

ANKYLOSING SPONDYLITIS

Ankylosing spondylitis is a chronic inflammatory disorder of unknown etiology that primarily affects the spine, axial skeleton, and large proximal joints of the body.

Also called as Marie- Strumpell or Bechterew's disease or Poker's Back.

The term ankylosing spondylitis is derived from the Greek word Ankylos (bent or crooked) and spondylitis (inflammation).

Therefore it means the spine that can lead to stiffness and fusion of the back.

Extra articular involvement is also seen in this condition.

- Idiopathic
- Rheumatoid factor absent
- HLA-B27 present in > 90% cases
- Disease usually begins in the second or third decade.
- M:F= 3:1

PATHOGENESIS

There are 2 theories:

Receptors theory – HLA B27 is a receptor for etiologic factor (bacteria, virus, etc.). The resulting complex provokes production of cytotoxic T-cells which cause damage to cells with HLA B27 molecule. So, urinary or bowel infection can be a trigger for AS.

Molecular mimicry theory – bacterial antigen (or other damaging factor) in complex with other HLA molecule gets similar to HLA B27 properties and is been recognized by cytotoxic T-cells as HLA B27 or decreases the immune reaction at pathologic peptide (immunological tolerance).

PATHOLOGY

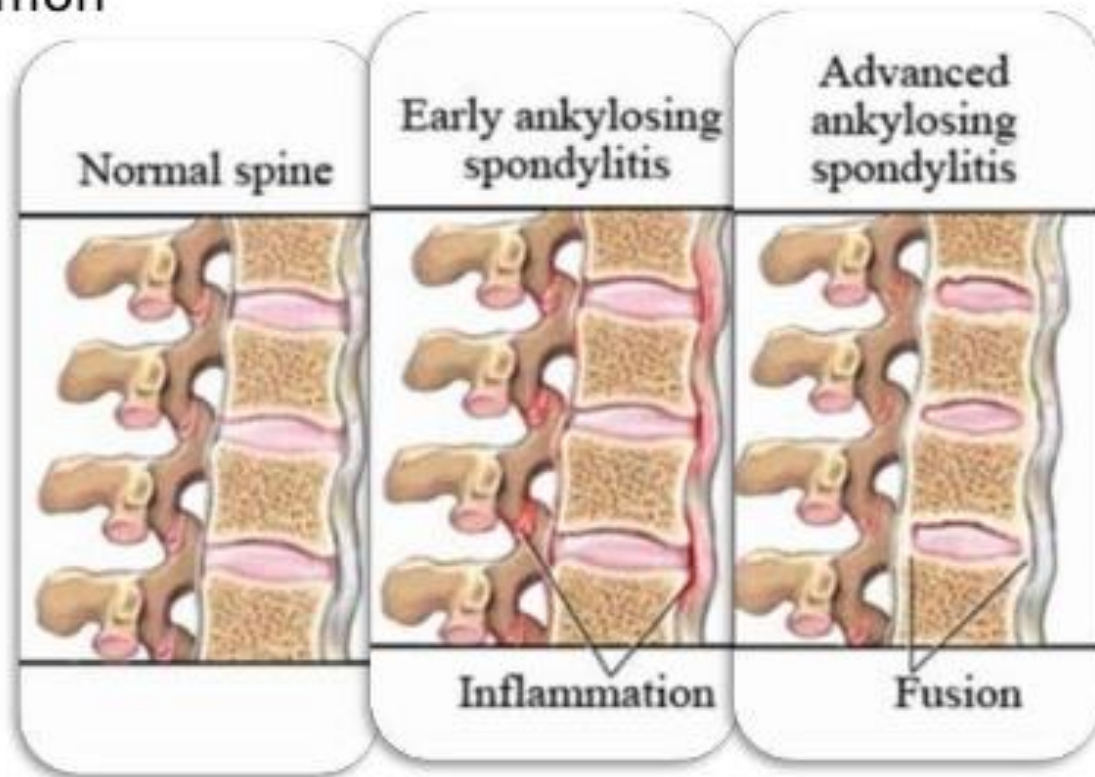
- The **enthesis**, the site of ligamentous attachment to bone, is thought to be the primary site of pathology
- **Enthesitis** is associated with prominent edema of the adjacent bone marrow and is often characterized by *erosive lesions* that eventually undergo ossification.
- **Sacroiliitis** is usually one of the earliest manifestations.

