BMJ Best Practice **Dupuytren contracture**

Straight to the point of care



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Summary

Dupuytren contracture is an inherited disease of progressive fibrous tissue contracture of the palmar fascia.

Predominantly affects men of northern European descent >40 years old who smoke, drink alcohol, or have diabetes.

Patients present with a small lump or multiple lumps with pits in the palm of the hand, progressing to contractures of the fingers.

Intralesional corticosteroid injections have been shown to reduce the need for surgery.

Surgical referral should be made when metacarpophalangeal joint contractures reach 30°, or if any degree of proximal interphalangeal joint contracture is present.

Percutaneous needle fasciotomy and collagenase injections are significant therapeutic alternatives to surgery.

Definition

An inherited disease of progressive fibrous tissue contracture of the palmar fascia, Dupuytren contracture is seen predominantly in men of northern European descent aged >40 years. Its inheritance pattern is believed to be autosomal dominant with variable penetrance.[1]

Epidemiology

There are differing reports on the prevalence of Dupuytren contracture depending on region and study design, however worldwide disease prevalence is 8.2%, with the highest rates per continent observed in Africa at 17.2%, and the lowest in America at 2.3%.[4] Other studies report higher prevalence among those of European descent than in other races, and within Europe it is higher in northern than in Mediterranean countries.[5] Prevalence in Western countries ranges from 0.6-31.6%.[6] For instance, in the same age group, prevalence has been reported as 22.0% (with progression to contractures in 4.2%), 2.6%, and 0.67% in Dutch, Swedish and UK cohorts respectively.[7] [8] [9] In Japan, disease prevalence is estimated to be 7.0%.[10]

Dupuytren disease is twice as common in men as in women, and incidence increases with age.[6] [7] [8] [9] [10]

Etiology

Dupuytren contracture is a progressive fibroproliferative disease that is believed to show autosomal dominant inheritance with variable penetrance.[1] The higher prevalence of the disease in men is probably due to the fact that androgen receptors are expressed in Dupuytren nodules.

The familial occurrence and its presence in identical twins suggest a genetic basis for the disease. The sibling recurrence risk ratio equals 2.9 (95% CI 2.6 to 3.3). Although the genetic factors involved have not been fully elucidated, in one family the gene has been mapped to chromosome 16q.[1] DNA microarray has demonstrated that >30 unique genes are upregulated and 6 unique genes downregulated by a factor of 4 or greater in affected patients.[11] A novel gene, MafB (musculoaponeurotic fibrosarcoma oncogene homolog B), is overexpressed in patients with Dupuytren contracture but not in those with normal fascia.[11]

A high number of mitochondria have been demonstrated in fibroblasts derived from diseased tissue, and a mitochondrial defect in the 16S ribosome RNA has been noted in 90% of affected patients compared with none in the control patients.[12] Thus, a mitochondrial etiology may be significant in some families, while a chromosomal defect may be important in others.

A number of growth factors, immunologic mediators, and free radicals are also implicated as causative factors, and it seems that an inciting factor triggers these mediators to stimulate myofibroblast proliferation.[13]

Diabetes mellitus has a strong association with Dupuytren contracture.[4] [10] [14] [15] [16] The prevalence of the disease in patients with diabetes ranges from 3% to 33% and increases with the duration of the diabetes. It is hypothesized that diabetes causes microvascular changes that produce local hypoxic tissue damage, inducing Dupuytren contracture. Patients with diabetes tend to have a mild form of the disease, with slow progression.

Greater alcohol intake is associated with an increased likelihood of Dupuytren contracture.[4] [7] [10] [17] However, it should be noted that most patients with the disease do not misuse alcohol, and not all studies have found a significant association.[18] For unclear reasons, smoking increases risk, although the microvascular changes associated with smoking and the effect of carbon monoxide on mitochondrial cytochrome c, released by mitochondria in response to pro-apoptotic stimuli, may play a role.[17]

Theory

Trauma was first proposed to be a cause by Dupuytren himself. Since the first report that linked trauma to the disease, the strength of this association has been controversial. It is unclear why the disease would be caused by heavy labor, and further studies are required to control for smoking and alcohol use.[19]

There have been reports of a connection between epilepsy and Dupuytren contracture. When discontinuing anticonvulsant drugs, findings such as palmar cords and knuckle pad thickening have been reported to regress, although large cohort studies have not found an association with epilepsy or epilepsy medication and the disease. It would seem that epilepsy is not a risk for the disease, but anticonvulsant drugs (e.g., phenytoin and carbamazepine) may trigger it.[20]

