

Dupuytren Disease: An Evolving Understanding of an Age-old Disease

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Abstract

Dupuytren disease, a clinical entity originally described more than 400 years ago, is a progressive disease of genetic origin. Excessive myofibroblast proliferation and altered collagen matrix composition lead to thickened and contracted palmar fascia; the resultant digital flexion contractures may severely limit function. The pathophysiology is multifactorial and remains a topic of research and debate. Genetic predisposition, trauma, inflammatory response, ischemia, and environment, as well as variable expression of proteins and growth factors within the local tissue, all play a role in the disease process. Common treatments of severe disease include open fasciectomy or fasciotomy. These procedures may be complicated by the complex anatomic relationships between cords (pathologic contracted fascia) and adjacent neurovascular structures. Recent advances in the management of Dupuytren disease involve less invasive treatments, such as percutaneous needle fasciotomy and injectable collagenase *Clostridium histolyticum*. Postoperative management focuses on minimizing the cellular response of cord disruption and maximizing range of motion through static or dynamic extension splinting.

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Dupuytren disease is a fibroproliferative disease of the hand that may lead to disabling flexion contracture. Originally described nearly 400 years ago by Plater, the disease became better recognized after descriptions by the renowned French surgeon Baron Guillaume Dupuytren in the 1830s.¹⁻⁴ Understanding of the pathophysiology, anatomy, and treatment of Dupuytren disease is constantly evolving.

Pathophysiology

Dupuytren disease results from the complex interplay between genetic predisposition, environmental factors, local and global protein expres-

sion, and the relationship between tissue histology and anatomy. Despite recent advances in understanding the pathophysiology and genetic basis of the disease, the mainstay of treatment remains palliative and not curative.

Genetics

There is a clear genetic predisposition in the development of Dupuytren disease. The disease is most prevalent in northern European white males. Many authors believe the genetic origin of the disease dates back to the Vikings between the years 500 and 800 CE.^{2,3} Work by McFarlane⁵ suggests that the disease originated in Germanic tribes be-

tween 1200 and 200 BCE and was spread throughout the world by international Viking conquest and Germanic tribal migration. As a result, Dupuytren disease is relatively rare in those of Middle Eastern, Asian, or African descent.

The mode of inheritance of Dupuytren disease is believed to be autosomal dominant with variable penetrance. Siblings of patients with Dupuytren disease are at least three times more likely to develop the disease than is the general population, and several identical twins with Dupuytren disease have been reported.⁶ Genomic studies have established a region of interest on chromosome 16q.⁷

Still, no single polymorphism or group of genes has been identified as the sole cause of the disease. Advances in DNA microarray technology have allowed researchers to determine which genes and their products of expression are dysregulated compared with normal tissues⁸⁻¹¹ (Table 1). Overall, genes regulating the natural breakdown of collagen are inhibited, and genes promoting structural development of collagen in the epidermis are upregulated.

Cellular Pathology, Pathophysiology, and Environment

Diseased tissue is classically described in two forms: nodules and cords. The term Dupuytren nodule is typically used in either a clinical or histologic context. Clinically, nodules describe palpable subcutaneous lumps that may be fixed to the skin and palmar fascia. In the histologic context, nodules are dense, fusiform, hypercellular, hypervascular masses of tissue that are much smaller than clinical nodules and typically not palpable. Cords are highly organized collagen structures arranged in parallel with a relatively hypocellular

matrix.¹² Cords are predominantly composed of collagen type III; normal palmar fascia is predominantly composed of collagen type I.

Previously, nodules were believed to be highly cellular palmar structures causing active contraction, and cords were thought to be static, acellular matrices located mainly in the digits and formed as the end products of nodule contraction. Verjee et al¹³ have determined that nodules are found throughout cords in varying degrees and that a range of cellularity exists within each cord. Cords are described as “nodular” and “non-nodular” based on the presence of localized collections of cells—non-nodular cords are the least cellular and most contracted. Verjee et al¹³ describe Dupuytren disease as a heterogeneous mix of static and dynamic contractile elements located throughout the fascia of the hand and digits. Contracture, therefore, occurs when actively contracting cellular tissue shortens and transforms into a static, acellular matrix. This cellular contracture leads to the development of fixed flexion deformities. Increased cellularity within diseased tissue predicts an increase in recurrence rates up to 50%.¹⁴

The major contractile element in Dupuytren disease is the myofibroblast, a cell that shares characteristics of both fibroblasts and smooth-muscle cells.¹⁵ Myofibroblasts generate significant contractile forces and transmit this force to surrounding collagen matrices. Transforming growth factor- β (TGF- β) is a mechanotransduction cytokine protein abundant in Dupuytren tissue and is largely responsible for myofibroblast proliferation and fibroblast differentiation.¹⁶ TGF- β also enhances the contractile behavior of Dupuytren myofibroblasts, causing faster and stronger contractions in response to mechanical stimuli.¹⁷ Other factors that influence the differentiation, growth, and

Table 1

Select Genes and Proteins Upregulated and Downregulated in Dupuytren Tissue

Upregulated

A disintegrin and metalloproteinase domain (ADAM) 12
 Alpha smooth muscle actin (α -SMA)
 β -1 integrin
 Cadherin 11 (CDH 11)
 Collagen I, collagen V, collagen VIII
 Contactin 1 (CNTN1)
 Fibronectin
 Heat shock protein 47 (HSP47)
 Laminin
 Leucine-rich repeat (LRR) domain-containing 17
 V-maf musculoaponeurotic fibrosarcoma oncogene homolog B (MafB)
 Periostin, osteoblast specific factor (POSTN)
 Postsynaptic density protein-95 (PSD-95)
 Tenascin C
 Tissue inhibitor of metalloproteinase (TIMP-1)
 Transforming growth factor- β 2 (TGF- β 2)
 Zonula occludens-1 protein (ZO-1)

