



Standard of Care: Ankylosing Spondylitis

Diagnosis: Ankylosing Spondylitis also known as **Marie-Strumpell disease** or **Bechterew's disease. (ICD-9: 720.0)**

Ankylosing Spondylitis (AS) is an inflammatory rheumatic disease, which primarily affects the axial skeleton manifesting in chronic back pain and spinal stiffness¹. The onset of the disease is gradual and characterized by aseptic inflammation at the sacroiliac (SI) joints. As the disease progresses it moves cephalad up the vertebral column. In advanced cases the stiffness progresses into spinal fusion and the normal lumbar lordosis is flattened and the thoracic kyphosis becomes exaggerated. Due to costovertebral involvement chest expansion can also become limited.

It is not uncommon to also find peripheral joints that are affected in 25% of the cases primarily involving the hips, knees, and shoulders. Acute anterior uveitis is also a common finding in 20-25% of patients with AS. Acute anterior uveitis may be a precursor to disease onset up to a few years prior. It presents as pain, photophobia, and increased lacrimation in the eye. In 5-10% of AS cases atrioventricular conduction defects, aortic insufficiency, and/or ascending aortitis will occur and is associated with spondylitic heart disease^{2,3}. AS is prevalent in approximately 0.1% of the general population and affects men greater than women with a 2-3:1 ratio^{4,5}. AS is found worldwide but is more prevalent in Caucasians.

A collection of laboratory findings can assist in the diagnosis of AS. The Erythrocyte sedimentation rate (ESR) will be elevated in 85% of the cases. C-reactive protein (CRP) is often elevated. Both are markers of inflammation. Human leukocyte antigen (HLA)-B27 is correlated with 90% of Caucasian patients and 50% of black patients with AS².

The earliest radiographic changes will be seen on x-rays of the sacroiliac joints with symmetrical blurring of the cortical margins or "postage stamp" serrated erosions in the lower third of the SI joints. Magnetic resonance imaging (MRI) and computed tomography (CT) can detect earlier stages of the disease and detect inflammation of the SI joints before plain radiographs³. Radiographically this appears as "squaring off" of the vertebrae⁶. Eventually, in the late stages of the disease x-rays demonstrating intervertebral fusion are known by the term "bamboo spine" due to their appearance⁶.

The etiology of AS is currently unclear, but it appears to be entirely genetic and immune system mediated. The entheses or site of ligamentous attachment to bone is implicated as the primary site of pathology in the SI joints and the spine in AS. The enthesitis is associated with local bone marrow edema and eventually bone erosions occur. The erosions are replaced by fibrocartilage and eventually ossification occurs. In the spine, syndesmophytes form from the outer annular portion of the intervertebral disc. Eventually these syndesmophytes grow vertically across the

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margin of the vertebral disc and bridge the adjacent vertebrae. This process is repeated up the spine⁶.

The pain is multifactorial in origin but is likely due to sacroiliitis, spondylitis, the formation of syndesmophytes leading to ankylosis, and is frequently associated with peripheral arthritis, enthesitis, and acute anterior uveitis¹

The diagnosis of AS is made based upon the modified New York Criteria⁷...

A. Diagnosis

1. Clinical criteria

- a. Low back pain and stiffness for more than 3 months which improves with exercise, but is not relieved by rest.
 - b. Limitation of motion of the lumbar spine in both the sagittal and frontal planes.
 - c. Limitation of chest expansion relative to normal values corrected for age and sex. (<5cm=Abnormal in young adult)
2. Radiologic criterion of sacroiliitis grade greater than or equal to 2 bilaterally or sacroiliitis grade 3-4 unilaterally. Grades are as follows...
- a. 0 = normal
 - b. 1 = suspicious changes
 - c. 2 = minimum abnormality (small localized areas with erosions or sclerosis)
 - d. 3 = unequivocal abnormality (moderate or advanced sacroiliitis with erosions, evidence of sclerosis, widening, narrowing or partial ankylosis)
 - e. 4 = severe abnormality (total ankylosis)

B. Grading

1. Definite ankylosing spondylitis diagnosis if the radiologic criterion is associated with at least 1 clinical criterion.
2. Probable ankylosing spondylitis if:
 - a. Three clinical criteria are present.
 - b. The radiologic criterion is present without any signs or symptoms satisfying the clinical criteria. (Other causes of sacroiliitis should be considered.)

