Medical Services

ANKYLOSING SPONDYLOSIS

1. General Information

One of the group of conditions known as the Spondyloarthropathies.

Ankylosing spondylitis (AS) is a chronic inflammatory disorder that preferentially affects the axial (central) joints. Sacroiliac joint involvement is the hallmark of the disease. Peripheral joints may also be involved, especially the hips, shoulders and temporomandibular joints. The disease also affects the areas of tendon and ligament attachments to bone (enthesopathy) causing achilles tendinitis and plantar fasciitis. In chronic progressive disease there is loss of lumbar lordosis with increased thoracic and cervical kyphosis which become fixed as a result of fibrosis and bony ankylosis.

1.1 Aetiology

The disease shows a strong association with HLA-B27. In Caucasians with AS 90% are found to be HLA-B27 positive, compared with a detected level in the general population of 8%. In black Americans the number with AS found to be HLA-B27 positive is less at 60%, but this accords with the reduced incidence in the general population of HLA-B27 of 4% [1].

(The association between AS and HLA B27 is very strong indeed. There are many other papers supporting this and HLA B27 is tested clinically in cases of suspected AS. See refs. 9 and 10).

As a group the spondyloarthropathies, including AS, can be triggered by certain bacterial and viral infections. Enterobacteria species have been variously implicated. It has been shown that antibodies to certain strains of Klebsiella react very specifically with cells from about 80% of B27-positive patients with AS, but not with B27-positive cells from unaffected individuals. This may be the result of a Klebsiella-derived factor modifying either B27 or some B27-associated receptor.

Other findings suggesting a gut-derived factor are the high frequency of subclinical ileitis (found on ileocolonoscopy) in patients with peripheral joint disease, elevation of serum IgA and a noted association of AS with ulcerative colitis, Crohn’s disease and Whipple’s disease [1].

In first degree relatives who are B27-positive the risk of developing AS is about 1 in 3, which contrasts with the risk in the general population of 2%. Although this increase supports a genetic basis there is lack of consistent concordance in twins, suggesting that environmental factors may also be operative.

Prevalence is variably quoted as between 0.02% and 0.22% of the population [2] with a male : female ratio of 6 :1 at age 16 yrs. decreasing to 2 :1 at age 30 yrs. It is, in general, much less common in dark skinned races.

2. Diagnosis

2.1 Common symptoms and signs

- The clinical diagnosis of AS is critically dependent on sacrolliac joint involvement and the radiological appearance of these joints. The X-ray grading is given below with the New York criteria. The ypical presentation in the young adult is insidious onset of back pain and stiffness. Initial involvement of the sacro-iliac joint often produces pain that radiates to one or both buttocks and sometimes to the back of the knee or thigh.

- Early morning low back pain is common. This often wakens the patient from sleep and continues to cause discomfort until the patient rises when relief is obtained with activity.

- Pain at other times of the day in the low back, buttocks and upper thighs, often from sacroiliitis, is similarly relieved by exercise and activity.

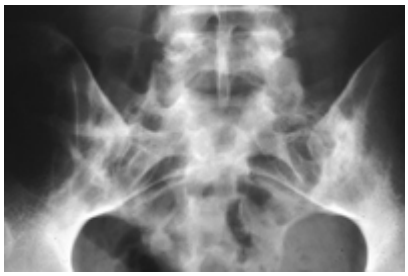
- Most patients with long standing AS have obvious spinal deformities and bend only from the hips and shoulders when attempting to touch their toes.

- AS occurs most commonly in young white males in their 20s, and onset is rarely seen over the age of 40 years.

- “Bamboo” spine is a late radiographic finding (2).

