Pathogenesis and Differential Diagnosis of the Swan-Neck Deformity

By James N. Dreyfus* and Thomas J. Schnitzer

THE MECHANISMS and clinical settings of the swan-neck deformity (SND) have interested physicians for more than a century. Archibald E. Garrod, son of the famed Sir Alfred Garrod, in his 1890 *Treatise on Rheumatism and Rheumatoid Arthritis*,[1] published an illustrated description of an arthritic deformity of the hand, characterized by "[flexion of] the terminal phalanges upon those of the second row . . . , [hyperextension of] the second phalanges upon the first, . . . and [flexion of] the first phalanges upon the metacarpal bones" (Fig. 1). This description, which he derived from the work of Charcot 30 years earlier,[2] would now be recognized as similar to the modern term SND. Garrod elaborated that

> the increased tonicity of the muscles is the cause of the grotesque distortions of the fingers and toes which are so commonly observed in the later stages of [rheumatoid arthritis]. . . . These muscular deformities are by no means peculiar to rheumatoid arthritis, being met with in the course of a number of lesions of the nervous system, . . . as well as in long-standing articular disease of other kinds, such as chronic gout, and the . . . chronic articular rheumatism [described] by Jaccoud. . . . The kind of deformity produced depends upon the relative strength of the different muscles, especially of the long flexors and extensors and the interossei.

Garrod's early observations on the etiology of the SND and his recognition that the deformity was not specific for rheumatoid arthritis (RA) remain valid and instructive today. Moreover, the diversity of disease categories that are associated with the SND (rheumatologic diseases, neurologic and vascular lesions, and congenital collagen abnormalities) has not been commonly appreciated.

The term SND refers in this review to the digital deformity, characterized by proximal interphalangeal joint (PIP) hyperextension (also termed *recurvatum*[3]) and distal interphalangeal joint (DIP) flexion. Metacarpophalangeal joint (MCP) flexion, although common in RA and other intrinsic-plus SND, need not be present. Hyperextension of the interphalangeal joint of the thumb, sometimes termed SND of the thumb, will not be discussed.[4] In this paper, we examine in detail the pathogenesis and differential diagnosis of the SND and discuss briefly appropriate treatment.

HISTORICAL PERSPECTIVE

Charcot and Garrod may not have been the first to describe the SND. In 1676, Thomas Sydenham wrote of a chronic arthritic condition in which the finger joints were "inverted . . . with the knots showing on the inner rather than the outer aspect of the fingers."[5] Short has claimed that this was in fact a description of the SND.[6] Moreover, others have discovered RA-like deformities, including the SND, in the early seventeenth-century paintings of the Flemish master Rubens.[7]

Although the deformity has been recognized clinically for perhaps 300 years, the actual term swan-neck has been in use only in the past 25 years. Prior to 1956, the standard orthopedic and rheumatology texts described the deformity without naming it.[8,9] Steindler, in 1951, divided all rheumatoid deformities of the hand into two broad categories: "pill-roller hand" (intrinsic-plus) and "claw hand" (intrinsic-minus).[10] He commented on the PIP hyperextension deformities resulting from the intrinsic muscle contractures. In 1957, Laine et al used the term SND in a paper on finger deformities in RA without identifying its inventor.[11] The subsequent edition of Bunnell's text on the hand refers to the "so-called swan-neck deformity." Thus, although the

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Fig. 1. **Rheumatoid hand deformity resembling the swan-neck deformity. (Reproduced by permission of Charles Griffin.[1])**

originator of the term may be obscure, his creative vision has been widely accepted (Fig. 2).

CLINICAL ANATOMY

The key to understanding the pathogenesis of the SND is a knowledge of the structural and dynamic factors that constitute the normal muscle and tendon balance of the digits.[3,12–15] Three sets of muscles and tendons provide digital equilibrium and control: (1) the extrinsic extensor group, (2) the extrinsic flexor group, and (3) the intrinsic muscles (IM).

The *extrinsic extensor* group of muscles includes the *extensor digitorum communis, extensor digiti minimi,* and the *extensor indicis.* These muscles originate in the dorsal forearm and insert via the *common extensor tendons* into the *extensor hood* (aka *dorsal expansion* or *aponeurosis*) and onto the dorsal surface of each digit (Fig. 3). The sites of insertion are multiple and include the dorsal base of the proximal and middle phalanges as well as the distal phalanx via the *lateral extensor tendons* (or *bands*). The nerve supply is a branch of the radial nerve. The extensor muscles act via the extensor mechanism to extend the digits at the MCP. In concert with the intrinsic muscles, they extend the PIP and DIP joints as well.