- The early lesions consist of **subchondral granulation tissue**, infiltrates of lymphocytes and macrophages in ligamentous and periosteal zones, and subchondral bone marrow edema.
- **Synovitis** follows and may progress to **pannus formation** with islands of new bone formation.
- The eroded joint margins are gradually replaced by **fibrocartilage regeneration** and then by **ossification**. Ultimately, the joint may be totally obliterated.

- Axial Arthritis (Eg, Sacroiliitis And Spondylitis)
- Arthritis Of 'Girdle Joints' (Hips And Shoulders)
- Peripheral Arthritis Uncommon

•Others:

- ☐Enthesitis
- ☐Osteoporosis
- ☐Vertebral Fractures
- ☐Spondylodiscitis
- ☐Costochondritis



The outer annular fibers are eroded and eventually replaced by bone, forming the beginning of a bony syndesmophyte, which then grows by continued enchondral ossification, ultimately bridging the adjacent vertebral bodies.

Ascending progression of this process leads to the “**bamboo spine**” observed radiographically.

Other lesions in the spine include diffuse osteoporosis, erosion of vertebral bodies at the disk margin, “**squaring**” of vertebrae, and inflammation and destruction of the disc-bone border. .



CLINICAL FEATURES

Initial symptom-

- Insidious onset dull pain in the lower lumbar or gluteal region
- Low-back morning stiffness of up to a few hours' duration that improves with activity and returns following periods of inactivity.
- Pain usually becomes persistent and bilateral. Nocturnal exacerbation +.
- Predominant complaint- Back pain or stiffness.
- Bony tenderness may present at- costosternal junctions, spinous processes, iliac crests, greater trochanters, ischial tuberosities, tibial tubercles, and heels.

Arthritis in the hips and shoulders (root joints) occurs in 25 to 35% of patients.

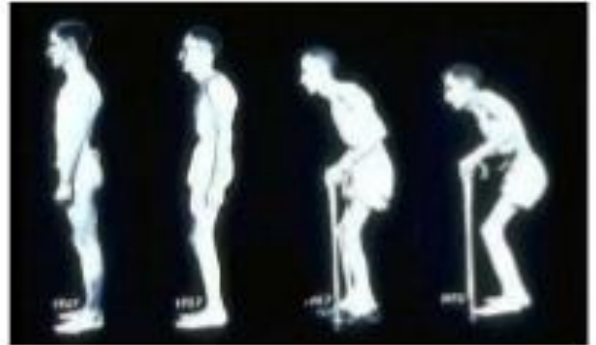
Arthritis of peripheral joints other than the hips and shoulders, usually asymmetric, occurs in up to 30% of patients and can occur at any stage of the disease.

Neck pain and stiffness from involvement of the cervical spine are usually relatively late manifestations.

Occasional patients, particularly in the older age group, present with predominantly constitutional symptoms such as fatigue, anorexia, fever, weight loss, or night sweats.

Flexion contractures of hip compensated by flexion at the knees.

DIAGNOSIS



- ▶ THE NEED, IS OF A **EARLY** DIAGNOSIS OF THE DISEASE ,WHICH WOULD HELP IN A BETTER PROGNOSIS AND THIS HAS BEEN MET BY THE TWO GROUPS NAMELY:
- ▶ **THE ROME CRITERIA** (1963)
- ▶ **THE NEW YORK CRITERIA** (1968)