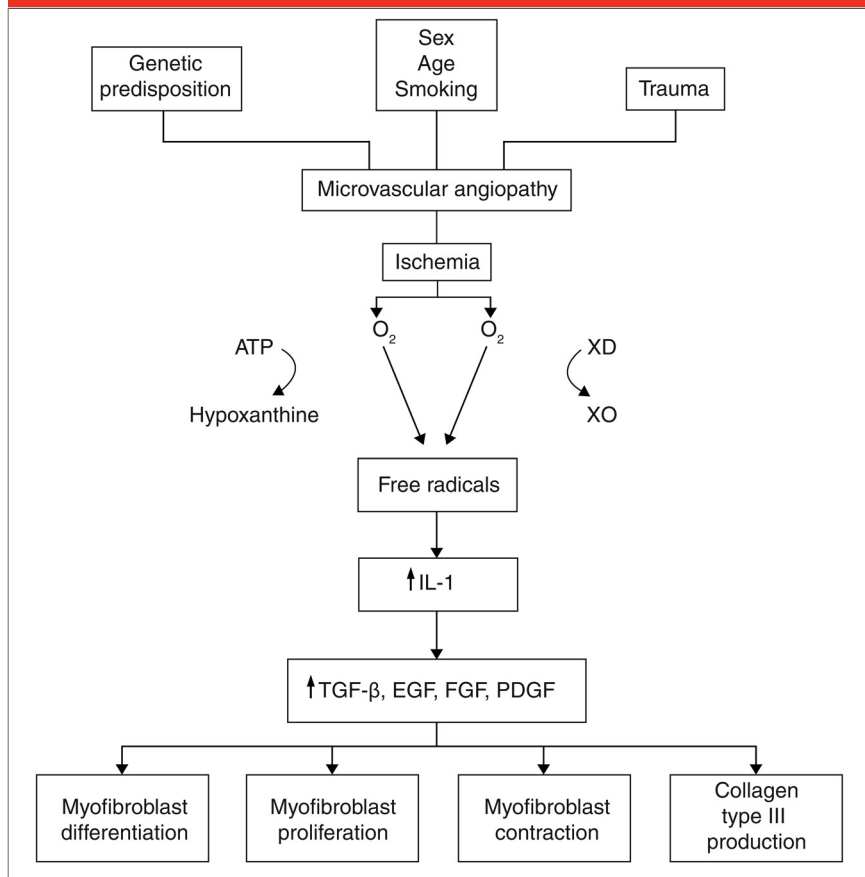
Downregulated

Chitinase 3-like 2
 Collagen XV
 Cornea-derived transcript 6 (CDT6)
 Cysteine dioxygenase 1 (CDO1)
 Matrix metalloproteinase 27 (MMP27)
 Matrix metalloproteinase-3 (MMP3)
 Superoxide dismutase (SOD)
 Superoxide dismutase 2 (SOD2)

contractility of myofibroblasts include the proteins platelet-derived growth factor, fibroblast growth factor, epidermal growth factor, and interleukin-1 (IL-1), as well as the cellular matrix proteins tenascin and periostin.^{14,18}

Trauma, microvascular angiopathy, and ischemia also play key roles in Dupuytren pathophysiology. Murrell et al¹⁹ believe that advanced age, genetics, and sex predispose patients to narrowed microvessels. Narrow-

Figure 1



Schematic of Dupuytren pathophysiology. Multiple factors may predispose patients to local tissue ischemia, leading to free radical release, which ultimately leads to myofibroblast differentiation, proliferation, and contraction, and to collagen dysregulation. ATP = adenosine triphosphate, EGF = epidermal growth factor, FGF = fibroblast growth factor, IL-1 = interleukin-1, O₂ = oxygen, PDGF = platelet-derived growth factor, TGF-β = transforming growth factor-β, XD = xanthine dehydrogenase, XO = xanthine oxidase

ing of the vessels is perpetuated through smoking, local trauma, and increased alcohol consumption. Microvascular disease causes local tissue ischemia, and through a complex molecular pathway, free radicals are released into surrounding tissue (Figure 1). These free radicals stimulate fibroblast proliferation and favor preferential production of collagen type III over type I. Free radicals also stimulate cytokines such as IL-1, which promotes myofibroblast proliferation, differentiation, and contraction through its effects on TGF-β, fibroblast growth factor, and

platelet-derived growth factor.²⁰ IL-1 is also released directly through microtrauma or disturbances of the palmar fascia secondary to infection. Thus, Dupuytren disease has been reported after trauma and/or infection.²¹

In population studies, patients who smoke or drink heavily are at increased risk to develop Dupuytren disease by odds ratios of approximately 1.3-2.8:1 and 1.6-1.8:1, respectively.^{22,23} The same holds true for males, those of increased age, and persons with diabetes mellitus (type I more so than type II). Loose

