RHEUMATOLOGY AND IMMUNOLOGY

SECOND EDITION

Edited by

ALAN S. COHEN, M.D.

and

J. CLAUDE BENNETT, M.D.

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PREFACE

Rheumatology is a young, rapidly developing medical subspecialty that has grown to maturity only in the past 20 years. The disorders that afflict the musculoskeletal system are multiple, varied, and often serious, and the number of persons with such disorders is enormous. Several studies have shown that musculoskeletal disorders are often the most common cause of symptoms that lead patients to physicians' offices. The fiscal impact of arthritis is estimated to be in the billions of dollars due to loss of wages, loss of productivity, and various social factors, as well as to the obvious direct medical costs. In addition to their prevalence and the cost of treating them, these disorders have a great impact on the quality of life because of pain, the loss of mobility, and general disability that may result. Further, because the subject matter of rheumatology—connective tissue, including bone, muscle, joints, and vessels—is of pervasive importance in the functioning of the human organism, and because the proper functioning of the immune system is crucial both to the maintenance of health and to recovery from any form of inflammation, rheumatology and immunology are of undeniable relevance to virtually every medical practitioner and investigator. They are also therefore a necessary component in the training of medical students and residents.

Although the two disciplines dealt with in this text are nominally distinct according to formal definitions, they are largely inseparable for the purposes of clinical practice and scientific research. Among both rheumatic diseases and immunologic disorders there are many whose etiology is unknown, as well as many for which no satisfactory clinical treatment has been discovered; in many of these cases the resolution of the problem depends on the resolution of one or more problems in the other discipline. Rheumatoid arthritis, for example, is a rheumatic disease of notoriously elusive etiology, but clearly it involves an infiltration of lymphocytes into the synovial membrane. Lymphocytes are part of the immune system, so the etiopathology of rheumatoid arthritis raises questions about immunology. By the same token, wherever immunologic disorders lead to pathologic effects on connective tissue, determination of diagnosis and appropriate treatment must depend on a knowledge of the many facets of rheumatology.

This interdependence between the clinical subspecialties and between basic and clinical science makes the dual emphases on pathophysiology and clinical practice that characterize this text particularly apt for the subject matter. At the same time a certain amount of flexibility in organization is demanded. The major divisions, rheumatology and immunology, are each subdivided into sections dealing with basic concepts, with differential diagnosis, and with specific diseases. In addition, because there is a large body of diagnostic procedures applicable to rheumatic diseases in general—much of it also applicable to immunology—a section on general diagnostic procedures precedes the differential diagnosis section of the rheumatology division. Some disorders, such as systemic lupus erythematosus, might plausibly have been included in either division while others, such as vasculitis, involve so many unanswered questions in both disciplines that it seemed appropriate to include a chapter in each division.

Isaac Newton once remarked that all of his scientific accomplishments were as a minute examination of a single grain of sand on a wide, unexplored beach stretching to both horizons. The student of rheumatology and immunology—and this includes practitioners and researchers—should understand well what Newton meant. Many great expeditions are underway now in these disciplines, and there is room for many more. All must begin with what is now known. It is hoped that this text will provide researchers and practitioners alike with a good basic grasp of the grain of sand that is known, so that the best possible patient care can be provided while new frontiers are being explored.

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CONTENTS

SECTION I. REGIONAL STRUCTURE AND FUNCTION: BASIC PRINCIPLES IN RHEUMATOLOGY

1
Articular Structures

HANDS, WRISTS, AND ELBOWS

Juan J. Canoso

HANDS AND WRISTS

The structural and functional complexity of the hand is reflected in a range of pathologic states that cannot be comprehensively discussed within the limits of this chapter. Only certain aspects of the normal and abnormal hand with particular relevance to the rheumatic diseases will be considered.