THE ROME CRITERIA

- ▶ ACCORDING TO THIS ,AS IS DIAGNOSED IF **B/L SACROILIITIS** IS SEEN ALONG WITH ONE OF THE FOLLOWING.
 - a) **LBA & STIFFNESS OF 3 MONTHS DURATION NOT RELIEVED BY REST**
 - b) **PAIN AND STIFFNESS IN THE THORACIC SPINE**
 - c) **LIMITED LUMBAR SPINAL MOVEMENT**
 - d) **LIMITED CHEST EXPANSION**
 - e) **HISTORY OR EVIDENCE OF IRITIS OR ITS SEQUELAE**

THE NEW YORK CRITERIA

▶ CLINICAL CRITERIA:

- a) LIMITATION OF LUMBOSACRAL MOVEMENT IN THREE PLANES
- b) HISTORY OF PRESENCE OF PAIN AT DL JUNCTION WITH OR WITHOUT LUMBAR SPINE PAIN
- c) LIMITED CHEST EXPANSION OF 2.5CM OR LESS AT 4TH INTERCOSTAL SPACE.

THE NEW YORK CRITERIA

- ▶ RADIOLOGICAL CRITERIA BASED ON SACROILIAC JOINT RADIOGRAPHS
- ▶ GR 0 : NORMAL
- ▶ GR 1: POSSIBLY NORMAL(minimal sclerosis)
- ▶ GR 2: DEFINITE MARGINAL SCLEROSIS
- ▶ GR 3: DEFINITE EROSION AND SCLEROSIS
- ▶ GR 4: COMPLETE OBLITERATION AND ANKYLOSIS



Modified New York Criteria for the Diagnosis of AS

• Clinical Criteria

- Low back pain, > 3 months, improved by exercise, not relieved by rest
- Limitation of lumbar spine motion, sagittal and frontal planes
- Limitation of chest expansion relative to normal values for age and sex

• Radiologic Criteria

- Sacroiliitis grade ≥ 2 bilaterally or grade 3 – 4 unilaterally

• Grading

- Definite AS if radiologic criterion present plus at least one clinical criteria
- Probable AS if:
 - Three clinical criterion
 - Radiologic criterion present, but no signs or symptoms satisfy clinical criteria



Physical Examination

- The most specific findings involve loss of spinal mobility, with limitation of anterior and lateral flexion and extension of the lumbar spine and of chest expansion.
- Limitation of motion is usually out of proportion to the degree of bony ankylosis, reflecting muscle spasm secondary to pain and inflammation.
- Pain in the sacroiliac joints may be elicited either with direct pressure or with manoeuvres that stress the joints..

TEST and MEASUREMENT for AS

Cervical mobility

- Occiput-to-wall distance
- Tragus-to-wall distance
- Cervical rotation

Thoracic mobility

- Chest expansion

Lumber mobility

- ☐ Modified schober index
- ☐ Finger-to-floor distance
- ☐ Lumber lateral flexion

Occiput To Wall Distance / Flesche Test

❑ The severity of cervical flexion deformity in ankylosing spondylitis can be assessed by measuring the occiput to wall distance

(Flesche test).

❑ With the patient standing erect, the heels and the buttocks are placed against a wall; the patient is then instructed to extend his or her neck maximally in an attempt to touch the wall with the occiput.

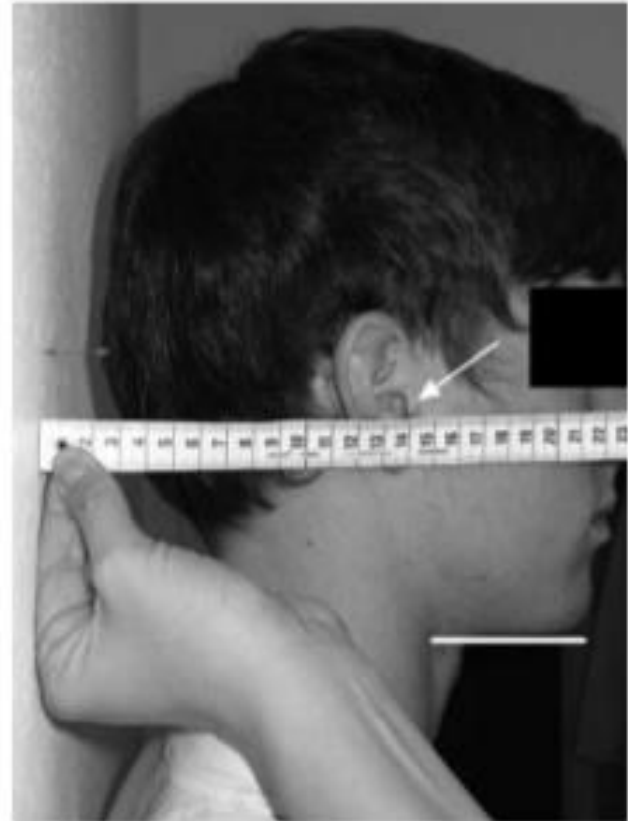
❑ The distance between the occiput and the wall is a measure of the degree of flexion deformity of the cervical spine.