2.2 Confirming diagnosis

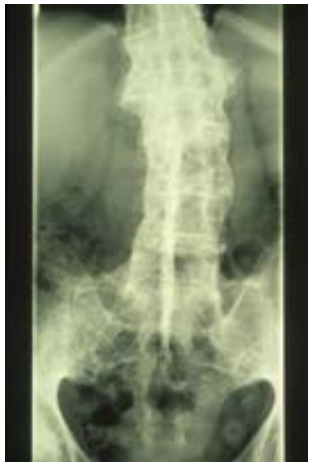
Diagnostic criteria were first proposed in Rome in 1961, and although they included sacro-iliac joint X-rays the interpretation of these was found not to be satisfactory.



A grading system was developed and the recommendations of this led to a revision of the Rome criteria in New York in 1966, with subsequent modifications again in 1984.

2.2.1 MODIFIED NEW YORK CRITERIA 1984 (1)

1. Low back pain of at least thress months duration improved by exercise and not relieved by rest.
2. Limitation of movement of the lumbar spine in saggital and frontal planes.
3. Chest expansion decreased relative to normal values for age and sex.
4. Bilateral sacroiliitis grade 2-4.
5. Unilateral sacroiliitis grade 3-4.



Definite AS if 4. or 5. and one of 1 – 3 is present.

(X-ray grading : 1 = suspicious
2 = minimal abnormality
3 = unequivocal abnormality
4 =severe abnormality, total ankylosis)

The ESR is mildly elevated in most patients as are other active phase reactants such as serum IgA levels. A positive test for HLA-B27 is usual but not specific.

Notably negative are tests for both rheumatoid factor and antinuclear antibodies.

2.3 Differential Diagnosis

2.3.1 Herniated intervertebral disc

The effects of this condition are limited to the spine and there are no systemic manifestations such as malaise or weight loss. All laboratory tests, including ESR, are normal. It may be confirmed by CT or MRI scanning.

2.3.2 DISH syndrome

This condition (diffuse idiopathic skeletal hyperostosis) may resemble AS clinically and on x-ray, and present with spinal pain, stiffness and loss of spinal movement. However, when it occurs in men over 50 years, the sacro-liliac joints are not involved and there is no link to HLA-B27. The ESR is also normal.

2.3.3 Reiter's syndrome

Criteria have been proposed for the definition of Reiter's syndrome as an episode of peripheral arthritis of more that one month's duration with nonbacterial urethritis or cervicitis.

2.3.4 Psoriatic Arthritis

Psoriatic arthritis affects both peripheral and axial joints. In general there is good correlation between kin activity and joint involvement. In more that half of the cases skin involvement appears first. Males and females are affected in equal numbers. Sacroiliitis is seen in 25 % to 40% of patients (“). The clinical course of psoriatic arthritis appears to be less disabling than that of AS and the presence of HLA-B27 does not appear to influence disease progression. Although patients with psoriatic arthritis have radiologic progression, this process often remains clinically silent and may not compromise spinal mobility.

3. Treatment

There is no specific therapy. Management is aimed at relieving pain and inflammation and maximising function through physical therapy and exercise. Dry and wet heat in combination with exercise directed towards the preservation of functional posture, joint mobility and muscular strength are useful. Despite medical management, the spine usually fuses in patients with ankylosing spondylitis and one of the goals of therapy is to ensure that fusion occurs in the most functionally acceptable position possible.

3.1 Nonsteroidal anti-inflammatory drugs

Are the cornerstone of drug therapy. Indomethicin (or another drug of the indole group), tolmetin or sulindac are the NSAIDS of choice. Entheses, particularly, become much less tender after one week's therapy.

3.2 Phenylbutazone

Has a place in AS resistant to other NSAIDs. Because of its potentially devastating side effect, haematologic parameters must be closely monitored.

3.3 Methotrexate

Has been used with promising results in patients with a more sever course of disease (2).

3.4 Sulphasalazine

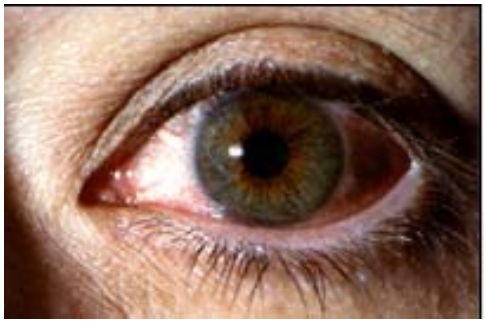
Is reserved for those with active peripheral joint involvement which has progressed despite other local measures, including intra-articular joint injections.