Anatomy

Bones and joints. *Wrist.* At the distal end of the ulna there is a tough fibrocartilaginous plate known as the triangular articular disk, which covers the ulnar head. This disk plate and the distal end of the radius articulate with the proximal row of carpal bones at the radiocarpal or wrist joint. This, like the midcarpal, is an ellipsoid joint in which all motions except rotation are possible. In contrast, little motion occurs at the carpometacarpal (CMC) joint, in which the articular surfaces are relatively flat. An exception is the CMC joint of the thumb, in which a saddle configuration of the trapezium corresponds to a saddle configuration of the metacarpal base, thus allowing motion in two planes that are oriented at 45 degrees to the planes of motion of the fingers. The functional value of the thumb, which has been estimated as at least one half the value of the entire hand, is dependent upon the motion and stability of the CMC joint.

Arthrography shows infrequent (10 to 20 percent) communication between the distal radioulnar joint and the radiocarpal joint and between the radiocarpal joint and the midcarpal joint. In contrast, communication between the midcarpal joint and the CMC is frequent, with the exception of the CMC of the thumb, which remains separate because it is enclosed in a tough capsule even when destructive processes of the carpus, such as rheumatoid arthritis, have produced confluence of all other wrist joints.

Fingers. The metacarpophalangeal (MCP) joints are ball-and-socket structures. Their capsule is reinforced anteriorly by a fibrocartilaginous plate, which is firmly attached to the proximal phalanx but loosely connected to the metacarpal. The transverse metacarpal ligament unites the fibrocartilaginous plates of the medial four MCP joints. Laterally, the capsule is reinforced by eccentrically placed collateral ligaments, which become taut in flexion and lax in extension, an arrangement that in normal persons prevents lateral movement of the finger when the MCP joint is flexed at 90 degrees. Finally, the interphalangeal (IP) joints of the fingers are hinges that only allow flexion and extension.

Muscles and innervation. Wrist motion is mainly exerted by ulnar and radial flexors and extensors of the carpus, whereas motion of the digits involves the coordinated action of long flexors, extensors, and small muscles of the hand (the thenar, hypothenar, interossei, and lumbrical muscles). Flexion of the digits results from simple pull of the flexor muscles, particularly the profundus, supplemented by the superficialis muscle when greater strength is needed. Extension is a much more complex function that requires coordinated action of long extensors, intrinsic muscles, and the retinacular ligament, a structure that links the anterior aspect of the proximal phalanx to the dorsal aspect of the distal phalanx and couples the passive motion of proximal and distal IP joints.

A consideration of the structure and function of the extensor hood is central to an understanding of the extensor mechanism in general and the action of the intrinsic muscles in particular. This roughly triangular membranous structure covers and reinforces the MCP joint dorsally, and near the proximal interphalangeal (PIP) joint it splits into three bands. The central band inserts at the base of the middle phalanx; the lateral bands, after merging at the dorsum of the middle phalanx, insert at the base of the distal phalanx. Tendinous contributions to the extensor hood include the tendon of the extensor digitorum, of which the hood itself is an expansion, and the tendons of the intrinsic muscles (interossei and lumbricals). As can be seen in Figure 1-1, the pull of the intrinsic muscles passes anteriorly to the MCP joints and posterior to the PIP and

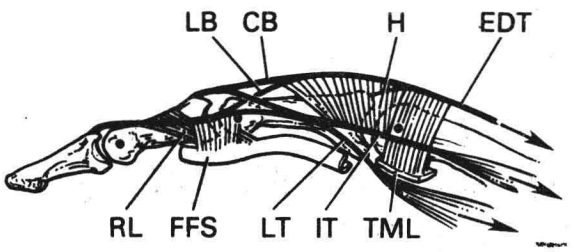

Fig. 1-1. Normal extensor mechanism of the digits: EDT, extensor digitorum tendon; H, extensor hood; LB, lateral band; CB, central band; TML, transverse metacarpal ligament; IT, interosseous tendon; LT, lumbrical tendon; FFS, fibrous flexor sheath; RL, retinacular ligament. (Modified from Tubiana R: Lésions traumatiques de l'appareil extenseur au niveau des doigts, in Verdan C (ed): Chirurgie des Tendons de la Main. Paris, Expansion Scientifique, 1976. With permission of the author and publisher.)