The *extrinsic flexor* group includes the *flexor digitorum profundus* and *superficialis* (the deep and superficial flexors), originating in the volar forearm and inserting respectively into the bases of the distal and middle phalanges (Fig. 3). The flexor tendons are housed within the synovial-lined flexor retinacular sheaths which stabilize and fix the tendons to the phalanges and to the *volar* (or *palmar*) plate (Fig. 4).[6,7] The extrinsic flexors, supplied by branches of the median and ulnar nerves, flex the fingers at the PIP and DIP

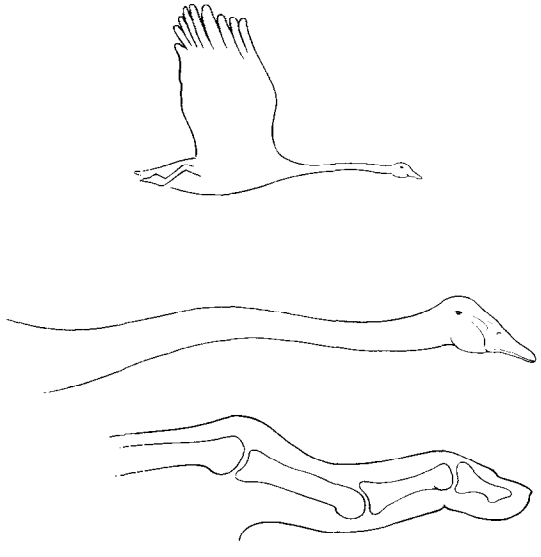

Fig. 2. **An artistic interpretation of the swan-neck deformity that is reminiscent of a swan in flight.**

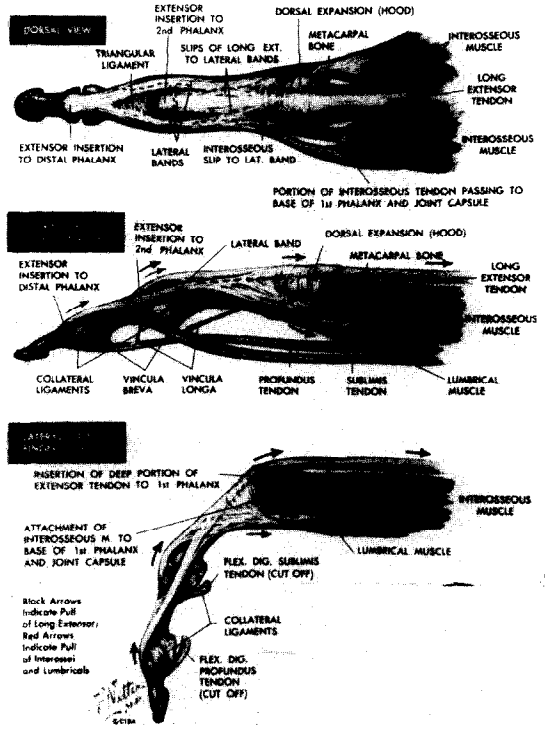

Fig. 3. **Anatomy of the extensor apparatus and flexor tendons of the finger. (Reproduced by permission of *Clinical Symposia.*[13])**

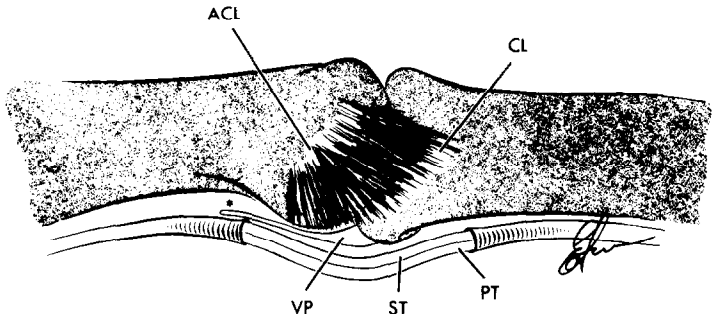

Fig. 4. Supporting structures of the proximal interphalangeal joint, including the collateral ligament (CL), accessory collateral ligament (ACL), superficialis tendon (ST), profundus tendon (PT), and volar plate (VP). (Reproduced by permission of C. V. Mosby.[67])

joints. Efficient MCP flexion requires the synergistic action of the intrinsic muscles.

The *intrinsic muscles* (IM) include the four *lumbricals* and the seven *volar* and *dorsal interossei.** The lumbricals arise from the flexor tendons and insert into the lateral extensor bands. The interossei arise between the metacarpals and have various insertions in the MCP joint capsule and proximal phalangeal base, the extensor hood, and the medial and lateral extensor bands. The usual nerve supply of these muscles is the ulnar nerve with the exception of the first and second lumbricals which are supplied by the median nerve. The mode of action of the intrinsic muscles is complex, a result of their multiple sites of origin and insertion and variable action depending on the initial position of the digit. However, from a simplistic perspective, the intrinsic muscles act as a functional unit with two major effects: flexion of the MCP joints and extension of the PIP and DIP joints. By virtue of insertions into the base of the proximal phalanx volar to the axis of the MCP joint, the intrinsic muscles flex the MCP. The tendons then cross dorsally and join the extensor apparatus over the PIP and thereby act to extend the PIP and DIP (Fig. 5).[68] The dual flexor/extensor action of the IM is central to the mechanism of one type of SND. Overactivity of these muscles produces a deformity with MCP flexion and PIP hyperextension, typical of SND.

