–15). If the original focus remains active and these abscesses remain untreated, they will rupture externally through the skin to form sinuses. The result is the inevitable secondary infection by pyogenic bacteria and complete destruction of the affected joint (Figs. 33–16 and 33–17).

CLINICAL FEATURES

Tuberculous arthritis is insidious in onset and often (90 percent of cases) monoarticular in involvement. The affected child appears generally ill, is easily fatigued, and has evident

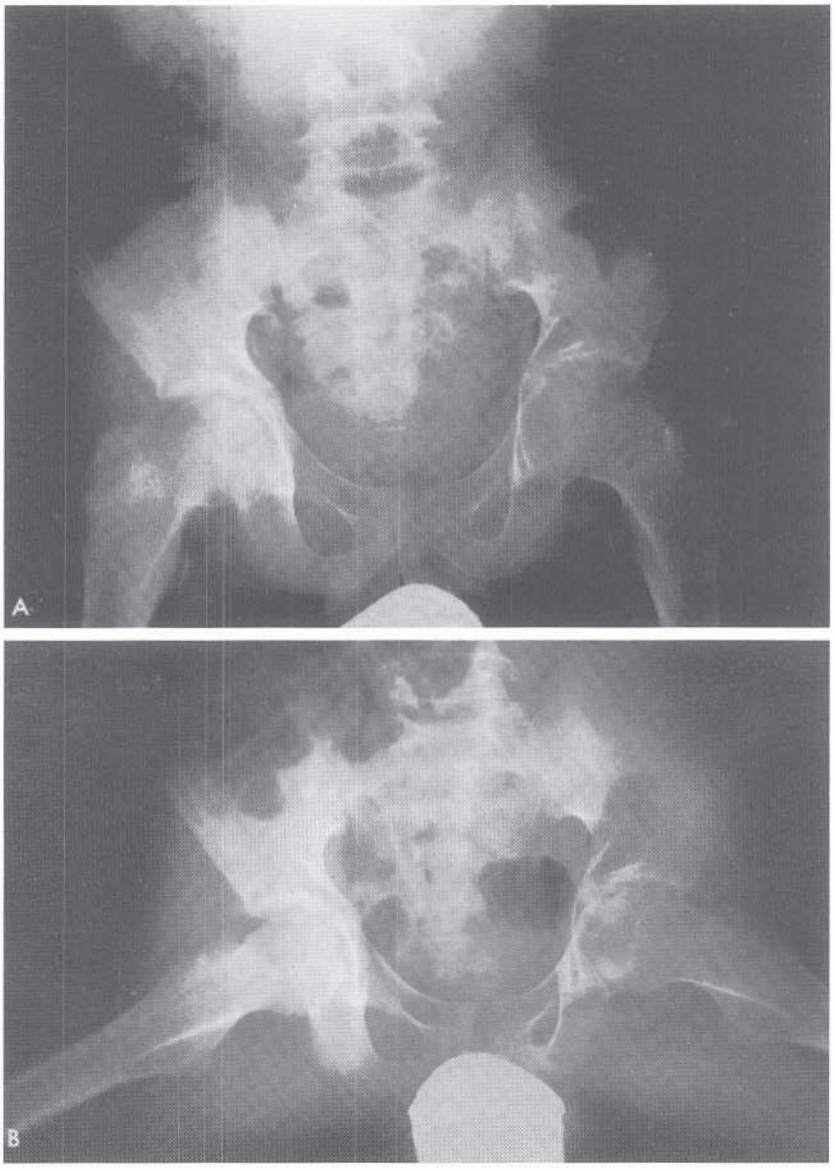

FIGURE 33–16 **A and B,** Tuberculous arthritis of the left hip. Note the regional bone atrophy.

weight loss. A family history of tuberculosis or a personal history of cervical adenitis or pleurisy may be obtained.

If the lesion is in the lower limb, for instance in the hip, the initial symptom may be a slight limp due to discomfort. The affected joint will be stiff, and soon the "night cries" develop: because irritation from the process is low grade, muscle spasm protects the part quite satisfactorily during the day, but when the child is asleep the protective action of the muscle is lost, and on motion, pain is produced; hence the cry.

Local physical signs vary according to the joint involved. The vertebral column is the most common site involved, the next in order of frequency being the hip, knee, ankle, sacroiliac, shoulder, and wrist joints. Almost any joint can be involved, and small joint involvement, although rare, may be confused with juvenile rheumatoid arthritis.[148,203,204] In tuberculous spondylitis the child usually presents with back pain and stiffness. The affected child will walk with a protective gait, keeping the back hyperextended while taking light steps. A kyphosis will develop if bone destruction continues and the vertebral bodies collapse. Untreated disease may progress to spinal cord compression with paralysis, which has been termed Pott's paraplegia. Caseous material collects in the front of the spine and may track inferiorly, producing a psoas abscess.

