The

Pocket Primer on the Rheumatic Diseases

Second Edition

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INTRODUCTION

For eight decades the *Primer on the Rheumatic Diseases* has been the standard text from which most medical students and house officers have learned rheumatology. I myself will never forget thumbing through an older edition of the *Primer* as a second-year resident, while waiting to review a perplexing patient with my tutor. Fortunately the tutor was running late with his own patients, so I had time to flip through the book – then much thinner – a couple of times. While turning the pages, perusing the features of those diseases whose names were still exotic to me, and considering my patient's history of conductive hearing loss and pulmonary nodules, a light went on when I stumbled eventually on a particular chapter. I still remember the jaw-dropping effect on my tutor of my announcement then that I had a patient with Wegener's granulomatosis. I think I became a rheumatologist that very moment!

Subsequent editions of the *Primer* have suffered from the inevitable "obesity creep," making it an outstanding reference textbook but virtually impossible to flip through quickly while awaiting one's tutor, and even more difficult to slip into the pocket of a white coat to carry on rounds. For this reason we have created the *Pocket Primer*, a mini version that cuts the larger book down to its essentials. Each chapter contains succinct descriptions of the diagnostic approach to and clinical manifestations, laboratory features, and strategies for the management and therapy of a particular rheumatic disease.

In editing the second edition of this book I am delighted to note the updating which has had to occur, not only of the first edition of the *Pocket Primer* but also of the more recent thirteenth edition of the larger *Primer on the Rheumatic Diseases*. The field of rheumatology remains lively, progressive, and fascinating to all who love the practice of medicine, regardless of specialty. Tutors, trainees, and practitioners will all find the *Pocket Primer* a handy guide to the rheumatic diseases and a superb overview of the latest advances in treatment.

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1 HISTORY AND PHYSICAL EXAMINATION

Conducting a careful history and physical examination to determine the onset and course of signs and symptoms, functional impairments caused by the arthritis, and the presence or absence of pathology or dysfunction in musculoskeletal structures is essential.

History

A thoughtful, detailed history is critical in determining the nature of the complaint and helps focus the clinical exam. Structure the history to answer these questions.

- Is the problem regional or generalized, symmetric or asymmetric, peripheral or central? Is it acute, subacute, or chronic? Is it progressive?
- Do the symptoms suggest inflammation or damage to musculoskeletal structures?
- Is there evidence of a systemic process? Are there associated extra-articular features?
- Has there been functional loss and disability?
- Is there a family history of a similar or related problem?

GALS (Gait, Arms, Legs, Spine) Screening

By asking three basic questions and systematically examining the patient's gait, arms, legs, and spine, the physician can rapidly screen for musculoskeletal disease.

- Have you any pain or stiffness in your muscles, joints, or back?
- Can you dress yourself completely without any difficulty?
- Can you walk up and down stairs without any difficulty?

Main Features of the Gait, Arms, Legs, Spine (GALS) Screening Inspection

Position/activity	Normal findings
Gait	Symmetry, smoothness of movement Normal stride length Normal heel strike, stance, toe-off, swing-through Able to turn quickly
Inspection from behind	Straight spine Normal, symmetric paraspinal muscles Normal shoulder and gluteal muscle bulk Level iliac crests No popliteal cysts No popliteal swelling No hindfoot swelling/deformity
Inspection from the side	Normal cervical and lumbar lordosis Normal thoracic kyphosis
"Touch your toes"	Normal lumbar spine (and hip) flexion
Inspection from the front Arms "Place your hands behind your head (elbows out)"	Normal glenohumeral, sternoclavicular, and acromioclavicular joint movement
"Place your hands by your side (elbows straight)"	Full elbow extension
"Place your hands in front (palms down)"	No wrist/finger swelling or deformity Able to fully extend fingers
"Turn your hands over"	Normal supination/pronation Normal palms
"Make a fist"	Normal grip power
"Place the tip of each finger on the tip of the thumb"	Normal fine precision, pinch
Legs	Normal quadriceps bulk/symmetry No knee swelling or deformity No forefoot/midfoot deformity Normal arches No abnormal callus formation
Spine "Place your ear on your shoulder"	Normal cervical lateral flexion

Examination of Specific Joint Areas

Any abnormalities detected through the GALS screening are followed with a more detailed examination.

