Juvenile Rheumatoid Arthritis: Early Diagnosis, Management, and Prognosis* **

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Rheumatoid arthritis (RA) is the major chronic rheumatic disorder of childhood. It affects as many as 250,000 American children and is slightly more common in girls than in boys.

Juvenile rheumatoid arthritis (JRA) is arbitrarily defined as RA beginning before the age of 16. It rarely begins before six months of age, and most cases appear between the ages of one and three with a second peak at 8-12 years of age.

Despite the inclination to link RA and JRA as identical diseases, one occurring in adults, the other in children, there are a number of striking differences between the two (1–5). In a comparative survey (Table 1), high fever and rheumatoid rash occurred far more frequently in children than in adults with RA. Another important difference is that chronic iridocyclitis developed in 8% of patients with JRA. Such ocular involvement is rare in adults with RA, if it occurs at all. Of all children with JRA, the most susceptible group are those with the mildest form of disease—those whose initial onset is monarticular and those whose course of disease is oligoarticular (pauciarticular); about one in five will develop this potentially serious eye inflammation. A monarticular onset, primarily of a knee, was found more often in JRA than in adult RA. This is difficult to explain, as is the relative infrequency among children of subcutaneous nodules. Fewer children (8%) had rheumatoid nodules than adults (20%). They occurred, however, in the same areas in both groups. Nodules are seen frequently at the elbow or may be found in any area of pressure or friction, such as the knuckles or at the back of the heels. Rheumatoid factor in the serum, as observed by either a positive sheep cell agglutination or latex fixation test, is present in only 10–25% of JRA patients, whereas in adults, it is found in 50-85%. Failure to appreciate the relative infrequency of rheumatoid factor in JRA constitutes one of the major pitfalls in early diagnosis.

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EARLY DIAGNOSIS

Clearly, it is precisely these major differences that have created much of the difficulty in the early diagnosis of JRA. The problem would be simpler by the recognition of three distinct modes of onset. These are *acute febrile* (*systemic*), *polyarticular,* and *monarticular*. The frequency, severity, and character of initial systemic and articular manifestations help to differentiate each.

An acute febrile or systemic onset is marked by systemic manifestations, including high fever, rash, generalized lymphadenopathy, splenomegaly, and heart involvement. Joint manifestations are variable; occasionally only arthralgia is present. In polyarticular onset, defined as the involvement of more than four joints, the arthritis predominates and is frequently generalized and symmetric, similar to RA in adults. Systemic manifestations are less prominent than in an acute febrile onset and fever is low grade. In a monarticular onset, arthritis is confined to a single joint, usually a knee; except for iridocyclitis, systemic manifestations are either absent or minimal.

TABLE 1

MAJOR DIFFERENCES BETWEEN 100 CHILDREN AND 100 ADULTS WITH RHEUMATOID ARTHRITIS

CONSECUTIVE REFERRALS TO AN ARTHRITIS CLINIC

Disease Feature	Frequency (%) Children	Adults
High Fever	20	3
Rheumatoid Rash	36	2
Chronic Iridocyclitis	8	0
Monarticular Onset	32	6
Subcutaneous Nodules	8	20
Rheumatoid Factor*	23	76

* By the latex fixation test, titer of 1:160 or greater.

Acute Febrile Onset. About 20% of all JRA patients present with an acute febrile or systemic onset. Recognition is easy when obvious arthritis is present in addition to several typical systemic manifestations of the disease (Fig. 1). Sometimes, however, only arthralgia is present and then the differential diagnosis can be difficult. In those without arthritis, the child's appearance may provide the first diagnostic clue. These children are irritable, listless, anorectic, and losing weight. They often wish to be left alone and assume a position of generalized flexion. Of the many systemic manifestations, fever and rash have the greatest diagnostic value. Both may be associated with generalized lymphadenopathy (particularly axillary and epitrochlear nodes), splenomegaly, hepatomegaly, pericarditis, myocarditis, pneumonitis, and a striking neutrophilic leukocytosis.