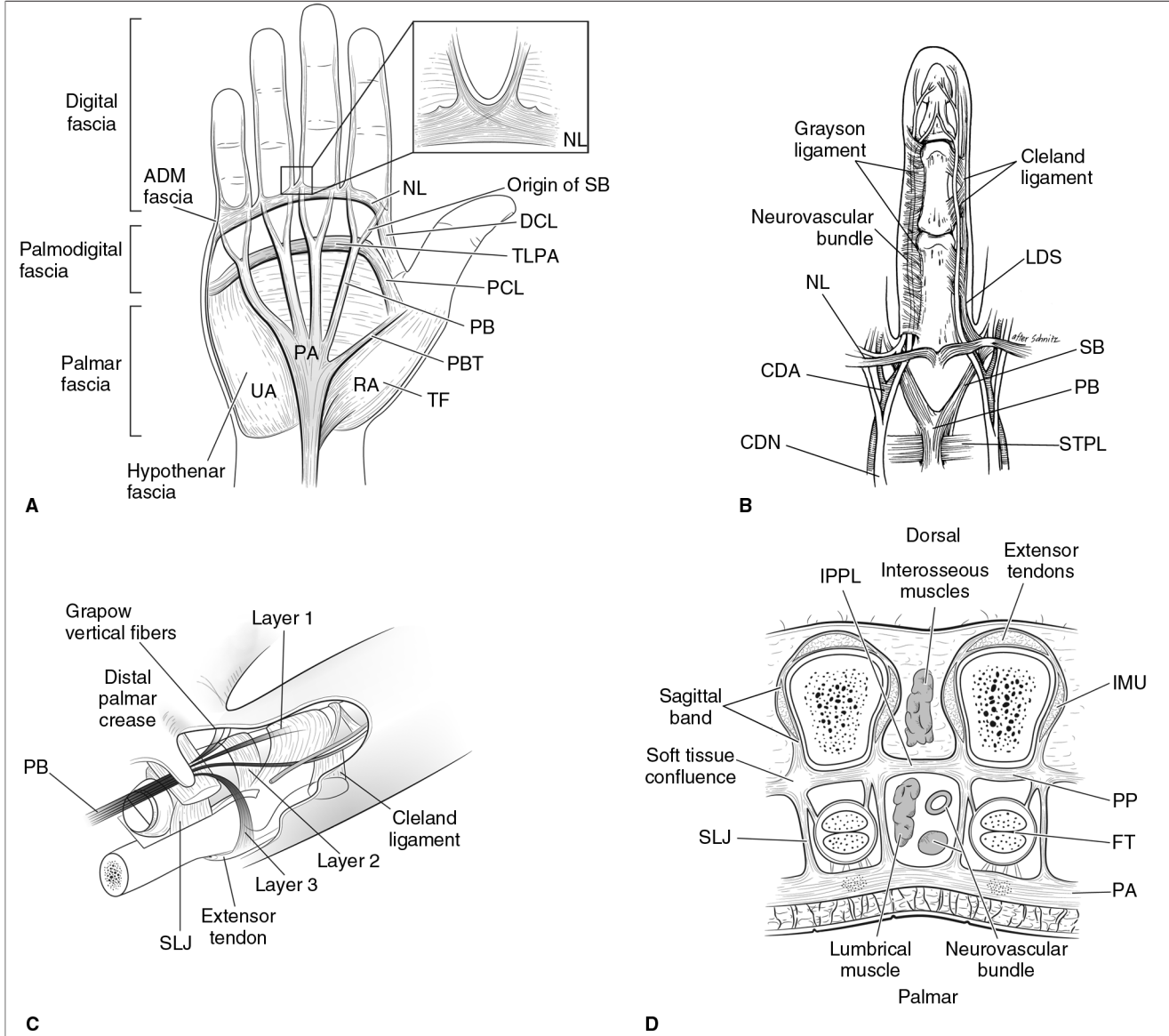
associations have also been made between Dupuytren disease and anti-epileptic medications and manual labor.

Anatomy and Pathoanatomy

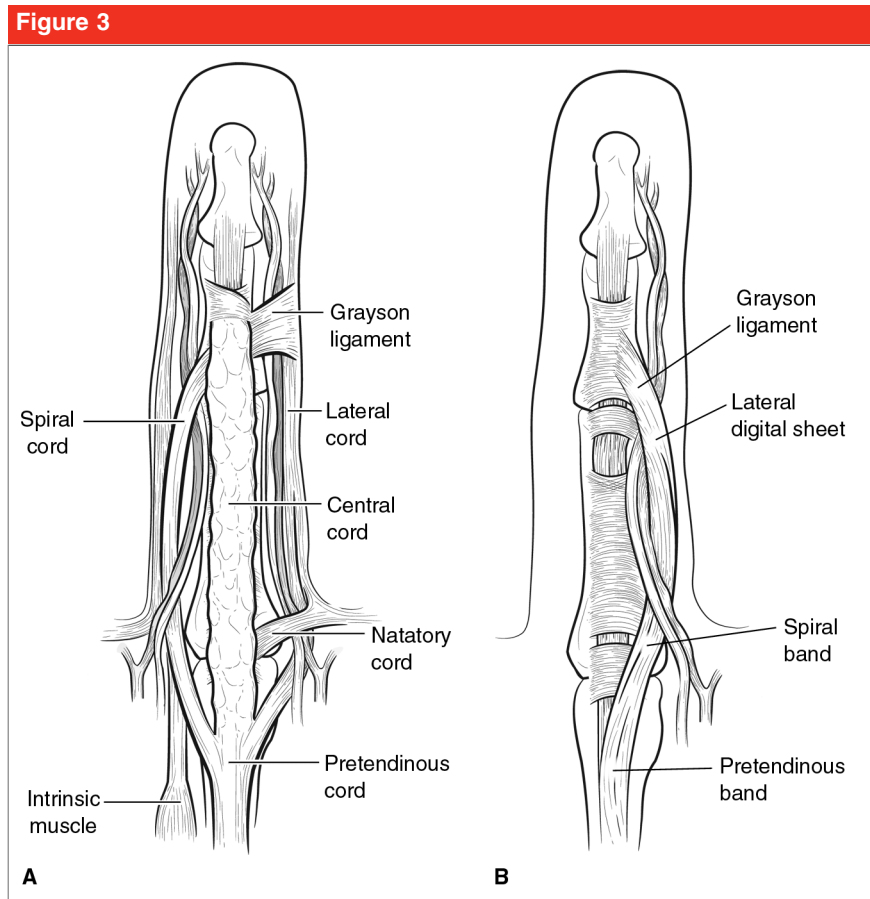
A thorough understanding of the complex anatomy of the palmar fascial structures is necessary in the management of Dupuytren disease. The palmar fascial complex may be divided into three zones—digital fascia, palmodigital fascia, and palmar fascia.²⁴ (Figure 2, A). Most proximally, the palmar fascia consists of the radial, ulnar, and central (ie, palmar) aponeuroses. The radial aponeurosis is subdivided into the thenar fascia, the pretendinous band to the thumb, the proximal commissural ligament (the thenar extension of the transverse ligament of the palmar aponeurosis), and the distal commissural ligament (the thenar extension of the natatory ligament [NL]). The ulnar aponeurosis is subdivided into the pretendinous band to the small finger (a large structure), hypothenar muscle fascia, abductor digiti minimi soft-tissue confluence, and pisiform ligamentous complex.