4. Prognosis

4.1 Life expectancy

Survival in patients with AS is close to normal. Deaths are usually due to cardiac involvement or fractures of the cervical spine. Mortality is estimated to be about 6%. Radiotherapy to the spine is no longer used as a treatment modality, but those who were treated in this way demonstrate a two-thirds excess mortality with a five fold increase indeaths due to leukaemia, and a 60% increase in carcinoma within the irradiated field.

4.2 Course of condition



The majority of patients do well, with about 90% either fully independent or minimally disabled. Overall 40% develop severe spinal restriction, with the early onset of peripheral arthritis or iritis being associated with more severe spinal restriction. Occasionally the course is severe and progressive, resulting in pronounces incapacitating deformities.

The longer-term prognosis is best determined after ten years of disease. 74% of those with mild restriction after ten years do not develop hip involvement within the first ten years appear to retain normal hips thereafter.

A review in 1986 of 100 patients with AS of at least five years duration (7) confirmed that 84% were in employment, with 24% reporting enforced changes of job due to the disease. Overall 9% considered they had required to take less satisfactory employment. 18% of those reviewed were registered as disabled.

Difficulties with six activities, bending, lifting, using a vacuum cleaner, gardening, painting walls and painting ceilings were assessed and graded 0-3 (where 0 = no difficulty, 3 = cannot do. A composite score of over 15 being accepted as evidence of gross impairment). On the basis 22% were considered disabled for day-to-day activities with peripheral joint involvement a significant factor.

The effects of loss of spinal mobility were most clearly demonstrated by activities required in driving. All round vision, reversing and reaching controls were reported as causing difficulties.

Other reported studies confirm inability to continue in work as affecting 30% of patients, and good functional progress after ten years of disease in 75% of those affected (8).

5. Main Disabling Effects

Ankylosing spondylitis affects primarily the axial skeleton although peripheral joints may be involved at sometime during the course of the disease. Once limitation of spinal movement begins, it progresses to involve movements in all planes, including lateral flexion as well as forward flexion and extension. There is frequently flattening of the lumbar lordosis. With more advances disease a thoracic kyphosis develops with restriction of thoracic rotation and chest expansion. In advanced and severe cases cervical spine movements are also restricted in all planes with dramatic limitation of lateral flexion.

Most patients with long standing disease have obvious spinal deformity and bend only from their hips and shoulders when attempting to touch their toes. The combination of the thoracic and cervical stiffness may lead to difficulties with forward vision or marker hyperextension of the cervical spine.

Peripheral arthritis generally involves the larger joints with the hip and shoulder most frequently involved followed by the knee, ankle and metatarsophalangeal joints. It occurs at some time in the course of the disease in 60% of patients [3]. This is an inflammatory arthritis with elevated white blood cell counts in the joint fluid.

X-rays may show erosive changes or new bone formation and bony ankylosis in 30% of those people who develop peripheral arthiritis. New bone formation or ankylosis does not occur in rheumatoid arthritis (4). Episodes of arthritis occur in flares which tend to persist for at least one month. However only 28% of the patients with arthritis had recurrent flares and in most instances the flares were many years apart (3).

Calcifications of the ligamentous bridges are the hallmark of AS in the cervical, thoracic and lumbar vertebrae. The apophyseal joints fuse and the normal curvature of the spine and configuration of the vertebral bodies is lost. This process leads to a rigid spine which is prone to injury, especially fracture.

The spondylitic spine also tends to be osteoporotic which results in an increased tendency to injury, even as the result of apparently trivial trauma.