Fever. First, it should be remembered that high fever may precede detectable signs of obvious arthritis by weeks, months, or rarely, by years (6). Rectal temperatures must be taken every four hours around the clock in order to disclose the characteristic quotidian or double quotidian febrile pattern (Fig. 2). Typically, there are one or two daily temperature peaks above 102°F, occasionally even to hyperpyrexic levels (fever to 105°F) (6). Diurnal ranges are wide, often as much as 8°F or 9°F, so that both hyperpyrexia and normal or subnormal temperatures occur within the same day. The fever usually, but not always, responds to aspirin provided large quantities, up to as much as 130 mg/kg (1 gr/lb) daily, are given. If the critical daily quantity of aspirin is then reduced, even by as little as 150 mg, the fever recurs promptly (6, 7). Eventually the fever pattern may become relapsing or even periodic (6), at which time other typical features of JRA become manifest, thereby facilitating a correct diagnosis.

Rash. The rheumatoid rash develops in up to 90% of children with an acute febrile onset (8) (Fig. 1). It consists of macular or slightly maculopapular lesions, usually discrete but sometimes confluent, that are found on the trunk and extremities and occasionally about the neck and face. Rarely is the eruption pruritic.

While it may be persistent, more frequently the rash tends to be fleeting or evanescent, with migratory macules appearing briefly in the late afternoon

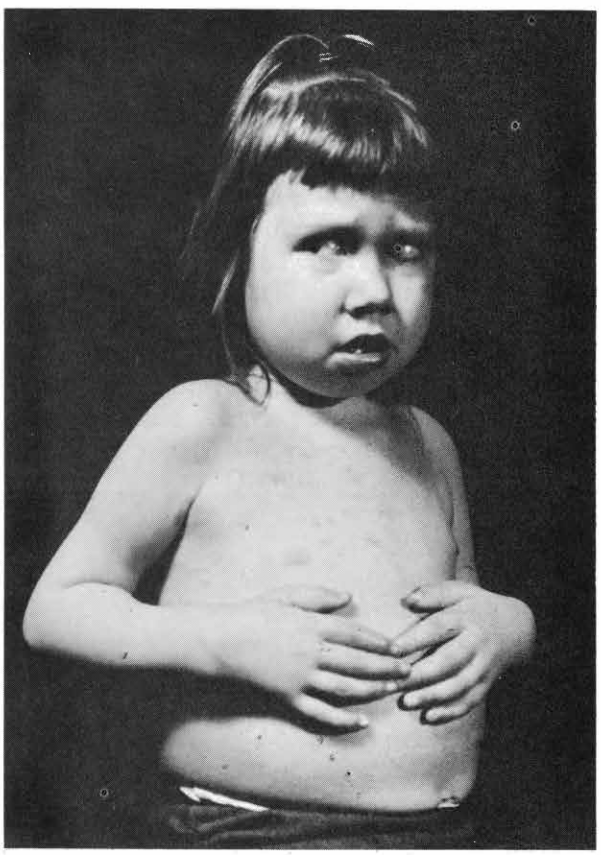

Fig. 1—The five-year-old girl above with acute febrile onset refuses to rotate her head because of cervical pain. Note typical anxious appearance, axillary prominence resulting from lymphadenopathy, symmetrical swelling of hand and wrist joints, and macular rash on the chest. (Reprinted by permission from *Med Clin N Amer* 25:567, 1968.)