Pathophysiology

Several hypotheses have been proposed for the underlying pathophysiologic mechanisms of Dupuytren contracture. Localized ischemia from diabetes, trauma, or other etiologies is thought to produce xanthine oxidase free radicals that may then damage the perivascular connective tissue and bring on a reparative response by surrounding fibroblasts, or triggering myofibroblasts. Fibroblast growth factor (FGF), platelet-derived growth factor (PDGF), and transforming growth factor-beta (TGF-b) may signal an overproduction of myofibroblasts, leading to the formation of nodules and contractures. It has also been demonstrated that, in Dupuytren contracture, the proportions of the different types of collagen are altered, with collagen type 1 replaced by collagen type 3, similar to the proliferative phase of wound healing.

Classification

Characteristics of fibrous tissue deformity and contracture[2]

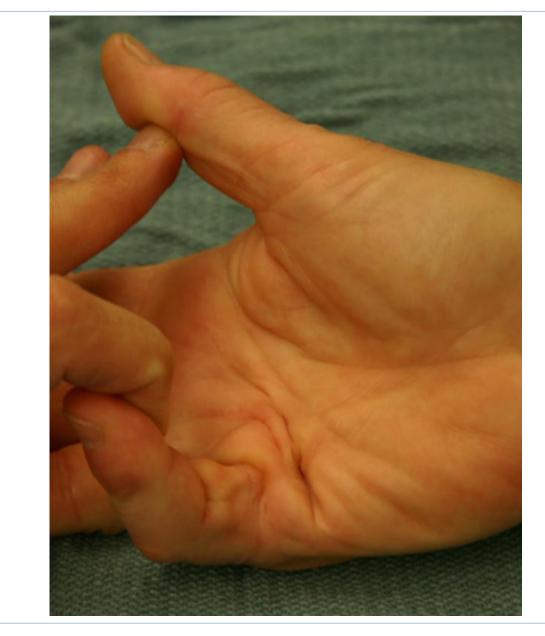
Three grades of Dupuytren contracture have been described, based on the characteristics of the fibrous tissue deformity and the presence of a contracture.

- Grade 1: thickened nodule and a band in the palmar aponeurosis that may progress to skin tethering, puckering, or pitting.
- Grade 2: peritendinous band with limited extension of the affected finger.
- Grade 3: presence of flexion contracture.



Preoperative view of a small finger flexion contracture with surgical indications From the collection of Dr C.M. Rodner; used with permission

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Preoperative view of a small finger flexion contracture with surgical indications From the collection of Dr C.M. Rodner; used with permission

Presence of nodules and degree of contracture[3]

Seven stages of Dupuytren contracture have been described, based on the presence of nodules and the severity of the contracture.

- Stage 0: no contracture
- Stage N: no contracture, palpable nodule
- Stage N/1:0° to 5° contracture, palpable nodule
- Stage 1:6° to 45° contracture
- Stage 2: 46° to 90° contracture
- Stage 3: 91° to 135° contracture
- Stage 4: >135° contracture

Case history

Case history #1

A 60-year-old man of Northern European descent presents with a several-year history of a mass on the ulnar side of his palm. He has no significant past medical history. The patient states this mass has increased in size over the past year and is now causing his small finger to bend down. He has no pain but is increasingly bothered by the flexion deformity of his small finger as it is hard for him to put on a glove, and his finger often gets caught on things. On physical examination there is a cord palpable over the small finger and ring finger rays, with extension across the metacarpophalangeal joint of the small finger producing a contracture of 50 degrees in this joint. There is no proximal interphalangeal joint involvement. Plain x-rays and laboratory studies are normal.

Approach

The diagnosis of Dupuytren contracture is clinical and based on a thorough physical exam of the hands supported by an indicative clinical history.

Clinical history

Dupuytren contracture is typically seen in men of northern European descent aged >40 years. As it is believed to show autosomal dominant inheritance with variable penetrance, there may be a family history of the disease.[1] The medical history may be positive for diabetes mellitus or epilepsy, and the social history may reveal that the patient is a smoker or a heavy drinker.

Patients describe difficulties with face washing, combing their hair, and putting their hands in their pockets or fitting them into gloves.

Physical exam

A detailed physical exam of the affected hand(s) reveals a number of characteristic findings, depending on the disease progression. Bilateral hand involvement is common, with one hand usually more severely affected than the other, although the handedness of the patient is not a predictor of severity.