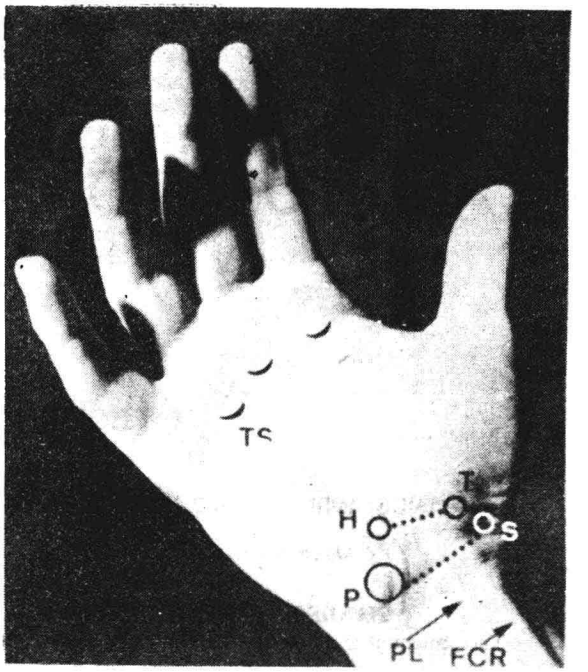

Fig. 1-2. Anterior view of the hand and wrist: TS, entrance of flexor tendon into the flexor synovial sheath; dotted lines outline the distal and proximal edges of the flexor retinaculum; H, hook of hamate; T, crest of trapezium; P, pisiform; S, tubercle of scaphoid; PL, tendon of palmaris longus—the median nerve runs deep to the PL; FCR, tendon of flexor carpi radialis.

distal interphalangeal (DIP) joints. Their contraction therefore will bring the MCP joints into flexion and the PIP and DIP joints into extension. In addition, the dorsal interossei muscles are inserted in such a manner that their contraction spreads the digits away from the third digit, while the palmar interossei muscles bring the digits together. In the thumb an expansion of the tendon of the extensor pollicis longus, joined laterally by the tendon of the abductor pollicis brevis and medially by the adductor pollicis, forms a triangular arrangement that resembles the extensor hood of the fingers.

All of the small muscles of the hand are supplied by the nerve root T1, whose fibers are carried by the ulnar and median nerves. The ulnar nerve innervates the hypothenar muscles, the interossei, lumbricals 3 and 4, the adductor pollicis, and sometimes part of the flexor pollicis brevis muscle. The abductor pollicis brevis, the opponens pollicis, and all or part of the flexor pollicis brevis are innervated by the median nerve. Ulnar nerve injury thus produces paralysis and atrophy of most of the intrinsic muscles of the hand. As a result of the loss of action of the intrinsic muscles, which, as explained above, flex the MCP joints and extend the PIP and DIP joints, ulnar palsy causes extension of the MCP joints and flexion of the PIP and DIP joints because of unbalanced action of long flexors and extensors.

Anatomic landmarks. Anteriorly, the flexor retinaculum, which bridges the concavity of the carpus and limits the carpal tunnel anteriorly, extends from the pisiform and the hook of hamate medially to the tubercle of the scaphoid and the crest of the trapezium laterally (Figure 1-2). The proximal edge of the flexor retinaculum corresponds to the distal crease of the wrist. Two tendons can be readily identified in the distal forearm and perpendicular to the crease—the lateral corresponding to the flexor carpis radialis and the medial to the palmaris longus (absent in 10 percent of persons). These two tendons allow the mapping of the median nerve, which as it enters the carpal tunnel runs medial to the flexor carpi radialis and deep to the palmaris longus tendons. Further down the palm, the proximal palmar crease corresponds to the entrance of the flexor tendon of the index finger into its synovial sheath, while the distal crease keeps a similar relation with the tendon sheaths of digits 3 and 4.