The central (ie, palmar) aponeurosis is most intimately involved in Dupuytren disease and may be divided into three layers with distinct orientations: longitudinal, vertical, and transverse. Longitudinally, the central aponeurosis is a triangular structure that fans out distally and becomes organized into distinct structures known as pretendinous bands (Figure 2, A and B). Distally, the pretendinous bands bifurcate and have three distinct layers of insertion (Figure 2, C). Superficially, the bands insert into the dermis and—most deeply—into the flexor and extensor mechanisms. The middle layer wraps around the metacarpal head by twisting 90° and traveling vertically ad-

Figure 2



A, There are three major divisions of fascia in the hand. Within the palmar fascia are the ulnar aponeurosis (UA), palmar aponeurosis (PA), and radial aponeurosis (RA). The transverse longitudinal palmar aponeurosis (TLPA) lies transversely before the bifurcation of the pretendinous bands. **B**, Within the digit, a portion of the pretendinous band wraps around the metacarpal head and twists around deep to the neurovascular bundle, emerging as the lateral digital sheet and located lateral to the neurovascular bundle. **C**, The three vertical layers of insertion of the pretendinous fibers: (1) the distal palmar dermis (Grapow vertical fibers) and pretendinous fibers to the proximal digit; (2) distally, as the lateral digital sheet from the spiral band; and (3) deeply into the flexor and extensor mechanisms. **D**, The vertical fibers of the central (palmar) aponeurosis help compartmentalize the hand. The neurovascular bundle runs with the lumbrical muscle and the flexor tendons run together. These are separated by the septa of Legueu and Juvara. ADM = abductor digiti minimi, CDA = common digital artery, CDN = common digital nerve, DCL = distal commissural ligament, FT = flexor tendons, IMU = intrinsic musculotendinous unit, IPPL = interpalmar plate ligament, LDS = lateral digital sheath, NL = natatory ligament, PB = pretendinous band, PBT = pretendinous band to the thumb, PCL = proximal commissural ligament, PF = pretendinous fibers, PP = palmar plate, SB = spiral band, SLJ = septa of Legueu and Juvara, STPL = superficial transverse palmar ligament, TF = thenar fascia. (Panels A and D redrawn with permission from Rayan GM: Palmar fascial complex anatomy and pathology in Dupuytren's disease. *Hand Clin* 1999;15:73-86. Panel B reprinted from Benson LS, Williams CS, Kable M: Dupuytren's contracture. *J Am Acad Orthop Surg* 1998;6[1]: 24-35. Panel C redrawn with permission from McGrouther DA: The palm, in McFarland RM, McGrouther DA, Flint MH, eds: *Dupuytren's Disease: Biology and Treatment*. New York, NY, Churchill Livingstone, 1990, vol 5, pp 127-135.)



A, A large central cord may cause significant proximal interphalangeal flexion deformity. Natatory cords may cause web space contractures. **B**, As a spiral cord contracts, the neurovascular bundle is pulled to the midline. The four major contributions to the spiral cord are the pretendinous band, spiral band, lateral digital sheet, and Grayson ligament. (Panel A redrawn with permission from McGrouther DA: Dupuytren's contracture, in Green DP, Hotchkiss RN, Pederson WC, Wolfe SW, eds: *Operative Hand Surgery*, ed 5. New York, NY, Churchill Livingstone, 2005, vol 1, pp 159-186. Panel B redrawn with permission from McFarlane RM: Patterns of the diseased fascia in the fingers in Dupuytren's contracture. *Plast Reconstr Surg* 1974;54[1]:31-44.)

adjacent to the metacarpophalangeal (MCP) joint capsule to form the spiral band. This layer continues deep (dorsal) to the neurovascular bundle and emerges distally as the lateral digital sheet—lying directly lateral to the neurovascular bundle.

The vertical fibers of the central aponeurosis consist of the Grapow vertical fibers and the septa of Legueu and Juvara (Figure 2, C and D). Grapow fibers are superficial and small, and they anchor the dermis primarily to the palmar aponeurosis.

Eight vertical septa of Legueu and Juvara create seven fibro-osseous compartments deep to the palmar fascia. Four of these compartments contain canals for the paired flexor tendons; the remaining three contain the neurovascular bundles and an associated lumbrical muscle.

The transverse fibers of the central aponeurosis include the transverse ligament of the palmar aponeurosis (also known as the superficial transverse ligament) proximally and the NL distally.

Distally, the lateral digital sheet is located on either side of the finger laterally and is a continuation of the spiral band and NL (Figure 2, B). The digital neurovascular bundle is surrounded by four major structures: Grayson ligaments palmarly, Cleland ligaments dorsally, the lateral digital sheet laterally, and the Thomine retrovascular fascia medially.

In Dupuytren disease, normal fascial bands and ligaments are transformed into contracted, diseased tissue referred to as cords. This transformation leads to joint contracture, stiffness, decreased skin mobility, and contracted fascial tissue that notably distorts surrounding anatomic structures. Skin thickening normally observed in Dupuytren disease is a result of the conversion of Grapow superficial fibers into small “micro-cords.”²⁴

The pretendinous cord (Figure 3, A) is the most commonly involved cord in the hand and results in MCP contracture (Table 2). This cord does not typically displace the neurovascular bundle. The superficial layer of the pretendinous cord inserts into the dermis and results in skin pitting. The vertical bands arising distally from the central aponeurosis and pretendinous cord transform into vertical cords, which are diseased septa of Legueu and Juvara. These less common cords surround the neurovascular bundle and flexor tendons in the hand and, when severe, can lead to stenosing tenosynovitis or painful triggering. The transverse ligament of the palmar aponeurosis is typically spared in Dupuytren disease. The NL develops from a U-shaped structure in the web spaces into the V-shaped natatory cord, leading to contractures of the second through fourth web spaces.