A firm and thickened palmar nodule over the metacarpal head at the level of the distal palmar crease, proximal to the metacarpophalangeal (MCP) joint, is often the earliest sign. This nodule may be associated with a band in the palmar aponeurosis. After nodule formation, palmar skin changes occur, with skin thickening, tethering, puckering, or pitting, as well as subcutaneous fat fibrosis.

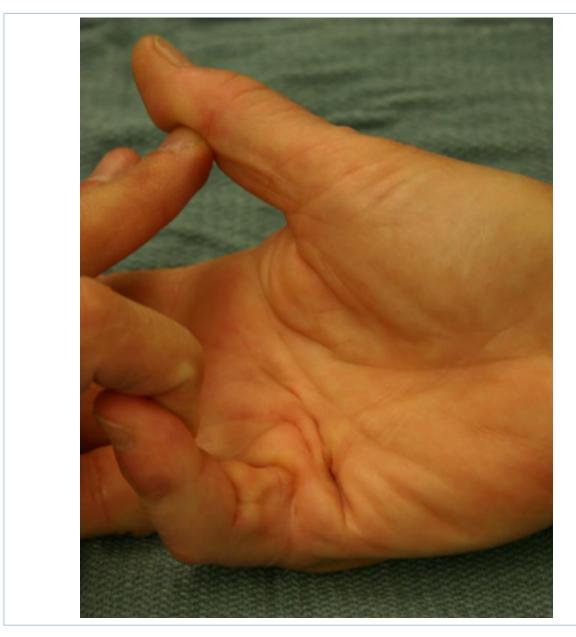
The formation of pretendinous cords usually occurs as isolated nodules coalesce, although both nodules and cords can be present simultaneously. The most commonly affected digit is the ring finger, followed by the small finger, thumb, middle finger, and index finger. As pretendinous cords in the palm progress, they may travel across the MCP joint and, over time, produce MCP joint flexion contractures, leading to limited extension of the affected finger.



Preoperative view of a small finger flexion contracture with surgical indications From the collection of Dr C.M. Rodner; used with permission

DIAGNOSIS

Diagnosis



Preoperative view of a small finger flexion contracture with surgical indications From the collection of Dr C.M. Rodner; used with permission

The degree of contracture is dependent on the severity of the disease. Digital cords that cross the proximal interphalangeal (PIP) joint may cause PIP joint contractures.

Exam of the dorsal aspect of the PIP joints may reveal areas of subcutaneous fibrosis, known as Garrod nodes or knuckle pads, which are indicative of systemic fascial disease and predictive of bilateral involvement. Garrod nodes are found in about one half of patients.

The term Dupuytren diathesis refers to patients with severe disease. These patients are usually younger, with very rapid disease progression involving both hands, and are more likely to have systemic fascial disease including Ledderhose disease affecting the plantar surface of the feet, and, in men, Peyronie disease, which affects the penis.[19]

The Hueston table-top test aids diagnosis and involves the patient attempting to lay the palm of the hand flat on a table surface. The test is positive if the patient is unable to flatten the hand on the table.

Investigations

As the diagnosis of Dupuytren contracture is predominantly clinical, ultrasound of the hand has limited usefulness in the diagnosis of the disease. It shows a mass lying between the flexor tendon below and the skin above. MRI and radiographs are not indicated in the diagnosis of the disease.

History and exam

Key diagnostic factors

male >40 years of age (common)

Dupuytren contracture is predominantly seen in men of northern European descent aged >40 years.[6]
 [7] [8] [9]

difficulties with manual activities (common)

• Patients describe difficulties with face washing, combing their hair, and putting their hands in their pockets or fitting them into gloves.[19]

palmar nodule (common)

- A firm and thickened palmar nodule over the metacarpal head at the level of the distal palmar crease, proximal to the metacarpophalangeal (MCP) joint, is often the earliest sign.
- Nodule may be associated with a band in the palmar aponeurosis.

palmar skin changes (common)

• After nodule formation, skin changes occur with skin thickening, tethering, puckering, or pitting, as well as subcutaneous fat fibrosis.

pretendinous cords (common)

- The formation of pretendinous cords usually occurs as isolated nodules coalesce, although both nodules and cords can be present simultaneously.
- The most commonly affected digit is the ring finger, followed by the small finger, thumb, middle finger, and index finger.

MCP joint contracture (common)

• As pretendinous cords in the palm progress, they may travel across the MCP joint and, over time, produce MCP joint flexion contractures leading to limited extension of the affected finger.