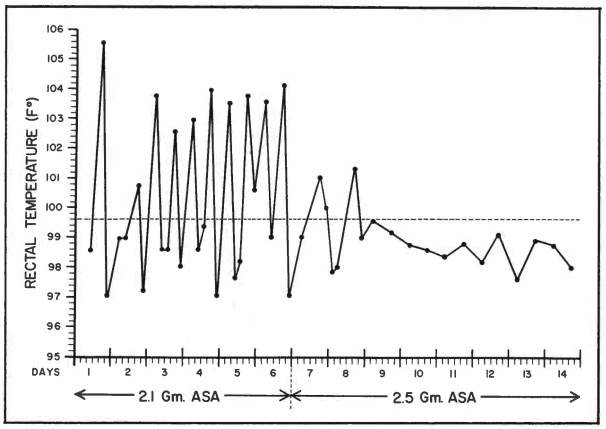

Fig. 2—The characteristic fever pattern of acute febrile onset, as shown above in a six-year-old boy, bears considerable diagnostic importance. High fever with its single and double daily peaks is suppressed only when the daily aspirin dosage is pushed to 110 mg/kg; patient's weight is 22.7 kg. (Reprinted by permission from *N Engl J Med* 276:11, 1967.)

or early evening, often in conjunction with fever spikes. The rash is most florid in areas where the skin has been rubbed or subjected to mild trauma, such as the light pressure of clothing. This manifestation is known as the Köbner phenomenon; it may be useful, diagnostically, when parents report rash that is not present at the time the child is being examined. The typical rash may be elicited by making scratch marks on the extremities or along the lower abdomen. Within several minutes, linear chains of isolated macules will appear that often persist for a day or two (Fig. 3).

Clearly, in these patients, it is the invariable combination of fever, rash, arthralgia, and other systemic features that leads one to an early diagnosis of acute febrile JRA. Otherwise, these patients are considered to have "fever of unknown origin." They will be hospitalized repeatedly, undergo exhaustive diagnostic workups, and receive needless courses of various antibiotics and even exploratory laparotomy. Differential diagnosis includes eliminating infection as well as other causes of high fever accompanied by rash and joint pain.

One of the most important disorders to rule out is systemic lupus erythematosus (SLE), which is unusual in a child younger than five years of age. Occasionally, the early skin lesions of SLE may resemble the rheumatoid rash, particularly when localized in the arms and trunk. Eventually, the rash of SLE takes on its typical *butterfly* distribution over the face and the diagnosis becomes apparent. Also, in SLE, the rash is not evanescent, is not timed to fever spikes, and does not produce an isomorphic response. Further, SLE may be confirmed by the presence of oral lesions, renal abnormalities, and LE cells. Finally, the absence of antinuclear antibodies virtually excludes a diagnosis of SLE.

Connective tissue disorders other than SLE must also be excluded. The rashes of Henoch-Schönlein (anaphylactoid) purpura, polyarteritis, and hypersensitivity angiitis are usually purpuric or ecchymotic. Also, each of these disorders is commonly associated with hypertension and renal manifestations which do not occur in JRA. In the child with arthritis and purpura, one must also suspect acute leukemia. Signs that suggest leukemia are severe anemia, leukopenia, and destructive lesions on x-rays of joints, each of which is distinctly unusual in the initial months of JRA (9).

Polyarticular Onset. This mode of onset, with arthritis of more than four joints, occurs in about half of all JRA patients. Most children appear ill and fail to grow. Rash, lymphadenopathy, and splenomegaly occur less frequently than in acute febrile onset, and the fever is low grade—peaking at 101-102°F once or twice daily. Polyarticular JRA is recognized easily when the pattern of joint involvement is generalized and symmetric and involves the hands (Fig. 4); however, when the arthritis is confined to large joints, is asymmetric, or is migratory, it may be confused with other

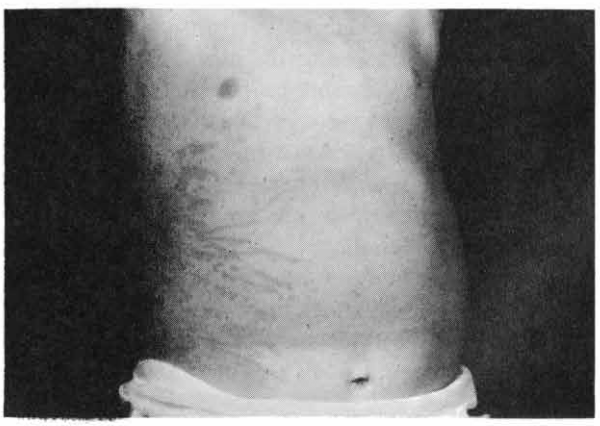

Fig. 3—Physician has evoked linear macules (Köbner phenomenon) by lightly scratching the patient's abdomen. (Reprinted by permission from *J Pediat* 72:611, 1968.)