Hand and Wrist

- **Alignment** – Inspect alignment of the digits relative to the wrist and forearm.
- **Nails** – Inspect nails for evidence of onycholysis or pitting suggestive of psoriasis. Inspect nail fold capillaries for redness and telangiectasia, indicative of a connective-tissue disease.
- **Skin** – Look for tightening of the skin around the digits, or sclerodactyly, typical of scleroderma.
- **Finger joints** – Inspect and palpate distal interphalangeal (DIP) and proximal interphalangeal (PIP) joints for swelling, which may signify bony osteophytes, synovitis, or an intra-articular effusion. Look for fullness in the valleys normally found between the knuckles (heads of the metacarpal bones), indicating swelling of the metacarpophalangeal (MCP) joints. Palpate individual hand joints to determine the presence of joint-line tenderness and effusion, important indicators of synovitis.
- **Palms** – Inspect the palmar aspect of the hands to identify atrophy of the thenar or hypothenar eminences, which can result from disuse due to articular involvement of the fingers or wrists or, in the case of the thenar eminence, carpal tunnel syndrome.
- **Wrist**
  - **Dorsum** – Note swelling on the dorsum of the wrist. Have patient gently wiggle the fingers to differentiate between synovitis of the wrist and tenosynovitis of the extensor tendons. Swelling tends to move with the tendons if it is a result of tenosynovitis. Palpate the dorsum of the joint to detect synovial thickening and tenderness suggestive of wrist joint synovitis.
  - **Ulnar styloid** – Check for swelling and tenderness in the area just distal to the ulnar styloid, where the extensor and flexor

carpi ulnaris tendons are directly palpable. This area is commonly involved in early RA.

- **Radial aspect** – Evaluate pain and tenderness confined to the radial aspect of the wrist, which are due most commonly to OA of the first carpometacarpal joint or to DeQuervain's tenosynovitis.
- **Function** – Evaluate global function of the hand by asking the patient to make a fist, and then fully extend and spread out the digits. Test pincer function of the thumb and fingers. Estimate grip strength by having the patient squeeze two of the examiner's fingers.

Elbow

- **Anatomy** – Identify the olecranon process, the medical and lateral epicondyles of the humerus, and the radial head. Locate the triangular recess formed in the lateral aspect of the joint between the olecranon process, in the lateral epicondyle, and the radial head. This recess is the point where the synovial cavity of the elbow is most accessible to inspection and palpation.
- **Bulging** – Look for bulging in this triangular recess to identify an effusion and synovitis. In contrast, swelling directly over the olecranon process is more suggestive of olecranon bursitis.
- **Contracture** – Have the patient extend the forearm as much as possible to detect the presence of a flexion contracture, an almost invariant feature of elbow synovitis.
- **Tennis elbow** – Palpate the lateral epicondyle for tenderness, suggesting lateral epicondylitis.

Shoulder

- **Observe** – Examine visually the entire shoulder girdle area, from the front and the back. Note shoulder height. Patients with rotator cuff tears often hold the affected shoulder higher than the other side.

- **Osteophytes** – Look for a prominent bump in the area of the acromioclavicular joint, often associated with osteophytes resulting from OA.
- **Atrophy** – Look for atrophy of the shoulder girdle musculature, an important sign of chronic glenohumeral joint pathology, as occurs in RA.
- **Effusion**s – Look for effusions in the shoulder joint, which are visible anteriorly just medial to the area of the bicipital groove and, if large enough, also are evident laterally below the acromion.
- **Motion** – Have the patient demonstrate active range of motion of the arms in several planes. Internal and external rotations of the shoulder are particularly sensitive to glenohumeral pathology. Test passive range of motion, particularly internal/external rotation and abduction.
- **Palpate** – Palpate the entire shoulder girdle, the cervical spine, and the thoracic wall.