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Preoperative view of a small finger flexion contracture with surgical indications From the collection of Dr C.M. Rodner; used with permission

• The degree of contracture is dependent on the severity of the disease.

proximal interphalangeal (PIP) joint contracture (common)

• Digital cords that cross the PIP joint may cause PIP joint contractures.

positive Hueston table-top test (common)

- Involves the patient attempting to lay the palm of the hand flat on a table surface.
- Result is positive if the patient is unable to flatten the hand on the table.

Other diagnostic factors

bilateral involvement (common)

- Bilateral hand involvement is common, with one hand usually more severely affected than the other, although the handedness of the patient is not a predictor of severity.
- Patients with Dupuytren diathesis have rapid disease progression involving both hands.

Garrod nodes (uncommon)

- Examination of the dorsal aspect of the PIP joints may reveal areas of subcutaneous fibrosis, known as Garrod nodes, or knuckle pads, which are indicative of systemic fascial disease and predictive of bilateral involvement.
- · Garrod nodes are found in about one half of patients.

involvement of plantar surface of the feet (uncommon)

• Patients with Dupuytren diathesis are more likely to have systemic fascial disease, including Ledderhose disease, which affects the plantar surface of the feet.

involvement of penis (uncommon)

• Patients with Dupuytren diathesis are more likely to have systemic fascial disease, including Peyronie disease in men, which affects the penis.

Risk factors

Strong

male sex

• Dupuytren contracture is twice as common in men as in women.[8] [9] It predominantly affects men of northern European descent aged >40 years.

age >40 years

• The incidence of Dupuytren contracture increases with age and it predominantly affects men of northern European descent aged >40 years.[6] [7] [8] [9]

family history

- Dupuytren contracture is believed to show autosomal dominant inheritance with variable penetrance. [1]
- The familial occurrence and its presence in identical twins suggest a genetic basis for the disease. The sibling recurrence risk ratio equals 2.9 (95% CI 2.6 to 3.3).
- Although the genetic factors involved have not been fully elucidated, in one family the gene has been mapped to chromosome 16q.[1]
- DNA microarray has demonstrated that >30 unique genes are upregulated and 6 unique genes downregulated by a factor of 4 or greater in affected patients.[11]
- A novel gene, MafB (musculoaponeurotic fibrosarcoma oncogene homolog B), is overexpressed in patients with Dupuytren contracture but not in those with normal fascia.[11]

Diagnosis

diabetes mellitus

- Diabetes mellitus has a strong association with Dupuytren contracture.[4] [10] [14] [15] [16] The prevalence of Dupuytren contracture in patients with diabetes ranges from 3% to 33% and increases with the duration of the diabetes.
- It is hypothesized that diabetes causes microvascular changes that produce local hypoxic tissue damage, inducing Dupuytren contracture. Patients with diabetes tend to have a mild form of the disease, with slow progression.

Weak

high alcohol intake

Greater alcohol intake is associated with an increased likelihood of Dupuytren contracture.[4] [7] [10]
 [17] However, it should be noted that most patients with the disease do not misuse alcohol, and not all studies have found a significant association.[18]

smoking

• For unclear reasons, smoking increases the risk of Dupuytren contracture, although the microvascular changes associated with smoking and the effect of carbon monoxide on mitochondrial cytochrome c, released by mitochondria in response to pro-apoptotic stimuli, may play a role.[17]

trauma

- Trauma was first proposed to be a cause by Dupuytren himself. Since the first report that linked trauma
 to the disease, the strength of this association has been controversial. It is unclear why the disease
 would be caused by heavy labor or vibrational exposure, and further studies are required to control for
 smoking and alcohol use.
- One theoretical explanation for the connection of trauma and Dupuytren contracture is the induction of transforming growth factor-beta (TGF-b) with trauma and the stimulation of myofibroblasts from the increased TGF-b.[21]

anticonvulsant medication

• There have been reports of a connection between epilepsy and Dupuytren contracture. When discontinuing anticonvulsant drugs, findings such as palmar cords and knuckle pad thickening have been reported to regress, although large cohort studies have not found an association with epilepsy or epilepsy medication and the disease. It would seem that epilepsy is not a risk for the disease, but anticonvulsant drugs (phenytoin and carbamazepine) may trigger it.[20]

Investigations

Other tests to consider

Test	Result
 ultrasound of hand As the diagnosis is predominantly clinical, ultrasound of the hand has limited usefulness. 	mass lying between the flexor tendon below and the skin above

Differentials

Condition	Differentiating signs / symptoms	Differentiating tests
Trigger finger	The finger can be fully extended with a notable click.	Ultrasound: thickening and hypoechogenicity, with possible increased vascularity of the A1 pulley.
Epithelioid sarcoma	Tends to be progressive and extend beyond localized digits.	• Biopsy: results can be difficult to determine but are often seen as well-defined eosinophilic cytoplasms and one or more atypical, eccentrically located nuclei, resulting in a plasmacytoid appearance.
Camptodactyly	 Presence of little finger contracture from an early age. 	No differentiating tests.
Traumatic finger contracture	 History of significant preceding trauma leading to injury of the proximal interphalangeal joint. 	X-ray of hand may show associated fracture.