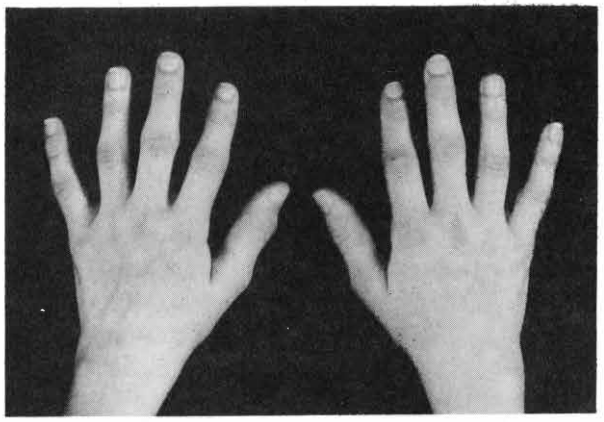

Fig. 4—Hands of a ten-year-old girl with polyarticular onset disclosing symmetrical swelling of proximal interphalangeal, metacarpophalangeal, and wrist joints. (Reprinted by permission from *Med Clin N Amer* 52:567, 1968.)

rheumatic disorders, particularly rheumatic fever (RF).

The individual macules of RF usually appear as open rings with distinct outer edges; there are only a few discrete macular rings with pale centers (Fig. 5). Typically, the erythema marginatum of RF extends centrifugally while the proximal skin returns to normal. It rarely lasts for more than a week or two, unlike the rash of JRA which persists for months.

Differential diagnosis is aided by close observation of the fever pattern; quotidian or double quotidian fevers suggest JRA, while remittent or sustained fevers point to RF. Several other features help to rule out RF. The most important include onset under four years of age, cervical involvement, a poor mucin clot on synovial fluid analysis, generalized lymphadenopathy, hepatosplenomegaly in the absence of cardiac failure, and arthritis lasting for more than 12 weeks.

Arthritis develops in 3% of all children who are vaccinated against rubella and involves primarily the knees or wrists. Developing within two-to-eight weeks after vaccination, the arthritis is self-limited, persisting for a few days to several weeks. The most important laboratory clue is a rising hemagglutination-inhibition antibody titer.

Another misdiagnosis is childhood dermatomyositis, primarily because the arthritis is so similar to that of JRA. Initially, loss of muscle strength is minimal and serum muscle enzyme levels may be normal while only arthritis predominates. Clues to an early diagnosis of dermatomyositis include a violaceous periorbital edema (heliotrope facies) and the presence of atrophic and scaly erythema of the skin over extensor surfaces of joints, particularly the knees, elbows, and hands.

Monarticular Onset. While the knee is the most common site of a monarticular onset (Fig. 6), the presentation may sometimes occur in other joints such as the ankle, elbow, wrist, or even a finger joint (10). Swelling, stiffness, and pain are usually minimal except when the hip is involved. Occasionally, painful tendinitis or bursitis, particularly of the heel, may be a presenting symptom. Symptoms or attention to the problem may be precipitated by trauma, a deceptive characteristic and a major pitfall in diagnosis of this mode of onset.

It is ironic that the mildest form of JRA, a monarticular onset, which comprises a third of all JRA patients, carries with it the serious threat of blindness from iridocyclitis (11). What makes this ocular manifestation particularly treacherous is that it is so often asymptomatic in its evolution, smoldering quietly for weeks or months until failing vision alone compels attention. It may be the initial manifestation of JRA or it may occur at any time in the course of the disease, even after the arthritis has remitted. If undetected and untreated, it may lead to blindness, primarily from band keratopathy and cataracts. While iridocylitis may occur in all children with JRA, those with monarticular onset are by far the most susceptible.