In the radial side the "anatomic snuffbox" is outlined anteriorly by the tendons of the extensor pollicis brevis and abductor pollicis longus, which run enclosed in a common tendon sheath above the radial styloid (Figure 1-3). Posteriorly, it is outlined by the tendon of the extensor pollicis longus. Ulnar deviation of the wrist allows palpation of the scaphoid. The trapeziometacarpal joint (CMC joint of the thumb) is identified immediately distal to the scaphoid.

Posteriorly, the head of the ulna and its styloid process and the dorsal tubercle of the radius (Lister's tubercle) can be readily identified (Figure 1-4). The latter is the point at which the tendon of the extensor pollicis longus turns laterally. Gentle flexion of the wrist facilitates the palpation of the radiocarpal and midcarpal joints.

Physical Examination

A brief review of activities of daily living concerning the hand, such as the ability to turn door knobs, hold a cup, button and unbutton clothes, and handle a fork and knife is of utmost importance in the patient's history. Questions should be asked about the presence of pain,

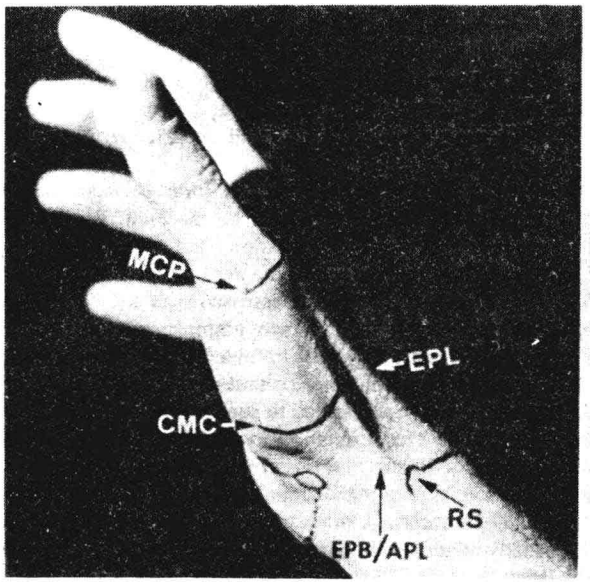

Fig. 1-3. Radial aspect of the wrist. The "anatomic snuffbox" is limited anteriorly by the extensor pollicis brevis (EPB) and abductor pollicis longus (APL) tendons in a common tendon sheath. The critical relation of the radial styloid (RS) to this sheath is shown. The extensor pollicis longus (EPL) tendon bounds the snuffbox posteriorly. CMC, first carpometacarpal joint; MCP, first metacarpophalangeal joint.

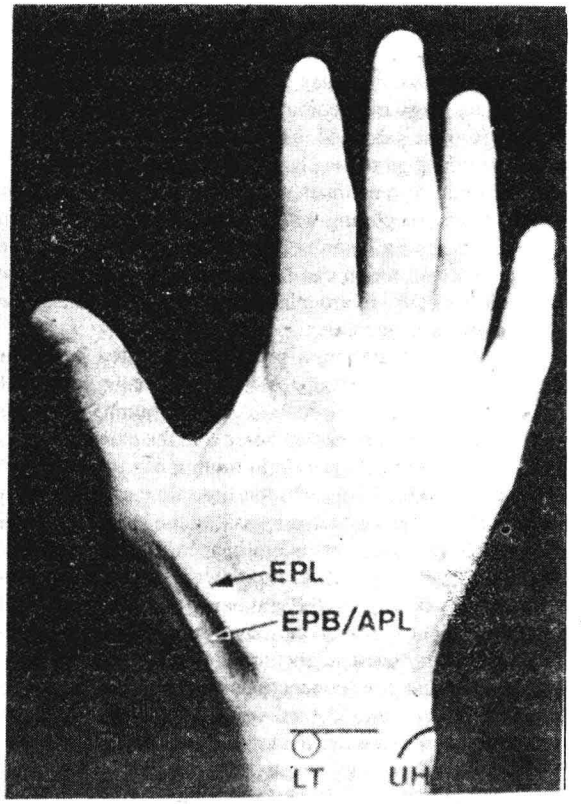

Fig. 1-4. Posterior aspect of the hand and wrist: EPB, extensor pollicis brevis; APL, abductor pollicis longus; EPL, extensor pollicis longus; LT, Lister's tubercle; straight line, the radiocarpal joint; UH, distal ulnar head. Radially, the tendinous boundaries of the "anatomic snuffbox" are shown.