Approach

Patients with early Dupuytren contracture can be managed expectantly, although the injection of nodules with a corticosteroid may be carried out if the Dupuytren lesions are bothersome. Those with metacarpophalangeal (MCP) joint contractures of 30 degrees or less and with no proximal interphalangeal (PIP) joint contractures can be treated with needle aponeurotomy, percutaneous fasciotomy, or corticosteroid injections. When function is impaired or there is a severe and disabling deformity present, surgery is recommended, and patients with MCP joint contractures >30 degrees or PIP joint contractures can be treated with either open partial fasciectomy, segmental aponeurotomy, or percutaneous fasciotomy.[19] [22] Partial fasciectomy has become the favored technique of hand surgeons in the surgical treatment of Dupuytren contracture due to the relatively low rate of recurrence seen with this procedure.[23] It is difficult, however, to compare the efficacy of the many different modalities used to treat Dupuytren contracture. A reason for this is the inconsistency in reporting of outcomes across studies evaluating treatments for Dupuytren contracture.[24] [25] Only 3 Level I studies have compared the different surgical techniques for the treatment of primary Dupuytren contracture, and current evidence does not support one procedure as being better than the other, apart from a particularly high recurrence rate after needle fasciotomy. More research will be needed in the future.[26]

All patients with contractures should receive hand therapy post-procedure, and postoperative splinting following open partial fasciectomy is used at the discretion of the surgeon. The effects of stretching on prevention of contracture development for up to 7 months have shown no added benefit, and no studies have been done for more than 7 months. There is therefore no current evidence to support the use of stretching to prevent contractures.[27]

Corticosteroid injections

Patients with early Dupuytren contracture who have evidence of the disease but have not yet developed contractures can be treated with corticosteroid injections if they are experiencing bothersome symptoms. Those with MCP joint contractures of 30 degrees or less with no PIP joint contractures who wish to avoid a more invasive procedure may also benefit from corticosteroid injections.

The injection of Dupuytren nodules with triamcinolone acetonide monthly for up to 5 months, or every 6 weeks for 3 injections, has been shown to produce significant regression of the disease, with an average of 3.2 injections per nodule required for improvement of function.[28] After corticosteroid injection, fewer patients progress to surgery than would be predicted with expectant management alone.[28]

Collagenase injection

Collagenase clostridium histolyticum is used to treat adult patients with a palpable cord along with MCP or PIP contracture, with a corresponding decrease in both fasciotomies and fasciectomies.[29] The efficacy and tolerability of injectable mixed collagenase subtypes as an alternative to surgical intervention in Dupuytren contracture has been examined in a phase 3, double-blind study, which found that collagenase safely and effectively restored normal finger extension in the majority (87%) of patients.[30] A mean of 1.4 injections was required to normalize affected joints, and clinical success was achieved within 29 days. Contracture recurrence was relatively low, occurring in just 5 joints (1 MCP, 4 PIP) between 6 and 24 months after treatment, and the recurrence severity ranged from 20 degrees to 40 degrees. Adverse events were localized to the injection site, generally of mild-to-moderate severity, and transient in nature. In another study, more cords that were injected with collagenase met criteria of a reduction in contracture to 0 to 5 degrees of full extension 30 days after the last injection (64.0% vs. 6.8%, P <0.001) than cords

injected with placebo.[31] The most commonly reported adverse events were localized swelling in the hand, pain, bruising, pruritus, and transient regional lymph-node enlargement and tenderness. Serious adverse events were seen in 2% of collagenase recipients, including tendon ruptures and complex regional pain syndrome.[31] Although there is some literature with small numbers of patients to suggest that recurrence is more common in PIP joint contractures than in MCP joint contractures, some guidelines recommend that collagenase treatment is limited only to clinical trials.[32][33]

One systematic review has found that having previous surgery did not affect the efficacy and safety of collagenase injections, making this an option in patients with recurrent Dupuytren contracture.[34]