Indications for Treatment:

Patients typically present in physical therapy with impairments of pain, loss of function, weakness and loss of muscle performance, fatigue, loss of flexibility and range of motion (ROM), and a knowledge deficit in self-management of symptoms and the nature of the disease process.

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Contraindications / Precautions for Treatment:

- A fused osteopenic spine is at great risk for fracture and the practitioner should use extreme caution when attempting to mobilize an AS spine ³.
- From the 1920's until the last few decades AS was treated effectively with spinal radiation. These patients carry a higher risk of cancer such as myeloid leukemias and hematologic malignancies. All AS patients should be asked if they have undergone this treatment ³.

Examination:

Medical History: Patients may report a history of acute anterior uveitis, diffuse pain in the lower back or SI joints described as deep and dull. They may report a history of morning stiffness, which improves with exercise. All results of imaging studies should be reported.

History of Present Illness: What symptoms are they having? How long have the symptoms been present? What mitigates or ameliorates symptoms? What worsens symptoms? Most patients complain of pain, stiffness, decreased spinal movement, and reduced energy. Were the symptoms insidious in onset? A careful and detailed history can be quite revealing and may prove useful when coupled with objective clinical information and imaging studies.

Social History: Suggested interview questions may include...

- Does patient live alone?
- Level of activity?
- Difficulty with ADLs?
- Frequency of exercise?
- Does the patient work?
- What behaviors have already been modified in order to accommodate the level of symptoms?
- What recreational activities would the patient like to do that currently is intolerable?
- What is/are the patient's goal(s)?

Medications:

Non-steroidal anti-inflammatory drugs (NSAIDs) appear to be effective with Indomethacin being the most effective. Sulfasalazine is useful for peripheral arthritis but not axial ². Anti-tumor necrosis factor (TNF) such as Infliximab or Etanercept has been extremely successful and show a 60% reduction in the Bath Ankylosing Spondylitis Disease Activity Index (BASDAI) ⁶.

In 2005 the ASsessment in AS (ASAS) International Working Group collaborated with the European League Against Rheumatism (EULAR) and compiled a group of evidence based medical recommendations for treatment of AS ⁸.

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- Optimal management of AS requires a combination of non-pharmacological and pharmacological treatments.
- NSAIDs are recommended as first line drug treatment for patients with AS with pain and stiffness. In those with increased GI risk, non-selective NSAIDs plus a gastroprotective agent, or a selective COX-2 inhibitor could be used.
- NSAIDs are insufficient, contraindicated, and/or poorly tolerated, however, analgesics, such as acetaminophen and opioids, might be considered for pain control.
- Corticosteroid injections directed to the local site of musculoskeletal inflammation may be considered. The use of systemic corticosteroids is not supported by evidence.
- There is no evidence for the efficacy of disease modifying anti-rheumatic drugs (DMARDs), including sulfasalazine and methotrexate, for the treatment of axial disease.
- Anti-TNF treatment should be given to patients with persistently high disease activity despite conventional treatments according to the ASAS recommendations. There is no evidence to support the obligatory use of DMARDs before, or concomitant with, anti-TNF treatment in patients with axial disease.

Examination: (Physical / Cognitive / applicable tests and measures / other)

This section is intended to capture the minimum data set and identify specific circumstance(s) that might require additional tests and measures.

Pain: Visual Analog Scale (VAS). Locus and nature of complaints, pattern am vs. pm, rest vs. activity. Patients may complain of waking from sleep and having to walk to relieve night pain, which is a rare complaint of mechanical low back pain³.

Muscle Performance: Perform specific manual muscle testing depending upon findings of upper and lower quadrant screening and patient's subjective reports of weakness.

Gait: Evaluate gait pattern and independence. Assess stair negotiation.

Neurological: Note the findings of deep tendon reflexes, sensory changes and/or muscle weakness. Assess reported numbness and/or paraesthesias. Note the locus of symptoms (dermatomal distribution) and level of severity associated with the defined level of activity. The most common neurological compromise is secondary to spinal fracture and neurological compromise affects one third of these cases. In patients with advanced AS, cauda equina syndrome can develop³.