Hip

- **Groin pain** – Pain in the groin (or less commonly the buttock) that tends to radiate down the anteromedial aspect of the thigh is often a result of hip arthritis.
- **Trochanteric pain** – Pain in the lateral trochanteric area is most often indicative of bursitis involving the trochanteric bursa.
- **Gait** – Note a "coxalgic" gait, quickly swinging the pelvis forward on the affected side to avoid weight-bearing on the affected hip, an indication of true hip disease.
- **Motion** – Test hip range of motion by having the patient actively flex, extend, abduct, and abduct the leg. Screen passive range of motion with the patient supine and "log rolling" the entire extended leg. The leg then is flexed maximally to assess completeness of this motion.
- **Internal rotation** – Note pain and loss of motion on internal rotation, particularly sensitive indicators of hip pathology.

Sacroiliac Joint

- **Palpate joint** – Palpate the sacroiliac (SI) joint with the patient lying flat on the abdomen. Find the SI joint by placing the palm of the examiner's hand around the iliac crest; the thumb tends to fall directly over the joint. Apply direct pressure with the thumb in this area to elicit tenderness in the SI joint.
- **Palpate sacrum** – Apply direct pressure over the sacrum to elicit pain in an inflamed SI joint.
- **Gaenslen's maneuver** – Perform Gaenslen's maneuver by having the patient hyperextend the leg over the edge of the examining table, thereby stressing the ipsilateral SI joint.

Spine

- **Observe** – Examine the spine initially with the patient standing and the entire spine visible. Evaluate the normal curvature of the spine, lumbar lordosis, thoracic kyphosis, and cervical lordosis by observing the patient from the back and the side, and noting any loss or accentuation of these curves.
- **Scoliosis** – Observe true scoliosis irrespective of the state of spinal flexion. A functional scoliosis due to leg-length discrepancy decreases with spinal flexion.
- **Motion** – Examine the range of motion of the entire spine in segments.
- **Lumbar** – The Schober test (movement of a 10-cm segment from the lumbosacral junction with spine flexion) is performed to specifically assess movement in the lumbar spine.
- **Lumbosacral** – Examine the lumbosacral area and perform a detailed neurologic examination of the leg in patients presenting with symptoms suggestive of a lumbar radiculopathy, such as pain and paresthesia shooting down the leg.
- **Thoracic** – Examine thoracic motion by measuring chest expansion at the level of the nipples. Ankylosing spondylitis can markedly reduce chest expansion from the normal 5–6 cm.

- **Cervical** – Test cervical range of motion with the patient upright and while lying down. Flex, extend, laterally flex (patient attempts to touch the ear to the shoulder), and laterally rotate (patient attempts to touch the chin to the shoulder) the head.

Knee

- **Observe** – Inspect the knee from both the front and the back, with the patient standing.
- **Atrophy** – Atrophy of the quadriceps usually indicates chronic knee pathology.
- **Suprapatellar bursa** – Inspect suprapatellar bursa for evidence of swelling due to synovial-fluid accumulation or synovial infiltration and thickening.
- **Varus deformities** – Varus deformities cause a bow-legged appearance, resulting most commonly from OA preferentially involving the medial compartment.
- **Valgus deformities** – Valgus deformities cause a knock-kneed appearance, more commonly associated with RA.
- **Palpate** – Palpate for tenderness the medial and lateral joint line with the patient lying supine and the knee in partial flexion.
- **Suprapatellar distension** – Distension of the joint in the suprapatellar area and in the medial and lateral compartments indicates large amounts of synovial fluid in the knee.
- **Ligaments** – Test the medial and collateral ligaments by gently applying varus and valgus stresses to the joint while the examiner firmly supports the joint with one hand or immobilizes the joint. Test the cruciate ligaments by using the drawer sign, in which anteroposterior stress is placed on the upper tibia with the knee in flexion; instability of the ligaments will result in the tibia moving back and forth relative to the femur.