The technique for collagenase injection is relatively straightforward. First, the skin is prepared with antiseptic solution. Without the use of any local anesthesia, an insulin needle (28-gauge) is inserted to a depth of 5 mm for MCP joint lesions and 3 mm for PIP joint lesions. The gristly structure of the cord is easily palpated with the end of the needle. A gentle amount of passive motion ensures the needle is not in the flexor tendon before the injection of collagenase into the MCP or PIP lesion. The injections are administered proximally-to-distally, repositioning the needle prior to injection of the next dose. After the injection is given, a dressing is placed on the hand. The patient is then instructed to return to the office the next day for the finger extension maneuver, in which the finger is extended to break the cord. This may be done with or without a local anesthetic. After the manipulation is successfully completed, there are no restrictions and early movement is encouraged. Although wearing a nighttime splint afterward has not been proven to decrease recurrence, it should be offered to the patient. The patient is followed to assess the need for up to 2 further injections every 4 to 6 weeks.[30]

Needle aponeurotomy

Needle aponeurotomy, also called percutaneous needle fasciotomy (PNF), is a minimally invasive technique that can be undertaken in the office. While needle aponeurotomy is typically used for early Dupuytren contracture, it has also been described for more advanced stages of the disease and can be used in those with MCP joint contractures of 30 degrees or less with no PIP joint contractures.

Needle aponeurotomy is usually successful in correcting the Dupuytren contracture, takes very little time to perform (usually 20 to 30 minutes), requires only local anesthesia, and is not very painful. In comparison with open surgical procedures, it results in similar resolution,[35] [36] minimal scarring, faster recovery, and can be repeated easily if the contracture recurs.[37] Needle aponeurotomy is thus an attractive option for patients with less aggressive and early disease.[38] One systematic review reported a tendency to greater patient satisfaction with needle aponeurotomy, with fewer adverse effects, compared with other procedures.[39] Recurrence rates of up to 58% have been reported during 3 to 5 years of follow-up, but long-term outcomes are not well reported.[40]

Randomized studies report no significant difference in treatment outcome between needle aponeurotomy and collagenase injection.[41] [42] In one trial, however, collagenase treatment led to more, mainly transient, complications than needle aponeurotomy.[42]

Percutaneous fasciotomy

Percutaneous fasciotomy is a similar procedure to needle aponeurotomy, but uses a scalpel to cut and release the band causing the digital contracture instead of a needle to weaken it.[43] [44] It is thus performed by a hand surgeon in the operating room. This technique is normally used in patients with MCP joint contractures of 30 degrees or less with no PIP joint contractures, but can also be used in more advanced stages of Dupuytren contracture.[22]

Like needle aponeurotomy, the finger is brought into full extension with a characteristic snap, and the goal of percutaneous fasciotomy is to promote greater extension and function of the affected digit.[22]

As with needle aponeurotomy, the major advantages of percutaneous fasciotomy are that it causes less pain and allows a faster recovery than traditional open fasciectomy interventions. However, like needle aponeurotomy, this procedure is associated with a risk of recurrence of up to 43%, as diseased fascia is unavoidably left behind. There is concomitant risk of flexor tendon or nerve injury.[43][44][45]

Segmental aponeurotomy

Segmental aponeurotomy is a compromise between percutaneous techniques and open fasciectomy, in which multiple small incisions are made in the palm and the digits to remove segments of the Dupuytren cord and achieve discontinuity between the segments of diseased tissue, with no effort made to remove all of the pathologic tissue. It is typically used in patients with MCP joint contractures >30 degrees, or in those with PIP joint contractures. The clinical results of this technique compare quite well with traditional open fasciectomy techniques, with a recurrence rate ranging from 20% to 35%.[46][47] [48]

Open partial fasciectomy

Although the timing of surgical intervention varies, the decision to operate is generally taken if the MCP joint contracture exceeds 30 degrees or a PIP joint contracture develops.[49] The presence of any PIP joint contracture alone is a relative indication for surgery.[50] Open partial fasciectomy is a successful treatment and is associated with a postoperative recurrence rate of 15%.[49] The risk of postoperative recurrence increases with the severity of the contracture, and the longer a deformity is present, the greater the chance of the joint contracture becoming irreversible.[49] [50]

Partial fasciectomy, also known as subtotal, limited, or regional fasciectomy, was first described by Goyrand in 1834 and involves opening up the hand and, under direct visualization, excising the diseased Dupuytren tissue. Excising only the pathologic cord(s) with a partial fasciectomy is now more commonly employed than total fasciectomy or dermatofasciectomy. Postoperative infection is countered with the use of perioperative antibiotics and careful soft tissue handling.