In superficial joints, such as the knee or elbow, synovial thickening and effusion present as a fullness or bogginess. This may be difficult to detect in the deep joints such as the hip. Local heat and redness are usually absent, and tenderness is minimal. Muscle atrophy is usually marked and is often present in the early stages. Joint motion is usually limited. Temperature elevation is ordinarily not marked.

RADIOGRAPHIC FEATURES

The earliest radiographic findings are regional bone atrophy, soft tissue swelling, and capsular distention (see Fig. 33–16). These changes are due to synovitis and are nonspecific. As a rule, the bone decalcification in tuberculous arthritis is widespread, extending 3 to 5 cm from the joint.

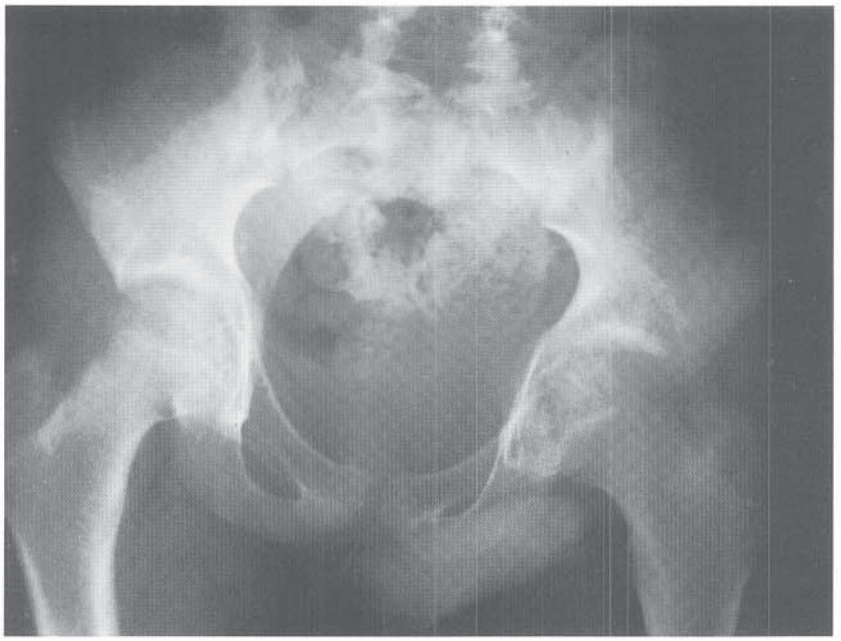

FIGURE 33–17 Tuberculous arthritis of the left hip. Note erosion of the hyaline articular cartilage.

The joint space is widened and is preserved until late in the course of the disease. Destruction of the hyaline cartilage by the tuberculous granulation tissue is a slow process. Eventually, with progression of the disease, the articular cartilage space gradually narrows. This is in contrast to what is seen in suppurative arthritis, in which the destruction of articular cartilage and joint space narrowing occur early in the course of the disease (see Figs. 33–16 and 33–17).

In joints such as the hip, in which there is a congruous and accurate fit of the opposing articular surface, the hyaline cartilage is eroded by the tuberculous granulation tissue in its periphery where there is little or no contact or pressure (see Fig. 33–17). In articulations with incongruous articular surfaces, such as the knee, contact areas are diffusely distributed and the tuberculous granulation tissue destroys articular cartilage wherever the noncontact areas are, centrally or peripherally.

Although tuberculous arthritis is a synovial disease with typical peripheral bony destruction, joints may also be involved when a focus of metaphyseal tuberculous osteomyelitis penetrates into the joint. In these cases the destruction of bone around the joint is more random and does not follow the typical pattern mentioned above (Fig. 33–18). At times, with tuberculous arthritis, both sides of the joint are involved and the two foci of tuberculosis will be directly opposite each other.

Reactive new bone formation is characteristically absent in the early stages of tuberculous arthritis; it is only in the late healing stages that it develops. Sequestra may occasionally be present.

If tuberculous arthritis remains untreated, the entire articular cartilage is eventually eroded and extensive destruction subjacent bone will take place, resulting in gross deformity of the joint (see Fig. 33–18). Abscesses are usually seen early in tuberculous spondylitis in the form of paravertebral or psoas abscesses.