Ankle and Hindfoot

- **Observe** – Examine the ankle and hindfoot as a unit.
- **Valgus deformities** – Detect valgus deformities of the ankle and hindfoot by inspecting the area from behind, with the patient standing.
- **Palpate** – Palpate the joint line of the ankle anteriorly. Boggy swelling and tenderness in this area are typical of ankle synovitis.
- **Achilles tendon** – Look for tenderness and swelling posteriorly, at the insertion of the Achilles tendon, usually indicating enthesitis.
- **Heel** – Look for tenderness in the heel, indicating plantar fasciitis, another enthesitis associated with spondyloarthropathies.
- **Talotibial joint** – Test for ankle synovitis by eliciting pain and limitation in the talotibial joint, capable only of dorsal and plantar flexion.
- **Subtalar joint** – Test the subtalar joint by rocking the calcaneus from side to side while holding the talus stable.

Midfoot and Forefoot

- **Arches** – Note pes planus (flat foot, collapsed arch) or pes cavus (high arch) with the patient standing.
- **Bunions** – Look for hallux valgus deformities, which cause bunions.
- **Daylight sign** – Observe a visible spreading of the toes caused by swelling of the metatarsophalangeal (MTP) joints.
- **Hammertoe** – Observe hammertoe deformity, which in cases of advanced RA results from the subluxation of the MTP joint.
- **Interphalangeal joints** – Observe inflammation of the interphalangeal joints of the toes, common with spondyloarthropathies.
- **Calluses** – Calluses tend to occur in conjunction with subluxation of the MTP joint, where the metatarsal head can be directly palpated subcutaneously.

2 LABORATORY ASSESSMENT

Laboratory tests can help confirm a diagnosis suggested by history and physical examination, but are not diagnostic on their own. In addition, laboratory tests can help monitor disease activity, but are meaningful only when they correlate with clinical findings.

Acute-Phase Reactants

Erythrocyte Sedimentation Rate (ESR)

Values

- **Normal for women** – ≤15 mm/h.
- **Normal for men** – ≤10 mm/h.
- **Adjust values** – Adjust normal values for age of patient. Upper limit for men = age divided by 2. Upper limit for women = age plus 10, divided by 2.

Interpretation

- **Elevated** – An elevated ESR reflects active inflammation and is used to determine or monitor disease activity in inflammatory and autoimmune forms of arthritis. Elevations of ESR may also be seen with infections and neoplastic diseases.
- **Factors that increase** – ESR increases with anemia, renal failure, hypergammaglobulinemia, and pregnancy.
- **Factors that decrease** – ESR decreases with changes in red blood cell morphology, hypofibrinogenemia, cryoglobulinemia, and congestive heart failure.

C-Reactive Protein

Values

- **Normal** – <1.0 mg/dl.

Interpretation

- **Elevated** – An elevated CRP reflects active inflammation and may be used to determine or monitor disease activity in inflammatory or autoimmune forms of arthritis.
- **Normal** – A normal CRP level does not necessarily indicate absence of inflammation.
- **Rapid changes** – CRP levels change rapidly after tissue injury, making this test a more timely indicator of disease activity than ESR.

Rheumatoid Factor

Values

- **Positive** – >1/20 by latex fixation method or ≥20 IU by nephelometry.

Interpretation

- **Healthy people** – 1–2% of healthy people have detectable serum RF; increases with age.
- **RA** – 75% of people with RA have positive RF ("seropositive RA").

Selected Diseases Associated with Elevated Serum Rheumatoid Factors

Chronic bacterial infections	**Parasitic diseases**
Subacute bacterial endocarditis	
Leprosy	**Chronic inflammatory disease of uncertain etiology**
Tuberculosis	Sarcoidosis
Syphilis	Periodontal disease
Lyme disease	Pulmonary interstitial disease
	Liver disease
Viral diseases	
Rubella	**Mixed cryoglobulinemia**
Cytomegalovirus	
Infectious mononucleosis	**Hypergammaglobulinemic purpura**
Influenza	

Modified from Koopman WJ, Schrohenber RE. Rheumatoid factor. In: Utsinger PD, Zvaifler MJ, Ehrlich GE (eds). Rheumatoid Arthritis: Etiology, Diagnosis and Therapy. Philadelphia, PA: JB Lippincott, 1985; pp 217–241.