The balance of flexor and extensor power about the digital joints depends not only on the muscles and tendons but also on the integrity of the joints and surrounding soft tissue structures. The *fibrous joint capsule,* the *collateral ligaments,* the *volar plate,* and the *flexor tendons* all act to stabilize the digital joints and prevent deformities such as MCP subluxation or PIP hyperextension (Fig. 4). The *retinacular system,* including the *ligament of Landsmeer,* is important in stabilizing the tendinous structures and facilitating the rhythm of digital flexion and extension. Destruction of any of these anatomic units by trauma or synovitis can alter the muscle forces around the joint and lead to deformity.

PATHOMECHANICS

Interrelation of PIP Hyperextension and DIP Flexion

As stated initially, the anatomic definition of SND is hyperextension or *recurvatum* of PIP associated with flexion of the DIP. Zancolli has emphasized that the two constituent deformities (PIP recurvatum and DIP flexion) are interrelated and are a logical consequence of the normal kinesiology of the digit.[16] Specifically, hyperextension of the PIP for any reason will secondarily produce flexion of the DIP. Conversely, a DIP flexion deformity alters the balance of forces around the PIP, leading to hyperextension. Fur-

*The intrinsic muscles also include the thenar and hypothenar muscles but these are not germane to our discussion of SND.

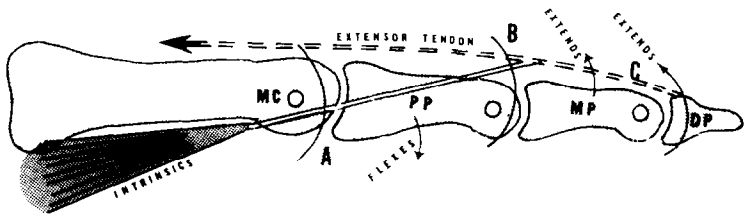

Fig. 5. Action of the intrinsic muscles (IM) and extensor muscles (EM). (A) The IM lie palmar to the MCP joint axis and thus flex this joint. (B) The IM pass dorsal to the PIP joint axis and extend this joint. (C) The IM join the extensor apparatus and thus help to extend to DIP. (Reproduced by permission of Lea & Febiger.[68])

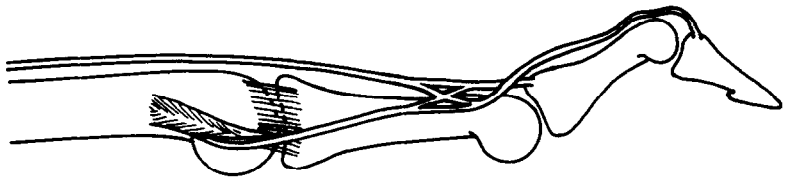

Fig. 6. Pathogenesis of the DIP flexion deformity in primary PIP hyperextension. When the PIP is hyperextended, the lateral bands become slack and allow the DIP to drop into flexion. (Reproduced by permission of C. V. Mosby.[69])

thermore, the deformities tend to vary equally in severity. The greater the recurvatum of the PIP, the greater is the flexion deformity of the DIP. The converse is also true.

Several mechanisms contribute to the secondary DIP flexion, occurring as a result of primary PIP hyperextension. PIP recurvatum relaxes the normal tension on the lateral extensor tendons, thereby leading to subsequent dorsal and central movement of these tendon bands. With relaxation, the lateral bands lose the ability to extend the distal phalanx. The DIP therefore drops into flexion (Fig. 6).[69] Recurvatum of the PIP also stretches the flexor digitorum profundus, thereby increasing its flexor action on the DIP. Another contributing factor is the loss of the mechanical advantage of the oblique retinacular ligament in extending the DIP.[17]

Conversely, in primary DIP flexion deformity or "mallet finger," the lesion is a disruption or stretching of the terminal extensor tendon or the lateral extensor bands. Thus, the ability to extend the DIP is lost, and the DIP is fixed in flexion, resembling a mallet. Because the lateral bands are now relaxed, all the power of the common extrinsic extensor is concentrated on the central slip that inserts into the middle phalanx. With time, the supporting structures of the PIP are weakened and the PIP is forced into hyperextension (Fig. 7).[70]

In summary, the SND can be a consequence of a primary deformity at only one joint with secondary changes at the other joint. Primary PIP hyperextension is more frequently a cause of SND that is seen by the rheumatologist.