MRI may be useful in making the diagnosis of tuberculosis of a joint. The diagnosis should be considered when the intra-articular synovial lesions show low or intermediate signal intensity on T2-weighted images.[194]

The general findings may reflect a chronic illness. Mild anemia is common, and the leukocyte count may be normal or mildly elevated. The ESR and CRP levels are almost always elevated. Pulmonary tuberculosis may be detected with chest radiography or CT. Renal tuberculosis may be detected through urinalysis, ultrasound, or plain radiography.

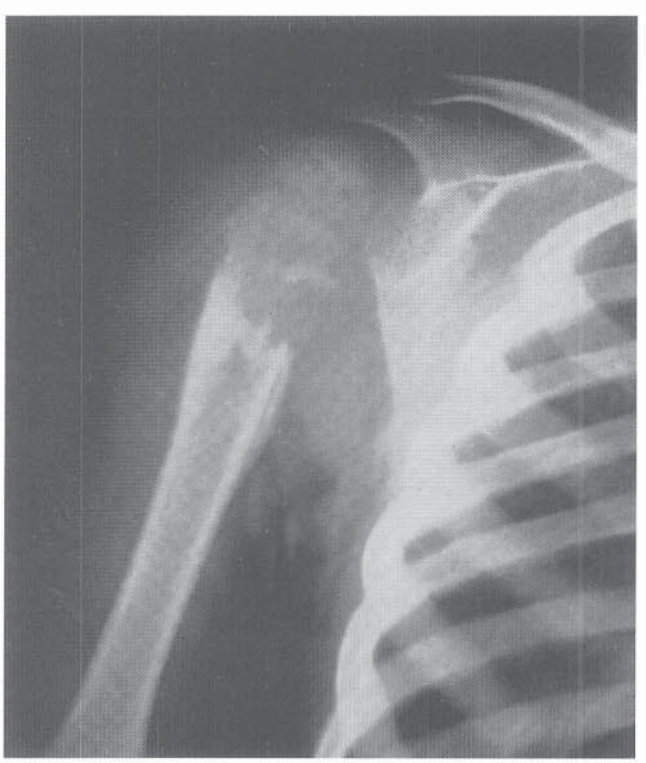

FIGURE 33–18 Tuberculosis of the right shoulder with extensive bone destruction.

The synovial fluid shows an elevated leukocyte count, a lowered sugar level, and poor mucin. The leukocyte count usually averages 20,000/mm^3, although it may vary between 3,000 and 100,000/mm^3. The differential leukocyte count will disclose a predominance of polymorphonuclear leukocytes(60 percent) with 20 percent lymphocytes and 20 percent monocytes. Tubercle bacilli may be seen on microscopic examination of sediment of the joint fluid. A finding of great help in establishing the diagnosis is the marked reduction in or absence of glucose in the synovial fluid. Cultures and specific polymerase chain reaction assays will usually be positive. The diagnosis is also confirmed by histologic examination of synovium obtained by open or needle biopsy or by biopsy of regional nodes.[98]

TREATMENT

In the past, patients with tuberculosis were treated with long periods of rest, fresh air, and joint immobilization. As the patient's immune system began to respond, the diseased joint was surgically fused and the disease could be arrested.

Current management is based on early diagnosis and the use of antimicrobial drugs to which the organism is sensitive. It is often necessary to remove diseased synovium by synovectomy and to debride bony lesions. Postoperative immobilization is probably not necessary, and early motion or continuous passive motion may help restore range of motion.

GENERAL MEDICAL MEASURES

Most patients are not debilitated and may continue reasonable activities while being treated for tuberculosis. Patients with active AIDS and those with visceral disease may need more intensive nutritional and medical management.

ANTITUBERCULOUS DRUGS

The emergence of drug-resistant organisms has prompted a reassessment of treatment protocols.* Current recommendations for treating active disease require a total of 6 months of drug administration. Most commonly a three-drug regimen of isoniazid (INH), rifampin, and pyrazinamide is used. One protocol suggests daily doses of all three for 8 weeks, followed by two to three times weekly doses to a total of 6 months. Another approach is to give daily doses for 2 to 3 weeks followed by bi- or triweekly doses to 6 months. Pyridoxine should be given with INH to prevent peripheral neuritis.[160] Streptomycin is the commonest fourth drug used. Its main adverse effect is cranial nerve VIII deafness, which is most likely to occur when serum concentrations are too high.[54] Ethambutol may be used as a fourth drug but is associated with the serious complication of optic neuritis, which may escape notice in younger children.