Except for the iridocyclitis, one cannot rely

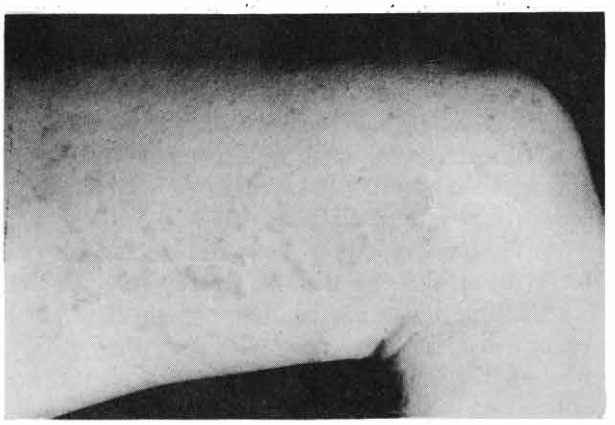

Fig. 5—In rheumatic fever, most macules appear as open rings with distinct outer borders and are larger than those observed in JRA. (Reprinted by permission from *J Pediat* 72:611, 1968.)

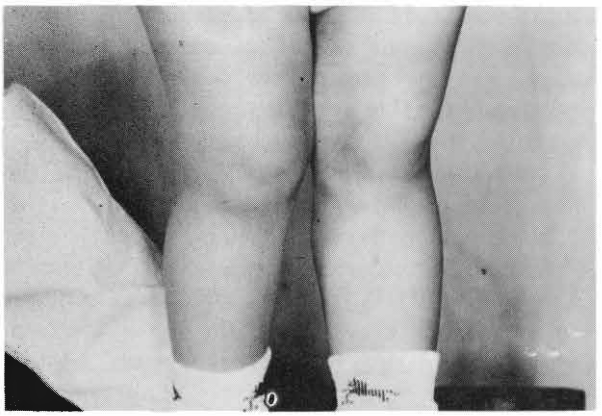

Fig. 6—Monarticular onset in a three-year-old girl with painless swelling of the right knee.

on systemic manifestations in this mode of onset. Lymphadenopathy and splenomegaly occur infrequently, while low-grade quotidian fever and rash are only occasionally present. Routine laboratory studies, such as the CBC and ESR, are frequently normal.

When one joint is involved, the single best test—if the disease is to be differentiated from acute infectious arthritis, which may rapidly destroy the joint—involves arthrocentesis and evaluating the synovial fluid. The first clues are provided by study of the gross appearance of the fluid. In JRA, the fluid will be clear-to-opalescent, in traumatic arthritis it may be clear-to-blood-tinged, while in infectious arthritis it will be cloudy or turbid. Examination of a drop of the fresh, uncentrifuged fluid under an ordinary microscope may reveal bacteria in infectious arthritis.

Next in order is inspection of the mucin clot. A few drops of synovial fluid are added to a small beaker containing about 10 ml of 5% acetic acid. If the sample is too small to allow this, the acetic acid can be drawn into the syringe, and even a slight amount of synovial fluid coating the syringe will suffice for the test. After allowing a minute for the clot to form, the container is shaken. A good clot forms a firm, ropy mass that does not fragment when shaken. A poor clot quickly flakes and shreds, clouding the solution. If the etiology of the arthritis is traumatic, the clot will be good; if rheumatoid, it will be good or poor; and if infectious, it will be poor.

The clinical laboratory is required for further synovianalysis. In traumatic involvement, the white cell count will be below 5,000, with fewer than 50% neutrophils; in JRA it will be between 15,000 and 25,000 and the neutrophils will range from 50-90%; in infectious arthritis, white cells will soar to counts of 50,000-100,000 and more than 90% will be neutrophils. Of particular interest are the paired determinations of serum and synovial fluid glucose levels; only in traumatic arthritis will these be about the same or show a disparity of less than 10 mg%. In JRA the difference will generally fall between 10-25 mg%, while in arthritis of infectious origin, it will usually be over 50 mg%

A negative tuberculin skin test usually rules out active tuberculous arthritis. If it is still necessary to pursue this possibility, synovial biopsy will generally show caseating granulomas and giant cells, while in JRA one will see nonspecific synovitis with hypertrophy, increased vascularity, and round-cell infiltration.