Causes of Primary PIP Hyperextension

Zancolli has classified the causes of PIP hyperextension into three general types:

1) *Extrinsic,* secondary to excess traction of the extrinsic extensor tendons;
2) *Intrinsic,* secondary to overactivity of the intrinsic muscles, also known as the intrinsic-plus deformity;
3) *Articular,* secondary to weakening or destruction of the normal stabilizing mechanisms of the PIP that ordinarily prevent hyperextension.

In general, in many cases of SND, especially long-standing SND, several etiologies of PIP hyperextension may be present and the type, therefore, mixed. We will examine the categories individually with special attention to clinical setting and diagnostic tests.

Extrinsic Type

Excessive traction on the extrinsic extensor tendons tightens the central slip, inserting at the base of the middle phalanx, and thereby pulls the PIP into hyperextension. Tightness of the extrinsic extensors can occur from a pathologic process such as muscle fibrosis or spasm or tendon adhesions. In addition, flexion of the wrist or MCP can produce extensor tightness by elongating the distance over which the extensor tendons must travel. In the normal hand, simple flexion of the wrist or MCP does not produce sufficient tension on the extensors to produce hyperextension of the PIPs. However, in patients with cerebral palsy, for example, the combination of general muscular spasticity with the commonly occurring wrist

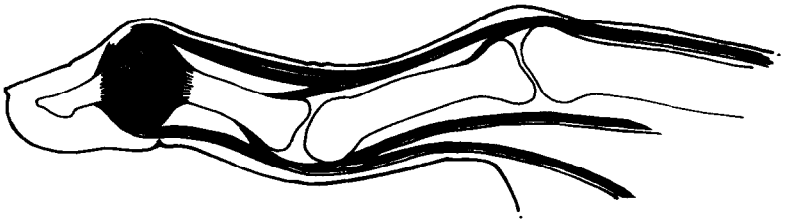

Fig. 7. Mallet-type SND. Attrition of the distal extensor tendon from synovitis (or trauma) leads to a SND. (Reproduced by permission of F. A. Davis.[70])

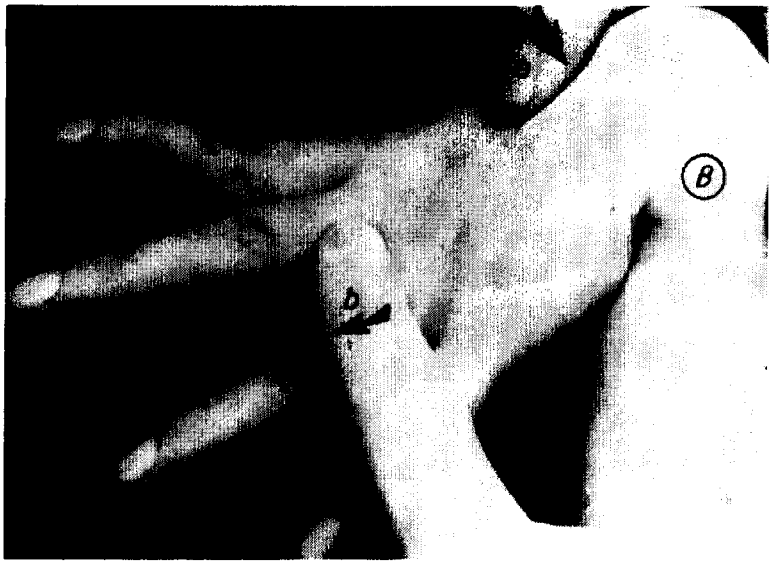

Fig. 8. Extrinsic type SND in a patient with cerebral palsy with an underlying flexion deformity of the wrist. Further wrist flexion (A) or MCP flexion (B) accentuates the deformity of the index and ring fingers. (Reproduced by permission of J. B. Lippincott.[3])

flexion deformities produces marked extensor traction with resulting PIP recurvatum (Fig. 8). Similarly in RA, where many factors may be present that contribute to SND (see below), flexion deformities at the wrist or MCP can exacerbate a tendency to PIP hyperextension.

The diagnostic test for extrinsic SND involves observing the deformity on motion of the wrist or MCP. A SND that appears only with flexion of wrist or MCP and disappears with extension of these joints has a component of extrinsic extensor tightness (Fig. 9). Similarly, if it becomes impossible to flex the PIPs when the wrist or MCP is held in flexion, then the *extrinsic-plus test* is positive.

Intrinsic Type

The combined action of the IM via the extensor mechanism is flexion at the MCP and extension at the PIP. Hyperactivity of the IM overpowers the opposing action of the extrinsic flexors and produces PIP hyperextension and ultimately the complete SND. The causes of IM hyperactivity are multiple; some will be discussed later. Further details are available in a superb discussion of intrinsic-plus deformities by Zancolli.*[18]

*Intrinsic tightness is also associated with deformities other than the SND. See Parkes A, "The lumbrical plus finger." J Bone Joint Surg 1971;53B:236–9.