If the person who is the source of the infection is known, it may be assumed that the child's organism will be susceptible to the same drugs while awaiting culture results. A resistant organism should be suspected when standard drugs have not controlled the infection. Also, resistance is likely if there has been relapse after a standard course or if the initial treatment has not been complied with. Finally, resistance should be anticipated if the organism is acquired in an area in which resistant organisms are indigenous.[54]

Newer trends in management center on more intense, directly observed protocols of shorter duration. Direct observation is proposed to ensure compliance with drug administration. Genetic studies are investigating the genetic basis of drug resistance in the hopes of finding more effective agents.[188] Infections caused by multiple-drug-resistant organisms require four times as long to treat and are more likely to be incompletely controlled. The treatment of resistant cases often involves the use of more toxic drugs.[151]

ORTHOPAEDIC TREATMENT OF THE TUBERCULOUS JOINT

The diagnosis is usually confirmed by joint aspiration and synovial biopsy and a positive tuberculin skin test. Drug treatment is begun once the diagnosis is made and should be continued for some 4 to 6 weeks to prevent systemic spread of disease. Synovectomy either by open or arthroscopic technique is then performed to remove the bulk of infected tissue. Bony foci of tuberculous osteomyelitis should be debrided. Early motion is begun; contrary to older teaching, it does not impair the healing process.

Arthrodesis is still occasionally necessary when the disease has destroyed the articular cartilage and underlying bony support. Ordinary techniques of arthrodesis are successful with continued drug treatment for the infection.[4,10,16,27,34,71,89,135]

Tuberculosis of the Spine

Tuberculosis of the vertebral column† was first described by Percivall Pott as a painful kyphosis of the spine associated with paraplegia.[157] The condition is since often referred to as Pott's disease. The spine is the most common site of skeletal tuberculosis, accounting for 50 percent of cases. Any level of the spine may be involved, the lower thoracic region being the most common segment; next in decreasing order of frequency are the lumbar, upper dorsal, cervical, and sacral regions.

In the past, tuberculous spondylitis was a disease of early childhood, usually affecting children between 3 and 5 years old. Recently, however, with improved public health measures, this age prevalence has changed, and adults are more frequently affected than children.

Tuberculous spondylitis warrants individual consideration because of certain significant differences between it and tuberculous arthritis of limbs.

PATHOLOGY

The initial focus of infection usually begins in the cancellous bone of the vertebral body and only occasionally in the posterior neural arch, transverse process, or subperiosteally, deep to the anterior longitudinal ligament in front of the vertebral body (Fig. 33–19). The area of infection gradually

*See references 50, 151, 162, 163, 183, 188, 208.

†See references 2, 3, 5, 11, 16–18, 32, 40, 46, 59, 67, 82, 90, 101, 107, 167, 181, 209–212.