An open partial fasciectomy remains the most common procedure used in the surgical management of Dupuytren contracture and is the preferred surgical procedure for advanced disease in patients with functional impairment who are surgical candidates. Using this technique, a small amount of potentially diseased fascia could theoretically be left behind, as a radical fasciectomy of the entire palmar fascia is not typically performed. Through a transverse palmar incision, usually overlying the distal palmar crease, the fascia that has formed pathologic cords is excised in a proximal-to-distal direction.



Preoperative view of the ring finger of a patient with a flexion contracture with surgical indications, showing the incision marking, demonstrating a transverse incision overlying the distal palmar crease, and oblique Brunner incisions coursing from it proximally and distally From the collection of Dr C.M. Rodner; used with permission

The choice of digital incision is surgeon-dependent and is most often between a Brunner incision (angled "zigzag" incisions made from the ulnar aspect of the MCP joint crease to the radial aspect of the PIP joint or vice versa) and a longitudinal incision (often combined with multiple Z-plasties). Brunner or longitudinal incisions are carried out along the middle of each affected digit, and skin flaps are carefully raised from the pathologic cord to avoid skin necrosis. Brunner "zigzag" incisions are made methodically in a proximal-to-distal direction from the transverse palmar incision following the path of the palpable Dupuytren cord.

In the palm, the neurovascular structures that lie deep to the involved fascia at this level are identified and, with particular attention paid to retracting these neurovascular bundles, the diseased fascia is excised and elevated in a proximal-to-distal direction.

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Intraoperative view of the ring finger of a patient with a flexion contracture, with the radial digital neurovascular bundle identified and isolated coursing volar over the Dupuytren cord, which is being held up by forceps as it is excised in a proximal-to-distal direction From the collection of Dr C.M. Rodner; used with permission

As the Dupuytren cord moves into the finger, great care is taken to identify the radial and ulnar digital arteries and nerves as they bifurcate from the parent common artery and nerve. As the proper digital neurovascular bundles may be displaced toward the midline in PIP joint contractures, careful dissection is required to identify them in such cases. The fascia must only be cut if it is directly visualized and the artery and nerve are protected. If a spiral cord is present, which is often the case with PIP joint contractures, dissection proceeds from both a proximal-to-distal and a distal-to-proximal direction so as to safely free the neurovascular bundle from the cord that is displacing it centrally.[51] When the small finger is involved, the insertion of the abductor digiti quinti muscle should be identified and excised in order to fully release the digit.

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Currently, most hand surgeons endeavor to dissect the normal skin away from the underlying diseased tissue.[46] If the palmar skin is adherent to the cord so that it cannot be saved and the defect is small, primary closure is often performed. Such defects may also occur as the result of straightening out a previously contracted digit. As the finger is extended, primary skin coverage in the palm may prove to be difficult. If the palmar defect is too large for primary closure, skin grafting or the McCash "open-palm" technique have both shown good clinical results.[52] When possible, direct primary closure of the palmar skin is performed over a Penrose drain to prevent hematoma, as this method of closure allows for early motion and good skin sensibility, avoiding the meticulous wound care required with an open wound.



Postoperative view of the ring finger of a patient with a flexion contracture, showing the closed wound over a Penrose drain, which is used to minimize subsequent hematoma formation From the collection of Dr C.M. Rodner; used with permission

The McCash "open-palm" technique, used by many surgeons, is an alternative to direct palmar wound closure, associated with a greater active range of motion without any increased risk of infection, although this technique has the disadvantage of leaving the wound open for about 1 month.[52]

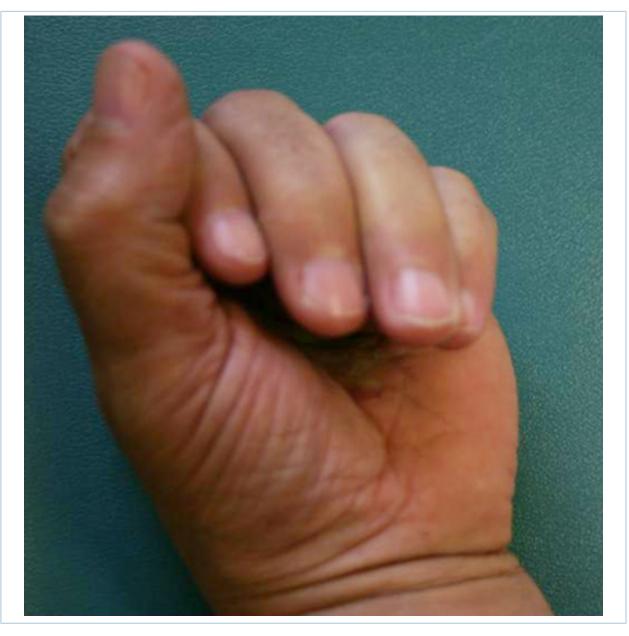
The fingers are then splinted in full extension, and the patient is followed up within a few days to pull out the drain and assess the wound. By the fifth postoperative day, patients are sent to the hand therapist for a forearm-based digital extension splint that is worn full-time between therapy visits. Flexion exercises begin after the wound has stabilized. Regaining digital flexion often proves more difficult than maintaining extension after fasciectomy, due to the postoperative extension splinting required.