- The occiput to wall distance should be zero



Tragus-to-wall distance

- Maintain starting position i.e. ensure head in neutral position (anatomical alignment), chin drawn in as far as possible. Measure distance between tragus of the ear and wall on both sides, using a rigid ruler. Ensure no cervical extension, rotation, flexion or side flexion occurs.



Cervical rotation

- Patient supine, head in neutral position, forehead horizontal (if necessary head on pillow or foam block to allow this, must be documented for future reassessments).
- Gravity goniometer / bubble inclinometer placed centrally on the forehead. Patient rotates head as far as possible, keeping shoulders still, ensure no neck flexion or side flexion occurs.



Normal ROM: 70-90°

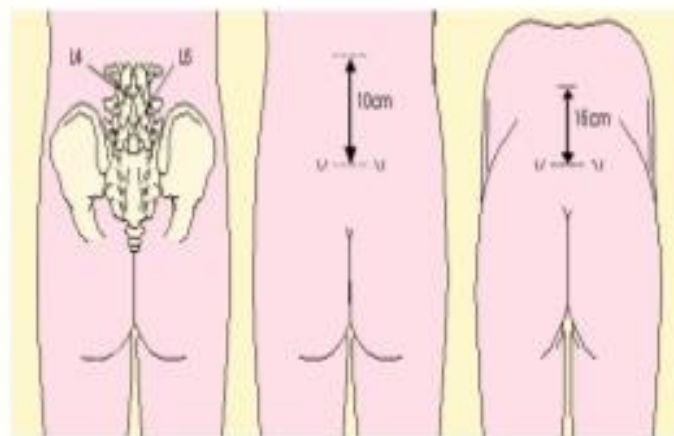
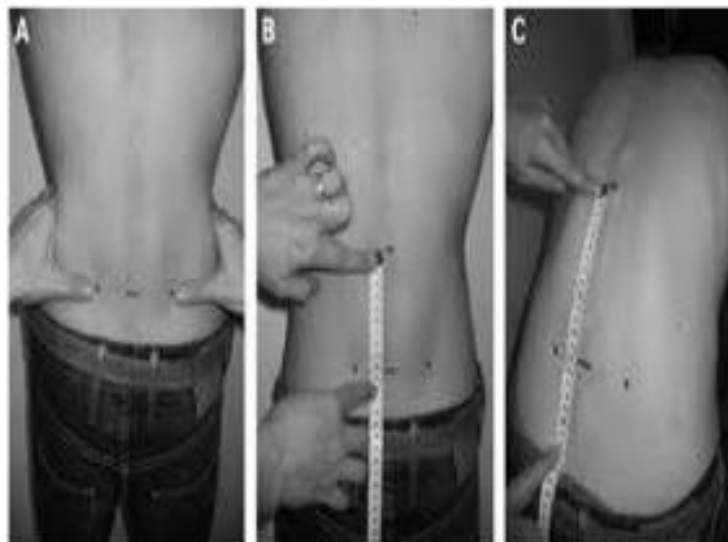
Chest expansion

- Measured as the difference between maximal inspiration and maximal forced expiration in the fourth intercostal space in males or just below the breasts in females. Normal chest expansion is ≥ 5 cm.