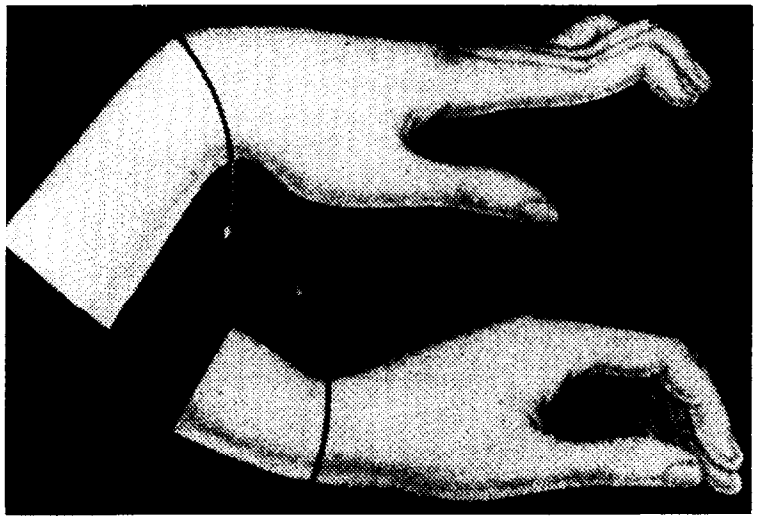

Fig. 9. Test for extrinsic-type SND. The deformity appears (or worsens) with wrist flexion and disappears (or improves) with extension. (Reproduced by permission of J. B. Lippincott.[17])

Ischemic contracture. Disruption of the vascular supply to the hand (trauma, burns, edema, external compression, vascular obstruction, radiotherapy) can result in fibrosis and shortening of the IM, particularly the interossei. The reason that the interossei are so sensitive to injury is that their blood supply travels within a tight compartment which does not expand when tissue edema is present. With severe fibrosis of the interossei, PIP hyperextension occurs. This etiology, first described by Finochietto in the 1920s and elaborated upon by Bunnell, has been the classic model for intrinsic deformities.[19,20]

Spasticity. Neurologic events with upper motor neuron dysfunction such as cerebrovascular accidents, cerebral palsy, encephalitis, and parkinsonism can be associated with marked spasticity of the IM, followed by PIP hyperextension. Spasticity, as opposed to fibrosis, can be demonstrated to be the contributing etiology if selective local anesthesia of the ulnar nerve abolishes the deformity.[21,22]

In RA, the intrinsic muscles are frequently abnormally tight. The mechanism (see below) has been proposed to be reflex spasm secondary to MCP synovitis.[23]

Intrinsic tendon adhesion. Wounds and fractures that are associated with the MCP joint may be followed by adhesions of the IM tendons, with subsequent intrinsic-plus deformity. Tendon adhesions secondary to synovitis have been described in RA.[24]

Volar subluxation of the MCP joints. Volar subluxation of the MCPs, a common occurrence in RA, shortens the distance between the proximal origins and distal insertions of the interossei. With time, these muscles, particularly the ulnar components, adjust to the shortened length with retraction, perhaps even fibrosis.

The classic test for intrinsic tightness, the intrinsic-plus or Bunnell test, is performed by observing the range of motion at the PIP when the MCP is alternately hyperextended and flexed (Fig. 10). Hyperextension at the MCP tightens the intrinsic muscles. In the normal hand, this maneuver does not interfere with the PIP flexion. However, when the IM are pathologically tight, MCP extension places the IM under exaggerated tension, causing powerful extensor force over the PIP which interferes with passive PIP flexion. Conversely, MCP flexion relaxes the IM and permits PIP flexion. Persistent stiffness of the PIP, regardless of MCP position, is diagnostic of primary PIP pathology, either articular or periarticular, as the cause of the deformity.

Fig. 10. Demonstration of the intrinsic-plus test. (*top*) SND in a female with polymyositis. PIP flexion is greater when the MCP is flexed (*middle*) than when hyperextended (*bottom*).

Articular Type

The final pathogenetic type of primary PIP hyperextension is that due to disease originating at the PIP. As previously discussed, several

structures are responsible for the normal stability and integrity of the joint: the collateral ligaments, joint capsule, palmar plate, superficial flexor tendon, and retinacular ligaments. Weakening, stretching, or destruction of any of these structures allows the *normal* extensor forces to cause *abnormal* hyperextension of the PIP. Once PIP recurvatum is present, a vicious cycle is created. Joint deformity produces secondary weakening of the articular structures and further deformity.

The causes of the articular type of SND, as delineated by Zancolli,[18] follow logically from this pathogenetic principle. Synovitis or trauma can disrupt or stretch the supporting structures (Fig. 11). In congenital joint laxity, either secondary to a collagen disorder or an idiopathic syndrome, the joint capsule and ligaments are already loose and easily permit a hyperextension deformity. Superficial flexor tendon rupture is another cause. Finally, a retractile scar in the skin overlying the extensor surface of the PIP may rarely exert such force that the joint is forced into hyperextension. Obviously, some secondary PIP articular laxity must occur in intrinsic and extrinsic SND.