The spiral cord (Figure 3, B) is derived from four main structures: the middle layer of the pretendinous band, spiral band, lateral digital

sheet, and Grayson ligament. In the hand, the cord is superficial to the neurovascular bundle until the level of the MCP joint, where it passes deep to the bundle. The spiral cord then shifts lateral to the bundle, where it begins to involve the lateral digital sheet. Distally, the cord is superficial to the neurovascular bundle as it fuses with the Grayson ligament. Cleland fascia is typically spared in Dupuytren disease. In its early form, the spiral cord wraps around the neurovascular bundle in a spiral fashion, but with further contraction, the cord straightens and is pulled toward the midline. This displaces the bundle superficially and toward the midline and therefore places the neurovascular bundle at risk of iatrogenic injury during surgery. In the little finger, the cord may be isolated to the digit and arise from the musculotendinous junction of the abductor digiti minimi muscle. This is referred to as the abductor digiti minimi cord, which is generally superficial to the neurovascular bundle and therefore does not frequently displace it. In addition to the little finger, other digits may develop isolated cords.^{24,25}

The central cord (Figure 3, A) is a midline structure in the finger that arises as an extension of the palmar pretendinous cord and attaches to the base of the middle phalanx and often the tendon sheath. Typically, this cord does not displace the neurovascular bundle but does cause proximal interphalangeal (PIP) flexion contracture. The lateral cord, which arises from the lateral digital sheet, may also cause PIP flexion contracture as well as distal interphalangeal contracture. The retrovascular cord, which arises from digital fascia dorsal to the neurovascular bundle, may combine with the lateral cord in recurrent Dupuytren disease to cause distal interphalangeal hyperextension contracture.²⁴

Table 2**Cords and Associated Deformities**

Dupuytren Cord	Deformity
Pretendinous cord	MCP contracture + skin pitting
Natatory cord	Web space contracture
Spiral cord	MCP + PIP contracture; displaces neurovascular bundle both superficially and toward midline
Central cord	PIP flexion contracture
Lateral cord	PIP or DIP flexion contracture
Retrovascular + lateral cord	DIP hyperextension contracture in recurrent disease

DIP = distal interphalangeal, MCP = metacarpophalangeal, PIP = proximal interphalangeal

Clinical Presentation and Diagnosis

Dupuytren disease is more common in males and in patients aged >40 years. The sex-specific ratio of males to females is as high as 7:1 in Europe. Recent epidemiologic studies in the United States suggest that the overall disease ratio is approximately 1.7:1 and that this ratio approaches 1:1 with increasing patient age.²⁶

Early in the disease, fibrosis of Grapow fibers leads to palmar skin pitting and thickening.²⁴ Patients may discover a painless nodule or cord in the palm as well as the development of a progressive flexion contracture. Care must be taken to distinguish early Dupuytren disease from other disease entities. Trigger finger, or stenosing tenosynovitis, presents with symptomatic locking or triggering and tenderness in the region of the A1 pulley. Soft-tissue masses and tumors usually do not present with characteristic skin thickening and pitting.

As the disease advances, nodules progress into cords through myofibroblast contraction and the conversion of cellular components into relatively acellular collagen matrices and cords. These cords become fixed to

the underlying skin and may resemble flexor tendons. Patients with notable disease (usually >30° of MCP contracture) often cannot place the palm of their diseased hand flat on a table, indicating a positive tabletop test.

Not all nodules transform into cords and result in clinically relevant flexion contracture. Studies estimate that approximately 50% of patients with palpable nodules will develop cords, and only a fraction of these will necessitate surgery.²⁷

The most commonly affected digit is the ring finger, followed by the small finger, long finger, index finger, and thumb. Flexion contracture of the MCP normally appears before PIP contracture does. Dupuytren disease commonly begins in the palm and spreads distally into the digits, but it is also possible for the disease to be isolated to the palm or digits.

Ectopic disease may be located distant to the palmar fascia. Patients with bilateral disease commonly present with Garrod nodes, that is, knuckle pads located over the dorsal PIP joint. These patients have higher incidences of concurrent ectopic disease, including Peyronie disease (ie, penile fibromatosis) and Ledderhose disease (ie, plantar fibromatosis). Dupuytren disease has also been de-

scribed isolated to the wrist.²⁸

Dupuytren diathesis describes a combination of features in patients with increased disease severity and recurrence risk. These criteria have recently been updated and include male sex, onset at <50 years of age, bilateral disease, one or more affected siblings/parents, Garrod pads, and Northern European descent. The presence of one or more of these features generally portends a worse prognosis and increased recurrence rates. When all of these factors are present, patients have a recurrence risk of 71% compared with 23% in those with no risk factors.²⁹

Nonsurgical Management

The role of nonsurgical management in Dupuytren disease has been evolving in recent years. Observation is a key form of nonsurgical treatment because isolated nodules without contracture do not always develop into cords, and cord contracture may be nonprogressive. Many patients with nodules that develop into contracted cords, however, progress to flexion contractures, which eventually limit function and warrant more aggressive treatment.²⁷ Physical therapy and splinting as therapies alone are generally not effective, but they are helpful in the postsurgical period. Ketchum and Donahue³⁰ have reported success with intralesional triamcinolone injection of Dupuytren nodules, with most patients experiencing softening or flattening of the nodules. In this study, however, 50% of patients experienced recurrence.

Percutaneous Fasciotomy

In percutaneous fasciotomy (or percutaneous needle aponeurotomy), a needle is typically used to percutaneously section the contracted cord at multiple levels, followed by early active and passive range-of-motion

(ROM) exercises. Proponents of this method cite early return of motion, low complication rates, and improvement of joint contractures, especially at the MCP joint. However, published recurrence rates have been high, and patients with more severe disease have less improvement than do those undergoing surgical release.

van Rijssen and colleagues^{31,32} directly compared percutaneous needle fasciotomy with limited fasciectomy (ie, surgical release) in a level I study. Patients' extension deficits improved between 61% to 76% with needle fasciotomy, and patients with less severe disease (<90° total passive extension deficit) showed initial improvements comparable to those of surgical fasciectomy. The authors reported high early satisfaction rates and low long-term complication rates (including no flexor tendon injury). However, at 5-year follow-up, 85% of patients who underwent needle fasciotomy developed a recurrence, compared with 24% of fasciectomy patients. The average time to recurrence was 3.7 years.

Based on these and other results, many surgeons believe percutaneous fasciotomy to be an acceptable preliminary treatment of early disease with less severe contracture (ie, total passive extension deficit <90°). It may also provide a useful tool in treating elderly, sicker patients who cannot tolerate surgery and for patients in whom immediate improvement would be useful.