Lumbar flexion (modified Schober)

- With the patient standing upright, place a mark at the lumbosacral junction (at the level of the dimples of Venus on both sides). Further marks are placed 5 cm below and 10 cm above. Measure the distraction of these two marks when the patient bends forward as far as possible, keeping the knees straight



The distance less than 5 cm is abnormal

Finger to floor distance

- Expression of spinal column mobility when bending over forward; the dimension that is measured is the distance between the tips of the fingers and the floor when the patient is bent over forward with knees and arms fully extended.



Lateral spinal flexion

Patient standing with heels and buttocks touching the wall, knees straight, shoulders back, outer edges of feet 30 cm apart, feet parallel.
Measure minimal fingertip-to-floor distance in full lateral flexion without flexion, extension or rotation of the trunk or bending the knees.



Greater than 10cm is normal.



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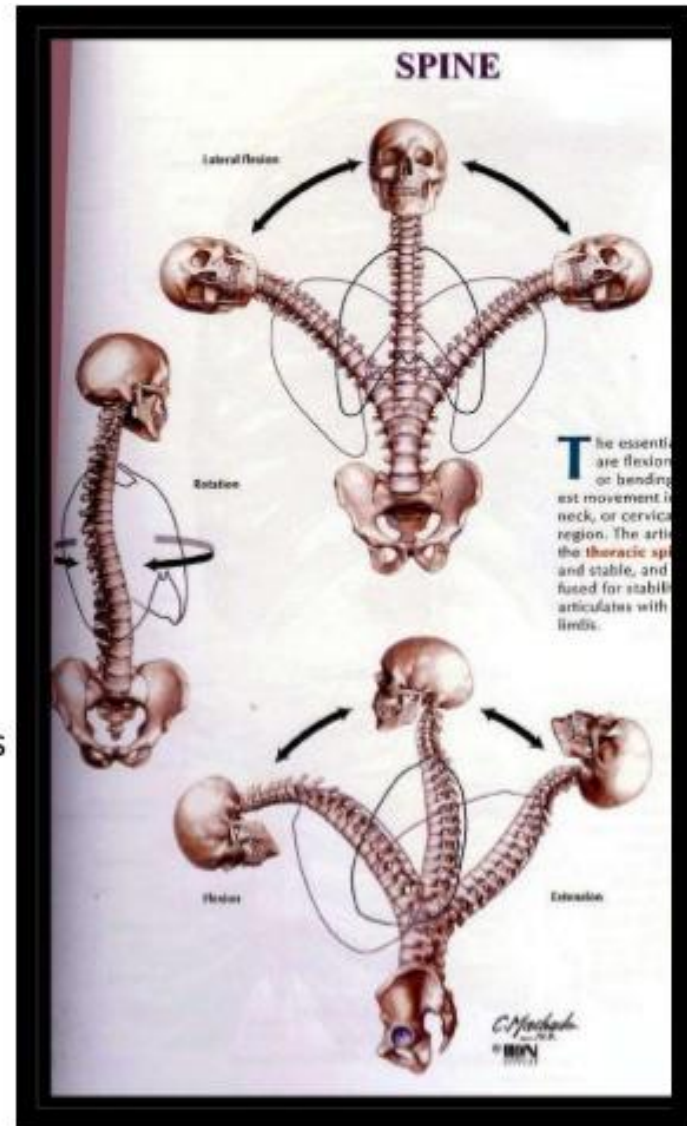
Range of motion

Cervical Spine

- Forward flexion: 0 to 45 degrees
- Extension: 0 to 45 degrees
- Left Lateral Flexion: 0 to 45
- Right Lateral Flexion: 0 to 45
- Left Lateral Rotation: 0 to 80
- Right Lateral Rotation: 0 to 80

Thoracolumbar spine

- Forward flexion: 0 to 90 degrees
- Extension: 0 to 30 degrees
- Left Lateral Flexion: 0 to 30
- Right Lateral Flexion: 0 to 30
- Left Lateral Rotation: 0 to 30
- Right Lateral Rotation: 0 to 30



EXTRASKELETAL MANIFESTATION

- Most common is **acute anterior uveitis**- typically unilateral, causing pain, photophobia, and increased lacrimation.
- Cataracts and secondary glaucoma are not uncommon sequelae.
- Inflammation in the colon or ileum.
- Aortic insufficiency.
- Third-degree heart block.
- Subclinical pulmonary lesions.
- Slowly progressive upper pulmonary lobe fibrosis.
- Retroperitoneal fibrosis.
- Prostatitis.