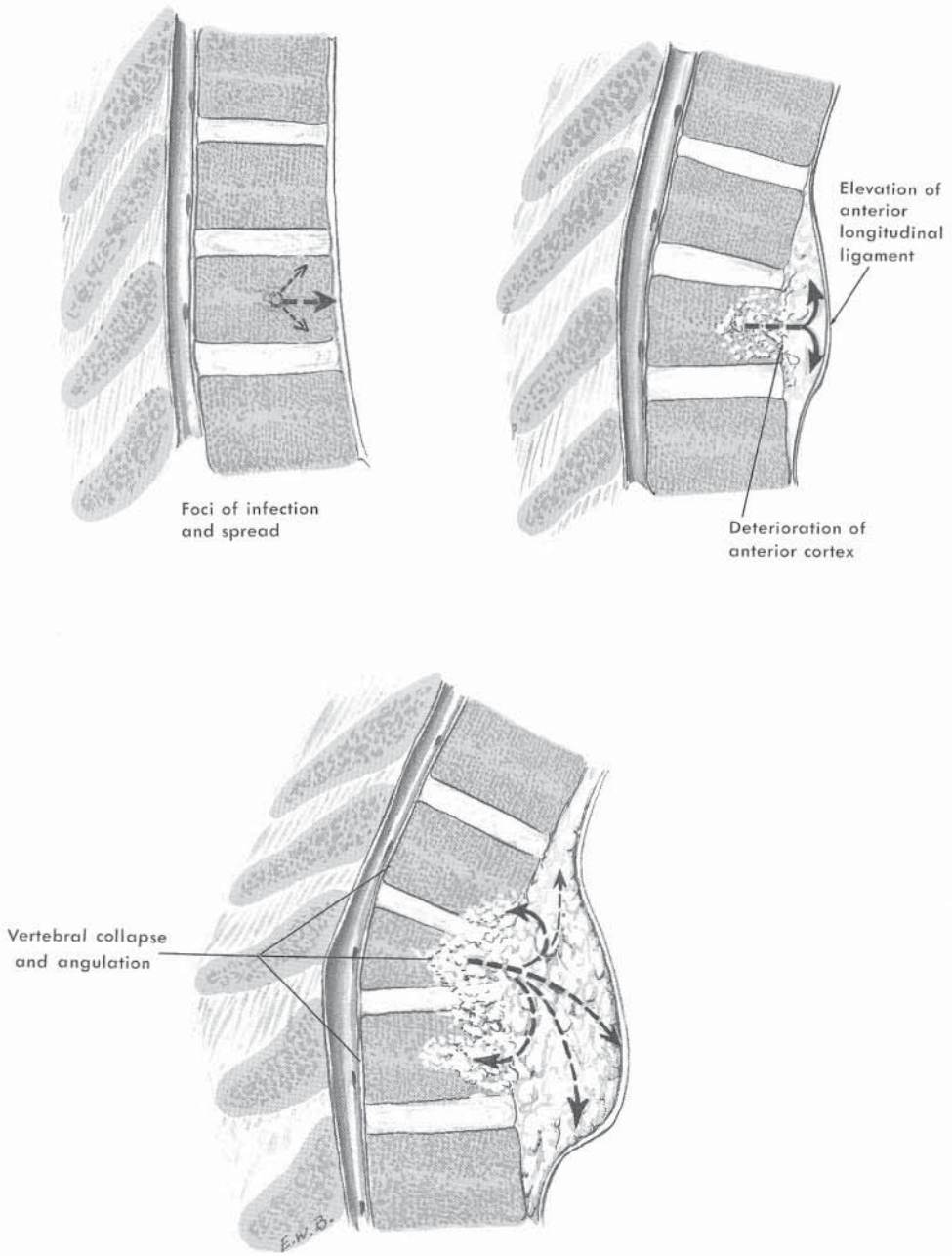

FIGURE 33–19 Pathogenesis of tuberculous spondylitis.

enlarges and spreads to involve two or more adjacent vertebrae by extension beneath the anterior longitudinal ligament or directly across the intervertebral disk. Occasionally there may be multiple foci or involvement separated by normal vertebrae, or the infection may be disseminated to distant vertebrae via the paravertebral abscess.

The vertebral bodies lose their mechanical strength as a result of progressive destruction under the force of body weight and eventually collapse, with the intervertebral joints and the posterior neural arch intact; thus, an angular kyphotic deformity is produced, the severity of which depends on the extent of destruction, the level of lesion, and the number of vertebrae involved. In the thoracic region the kyphosis is most marked because of the normal dorsal curvature; in the lumbar area it is slight because of the normal lumbar lordosis in which most of the body weight is transmitted posteriorly and collapse is partial; and in the cervical spine, collapse is minimal, if present at all, because most of the bony weight is borne through the articular processes.

Healing takes place by gradual fibrosis and calcification of the granulomatous tuberculous tissue. Eventually the fibrous tissue is ossified, with resulting body ankylosis of the collapsed vertebrae.

Paravertebral *abscess formation* occurs in almost every case. With collapse of the vertebral body, tuberculous granulation tissue, caseous matter, and necrotic bone and bone marrow are extruded through the bony cortex and accumulated beneath the anterior longitudinal ligament. These cold abscesses gravitate along the fascial planes and present externally at some distance from the site of the original lesion

(Fig. 33–20). In the lumbar region the abscess gravitates along the psoas fascial sheath and usually points into the groin just below the inguinal ligament. In the thoracic region the longitudinal ligaments limit the abscess, which is seen on the radiograph as a fusiform radiopaque shadow at or just below the level of the involved vertebra; if under great tension, it may rupture into the mediastinum, where it may be walled off to form the "bird's nest" type of paravertebral abscess. Occasionally, a thoracic abscess may reach the anterior chest wall in the parasternal area by tracking via the intercostal vessels. The prevertebral fascia limits the cervical abscess, which may burst into the retropharyngeal area or gravitate laterally on each side of the neck.

Paraplegia results from compression of the cord by the abscess, by the caseating or granulating mass, or by the posteriorly protruding border of the intervertebral disk or edge of bone. Other contributory factors may be thrombosis of the local vessels and edema of the cord. It occurs most often in the mid- or upper thoracic region where the kyphosis is most acute, the spinal canal is narrow, and the spinal cord is relatively large. In the literature it is described as occurring in 6 to 25 percent of reported cases, but recently, with early diagnosis and effective treatment, the incidence has become greatly diminished.

CLINICAL FEATURES

The onset of Pott's disease is usually insidious and of slow evolution. Initial symptoms are vague, consisting of generalized malaise, easy fatigability, loss of appetite and weight, and loss of desire to play outdoors. There may be an afternoon or evening fever. Backache is usually minimal; it may be referred segmentally.