Collagenase

Collagenase is a mixture of two enzymes derived from *Clostridium histolyticum* that lyses contracted collagen cords. This relatively new and less invasive treatment is delivered by injection into a diseased cord and may be performed in the office without general anesthesia. The authors of a recent phase III prospective,

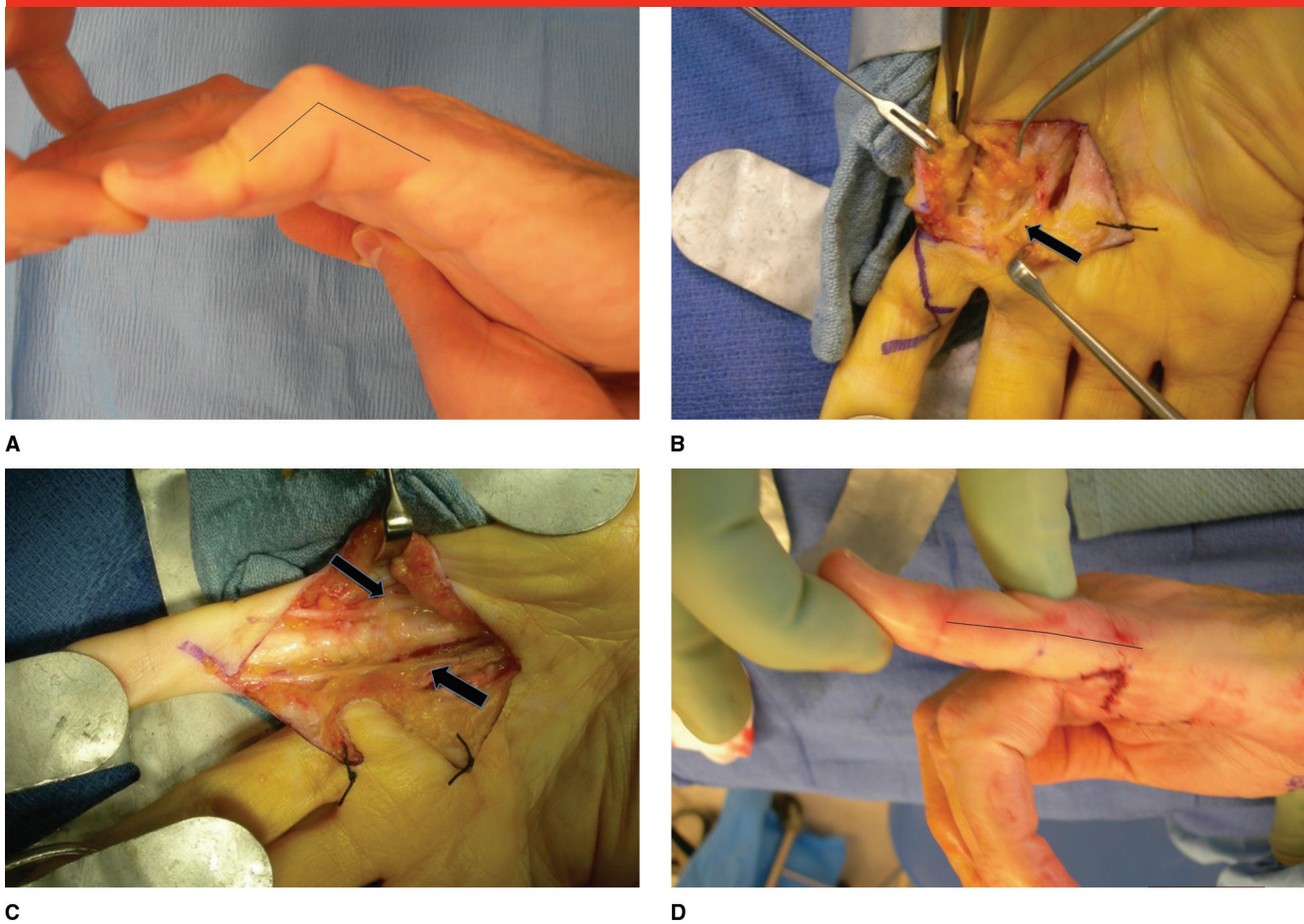
multicenter, double-blind, randomized, placebo-controlled study³³ studied the efficacy of collagenase injection in more than 300 patients with contractures of $\geq 20^\circ$. Cords detected by physical examination were injected and subsequently manipulated to facilitate release. This cycle was repeated up to three times per cord in 30-day intervals. The primary end point was a reduction of contracture to $< 5^\circ$ in a particular joint. The authors discovered a notable decrease in the level of contracture for patients in the treatment arm: 77% of patients with MCP contracture and 40% of patients with PIP contracture met the primary end point compared with 7.2% and 5.9%, respectively, of patients in the placebo group. Overall joint ROM also improved by an average of 41° in MCP joints and 29° in PIP joints over baseline compared with $< 5^\circ$ in placebo. After injection, 92% of MCP contractures were $< 30^\circ$. Long-term data are lacking with respect to follow-up ROM examinations and recurrence.

The most common complications of collagenase injections are edema, contusion, pain (resulting from injection and manipulation), lymphadenopathy, and skin laceration (resulting from manipulation). Major complications (eg, complex regional pain syndrome, flexor tendon rupture) are rare ($< 1\%$).

Surgical Treatment

Surgical intervention is often indicated in cases of $\geq 30^\circ$ of MCP contracture or any PIP contracture (usually $> 15^\circ$). Surgery usually involves a form of fascial excision. The method of excision, extent of intraoperative correction, and planned skin incisions and skin coverage are all essential considerations in optimizing outcomes.

Figure 4



Limited fasciectomy for proximal interphalangeal contracture. **A**, Preoperative clinical photograph of flexion contracture measuring approximately 60° in the small finger. **B**, Intraoperative photograph identifying the neurovascular bundle proximally (arrow) and clamp on the proximal aspect of the cord. **C**, Intraoperative photograph. The cord has been removed and the flexor tendon is shown; the flexor pulley system is not disrupted. The neurovascular bundles (arrows) are preserved. **D**, Immediate postoperative photograph demonstrating $<5^\circ$ flexion contracture.