weakness, stiffness, paresthesias, and color changes, as well as their exact location and circumstances.

A careful inspection should be made of the following: nails; skin; alignment of the forearm, wrist, and fingers; normal contours; and muscle bulk in the interosseous spaces and the thenar and hypothenar eminences. Pitting or detachment (onycholysis) of the nails, ulcerations, scars, rashes, swelling, atrophies, and deformities should be recorded.

A functional assessment of the thumb and fingers is more important than detailed goniometric measurements. Precise goniometry is important in the wrist and, under certain circumstances, in the thumb and fingers (Table 1-1). In general, motion of the finger and thumb is normal when all can reach the palm in flexion and can be returned to their extended position, and when the tip of the thumb can reach the tip of all other digits (opposition). This is also an excellent estimation of the precision motion of the hand. Impaired thumb function is expressed as the distance in centimeters from its tip to the MCP joint of the little finger.

Flexion and extension of the fingers occur in the sagittal plane, but similar motions of the thumb are recorded in the frontal plane (despite the fact that anatomically the axis of flexion at the thumb CMC joint is not at 90 degrees with the axis of flexion of the MCP joints of digits 2–5, but at 45 degrees). In extension, the thumb thus is maximally deviated toward the radial side in the plane of the hand, and flexion brings its tip close to MCP 5. In abduc-

Table 1-1
Range of Motion of Wrist, Thumb and Fingers

Joint	Maneuver	Degrees
Wrist	Flexion	75
	Extension	−70
	Abduction (radial deviation)	20
	Adduction (ulnar deviation)	35
Thumb		
CMC	Flexion	15
	Extension (radial abduction)	20
	Abduction (palmar abduction)	60
	Adduction	0
MCP	Flexion	50
	Extension	−15
IP	Flexion	70
	Extension	−30
Fingers		
MCP	Flexion	90
	Extension	−45
	Abduction/adduction*	
PIP	Flexion	100
	Extension	0
DIP	Flexion	80
	Extension	0

*Variable, 0 at 90 degrees flexion.

tion, the thumb moves anteriorly and becomes almost vertical to the plane of the hand, while adduction returns it to the radial side of the index finger.

Flexor lags are recorded in centimeters from the tip of the digit to the palm and extensor lags in centimeters from the tip of the digit to the plane of the dorsum of the hand. It is important to remember that lags resulting from tendon rupture, loss of gliding within sheaths, or peripheral neuropathy lead to a disproportionate loss of active motion, while passive motion can be normal. In contrast, loss of motion as a result of articular disease leads to equal loss of active and passive motion.

The flexor digitorum profundus is tested by having the patient attempt flexion of the digit while the middle phalanx (the proximal phalanx in the thumb) is held in extension. It must be remembered that the extensor indicis proprius and the extensor digiti minimi can by themselves extend the index finger and the little finger, respectively. Their integrity is evaluated by asking the patient to extend the finger while the others are bent in a fist. Ruptures of extensor tendons produce a variable degree of extension lag, which is related to the number of tendons ruptured. If rupture of only one tendon has occurred (usually in the dorsum of the wrist in rheumatoid and other synovitis), distal intertendinous connections tend to mask the loss by transferring pulling forces from neighboring tendons. Rupture of two or more tendons negates this phenomenon and obvious deformity results.

The extensor pollicis longus, which frequently is ruptured in rheumatoid arthritis, is evaluated by having the patient place the hand flat on the table and asking the patient to raise the thumb off the surface. Even when a stiff MCP prevents such motion, the tensed tendon can be seen (or felt) under the skin.