Ankylosis of the thoracic spine can lead to fixation of costovertebral junctions and kyphosis. This usually causes slight changes in pulmonary function. Total lung capacity is usually preserved but vital capacity is frequently decreased to 65% to 88% of predicted normal range (5). Diaphragmatic compensation, even in advanced cases of thoracic fixation, maintains relatively normal pulmonary function. Patients with AS have decreased exercise tolerance that correlated with the decrease in vital capacity. As patients' lifestyle and level of activity also correlates with exercise tolerance, affected individuals should participate in a regular exercise programme to maintain exercise tolerance. Treatment

with anti-inflammatory agents does not alter pulmonary function.

Fibrotic changes in the upper lung lobes is reported in 30% of patients (2). This abnormality generally appears late in the course of the disease, often in the absence of any painful symptoms of underlying skeletal disease and is usually asymptomatic. Its appearance does not seem to correlate with the severity of ankylosis [5]. In 1.3% of a large study, progression to cavitation or bulla formation was seen with secondary infection with aspergillus leading to cough and significant haemoptysis.

Enthesopathy occurs particularly at the heel at the insertions of the plantar fascia, and the Achilles tendon to the calcaneum. This enthesopathy is associated with pain, tenderness and swelling. It can also occur at the iliac crests, ischial tuberosities and greater trochanters.

Acute anterior uveitis occurs at any time in the course of the disease and is seen in up to 20% of patients. It manifests as a painful; red eye with altered vision. Either eye may be affected in separate episodes but simultaneous involvement is uncommon.

Asymptommatic cardiac disease may occur in 20% to 30% of affected patients as a sub-clinical aortitis or ECG documented abnormality in the aortic root. However, symptomatic and detectable aortic valve incompetence or dilatation of the ascending aorta occurs in up to 10% of affected patients with the incidence gradually increasing with duration of disease (5).

Neurological problems, other than those associated with fractures, may result from atlantoaxial subluxation. Chronic arachnoiditis may accompany cauda equina syndrome with loss of function in the legs and disturbance, and incontinence, of bladder and bowel function.

6. References

[1] Khan MA et al.
Ankylosing spondylitis and other spondyloarthropathies.
Rheum Dis Clin North Am. 1990 Aug; 87: 140-147
[2] Espinoza LR.
Spondyloarthropathies
Lippincotts Prim Care Pract. 1998 Jan; 2(1): 81 – 86
[3] Ginsburg WW. et al.
Peripheral arthritis in ankylosing spondylitis. A review of 290 patients followed up for more than 20 years.
Mayo Clin Proc. 1983 Sept; 58(9): 593 - 596
[4] Lisse JR.
Ankylosing spondylitis. An optimistic outlook.
Postgrad Med. 1989 Jul; 86(1): 147 –148
[5] Tanoue LT. Pulmonary involvement in collagen vascular disease: a review of the pulmonary manifestations of the Marfan syndrome, ankylosing spondylitis, Sjögren's syndrome and relapsing polychondritis.
J Thoracic imaging. 1992 Mar; 7(2): 62 –77
[6] Bakker C. et al.
Measures to assess ankylosing spondylitis: Taxonomy, review and recommendations.
J Rheumatol. 1993 Oct; 20(10): 1724 – 1730
[7] Wordsworth BP. et al.
A review of 100 patients with ankylosing spondylitis with particular reference to socio-economic effects.
Br J Rheumatol. 1986 May; 25(2): 175 – 180
[8] Rigby AS.
Review of U.K. data on the rheumatic diseases - 5 Ankylosing spondylitis.
Br J Rheumatol. 1991 Feb; 30(1): 50 – 53
[9] Brown MA, Pile KD, et al
HLA class I associations of ankylosing spondylitis in the white population in the United Kingdom
Ann Rheum Dis 1996 Apr 55:268-70
[10] van der Linden SM, et al
The risk of developing ankylosing spondylitis in HLA-B27 positive individuals. A comparison of….
Arthritis Rheum 1984 Mar 27:241-9