Causes of Primary DIP Flexion

Disruption of the terminal extensor tendon or the lateral extensor bands produces a flexion deformity of the DIP, known as the mallet or baseball finger[25] (Figs. 7 and 12). As previously noted, the complete SND with secondary PIP hyperextension can be a consequence. Trauma is a frequent cause of laceration or avulsion of the distal tendons. Loss of ability to extend the distal phalanx when the middle phalanx is held in extension is indicative of a mallet finger mechanism. Although not emphasized in the literature, DIP synovitis can cause secondary distal extensor tendon weakening and rupture, followed by

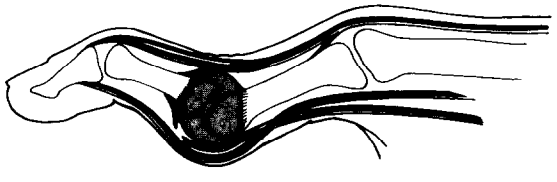

Fig. 11. Articular type SND. In this example, synovitis of the PIP has disrupted the paraarticular structures, including the superficial flexor tendon. (Reproduced by permission of F. A. Davis.[70])

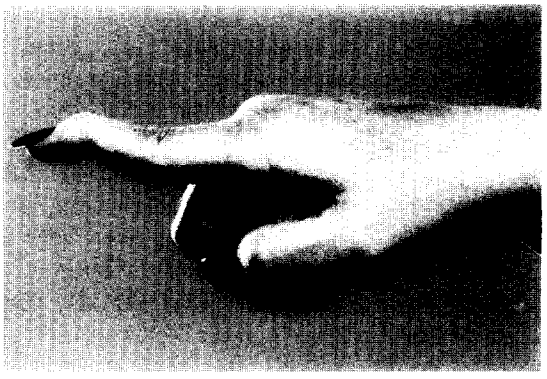

Fig. 12. Mallet-type SND in a patient with psoriatic arthritis. Synovitis was restricted to the DIP.

SND. Associated diseases include psoriatic arthritis, gout, and rarely RA.[11,26]

CLINICAL SETTINGS FOR SND

The four pathogenetic types—extrinsic, intrinsic, articular, and mallet finger—constitute the final common pathways by which different diseases produce the SND. The diversity of diseases deserves emphasis and is summarized in

Table 1. Disorders Associated With SND

Clinical Condition	Mechanism
Rheumatologic diseases	
RA	E, A, I, M
SLE	I, A
Postrheumatic fever (Jaccoud arthropathy)	I, A
Psoriatic arthritis	M, A, I
Gout	M, A, I
Polymyositis	I
Neurologic disorders	
Cerebral palsy	E, I
Cerebral vascular accident	I
Postencephalitis	I
Post-head trauma	I
Parkinsonism	I
Hypermobility syndromes	
Ehlers-Danlos	A
Marfans	A
Benign familial joint hypermobility	A
Traumatic causes	
Ischemic injury of the hand	I
Rupture of the flexor tendon	A
Posthand tendon surgery	E, I, A
Rupture of the distal extensor tendon	M

Note. A = Articular type, E = Extrinsic type, I = Intrinsic type, and M = mallet finger type.

Table 1. We have divided the types of disorders into four major categories, including rheumatologic diseases, neurologic problems, hypermobility syndromes, and traumatic causes. In this section, we focus on the mechanism and incidence of SND, where known, in nontraumatic diseases. The traumatic causes are adequately reviewed in the orthopedic literature.[3] The reader is also referred to two excellent reviews of hand deformities that mimic those seen in RA.[27,28]

Rheumatologic Causes

Rheumatoid Arthritis

The association of SND with RA is the best known clinical setting of this deformity, dating back to clinical recognition of RA as a separate entity from other forms of chronic arthritis.[1] Two published series have examined the incidence of SND in RA. Brewerton examined 300 men and women with RA of varying duration.[29] Thirteen percent had hyperextension deformities of one or more digits. Functional disability ranged from negligible to severe, depending on the resulting range of motion in the PIP. In patients followed longitudinally, intrinsic muscle spasm frequently, but not universally, preceded the SND. Laine et al.[11] studied 305 patients and found SND in 44 (14%). Mechanisms that were noted included intrinsic tightness and primary articular laxity. One patient had rupture of the superficial flexor tendon. In addition, a mallet ("nontraumatic baseball") finger was noted in 10 patients. Whether there was progression to the complete SND was not described.