Extensor lags can occur in the absence of ruptured extensor tendons in three situations. The first results from MCP synovitis, leading to volar subluxation and contracture of the intrinsic muscles. The second is also caused by MCP synovitis, but here the extensor tendons slip into the ulnar valleys, on occasion deeply enough to become weak flexors. Finally, posterior interosseous nerve palsy (discussed below) can be confused with extensor tendon rupture.

Abnormal intrinsic muscle tension is tested by holding the proximal phalanx in extension (thereby stretching the intrinsic muscles further) while the middle phalanx (distal in the thumb) is passively brought into flexion. Tight intrinsic muscles make the flexion difficult or impossible.

Finally, to test the integrity of the collateral ligaments of the MCP joints, the observer attempts to abduct and adduct the digit while the proximal phalanx is kept at 90 degrees flexion. Only tears of one or both ligaments will allow such motion to occur.

Synovitis is identified by pain on motion (particularly at the extremes of motion), weakness, swelling, increased warmth, local tenderness, and decreased range of motion. In IP joints the sole evidence of synovitis may be pain on lateral compression of the joint. In IP and MCP joints, small effusions are best appreciated by four-finger palpation—two at the sides, one dorsal, and one palmar to the joint. Swelling can be inapparent at the wrist and the only finding may be a weak grip and deep tenderness at the radiocarpal joint.

Pain resulting from eroded joint surfaces occurs throughout the range of motion of the joint, is generally associated with bony crepitation, and is aggravated by longitudinal compression of the joint.

The power grip can be assessed with a dynamometer or tested with an ordinary sphygmomanometer, in which the rubber part of the cuff is folded twice and the cotton flap rolled around it. The apparatus is calibrated at 20 mm Hg and the patient is asked to squeeze the cuff maximally three times. The results are averaged and noted as grip strength.

Abnormal Findings

Deformities. *Ulnar drift.* In ulnar drift, which is usually bilateral, the digits have undergone an ulnar angulation at the MCP joint. A common association is volar (anterior) subluxation of the base of the proximal phalanx. The pathogenesis of ulnar drift is a subject of controversy. Some important factors include (1) weakening of the capsule and ligaments secondary to chronic MCP synovitis; (2) normally greater ulnar and volar strain exerted by the flexor tendons; (3) ulnar pull along the line of the transverse metacarpal ligament (which is connected to the base of digits 2–5) upon completion of tightening of power grip; (4) ulnar deviation of the digits to offset radial deviation of the wrist (the "Z" compensatory mechanism); and (5) spontaneous ulnar deviation of the digits brought about by tense MCP effusions.

Rheumatoid arthritis is the most common cause of ulnar drift, but the abnormality has also been observed in systemic lupus erythematosus, rheumatic fever, Parkinson's disease, congenital deformities, and as a result of certain occupations such as carpentry.

Boutonnière deformity. Boutonnière deformity is characterized by flexion at the PIP joint and hyperextension at the DIP joint (Figure 1-5). It is caused by chronic synovitis of the PIP joint, which causes weakening of the central slip of the extensor tendon and palmar dislocation of the lateral bands. When this occurs, the lateral bands act as flexors rather than extensors of the PIP joint. Boutonnière deformity is seen in 15 to 30 percent of patients with rheumatoid arthritis, in other chronic synovitis such as psoriatic arthropathy, and as a result of trauma or burns.

Swan-neck deformity. Swan-neck deformity consists of hyperextension of the PIP joint and flexion of the DIP joint (Figure 1-6). Unlike ulnar drift and boutonnière deformity, which result from disease in one joint, swan-neck deformity can occur as a result of disease at the DIP, PIP, MCP, or extensor and flexor tendon, muscle imbalance as seen in certain neurologic disorders, and heritable disorders of the connective tissue, particularly the Ehlers-Danlos syndrome.

Rupture or stretching of the extensor tendon as it in-