FIGURE 33–20 Abscess formation in Pott's disease. Diagrammed are the various courses a tuberculous abscess arising from the thoracolumbar spine may take. (After Calott.)

FIGURE 33–21 A child with tuberculous spondylitis. When picking up an object from the floor, because of rigidity of the spine she flexes her hips and knees, keeping the spine in extension.

Muscle spasm in the affected region of the spine is a constant finding. The spine is held rigid. When picking an object up from the floor, the child flexes the hips and knees, keeping the spine in extension (Fig. 33–21). Motion of the spine is limited in all directions. Spasm of the paravertebral muscles in the lumbar region is also elicited by passive hyperextension of the hips with the patient prone; this also puts stretch on the iliopsoas muscle, which is in spasm and contracture owing to psoas abscess.

A kyphosis in the thoracic region may be the first noticeable sign. As the kyphosis increases, the ribs crowd together and a barrel chest deformity develops. When the lesion is situated in the cervical or lumbar spine, a flattening of the normal lordosis is the initial finding.

On gentle percussion or pressure over the spinous process of the affected vertebrae, tenderness is often present. The abscesses may be palpated as fluctuant swellings in the groin, iliac fossa, retropharynx, or on the side of the neck, depending on the level of the lesion.

The gait of the child with Pott's disease is peculiar, reflecting the protective rigidity of the spine. The child takes short steps, as he or she is trying to avoid any jarring of the back. In tuberculosis of the cervical spine, the child holds the neck in extension and supports the head with one hand under the chin and the other over the occiput. If the level is in the lumbodorsal area and a psoas abscess is present, the child walks the knees and hips in flexion and supports the spine by placing the hands over the thighs. If a paraplegia develops, there will be spasticity of the lower limbs with hyperactive deep tendon reflexes, a spastic gait, a varying degree of motor weakness, and disturbances of bladder and anorectal function.

RADIOGRAPHIC FEATURES

Radiographic studies of spinal tuberculosis are highly suggestive of the diagnosis and often are definitive. Plain radio-

graphs initially show radiolucency of the vertebral body. As the disease advances, the vertebral body expands and the borders become indistinct. With progressive destruction of bone the vertebral body collapses (Figs. 33–22 and 33–23). At first the intervertebral disk space narrows; later it is obliterated. Paraspinal abscesses appear as fusiform shadows along the vertebral column on the AP radiograph and as an anterior soft tissue mass on the lateral radiograph. Late involvement shows a sharply angled kyphosis or gibbus at the level of vertebral destruction.

MRI is useful in delineating the extent of disease and differentiating it from other types of vertebral osteomyelitis. Vertebral and disk destruction and the location of paravertebral abscesses are graphically demonstrated with MRI.[14,109,124] Gadolinium studies show rim enhancement of lesions, more typical of tuberculosis than septic conditions.[9,109] With tuberculosis of the spine more vertebrae are involved and paravertebral abscesses are larger than in vertebral osteomyelitis.[9] Brucellar spondylitis may be differentiated from tubercular spondylitis in that brucellosis more commonly involves lumbar vertebrae, while tuberculosis more often affects thoracic levels.[42]

A gallium scan may add significant information in the evaluation of a patient with tuberculous spondylitis. In one study this scan was more likely than a technetium scan to delineate paraspinal abscesses and unsuspected levels of vertebral and other bony and soft tissue involvement.[123]

Entities to be ruled out in the diagnosis of spinal tuberculosis include suppurative spondylitis, leukemia, Hodgkin's disease, eosinophilic granuloma, aneurysmal bone cyst, and Ewing's sarcoma. All of these conditions result in destruction and collapse of the vertebral bodies, narrowing and obliteration of disk spaces, and paraspinal soft tissue swelling.

TREATMENT

Immobilization and drug therapy are the mainstays of treatment of spinal tuberculosis.

Drug therapy is initiated and the patient is placed in a spinal cast or brace as soon as the diagnosis is made. Immobilization itself may enhance healing of the lesion and prevent further bony destruction. In addition, immobilization is helpful in preventing progressive kyphosis. In the lumbar spine it has been recommended that the hips be included in the cast or brace. When there is minimal spinal deformity, nonoperative therapy results in a satisfactory outcome with solid bony fusion of the spine in 75 percent of cases.[216]

When there is a large anterior caseous abscess or an unacceptable kyphotic deformity, surgical debridement and stabilization should be considered.[92–95] The surgical approach entails removal of necrotic and caseous material followed by grafting. Autogenous graft is preferred when feasible, and strut grafts may provide anterior stability.[128] Posterior instrumentation and fusion may allow further correction of the kyphotic deformity and may reduce or eliminate the need for postoperative immobilization.