The literature on the underlying cause of rheumatoid SND has been prolific and often contradictory.[3,4,10,17,23,30–32] The traditional explanation for SND has been that intrinsic muscle tightness, demonstrated by a positive intrinsic-plus test, is the cause. But the etiology of the tightness is controversial. The older literature refers to "spasm" of the intrinsic muscles, implying continuously contracting muscle.[33] The cause of the spasm was said to be "irritation" from adjacent MCP synovitis. Attempts to document spasm by electromyographic studies of the interossei have yielded conflicting data. Wozny and Long found no electrodiagnostic evidence of continuing IM contracture.[34] However, Nakamura et al. found that at least in early RA there was persistent electrical stimulation of the interossei.[35] They and others[36] have proposed that afferent pain stimuli from the inflamed MCP joint capsule triggers a "protective pain reflex" which produces spasm in the IM that are supplied by the same spinal cord segment (Fig. 13). Secondary fibrosis would then occur in the persistently contracting muscle. Myositis involving the IM has also been proposed as a cause[37] but has not been confirmed.[38] Others have challenged this concept, stating the IM tightness resides not at all in the muscles but in adhesions in the tendons to the joint capsule with subsequent joint stiffness.[23,24]

Synovitis of the PIP and flexor tenosynovitis[39] predispose to PIP stiffness which can contribute to a hyperextension deformity. As noted earlier, extrinsic causes with wrist and MCP flexion contractures and primary DIP disease, which is rare, are sometimes present in the rheumatoid SND.

SLE

Deforming, nonerosive arthritis, including the SND has been commonly recognized as a complication of SLE.[40–48] The frequency of SND ranges from 3% to 38%, depending on the series. Bleifeld and Inglis, in an orthopedic study, examined the hands of 50 consecutive patients with SLE of greater than one year's duration.[43] Generalized ligamentous laxity was present in 50% of the patients. SNDs, commonly involving two or more fingers, were present in 38% of patients.

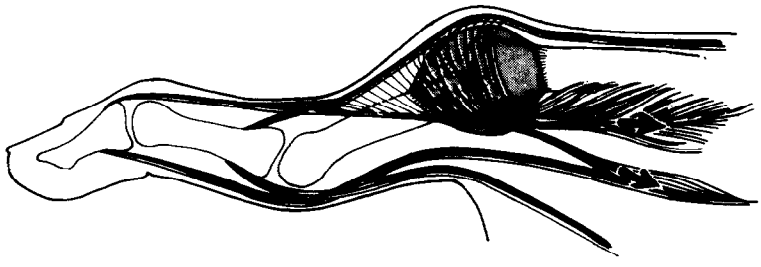

Fig. 13. Intrinsic SND in RA. MCP synovitis is associated with increased intrinsic muscle tension, producing PIP hyperextension. (Reproduced by permission of F. A. Davis.[70])

The mechanism in all patients was articular secondary to stretching of the volar plate. No patient had intrinsic muscle tightness. PIP synovitis was uncommon. Radiographs showed no erosions or joint space narrowing. Numerous case reports have confirmed that ligamentous laxity and capsular weakness are present. The PIP hyperextension deformity is usually reversible, at least initially. The intrinsic tightness is probably common as a secondary phenomenon but has not been documented rigorously in the literature.[45]

The pathogenesis of the articular laxity is poorly understood. Does laxity develop from simple stretching of the supporting structures by underlying synovial distension[41] or are there primary inflammatory changes in the soft tissues as has been proposed?[43,44] Despite frequent speculation, there have been few observations on the pathology of the joint capsule. Most studies have focused on the minimal inflammatory changes of the synovium[40] rather than the periarticular structure. Flexor tenosynovitis has been documented pathologically[49] and could predispose to joint deformity.

Chronic Post-Rheumatic Fever Arthritis (Jaccoud Arthropathy)

The recognition during the past 30 years of the association between rheumatic fever and chronic joint deformity is largely due to the work of Bywaters[50] and Zvaifler.[51] Criteria for diagnosis include a history of rheumatic fever attacks, absence of marked synovitis, reversible ulnar deviation, and occasionally a characteristic "hooklike" erosion of the metacarpal.[44] PIP hyperextension with a full SND may be present (as it was in Jaccoud's original case in 1869).

The mechanism of the arthropathy, like that of SLE, is incompletely understood. Capsular thickening and fibrosis with minimal synovitis have been observed in biopsy and necropsy specimens.[50,52] Presumably, the SND arises from a combination of articular laxity from capsular distension and progressive intrinsic tightness following ulnar subluxation of the intrinsic tendons. Tendon contracture secondary to primary inflammation of the tendons, associated with nodules and crepitus on exam, may also be a factor.

Miscellaneous Rheumatologic Causes

Both gout[28] and psoriasis,[53] diseases that are associated with marked synovitis, can lead to the SND as a result of joint destruction, compounded by the resulting imbalance of muscular forces. The precise mechanism depends on the location of predominant synovitis, that is, MCP, PIP, or DIP.

The SND has also been described in mixed connective tissue disease[54] and rarely in polymyositis.[55] The lack of historic data and the controversy over nosology make it impossible to determine whether the pathogenesis of the SND and other deformities in these overlap syndromes differ in any way from the arthropathy of SLE.