Methods of Surgical Release

Management options for diseased palmar fascia include fasciotomy, local or segmental fasciectomy, radical fasciectomy, limited fasciectomy, and dermofasciectomy. Fasciotomy is local division of the diseased cord without its removal. This can be done with a needle or with small open incisions. Local fasciectomy is a limited removal of diseased fascia through one or more incisions. Although less invasive and providing earlier functional benefit than other options, both procedures tend to

have higher recurrence rates and earlier postoperative contractures.^{25,34}

Conversely, radical fasciectomy is an extensive removal of both healthy palmar fascia and diseased fascial cords. The aim of removing healthy fascia is to remove tissue that may appear to be healthy but that has the possibility of causing recurrent disease. This method has been largely abandoned because of increased complication rates and recurrence rates similar to those of other methods of excision.³⁴

Limited fasciectomy (Figure 4) is the most widely performed method

of surgical excision and involves a longitudinal dissection of diseased tissue. Skin is sharply divided in the layer between fascia and fat, and fascia is lifted off proximally to distally. Typically, only diseased tissue is removed; adjacent normal fascia is spared. Great care must be taken to identify and protect the neurovascular bundle in the digit because the contracted cord may displace it toward the midline. The surgeon must be meticulous about hemostasis, because hematoma formation can be a notable complication.

Dermofasciectomy is similar to

limited fasciectomy but involves the excision of overlying contracted skin and coverage with skin graft (Figure 5). Proponents of this method argue that diseased tissue is not isolated within the cord itself but involves a substantial amount of overlying skin and soft tissue that is typically left behind in traditional closures.³⁴

Despite many studies detailing the outcomes of each of these methods, few studies exist that directly compare treatment modalities.³⁵ Overall, however, surgery has been validated by objective outcome measures to notably improve hand function.³⁶ In addition, one survey of more than 1,100 patients undergoing surgical release described excellent self-reported outcomes measures. Seventy-five percent of patients reported “full or almost full” correction of deformity 27 months postoperatively, with only 15% reporting disease recurrence.³⁷

The survey by Dias and Braybrooke³⁷ also detailed two key points that are prevalent throughout the surgical outcomes literature: higher degrees of initial deformity and the location of deformity in the PIP joint rather than the MCP joint both portend worse prognoses. Patients with larger initial deformities may have decreased rates of correction,³⁸ higher complication rates,³⁷ worse grip strength, and possibly worse functional outcomes postoperatively.³⁶

The MCP joint is notably more forgiving than the PIP joint in Dupuytren disease, both in surgical and nonsurgical management. MCP joints are much more likely than PIP joints to achieve full intraoperative correction and to remain this way 6 months postoperatively.³⁹ Patients whose PIP joints achieved full intraoperative correction are also more likely to lose this correction by 6 months compared with patients with similar MCP contractures.

Higher initial degrees of contraction (especially those >60°) and non-

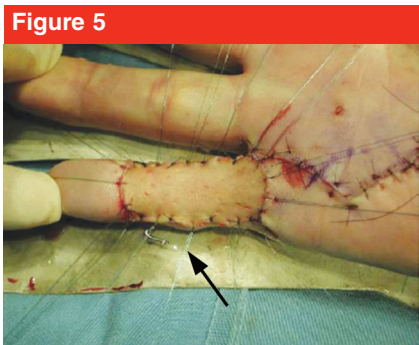


Figure 5
Clinical photograph of a full-thickness skin graft in dermofasciectomy. The proximal interphalangeal joint has been pinned with a Kirschner wire (arrow). (Reprinted with permission from Abe Y, Rokkaku T, Kuniyoshi K, Matsudo T, Yamada T: Clinical results of dermofasciectomy for Dupuytren’s disease in Japanese patients. *J Hand Surg Eur* 2007;32[4]:407-410.)

compliance with therapy increase the risk of recurrent PIP joint deformity.^{39,40} However, improvements in PIP joint contracture are correlated with statistically significant improvements in hand function.⁴¹ It is for these reasons that some surgeons advocate earlier and more aggressive releases for less deformed PIP joints to minimize recurrence and maximize chances for functional improvement. However, aggressive manipulation in severe PIP contracture may lead to digital artery spasm and digital ischemia. This situation must be identified immediately and a lesser degree of correction accepted.

Release of the contracted PIP joint capsule remains controversial; no overwhelming body of data supports or rejects the concept.⁴⁰ Many surgeons believe that release of the contracted PIP joint is sometimes the only way for a patient to regain and keep full extension. However, exposure requires violation of the flexor-tendon pulley system and a more extensive dissection. This method therefore confers added morbidity and the possibility of postoperative complications, including

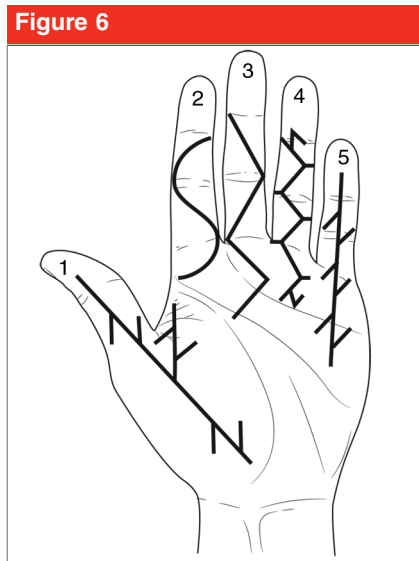


Figure 6
Illustration of types of incisions and possible closure methods for fasciectomy. Type 1, combination of longitudinal incision with multiple Z-plasty. Type 2, curvilinear incision. Type 3, Brunner incision. Type 4, combination of V-Y incision with Z-plasty. Type 5, A midline longitudinal incision closed with Z-plasty. (Redrawn with permission from McGrouther DA: Dupuytren’s contracture, in Green DP, Hotchkiss RN, Pederson WC, Wolf SW, eds: *Operative Hand Surgery*, ed 5. New York, NY, Churchill Livingstone, 2005, vol 5, pp 159-186.)

wound breakdown, swelling, and pain and stiffness if a therapy regimen is not followed. Some surgeons prefer to temporarily immobilize the PIP joint after release (ie, using a Kirschner wire), although the efficacy of this has not been studied in detail.