PARAPLEGIA IN TUBERCULOUS SPONDYLITIS

Paraplegia is the most serious complication of tuberculosis of the spine. The incidence is reported at between 10 and 29 percent.[137,174] The current incidence is much lower, owing to better diagnosis and early treatment. Younger children are more likely to become paraplegic.

Hodgson classified paraplegia into four types. The first is paraplegia of active disease resulting from external com-

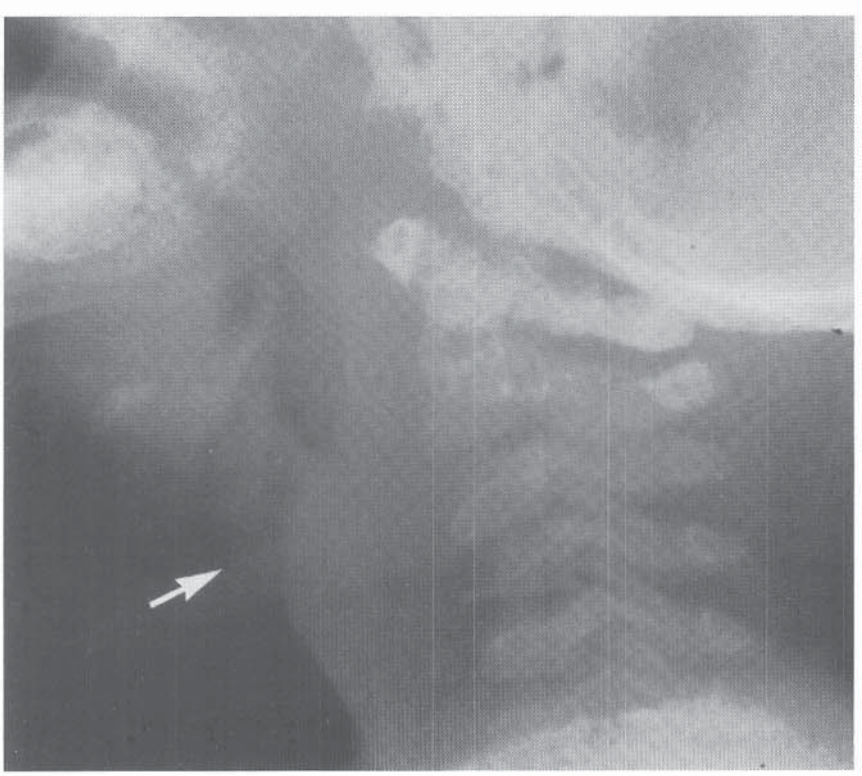

FIGURE 33–22 Tuberculosis of the cervical spine. Note the destruction of the fourth cervical vertebra and the retropharyngeal abscess.