Neurologic Causes

A variety of neurologic disorders that share a common denominator of spasticity can have SND as a sequela. The best studied subgroup is that of patients with cerebral palsy.[21,56] The mechanisms that are implicated have included extrinsic, intrinsic, and articular. The relative contribution of each mechanism was elegantly studied by Swanson who performed selective motor nerve blocks of the extensor, flexor, and intrinsic muscles in 14 patients. Relaxation of the intrinsic muscles decreased the SND partially. However, inactivation of the extensor muscles with a radial nerve block essentially eliminated the SND. Simultaneous passive flexion of the wrist (extrinsic-plus test) caused recurrence of the SND. Thus, the combination of extrinsic muscle spasticity, flexion deformity of the wrist, and slight IM spasticity all contributed to the SND.

In contrast, in other neurologic causes of SND, such as parkinsonism, CVA, encephalitis, and head trauma, intrinsic muscle hyperactivity seems to be predominant. Anesthesia of the ulnar nerve that supplies the IM leads to improvement in the deformity. In parkinsonism, moreover, L-dopa and thalamic surgery have been shown to reverse early deformities.[22,57]

Hypermobility Syndromes

Joint hypermobility secondary to congenital ligamentous and capsular laxity is a feature of several disorders, including Marfan's syndrome, Ehlers-Danlos' syndrome, osteogenesis imper-

fecta, homocystinuria, hyperlysinemia, and idiopathic hypermobility syndrome.[58–62] Patients can have multiple articular abnormalities such as recurrent dislocations, effusions, osteoarthritis, and deformities, including reversible SND[59] (Fig. 14). The association with SND has not been emphasized in the literature, perhaps because the standard screening tests for hypermobility do not include assessment of PIP hyperextensibility.

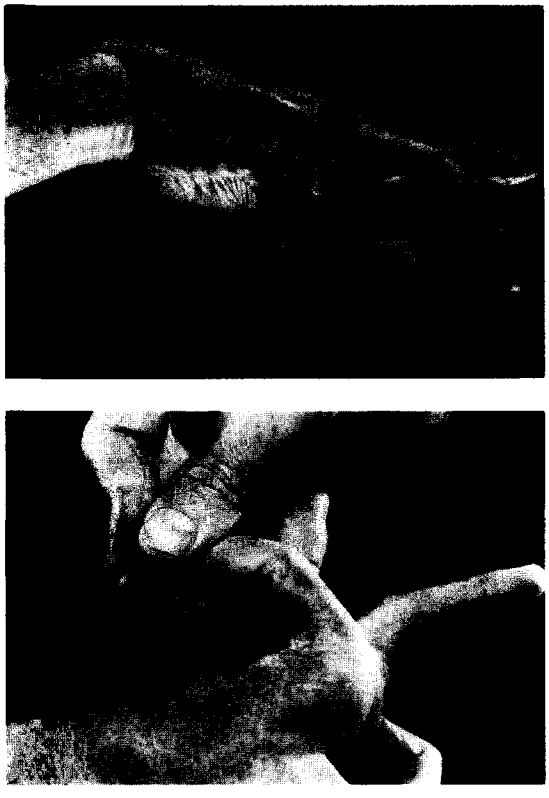

Fig. 14. Reversible articular type SND (*top*) in a patient with idiopathic hypermobility (*bottom*).

Diagnosis and Treatment of SND

Recognition of the underlying clinical diagnosis that is responsible for the SND is not difficult in most patients. The disorders listed in Table 1 have characteristic clinical histories and manifestations. Usually, a well-developed clinical syndrome will have been present for some time before the SND develops.

The pathogenetic mechanism of the SND can usually be determined by a physical examination. The hand should be evaluated for the presence of contributing deformities at the wrist, MCP, and DIP besides the obvious PIP hyperextension. Extensor and flexor tendon ruptures should be excluded. PIP flexibility during the extrinsic and intrinsic tests should be carefully observed. Hand radiographs are a useful adjunct in determining the presence and severity of primary PIP disease.

A detailed description of the surgical therapy of SND is beyond the scope of this review but is discussed in a number of reports.[63–66] The key to selection of appropriate surgical treatment depends exclusively on identification of the pathogenetic mechanism. Intrinsic muscle tightness can be relieved by various intrinsic release procedures that center on the extensor apparatus. Coexistent MCP involvement that contributes to IM tightness must be repaired at the same time for the intrinsic release to be effective. In cerebral palsy with pure intrinsic muscle spasticity, ulnar nerve section may be sufficient. Correction of wrist and MCP flexion contractures is an essential part of the treatment of extrinsic type SND. Finally, in articular type SND, flexor tendon tenodesis and retinacular ligament reconstruction are necessary to correct joint laxity and tether the PIP in flexion rather than hyperextension. Arthroplasty with silastic implants may be necessary in patients with severe articular destruction.

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