Laboratory and X-Ray Aids. Low-grade anemia and an elevated ESR are frequent, except in patients with a monarticular onset in whom values for hemoglobin, hematocrit, and the ESR are often normal. In acute febrile onset, there is a neutrophilic leukocytosis, usually between 20,000 and 30,000 cells, but sometimes as high as 50,000 cells/mm^3. A less striking leukocytosis occurs in a polyarticular onset, while white cell counts are often normal in a monarticular onset. Leukopenia rarely occurs in JRA; its presence should lead one to suspect leukemia or SLE.

The latex fixation test is positive in only 10-25% of children with JRA. Elevated titers are found primarily among children whose disease begins at 12-16 years of age. Antinuclear antibodies (ANA) are present in 10-30% of patients with JRA. Titers, however, are much lower than those found in children with SLE. Miller (12) has shown that the incidence of ANA is highest in girls and in patients under six years of age. ANA also seems to correlate with the presence of iridocyclitis (2). As for serum immunoglobulins, persistently elevated levels of IgG, IgA, and IgM are generally associated with an increased incidence of hip involvement and a poorer functional status. Serum electrophoresis may reveal low albumin and elevated β- and γ-globulins. It also serves to detect underlying hypogammaglobulinemia.

Antistreptolysin-O (ASO) titers, prolonged and moderately elevated, occur in about 30% of patients with JRA. The titers appear to be nonspecific and are also found in childhood tuberculosis, nephrosis,

and hepatitis (5). They can be inhibited by adding albumin to the test procedure, in contrast to specific ASO titers, which indicate recent infection with Group A β-hemolytic streptococcus. Intramuscular monthly injections of benzathine penicillin over a period of 12 months failed to lower titers of ASO in six of our patients with JRA, seemingly confirming the nonspecific nature of these titers (5).

Early x-ray findings are also nonspecific and include soft tissue swelling, early closure of epiphyses, and periosteal proliferation (5). Osseous erosions are only late findings, and their early presence should lead one to suspect leukemia or other forms of malignancy (9).

COURSE OF DISEASE

The subsequent course of disease is largely determined by the mode of onset (1) (Table 2). Three patterns of disease course may evolve: polycyclic acute febrile, polyarthritis, and oligoarthritis (also known as pauciarticular arthritis).

Ten of our 20 patients with an acute febrile onset have had recurrent flare-ups of systemic features, primarily high fever and rash, and little or no arthritis—a disease course designated as *polycyclic acute febrile*. The number of acute febrile attacks has varied from one to as many as ten in a single year. None of these ten patients has ever developed chronic arthritis. The other ten patients developed *polyarthritis* or the simultaneous arthritis of more than four joints. This occurred either at the onset of disease or only after months-to-years of recurrent acute febrile attacks. One girl had periodic fever that recurred regularly every three months for as long as nine years (from age 3-12) before she developed polyarthritis (6).

Of the 32 patients with a monarticular onset, 22 became *oligoarthritic*, with chronic, often recurrent, arthritis of only one-to-four joints; the ten remaining children developed polyarthritis.

Children with a polyarticular onset remained polyarthritic. Altogether then, 68 patients (48 with a polyarticular onset, ten with an acute febrile onset, and ten with a monarticular onset) eventually developed polyarthritis (Table 2). The course of polyarthritis took two forms, one characterized by exacerbations and remissions (58 patients), the other by an unremitting and progressively downhill course (13 patients)—resulting in marked joint deformities and an enhanced susceptibility to secondary amyloidosis. Amyloidosis should be suspected when there is progressive hepatosplenomegaly or when proteinuria develops that is unrelated to gold therapy.

MANAGEMENT

The aim of treatment is to provide a setting that will give the young patient a reasonably normal childhood. Management must include an active home program and regular follow-up care, because therapy may have to be changed at any time during the long course of the disease (13).

