ENTITLEMENT ELIGIBILITY GUIDELINES

ANKYLOSING SPONDYLITIS

MPC 00305
ICD-9 720

DEFINITION

Ankylosing Spondylitis is a chronic, progressive, inflammatory disease of unknown etiology. It mainly involves the sacroiliac joints, and the axial skeleton. It may also involve peripheral joints and extra-articular structures.

Extraarticular manifestations include the following:

- eye disease
- cardiovascular disease
- pulmonary disease
- neurologic involvement
- renal involvement

Ankylosing Spondylitis is also known as Marie-Strumpell Disease, or Bechterew's Disease.

DIAGNOSTIC STANDARD

Diagnosis by a qualified medical practitioner is required, including copies of relevant test reports and other supporting documentation.

Although no laboratory test is specific for, or diagnostic of, Ankylosing Spondylitis, investigations are necessary as the diagnosis cannot be made on clinical grounds alone.

X-ray findings usually demonstrate sacroiliitis. The changes seen on x-ray begin with blurring of the cortical margins of the subchondral bone, followed by bone erosions, and eventually by sclerosis. The HLA-B27 gene is found in approximately 90% of persons with Ankylosing Spondylitis. Elevated erythrocyte sedimentation rate (ESR) and an elevated level of C-reactive protein, both of which are not specific for Ankylosing Spondylitis, are often present when the disease is active. Individuals with severe disease may have elevated serum alkaline phosphatase. Rheumatoid factor and antinuclear antibodies are negative unless coexistent disease is present. When Ankylosing Spondylitis results in restricted chest wall motion, pulmonary function

studies show decreased vital capacity and increased functional residual capacity.

ANATOMY AND PHYSIOLOGY

Ankylosing Spondylitis has distinct pathological changes at the enthesis, i.e. the area in which a ligament or joint capsule attaches to bone.

The inflammatory component of this disease causes bony erosions at the enthesis. The body seeks to repair these erosions, and this repair activity results in ossification at these sites. It is this inflammation-repair process that is the basis for many of the findings in Ankylosing Spondylitis.

Ankylosing Spondylitis is usually diagnosed during the second to fourth decade of life. It is more common in first degree relatives of those with Ankylosing Spondylitis. While a strong affiliation with the HLA-B27 antigen supports a genetic predisposition, the evidence suggests that other factors also play a role in susceptibility to Ankylosing Spondylitis. The HLA-B27 antigen is a generic maker found in 8% of North Americans of European (white) heritage. Of those who are HLA-B27 positive, the actual risk of developing Ankylosing Spondylitis is estimated to be 1 to 2 %. Only 20% of HLA-B27 positive first degree relatives of individuals with Ankylosing Spondylitis and who are positive for HLA-B27 will develop the disease.

CLINICAL FEATURES

Ankylosing Spondylitis is a progressive disorder, characterized by alternating flare-ups of active spondylitis and periods of inactive inflammation. Occasionally, the course is severe and unremitting.

The most frequent presenting complaint is recurrent low back pain which has persisted more than 3 months. It is often nocturnal in nature. Individuals will develop early morning lumbar spine stiffness, which is typically relieved by activity.

The hallmark of Ankylosing Spondylitis is sacroiliitis; it is often bilateral. Sacroiliitis develops early in the progression of the disease but, although symptomatic, changes may not become evident on x-ray for 7 to 10 years.
Spinal deformities related to Ankylosing Spondylitis may evolve for up to 10 years. They result in marked immobility and, subsequently, generalized osteoporosis. In extreme cases the entire spine may become fused in a forward flexed position. Untreated persons may develop kyphosis due to the flexed posture assumed because of pain.

Spinal stiffening may produce the characteristic manner of body movement, where the torso is moved as one block.

Decreased chest expansion (<4cm) may result as the axial skeleton becomes involved. The chest restriction results in a restrictive lung disease. This does not include asthma, as a restrictive lung disease is due to an abnormality of the chest wall which limits expiratory and inspiratory movements.

In some cases, Ankylosing Spondylitis also involves the peripheral joints. This involvement is typically asymmetric and usually affects the large joints of the lower limbs. Peripheral joint disease may be readily apparent from the onset and, in some cases, may dominate the clinical picture.

Approximately one-third of those with Ankylosing Spondylitis also develop systemic manifestations, of which uveitis is most common.

PENSION CONSIDERATIONS

A. CAUSES AND/OR AGGRAVATION

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

1. Idiopathic

   The precise etiology is unclear.

2. Genetic

   Although the precise cause of Ankylosing Spondylitis is unknown, there is a strong genetic component, i.e. HLA-B27.

3. Significant physical trauma: aggravation only

   Significant physical trauma will produce aggravation only in the site that is affected by significant physical trauma.

   For significant physical trauma to produce aggravation of Ankylosing Spondylitis, the following should be evident:

Significant physical trauma must occur to an area of the body where Ankylosing Spondylitis is active; *and*

Increased signs/symptoms of Ankylosing Spondylitis must be present on a continuous or recurrent basis for at least 6 months.

Significant physical trauma is a discrete injury that causes, within 24 hours of the injury being sustained, the development of acute symptoms and signs for which medical attention would normally or reasonably be sought.

4. Inability to obtain appropriate clinical management

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

- Chronic Mechanical Lumbar/Thoracic (Dorsal)/Cervical Pain
- Peripheral arthritis due to ankylosing spondylitis
- Enthesitis

C. COMMON MEDICAL CONDITIONS WHICH MAY RESULT IN WHOLE OR IN PART FROM ANKYLOSING SPONDYLITIS AND/OR ITS TREATMENT

- Restrictive lung disease
- Cauda equina syndrome
- Post-traumatic intervertebral fractures (the C5-C6 or C6-C7 level is the most commonly involved site)
- Osteoporosis
- Uveitis
- Aortic regurgitation
- Cardiac conduction abnormalities
- Atlantoaxial subluxation
- IgA nephropathy

REFERENCES FOR ANKYLOSING SPONDYLITIS

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    2. Ibid pp1666-67
    3. Ibid p 1667

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