Antinuclear Antibody (ANA)

Values

- **Positive** – >1/40. Many healthy individuals have low titers of ANA. The higher the ANA titer the greater the likelihood that it is associated with an autoimmune condition. However, in all cases, serological findings such as ANA assays require careful clinicopathologic correlation.

Interpretation

- **Systemic lupus erythematosus (SLE)** – >95% of people with SLE have ANAs.
- **Healthy** – 5% of healthy people, particularly women, have ANAs.
- **Usefulness** – ANA test has high sensitivity, low specificity.

Specific Autoantibodies

- **Presence** – Many autoantibodies correlate with specific rheumatic diseases.
- **Outcomes** – In some instances, autoantibodies help predict disease prognosis or the occurrence of certain kinds of organ involvement. More often, however, these antibodies are markers for diagnosis and correlate only roughly (if at all) with disease activity or severity.

Complement

Values

- **Units** – U/ml.
- **Normal** – Depends on the reference ranges of the laboratory.

Interpretation

- **Decreased serum complement** – Often reflects active immune complex-mediated diseases (SLE).
- **Persistently low total hemolytic complement** – Suggests an inherited deficiency of a complement component.

- **Deficiencies of C1, C2, C3, or C4** – Increased susceptibility to SLE.
- **Acute-phase reactants** – Several complement components are acute-phase reactants. As such, their serum levels may rise during active inflammation.

Guidelines for Clinical Use of the Antinuclear Antibody Test

Conditions Associated with Positive IF-ANA Test Results[a]

Disease	Frequency of positive ANA result, %
Diseases for which an ANA test is very useful for diagnosis	
SLE	95–100
Systemic sclerosis (scleroderma)	60–80
Diseases for which an ANA test is somewhat useful for diagnosis	
Sjögren's syndrome	40–70
Idiopathic inflammatory myositis (dermatomyositis or polymyositis)	30–80
Diseases for which an ANA test is useful for monitoring or prognosis	
Juvenile chronic oligoarticular arthritis with uveitis	20–50
Raynaud's phenomenon	20–60
Conditions in which a positive ANA test result is an intrinsic part of the diagnostic criteria	
Drug-induced SLE	~100
Autoimmune hepatitis	~100
MCTD	~100

ANA, antinuclear antibody; IF, immunofluorescent; MCTD, mixed connective-tissue disease; SLE, systemic lupus erythematosus.

[a] Values are titers. Prevalence of positive ANA test result varies with titer. Female sex and increasing age tend to be more commonly associated with positive ANA.

Disease	Frequency of positive ANA result, %
Diseases for which an ANA test is not useful in diagnosis	
Rheumatoid arthritis	30–50
Multiple sclerosis	25
Idiopathic thrombocytopenic purpura	10–30
Thyroid disease	30–50
Discoid lupus	5–25
Infectious diseases	Varies widely
Malignancies	Varies widely
Patients with silicone breast implants	15–25
Fibromyalgia	15–25
Relatives of patients with autoimmune diseases (SLE or scleroderma)	5–25
Normal persons	
≥1:40	20–30
≥1:80	10–12
≥1:160	5
≥1:320	3

Autoantibodies in Rheumatic Diseases

Type	Description
Anti-dsDNA	Antibodies to double-stranded DNA; greater specificity than those to single-stranded DNA
Anti-histone	Most assays do not differentiate the antibodies to the five major types of histones
Anti-ENA	Typical assays test for antibodies to two extractable nuclear antigens (ENAs): Sm (Smith) RNP (ribonucleoprotein)
Anti-SSA/Ro	Ribonucleoproteins
Anti-SSB/La	Ribonucleoproteins
Anti-centromere	Antibodies to the centromere/kinetochore region of the chromosome
Anti-Scl 70	Antibody to DNA topoisomerase I
Anti-Jo-1	Antibody to histidyl transfer RNA synthetase. The anti-Jo-1 antibody is the most common representative of a class of "myositis specific autoantibodies" known as anti-synthetase antibodies
Anti-PM-Scl	Antibodies to nucleolar granular component
Anti-Mi-2	Antibodies to a nucleolar antigen of unknown function