From the outset, the physician must recognize that he can only initiate and supervise therapy. The actual treatment will be given in the home, and the results will depend on establishing a working relationship with the parents. Therefore, the physician must educate and motivate the parents. He must teach them all about their child's disease—what they must do, and what they may expect. This is clearly the best way to reassure the parents and to allay their anxieties.

TABLE 2

THE ONSET, COURSE, AND OUTCOME OF 100 PATIENTS WITH JUVENILE RHEUMATOID ARTHRITIS OBSERVED FOR 15 YEARS*

				Outcome (%) After 15 Years		
Modes of Onset	%	Course of Disease	%	Still Active	Classes III-IV	Iridocyclitis
Acute Febrile	20	Polycyclic AcFeb	10	1	0	0
Polyarticular	48	Polyarthritis	68	28†	13†	2
Monarticular	32	Oligoarthritis	22	7	0	6‡

* During these 15 years, 88 of our 100 children were maintained only on aspirin. Only four children have received oral steroids for more than one year, two because of protracted iridocyclitis; eight are currently on gold therapy.

† Including three deaths, one from postsurgical sepsis, one from secondary amyloidosis, and one of suicide.

‡ Two children are blind in one eye.

Drug Therapy. *Salicylates.* While physical therapy is the most important part of home care, it can only be made possible by the use of antirheumatic drugs that reduce joint inflammation and increase mobility. Whatever the mode of onset, aspirin, in individualized doses, is the drug of choice. Active disease can be suppressed in most children by four-to-six daily doses, totaling 90-130 mg/kg (2/3-1 gr/lb) daily. Acute febrile disease requires the more frequent and higher doses.

Early signs of chronic salicylate intoxication can easily be overlooked. Parents must be instructed to watch for lethargy and episodic hyperpnea, changes that are especially important in a child who is too young to complain of tinnitus. When these symptoms occur, aspirin should be stopped for 24 hours, and then reinstituted at slightly lower doses.

Adrenocorticosteroids. These drugs are administered in heart failure due to myocarditis as well as in pericarditis, vasculitis, and protracted iridocyclitis. For short periods, they may also be given in acute febrile disease or to patients who either do not tolerate or who fail to respond to aspirin. Intra-articular injections of steroid may also prove beneficial, especially when one or two joints are so seriously inflamed that exercise and other rehabilitative efforts are being compromised.

Chrysotherapy. Children with polyarthritis who respond poorly to aspirin should be given a trial of gold therapy; however, only physicians experienced in its use and potentially serious side effects should prescribe and administer gold. Gold salts are injected intramuscularly at weekly intervals in a dose of 0.5-1 mg/kg. Complications are more frequent during the early months of therapy and in children less than six years old. Renal and hematopoietic side effects are potentially fatal, and it is, therefore, necessary to monitor the patients with weekly blood and urine tests. These tests must be evaluated before each injection.

Other Drugs. The antimalarial drugs are not recommended for routine use in children. Chloroquine is especially dangerous; the accidental ingestion of as little as one gram (four tablets) may cause rapid cardiorespiratory arrest. Serious toxic effects to phenylbutazone and oxyphenbutazone include hepatitis, thrombocytopenia, and agranulocytosis, which seem to occur more frequently in children than adults. For this reason, these drugs are currently contraindicated in children 14 years of age or under. So, too, is indomethacin. The place of immunosuppressive or cytotoxic agents has not yet been evaluated in JRA.

Eye Care. Susceptibility to iridocyclitis is enhanced in all patients with JRA. The recognition and therapy of iridocyclitis should be entrusted to an ophthalmologist. In those with minimal arthritis, the most susceptible group, slit-lamp examinations are required every three months, even if the arthritis is in remission. Patients who have had previous iridocyclitis should be examined on a monthly basis. All other JRA patients should be examined regularly by slit lamp at six-month intervals. Ophthalmologic screening should be routine until patients reach adulthood, at which time attacks tend to become acute and routine screening for silent iridocyclitis is no longer necessary.