Soft-tissue Considerations

A large number of surgical incisions have been used in performing a fasciectomy. A midline incision with Z-plasty closure, a Brunner incision with primary closure, or a zigzag incision with V-Y advancement flaps are examples (Figure 6). The benefit of a midline incision is that the neurovascular bundle will almost never

be drawn to the exact midline of the palmar digit even in severe contracture, and it can easily be converted to a lengthened Z-plasty for closure. One study examining surgical approaches randomly assigned either a Brunner incision closed with V-Y advancement flaps or a longitudinal incision closed with Z-plasty and found no statistically significant differences in wound complications or recurrence rates.⁴²

It is important to remember to maintain adequate skin bridge thickness to prevent flap necrosis. In addition, adequate exposure is essential, and the surgeon should not hesitate to extend the incision if necessary.

Skin closure may occur by direct suture or flap advancement closure. Skin deficits may be treated by skin grafting, or portions of the wound may be left open to heal by secondary intention—the open palm technique. Grafting may also be part of the planned procedure, with the aim of removing diseased skin (dermofasciectomy). Many surgeons believe removing diseased skin is essential to preventing recurrence, especially in recalcitrant disease, recurrent disease, disease involving the skin, or in patients with Dupuytren diathesis. Ullah et al⁴³ randomized patients to primary wound closure or skin removal with full-thickness grafting. There were no statistically significant differences in outcomes or recurrence rates between the two groups. The study reported a nonstatistically significant trend toward increased rates of infection and wound dehiscence in the dermofasciectomy group ($P = 0.08$ and $P = 0.1$, respectively).

Complications

Overall physician-reported surgical complication rates range from 3.6% to 39.1% in the literature, with major complications occurring in 15% of patients on average.⁴⁴ Wound-

healing complications (22.9%) and pain (18%) are most common in those with primary disease. Digital artery and nerve injury occur in approximately 2.0% and 3.4% of surgeries, respectively. Infection rates are approximately 2.4%, and complex regional pain syndrome occurs, on average, in 5.8% of patients. Patients' self-reported complications may be as high as 46% in those undergoing surgery, the most common being numbness lasting >2 days postoperatively (36%), followed by wound infection (19%).³⁶ Complication rates increase substantially when correcting deformity >60°.

Recurrence

Although surgical results have been shown to be satisfactory, it is important that the patient be educated about the possibility of disease recurrence and spread. A complex interplay exists between genetic predisposition, environmental effects, and global and local disease pathophysiology; therefore, surgical excision cannot be completely curative. Although many surgeons define recurrence as Dupuytren disease returning to an area of prior surgical removal, and define extension as contracture resulting in disease in a nonoperated area, these terms can be difficult to differentiate in practice.

Recurrence rates of Dupuytren disease increase with time from surgical release, and recurrent disease is more common in patients with higher preoperative flexion contractures, especially of the PIP. In the literature, as many as 50% of patients may have recurrent flexion contractures at 5 years, although rates vary by case series and the respective definitions of recurrence.³⁴

Surgical dissection is notably more difficult in recurrent Dupuytren disease because previous landmarks and tissue planes are more obscure. Com-

plication rates are also higher; digital nerve and artery injuries may be up to 10 times more common in patients with recurrent disease.⁴⁴ Many surgeons prefer dermofasciectomy in recurrent disease because the skin is frequently involved in the contracture. Other surgical options include PIP joint fusion, extension force external fixators, and, rarely, amputation for painful or dysvascular digits.

Postoperative Considerations

Most surgeons believe that some form of postoperative rehabilitation is essential to maintain flexibility and prevent further contracture after release. Patients are typically immobilized for 2 or 3 days postoperatively (up to 10 days with skin grafting) before beginning therapy. Skin grafts must be protected from shear forces for up to 1 month postoperatively. Hand therapists focus on wound healing, managing scar tissue, and maximizing ROM, often through use of splinting therapy.

Splinting techniques are variable but tend to focus on continuous low-load extension forces to the affected digits. Although static extension splinting may play a role in decreasing flexion contractures, static extension splinting alone may increase fibroblastic scar response and lead to lower total finger flexion and worse upper extremity function scores because of stiffness.⁴⁵ Dynamic splinting incorporates active movement in the therapy program with the hope of preventing postoperative stiffness in flexion. Both splinting techniques can be combined at different periods of the day (static at night and dynamic during the day) to maximize gain. ROM exercises should persist into the scar maturation period (up to 1 year postoperatively), and resistive exercises may begin approximately 4 weeks postoperatively.

Summary

Dupuytren disease, believed to have originated at least 2,500 years ago and first described more than 400 years ago, is a fibroproliferative disease of abnormal collagen production (collagen type III) and contraction. The complex interplay between genetic predisposition, genetic and protein dysregulation, environmental factors, and local tissue environment is not completely understood. Transmission is autosomal dominant with variable penetrance, and the myofibroblast is the predominant cell type involved in soft-tissue contraction and collagen deposition. Management is based on level of contracture (usually $>30^\circ$ MCP and $>15^\circ$ PIP contracture) and may be surgical or nonsurgical. Neither option, however, is curative, and recurrence is common with time. Understanding surgical anatomy is key in disease resection because cords are often intimately involved with critical anatomic structures. Postoperative therapy typically focuses on scar maturation and minimizing residual flexion and/or extension contractures with formal ROM therapy.

References

Evidence-based Medicine: Levels of evidence are described in the table of contents. In this article, references 31-33, 42, and 43 are level I studies. References 6, 16, 22, 29, 30, 36, and 39-41 are level II studies. References 8-11, 14, 17, 19, 23, 26, 27, 37, and 38 are level III studies. References 7 and 28 are level IV studies. References 5 and 12 are level V expert opinion. References printed in **bold type** are those published within the past 5 years.

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