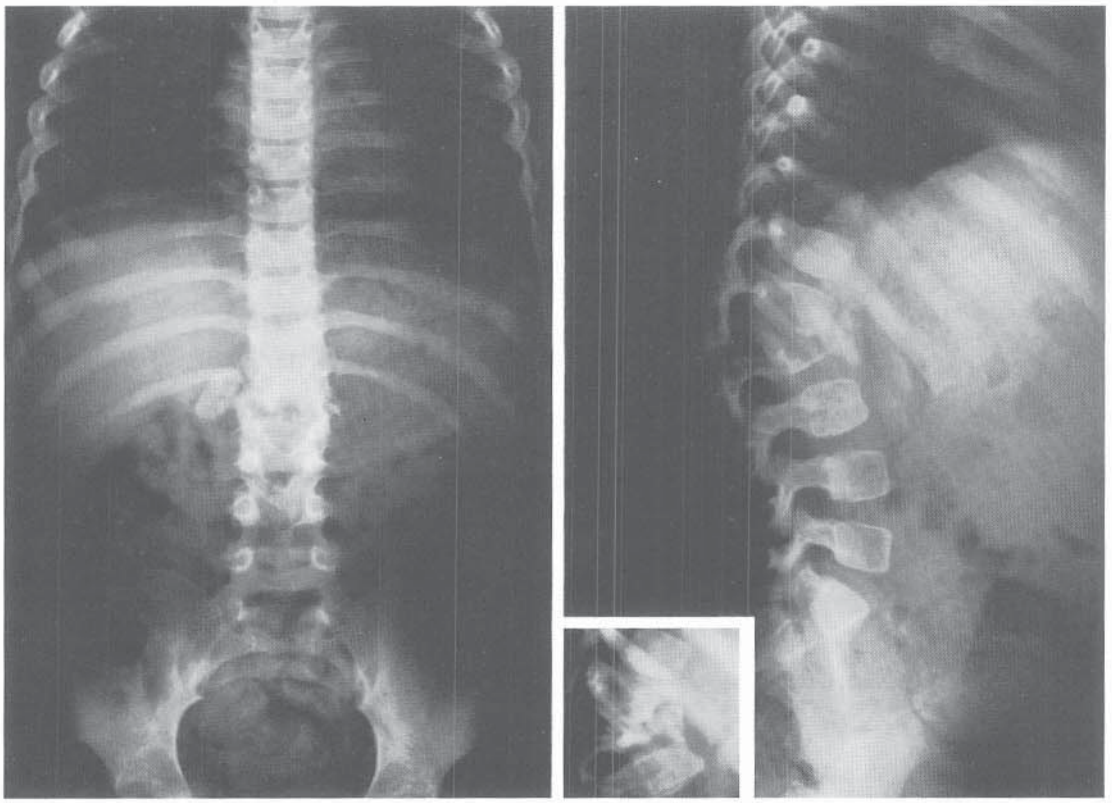

FIGURE 33–23 Tuberculous spondylitis. First and second lumbar vertebrae are involved. Note collapse of the vertebral bodies, obliteration of intervertebral disk interspaces, and localized kyphosis.

pression of the cord and dura. The compression comes from caseating pus, sequestra of bone and disk, dislocation of vertebrae, and granulation tissue within the spinal canal. Clinically, these patients have varying degrees of spasticity of the lower limbs but do not have involuntary muscle spasms and withdrawal reflex. This type of paraplegia carries a good prognosis for full recovery after decompression and stabilization.

The second type is paraplegia due to direct tuberculous involvement of the spinal cord. In these cases tuberculous meningitis and myelitis are present. These patients have more severe spasticity with involuntary muscle spasm and withdrawal reflex. This type of paraplegia is associated with a poor prognosis for recovery.

The third type of paraplegia occurs after healing and is due to fibrosis of the meninges and granulation tissue causing cord compression. The fourth type is due to rare causes such as thrombosis of vessels supplying the cord.

When paraplegia occurs, the level and type of lesion are determined by radiography, MRI, and myelography. Spinal fluid cell count and total protein determination will determine the extent of intradural infection. Early anterior decompression is strongly recommended, followed by spinal stabilization. Delay in treatment may result in permanent paraplegia.[95]

Gonococcal Arthritis

Gonococcal arthritis is caused by metastatic invasion of the joint by the gonococcus, usually from a recent or inadequately treated gonorrheal urethritis. The arthritis, which usually develops 2 to 4 weeks following the initial infection, may be polyarticular or monoarticular. The knees, ankles, wrists, and sternoclavicular joints are the most frequently affected sites. The disease usually begins as fleeting pains in multiple joints, accompanied by fever and malaise—quite similar to what is seen at the onset of rheumatic fever. In a few days the obvious infection settles into a single joint, which becomes hot, red, extremely tender, swollen, tense, and very painful on motion. The acute inflammation may spread to the adjacent tendons and bursae. Often there is a history of gonorrheal infection or a concomitant urethritis. In the more chronic cases, systemic reaction is usually minimal and multiple joints are involved. Gonorrheal arthritis may be associated with dermatitis.[120,141]

A mother may have primary gonococcal infection of her genitourinary tract that may be so slight as to go unnoticed. Such maternal infection may be transmitted from mother to infant. Gonococcal arthritis-dermatitis may manifest as a form of erythematous papules surrounded by a hemorrhagic or vesiculopustulous lesion that may precede the joint involvement. When both skin and joint are involved, the condition is referred to as gonococcal arthritis-dermatitis.[146]

Diagnosis is made by bacteriologic study of the aspirated joint fluid and of the urethral or vaginal discharge. The gonococcal organism can often be identified in the joint fluid within the first week of infection; during the course of the disease, however, joint cultures are negative. In such subacute or chronic cases, immunofluorescent methods for the detection of gonococcal antibodies and gonococcal complement fixation tests are of some aid in diagnosis. Gonor-

rheal arthritis should be distinguished from Reiter's syndrome, which consists of the triad of polyarthritis, urethritis, and conjunctivitis. Reiter's syndrome is rare in childhood; it is a form of nongonococcal urethritis, probably caused by a virus.

In gonococcal arthritis, destruction of articular cartilage is rapid, as shown by the disappearance of articular cartilage space on the radiograph.

Treatment should be instituted immediately to prevent permanent damage to the joint. Penicillin is specific and effective, and may be given intravenously or intramuscularly initially, followed by oral administration when the infection is controlled. Unlike in other types of septic arthritis, joint drainage is usually unnecessary.

ACKNOWLEDGMENT

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