	Clinical association
	High specificity for SLE; occasionally appears in other illnesses and in normal people
	SLE, drug-induced lupus, other autoimmune diseases
	High specificity for SLE MCTD, SLE
	SLE (especially subacute cutaneous lupus), neonatal lupus, Sjögren's syndrome
	Sjögren's syndrome, SLE, neonatal lupus
	Limited scleroderma (i.e., CREST syndrome)
	Diffuse scleroderma
	Poly/dermatomyositis; especially in patients with interstitial lung disease, Raynaud's phenomenon, cracked skin on hands ("mechanic's hands"), arthritis, and resistance to treatment
	Polymyositis/scleroderma overlap syndrome
	Dermatomyositis

3 ARTHROCENTESIS AND SYNOVIAL FLUID ANALYSIS

Arthrocentesis and synovial fluid (SF) analysis yields valuable information that is important for the diagnosis of arthritis; arthrocentesis may help relieve signs and symptoms of arthritis, particularly if large joint effusions are present.

Indications

- **Infection** – Arthrocentesis must be performed immediately if there is any suspicion of infection. An inflammatory monarticular arthritis should be considered infectious until proven otherwise.
- **Crystal-induced disease** – Arthrocentesis and SF analysis are the only way to identify unequivocally.
- **Post-traumatic** – Analysis of joint fluid is the only way to distinguish post-traumatic hemarthrosis from post-traumatic arthritis with bland SF.
- **Inflammatory versus noninflammatory arthritides** – SF analysis enables the clinician to differentiate inflammatory and noninflammatory arthritides.
- **Therapeutic** – Arthrocentesis can be therapeutic, and can increase the efficacy of intra-articular glucocorticoids. Therapeutic arthrocentesis is indicated in any patient with a hemarthrosis.

Technique

- **Anesthesia** – Local anesthesia with 1% lidocaine without epinephrine significantly reduces discomfort associated with the procedure. A number 25 or 27 needle should be used to infiltrate the skin, subcutaneous tissue, and pericapsular tissue.
- **Needle choice** – After the periarticular tissues have been anesthetized, a 20- or 22-gauge needle can be used to aspirate small- to medium-sized joints. An 18- or 19-gauge needle should be used

for aspirating large joints, if there is a suspicion of infection or intra-articular blood, or if there is a likelihood of viscous or loculated fluid.

- **Landmarks** – Typical landmarks often are obscured around a swollen joint. Therefore, after a thorough physical examination and before anesthetizing the skin, it is often helpful to mark the approach.
- **Position** – Unlike injection, aspiration is best done when a joint is in a position of maximum intra-articular pressure.
- **Radiographic assistance** – Although most joints can be aspirated without radiographic assistance, some joints, such as the hips, sacroiliac joints, or zygoapophyseal joints, require aspiration by an interventional radiologist under computed tomography guidance.
- **Collection** – SF should be collected in an EDTA or sodium heparin tube for cell counts, and a sterile tube for Gram stain and microbiology culture studies.

Classes of Synovial Fluid

Characteristic	Class I (noninflammatory)
Color	Clear/yellow
Clarity	Transparent
Viscosity	High
Mucin clot	Firm
WBC count	>2,000
Differential	<25% PMNs
Culture	Negative

PMNs, polymorphonuclear cells.

Anatomic Approach to Aspiration

Joint
Knee
Shoulder
Ankle
Subtalar
Wrist
First carpometacarpal
Metacarpophalangeal and interphalangeal
Metatarsophalangeal and interphalangeal
Elbow