Inspection/Posture: In cases of advanced disease a marked loss of the normal lumbar lordosis and a marked increase of the normal thoracic kyphosis may be seen.

Balance: According to Murray et al⁹ poor posture, decreased ROM and pain lead to a significant number of AS patients having impaired balance with eyes open or closed.

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The Berg balance test may be administered for objective measurement of balance impairment.

Function: The BASDAI^{10, 11} (see appendix A) has established content validity and has been used as an index to evaluate patients on a visual analog scale (0-10) quantifying:

- Fatigue
- Axial pain
- Peripheral pain
- Stiffness
- Enthesopathy

Palpation: Maneuvers or direct pressure over the SI joints can be used to stress the SI joints⁶.

ROM: A lack of spinal mobility is the hallmark of AS. All planes should be measured for active and passive ROM of the spine, shoulders, hips, knees and ankles as appropriate.

Special Tests:

Cervical Rotation (CROT): distance between tip of nose and the acromioclavicular joint in neutral and maximal ipsilateral rotation. The difference between the two positions is calculated for right and left rotation. Smaller differences indicate a more restricted range of motion. Measured with a tape measure¹².

Tragus-to-wall distance (TWD): measures the horizontal distance between the right tragus and the wall, standing with the heels and buttocks against the wall, knees extended and chin tucked in. The large the distance indicates worse spinal/upper cervical posture. Measured with a tape measure. This distance has been correlated with radiographic change in the cervical spine¹².

Fingertip-to-Floor distance (FFD): measures the distance between the tip of the right middle finger and the floor following maximal lumbar flexion, while maintaining heel contact with the floor and without trunk rotation. A smaller distance indicates greater movement. Measured with a tape measure¹².

Modified Schober index (MSI): is a useful measure of lumbar spine flexion. The patient stands erect, with heels together, and marks are made directly over the spine 5 cm below and 10 cm above the lumbosacral junction (identified by a horizontal line between the posterosuperior iliac spines.) The patient then bends forward maximally, and the distance between the two marks is measured. The distance between the two marks increases by greater than or equal to 5 cm in the case of normal mobility and by <4cm in the case of decreased mobility. Measured with a tape measure¹².

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Lumbar Lateral Flexion (LLF): distance between the tip of the ipsilateral middle finger and the floor following maximal LLF maintaining heel contact with the floor and without trunk rotation. Smaller distance indicates greater movement. Measured with a tape measure¹².

The literature suggests a strong correlation between the Modified Schober Index and the measurement of Lumbar Lateral Flexion (LLF) and AS specific lumbar spine radiographic changes¹².

Chest expansion is measured as the difference between maximal inspiration and maximal forced expiration in the fourth intercostals space in males or just below the breasts in females. Normal chest expansion is greater than or equal to 5 cm and any measurement less indicates impairment. Assess for limitation of motion in the hips or shoulders⁶. Due to restriction in the chest wall pulmonary function can suffer minor impairment with slight reduction in vital lung capacity, total lung capacity, and normal diffusion capacity³.

The CROT and/or the FFD are recommended for use in measuring reversible short-term change in spinal mobility. The MSI is recommended for use in measuring long-term irreversible change in spinal mobility. The ASAS recommends that chest expansion, MSI, and occiput (tragus)-to-wall be used to measure change in spinal mobility. After study and structured review of the literature Haywood et al recommends the use of the MSI, CROT and FFD for measurement of spinal mobility in AS¹².

Differential Diagnosis:

Sacroiliitis can be present in the following disease processes³...

- Hyperparathyroidism: if prolonged (can be induced by hemodialysis)
- Familial Mediterranean fever: can create sacroiliac erosions.
- Whipple disease: can create sacroiliac erosions.
- Paget's disease: can create sacroiliac erosions.
- Paraplegia: rarely can create sacroiliac erosions.
- Bechet disease: debatable.
- Tuberculosis: has an affinity for settling in the SI joints.
- Brucellosis: has an affinity for settling in the SI joints.
- Pyogenic sacroiliitis
- Malignancy: rare
- Retinoid treatment
- Synovitis-acne-pustulosis-hyperostosis-osteitis (SAPHO) syndrome

Vertebral hyperostosis can be present in the following disease processes³...

