

ENTITLEMENT ELIGIBILITY GUIDELINES

RHEUMATOID ARTHRITIS

MPC 00302
ICD-9 714.0, 714.1, 714.2, 714.81

DEFINITION

Rheumatoid Arthritis (RA) is a chronic multisystem disease primarily involving the joints. It is characterized by inflammatory synovitis, joint destruction, muscle atrophy and bone destruction. Other areas of the body which may be affected include the lungs, eyes, blood vessels, and skin.

The guideline **excludes** Juvenile Rheumatoid Arthritis.

DIAGNOSTIC STANDARD

Diagnosis from a qualified medical practitioner is required. Appropriate history and physical examination data, and reports of relevant investigations must be provided.

Specialist consultation may be necessary in some cases. While investigations may include x-rays, serum rheumatoid factors, serum IgA, and antinuclear, antiperinuclear, or anti-stratum corneum antibodies, no specific laboratory tests are diagnostic of RA.

A positive rheumatoid factor is found in more than two-thirds of adults with the disease. The predictive value of the presence of rheumatoid factor in determining a diagnosis of RA is poor. The test is not useful as a screening procedure, but can be employed to confirm a diagnosis in individuals with a suggestive clinical presentation and, if present in high titer, to designate those at risk for severe systemic disease. Rheumatoid factor may appear transiently in normal individuals after vaccination or transfusion.

ANATOMY AND PHYSIOLOGY

The earliest pathology in rheumatoid synovitis is microvascular injury and an increase in the number of synovial cells. The rheumatoid synovium is characterized by the presence of a number of secreted products of activated lymphocytes, macrophages, and fibroblasts. The local production of these cytokines and chemokines appears to account for many of the pathological and clinical manifestations of RA.

CLINICAL FEATURES

The prevalence of Rheumatoid Arthritis is approximately one percent of the general population, with a three-fold female preponderance. First degree relatives of persons with RA have four times the expected rate of disease.

The incidence of RA rises dramatically during adulthood, the exception being men in their forties through sixties.

Some ethnic groups, such as North American Indians, have a much higher incidence of RA.

Diagnosis may be made primarily on clinical features. These include the presence of four of the following criteria of the 1988 Revised Rheumatism Association Criteria of for Classification of Rheumatoid Arthritis:

- *1) morning stiffness in and around joints lasting at least one hour before maximum improvement
- *2) arthritis of three or more joint areas
- *3) arthritis of hand joints
- *4) symmetric arthritis
- 5) rheumatoid nodules
- 6) serum rheumatoid factor
- 7) typical radiographic changes - erosions or unequivocal bony decalcification localized to, or most marked adjacent to, the involved joints of the hand and wrist

*(Conditions 1 through to 4 must be present for at least 6 weeks for criteria to be met.)

The clinical course of RA is variable. Some individuals experience mild self-limiting disease, while others have progressively severe disease with extra-articular manifestations which can be life-threatening.

While approximately 10 percent of those affected have an acute onset with severe symptoms and polyarticular involvement developing within a few days, the typical pattern is one of insidious onset with progressive joint involvement occurring over a period of months to years.

In establishing onset, it is to be noted that the initial symptom may have been a minor complaint. Medical attention may not have been sought for a period of time after symptoms commenced, but continuity of symptoms would be evident.

The disease course may be slow or rapid, and fluctuate over years. Partial or complete remission may occur, although symptoms inevitably return and involve previously unaffected joints.

Early symptoms are often non-specific and may include malaise, fatigue and generalized musculoskeletal pain. Pulmonary manifestations (diffuse interstitial fibrosis of nodules in lung parenchyma) may precede more typical manifestations by months to years. Only after several weeks or months do joints become involved, with initial minimal limitation of motion that progresses in severity. Pain, swelling, and tenderness of the joint regions are typical of established RA. Involved joints are swollen, warm, painful and stiff on arising or after inactivity. The pattern of joint involvement varies, but generally the small joints are affected before the larger ones. Symptoms usually appear in the small bones of the feet and hands (metacarpophalangeal and proximal interphalangeal joints), followed by the wrists, ankles, elbows and knees. The upper spine, lumbosacral region and hips are rarely involved.