Supportive Care. Appropriate rest, splinting, and exercise are fundamental to the prevention and correction of deformity. An increase in the hours of sleep at night and a nap during the day facilitate the resolution of synovial inflammation. Complete bed rest must be avoided, however, since this may cause undue flexion contractures and muscle atrophy.

Bivalved splints, made of plaster of paris or of plastic, are used to rest inflamed joints or to correct deformities. Traction, sandbags, or other means of keeping joints in proper alignment may be used in place of resting splints. For sustained deformities—a flexion contracture of a knee, for example—serial splinting may be useful. New bivalved splints are applied every one-to-two weeks as the range of motion improves. Splints may be removed daily to permit prescribed exercises and then replaced.

A physiatrist or physical therapist must teach the patient or parents a program of regular daily exercises that can be performed in the home. Daily activities, in addition to formal exercise, should be those that maintain strength and move joints primarily through motions of extension. Games and sports can help to achieve these goals, but those which involve sharp impact to the joints, such as basketball or football, must be avoided. Swimming is an excellent sport, since it promotes mainly extension-type movement and has the added advantage of the positive buoyancy of water. Keeping the child in school also assures that mental activities are maintained. Provisions must be made, however, to allow the patient more freedom than other children.

Surgery. Established deformities can be successfully corrected by a variety of procedures, even in

the presence of active disease. Early synovectomy, or the removal of granulation tissue early enough to prevent erosion of cartilage and bone, however, is an area of current controversy. The correct timing of the procedure and the proper selection of patients are the issues at the moment. Generally, children six years or younger are poor surgical risks because of their inability to cooperate fully with important postoperative measures.

PROGNOSIS

Generally, the prognosis for children is quite favorable (1, 2). Of our 100 patients now observed prospectively for 15 years, 64 are in remission (Table 2)—they are not taking any drugs and have no evidence of active articular or systemic disease. All 64 patients are in functional classes I and II and therefore capable of all ordinary activity.

Active disease was present in 36 patients, including the three who died. Of the 36, 28 have had a course of polyarthritis. Only one of ten patients with a polycyclic acute febrile course and seven of 22 with oligoarthritis continue to be active, affirming the benign nature of these two patterns of disease course.

Only 13 patients are in the unfavorable American Rheumatism Association functional classes III and IV and are capable of little or no activity. Each has had a course of unremitting polyarthritis and ten have had progressive hip involvement. Three of these patients died, one of staphylococcal bacteremia following knee synovectomy, one from secondary amyloidosis, and the third of suicide. Permanent stunting of growth occurred in seven of these 13 patients, six of whom also had pronounced micrognathia and progressive cervical involvement.

One of our most striking observations is that chronic iridocyclitis appeared and recurred primarily in patients with the least amount of joint involvement (Table 2). Of the eight patients with iridocyclitis, six had oligoarthritis, two of whom lost vision in one eye. Iridocyclitis subsequently recurred in three of these six children and in two when their arthritis had been in remission for as long as four years.

CONCLUSIONS

Varying degrees of systemic and articular involvement characterize the three modes of onset observed in JRA. Systemic features, notably high fever and rash, characterize an acute febrile onset which may be observed weeks, months, and even years before objective arthritis develops. In polyarticular onset, the arthritis predominates and is frequently generalized and symmetric, similar to adult RA. Monarticular onset, with arthritis confined to a single joint, is the mildest form; it poses, nevertheless, the major threat of potential blindness from chronic iridocyclitis.

Treating the patient with JRA is a long-range collaborative effort, the success of which will largely depend on early diagnosis along with home care and parental cooperation. For all modes of onset, and subsequently during active arthritis, aspirin continues to be the drug of choice. Management must include regular follow-up and comprehensive care to prevent joint deformity and blindness due to asymptomatic iridocyclitis. The prognosis is generally favorable, as gleaned from our 15-year prospective study of 100 patients, and this allows us to discard old fears that JRA is inevitably unremitting and largely intractable to treatment.

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