- Ankylosing hyperostosis or diffuse idiopathic skeletal hyperostosis (DISH):

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there exists exuberant osteophyte formation but they are more anterior. The SI joints are not affected and they do not have inflammatory back pain ².

- Ochronosis
- SAPHO syndrome
- Retinoid treatment

Enthesopathy can be present in the following disease processes ³...

- Gout
- Disseminated gonococcal infection
- SAPHO syndrome
- Retinoid treatment
- Bacillus Calmette-Guerin (BCG)-induced

Other related disease processes....

- Degenerative joint disease
- Rheumatoid arthritis: primarily affects multiple small peripheral joints. RA spares the SI joints and the majority of the spine except C1-C2. Pt's with RA usually have a positive rheumatoid factor present of lab results ².
- Osteitis condensans ilii
- Chondrocalcinosis
- Gout

Evaluation / Assessment:

Establish Diagnosis and Need for Skilled Services

Problem List (Identify Impairment(s) and/ or dysfunction(s))

- Knowledge deficit re: understanding of diagnosis, relationship of posture and upright activity on symptoms, correct use of joint protection techniques, modification(s) of activity level, proper positioning and stretching techniques, use of assistive device(s) and posture cues, and use of cold/ heat, massage and other comfort measures.
- Pain - management with conservative measures of positioning, pacing and/ or modification of functional activities, therapeutic exercise, and conditioning activities.
- Impaired muscle performance
- Impaired function.
- Impaired ROM (active and/or passive restrictions)

Prognosis

Some studies show that onset of the disease in adolescence correlates with a worse prognosis and early severe hip involvement is an indication of progressive disease. The disease in women tends to progress less frequently to total spinal ankylosis, however, there is evidence of isolated

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cervical ankylosis and peripheral arthritis⁶. The extent of spinal involvement has a direct correlation with the patients' functional status³.

Goals (with measurable parameters and with specific timelines)

- Independent home program and avoidance of provoking postures and activities; progressive independence in advancing home program over 6-8 treatments.
- Independent pain management with proper use of joint protection methods including posture, positioning, use of assistive device(s), pacing of activities, modification of activities, body mechanics and, use of comfort measures as needed (heat, ice, massage, relaxation techniques); 4-6 treatments
- Improve function including safe and proper transfers and ambulation with or without ambulatory device(s) and/or frequency or distance of walking; 2-4 treatments.
- Improve muscle performance; progressive improvement in quality of performance, number and nature of exercises, MMT and/or number of repetitions tolerated over 6-8 treatments
- Improve flexibility of identified tight soft tissue structures; measurable decrease per particular measures over 6-8 treatments. E.g. 25% decrease in hip flexor tightness as measured by the Thomas Test.
- Improve level of fitness; patient to return to conditioning activities or recreational activities. 6-8 treatments
- To maintain the patient's maximal potential movement, prevent postural deformities, improve muscle strength and fitness, and relieve pain⁵.

Age Specific Considerations:

The average age of diagnosis is 24 and the median age is 23. The most active disease phase occurs between the ages of 20 and 50 years⁵. Diagnosis after the age of 40 is uncommon³.

Treatment Planning / Interventions

Established Pathway ___ Yes, see attached. X No

Established Protocol ___ Yes, see attached. X No

Interventions most commonly used for this case type/diagnosis.

This section is intended to capture the most commonly used interventions for this case type/diagnosis. It is not intended to be either inclusive or exclusive of appropriate interventions.

Joint Protection Techniques: Body mechanics for transfers, lifting and carrying methods, positioning techniques, posture awareness and cues for maintaining pelvis in

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neutral, pacing and planning activities, modifications of activities, use of assistive device(s).

Therapeutic exercises: A progressive therapeutic exercise program with extension bias, stretching of identified tight muscles; lumbar and lower extremity stabilization exercise techniques; strengthening of any identified muscle weaknesses; postural reeducation, conditioning activities (recumbent bike, stationary bike, walking program, treadmill use), return to recreational sport activities (for example, swimming with use of modified strokes to avoid trunk extension). Breathing exercises and postural exercises should also be incorporated ².

Transfer and gait training: Balance, safety, pacing of cadence and planning for distance tolerated. Appropriate assessment for and use of any assistive device(s) should be addressed. Consider the use of 1 or 2 canes or a rolling walker for patients who require an assistive device(s). Some patients may benefit from a specialized walker with a seat option. Consider adjusting the height of the assistive device just slightly lower than usual to help patient achieve the postural correction needed for symptom management.