Associated deformities are caused by destruction of tendons, ligaments and joint capsules. The most common deformities (which may also be produced by other conditions or injuries) include:

Fingers/Thumb

- boutonniere deformity
- swan neck deformity
- ulnar drift (MCPs)
- subluxation (thumb MCP)

Wrist

- radial deviation
- subluxation

Elbow

- valgus deformity
- proximal subluxations

Ankle/Hindfoot

- valgus deformity

Toes

- metatarsophalangeal joint subluxations
- hammer toes
- claw toes
- hallux valgus
- hallux rigidus

The deformed joints have reduced stability and restricted range of motion. Large synovial cysts may develop.

Radiological findings include joint effusions and peri-articular osteopenia. Erosions and narrowing of the joint base with loss of articular cartilage are also seen on x-ray.

RA in the cricoarytenoid joints may cause hoarseness or life-threatening upper airway obstruction if the joints become fused in adduction. Involvement of the temporomandibular joint may interfere with mastication.

Extra-articular manifestations are common and vary with the duration and severity of the disease. It is important to recognize that some of these manifestations may be caused by factors other than RA. The following are some of the extra-articular manifestations produced by RA:

- rheumatoid vasculitis - often manifesting as distal gangrene and/or skin ulceration of lower extremities
- pulmonary disease - diffuse interstitial fibrosis or nodule(s) in lung parenchyma
- rheumatoid nodule(s)
- eye involvement - keratoconjunctivitis sicca (Sjogrens syndrome); uveitis is seen occasionally
- nerve entrapment - depending on the particular joint involved
- osteoporosis - in the area of affected joints, or generalized, if RA results in significant and prolonged immobility

PENSION CONSIDERATIONS

A. CAUSES AND/OR AGGRAVATION

THE TIMELINES CITED BELOW ARE NOT BINDING. EACH CASE SHOULD BE ADJUDICATED ON THE EVIDENCE PROVIDED AND ITS OWN MERITS.

1. Idiopathic

Only modest progress has been made in determining the cause of RA. The vast majority of persons with RA have no known external cause. A variety of studies suggests that a blend of environmental and genetic factors is responsible.

2. Genetic susceptibility prior to clinical onset

There are a number of genetic variables which affect the development of RA.

The HLA-DR4 gene product is found in the majority of persons with classic symptoms; however, this gene type is also found in the population not affected by RA.

3. Exposure to silica dust prior to clinical onset or aggravation

Crystalline silica, or quartz, is an abundant mineral found in sand, rock (mainly granite), and soil. High-level exposure to respirable silica dust can cause chronic inflammation and fibrosis in the lung and other organs. The pathophysiological role of silica dust in RA may be based on its effects on the immune system.

For silica dust exposure to cause or aggravate RA, the following should be evident:

Heavy exposure to silica dust, such as might occur in an environment where granite or quartz is being drilled or excavated, should occur over an approximate 10 year period; *and*
Signs/symptoms of RA should develop during the exposure or within 25 years of cessation of the exposure.

Occupations associated with silica dust exposure include sandblaster and smelter.

2. Inability to obtain appropriate clinical management

Exclusions:

Despite research efforts to date, there is a lack of sufficient evidence at this time to establish for pension purposes causation and/or aggravation between the following and RA:

- infectious etiology
- immunization
- physical trauma
- stress

B. MEDICAL CONDITIONS WHICH ARE TO BE INCLUDED IN ENTITLEMENT/ASSESSMENT

- rheumatoid nodules

C. COMMON MEDICAL CONDITIONS WHICH MAY RESULT FROM RHEUMATOID ARTHRITIS AND/OR ITS TREATMENT

- Nerve entrapment where caused by RA, for example:
 - wrist joint: median nerve resulting in carpal tunnel syndrome
 - elbow joint: ulnar nerve entrapment
 - knee joint: peroneal nerve entrapment
- Aortic regurgitation
- Cardiac conduction abnormalities
- Interstitial fibrosis
- Nodular lung disease
- Systemic rheumatoid vasculitis:
 - Distal arteritis (ranging from splinter hemorrhage to gangrene)
 - Cutaneous ulceration (including pyoderma gangrenosum)
 - Peripheral neuropathy
 - Arteritis of viscera, including heart, lungs, bowel, kidney, liver, spleen, pancreas, lymph nodes, and testis
 - Palpable purpura
- Anemia
- Osteoporosis/stress fractures
- Scleritis/episcleritis
- Felty's syndrome

REFERENCES FOR RHEUMATOID ARTHRITIS

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