INVESTIGATIONS

- ▶ RADIOLOGICAL
- ▶ LAB INVESTIGATIONS
- ▶ OTHER INVESTIGATIONS

LAB. TESTS

- HLA B27: present in $\approx$ 90% of patients.
- ESR and CRP – often elevated.
- Mild anemia.
- Elevated serum IgA levels.
- ALP & CPK raised.

Lumbar spine:

- Loss of lordosis/ straightening
- Diffuse osteoporosis
- Reactive sclerosis- caused by osteitis of the anterior corners of the vertebral bodies with subsequent erosion (**Romanus lesion**), leading to “**squaring**” of the vertebral bodies.
- Ossification of supraspinous & interspinous ligaments “ **dagger Sign**”.
- Formation of marginal **syndesmophytes**,
- Later **Bamboo spine appearance** when ankylosis of spine occurs.
- **Odontoid erosion.**



JOINTS



DD

- The inflammatory back pain of AS is usually distinguished by the following five features:
 1. age of onset below 40
 2. insidious onset
 3. duration 3 months before medical attention is sought
 4. morning stiffness
 5. Improvement with exercise or activity.
- The most common causes of back pain other than AS are primarily mechanical or degenerative rather than inflammatory and do not show these features.

- Rheumatoid Arthritis.
- Paget's disease
- Metastatic diseases
- Gout
- Infections (brucellosis, staphylococcal, etc.) – Unilateral involvement
- Ochronosis.
- **Diffuse idiopathic skeletal hyperostosis (DISH):** Marked calcification and ossification of paraspinous ligaments, usually most prominent in the anterior spinal ligament and give the appearance of “flowing wax” on the anterior bodies of the vertebrae. Intervertebral disk spaces are preserved, and sacroiliac and apophyseal joints appear normal, helping to differentiate. (fine calcification of annulus fibrosus in AS).

TREATMENT MODALITIES

- ▶ DRUG THERAPY
- ▶ PHYSICAL THERAPY
- ▶ SURGERY

DRUG THERAPY : MAINSTAY

▶ IT IS TARGETED :

- a) GIVING SYMPTOMATIC RELIEF TO THE PATIENT.
- b) TO PRODUCE IMMUNOSUPPRESSION.
- c) SLOW DOWN THE DISEASE PROGRESS.

NSAIDS **Indomethacin** (up to maximum of 50 mg PO tid)
COX-2 inhibitors

Sulfasalazine, in doses of 2 to 3 g/d- Effective for axial and peripheral arthritis

Methotrexate, in doses of 10 to 25 mg/wk- primarily for peripheral arthritis

Local Corticosteroids injection- for persistent synovitis and enthesopathy
6. Medications to avoid- Long term Systemic Corticosteroids, gold and Penicillamine

Anti-TNF- α therapy - heralded a revolution in the management of AS.
Infliximab (chimeric human/mouse anti-TNF- α monoclonal antibody)
Etanercept (soluble p75 TNF- α receptor-IgG fusion protein)

have shown rapid, profound, and sustained reductions in all clinical and laboratory measures of disease activity.

Pamidronate, thalidomide, α -emitting isotope ^{224}Ra

Most common indication for surgery - severe hip joint arthritis, total hip arthroplasty.

- **Patient education:** Proper posture in standing , sitting and lying, sleeping on a firm mattress without pillow, stop smoking.
- **Physiotherapy:** The aim of exercise is to maintain joint movement and to build up muscles that oppose direction of the deformity with particular emphasis on the spinal extensors , intercostal and hip extensor muscles.