Manual therapy: Soft tissue and joint mobilization techniques to improve patient's level of symptoms and/or mobility. It may be inferred general fitness exercises will not isolate specific spinal segments that need to be mobilized. Manual treatments specifically designed to target a portion of the column are required ⁵. The effects of different hands-on techniques such as manual therapy, electrotherapy, and information on education programs need to be studied further with regard to patients suffering from AS.

Exercise Considerations: Studies show that patients with AS had some short term beneficial effects on function from individualized home exercise programs, but supervised group physical therapy programs were better than an individualized home exercise program (HEP). It has also been shown that patients who participated in group exercise (inpatient and outpatient) improved more than individual HEP due to non-physical factors such as mutual encouragement, increased motivation, and exchange of experiences with fellow sufferers of AS ⁵. When group exercise is combined with a three week thermal hydrotherapy, or spa treatment the intervention was better than weekly group physical therapy alone ¹.

Frequency & Duration: 6-8 treatments to achieve identified short-term goals over an 8 week period. Patients with lower tolerance levels may require more intensive intervention.

Patient / family education: It has been found that educating and advising patients about their condition is important because it enables the patients to manage their disease more effectively and know when to seek assistance at the appropriate time. It has been shown

that education improves motivation and reduces anxiety¹. The patient should understand the diagnosis and related basic anatomy, joint protection techniques including posture awareness, activity modifications, body mechanics, proper positioning and stretching techniques, use of assistive device(s) and heat/cold, relaxation techniques or massage.

Recommendations and referrals to other providers:

1. Occupational therapy- especially for ADL and additional training in joint protection methods if pain symptoms, loss of function and health status limit patient's independence and ease of function. Discuss with referring physician and explain your recommendations to the patient.
2. Additional support system/ counseling if patient has difficulty coping with the loss of independence and needs to modify activity level. Discuss with referring physician.
3. A referral will need to be made to an orthopedic surgeon if spinal alignment correction is needed to improve gait or field of vision. Typically this requires a large procedure involving a lumbar osteotomy³. In 5% of cases, patients with AS will require a hip replacement. These patients have an increased risk of peri-implant heterotopic ossification³. Referral to an orthopedic surgeon to perform total hip arthroplasty should be considered in patients with refractory pain or disability and radiographic evidence of structural damage, regardless of age¹. If vertebral fracture is suspected the patient should be immediately immobilized and treated with emergency medical management³. Referrals can be made for intra-articular corticosteroid injections to manage sacroiliitis⁸.

Re-evaluation / assessment

Standard Time Frame: every 30 days.

Other Possible Triggers: Worsening symptoms despite adhering to recommendations.

Discharge Planning

Commonly expected outcomes at discharge: Independence in home program of body mechanics, pain management with conservative measures, a routine stretching and strengthening program and independence in walking with or without an assistive device(s). According to Dr. Annelies Boonen the total cost-of-illness was \$6,720 per AS patient per year. Person's older in age at the onset of disease, performing manual jobs, having a lower educational level, or poor coping mechanisms are associated with withdrawal from the work force¹³.

Patient's discharge instructions: Continue prescribed home program.

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Appendix A
The Bath Ankylosing Spondylitis Disease Activity
Index.

PLEASE PLACE A MARK ON EACH LINE BELOW TO INDICATE YOUR ANSWER TO EACH QUESTION,
RELATING TO THE PAST WEEK.

(1) How would you describe the overall level of fatigue / tiredness you have experienced?

NONE _____ VERY SEVERE

(2) How would you describe the overall level of AS neck, back or hip pain you have had?

NONE _____ VERY SEVERE

(3) How would you describe the overall level of pain/swelling in joints other than neck, back or hips you have had?

NONE _____ VERY SEVERE

(4) How would you describe the overall level of discomfort you have had from any areas tender to touch or pressure?

NONE _____ VERY SEVERE

(5) How would you describe the overall level of morning stiffness you have had from the time you wake up?

NONE _____ VERY SEVERE

(6) How long does your morning stiffness last from the time you wake up?

0 hrs 1/2 1 1 1/2 2 or more hrs

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