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Management



One-month postoperative view of the ring finger of a patient with a flexion contracture, demonstrating full active digital extension From the collection of Dr C.M. Rodner; used with permission



One-month postoperative view of the ring finger of a patient with a flexion contracture, demonstrating active digital flexion From the collection of Dr C.M. Rodner; used with permission

By the third postoperative week, the splint is weaned, to be worn at night only, and nighttime extension splinting can continue for as long as 6 months. Some surgeons have abandoned postoperative splinting, favoring earlier mobilization in order to minimize difficulties with flexion. There is also evidence to suggest that splinting (including nighttime extension splinting) after surgery provides no additional benefit to standard hand therapy in maintaining finger extension, except perhaps for cases in which extension loss occurs postoperatively, whereby nighttime extension splinting may provide some benefit.[53] [54] Postoperative splinting may not be justified in all patients.

Other surgical techniques

Total fasciectomy and dermatofasciectomy are now seldom used but are described here for completeness.

The technique of total (or radical) fasciectomy via a transverse palmar incision and multiple digital Zplasties in which all of the palmar fascia is excised has not been found to have any clinical advantage over a more limited fasciectomy, and is associated with a higher complication rate.[55] [56] Although total fasciectomy should theoretically reduce recurrence rates by excising the entire palmar fascia responsible for Dupuytren contracture, this has not been supported by the literature.

Dermatofasciectomy, involving a simultaneous excision of the skin, which is subsequently replaced with full-thickness skin grafts, has produced fairly good results, although morbidity associated with skin grafting such as poor graft sensibility have made this technique less popular than partial fasciectomy.[57] [58][59][60]

Radiation therapy

Although its use in Dupuytren contracture is extremely limited, radiation therapy is described here for completeness, as it has been reported to be successful in the treatment of the disease. One study showed that the majority (77%) of lesions did not progress following a total radiation therapy dose of 30 Gy. Long-term studies have not found differences in disease progression 7 years after radiation therapy, compared with controls.[61]

Treatment algorithm overview

Please note that formulations/routes and doses may differ between drug names and brands, drug formularies, or locations. Treatment recommendations are specific to patient groups: <u>see disclaimer</u>

Ongoing		(summary)
no MCP joint or PIP joint contracture		
	1st	expectant management
	adjunct	corticosteroid injections
≤30 degrees MCP joint contracture with no PIP joint contracture		
	1st	collagenase injection
	1st	needle aponeurotomy
	1st	percutaneous fasciotomy
	1st	corticosteroid injections
>30 degrees MCP joint contracture and/or PIP joint contracture		
	1st	open partial fasciectomy + perioperative antibiotics
	adjunct	postoperative splinting
	1st	segmental aponeurotomy
	1st	percutaneous fasciotomy
	1st	collagenase injection

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Treatment algorithm

Please note that formulations/routes and doses may differ between drug names and brands, drug formularies, or locations. Treatment recommendations are specific to patient groups: <u>see disclaimer</u>

Ongoing

no MCP joint or PIP joint contracture		
	1st	expectant management
		» Patients with early Dupuytren contracture who have evidence of the disease but have not yet developed contractures may be managed expectantly with regular follow-up every 6 months to evaluate for disease progression. Use of the Hueston table-top test and appropriate staging of Dupuytren contracture are essential during follow-up sessions.[49] [50]
	adjunct	corticosteroid injections
		Treatment recommended for SOME patients in selected patient group
		Primary options
		» triamcinolone acetonide: consult specialist for guidance on dose
		» Patients with early Dupuytren contracture who have evidence of the disease but have not yet developed contractures can be treated with corticosteroid injections if they are experiencing bothersome symptoms.
		 The injection of Dupuytren nodules with triamcinolone acetonide monthly for up to 5 months or every 6 weeks for 3 injections has been shown to produce significant regression of the disease, with an average of 3.2 injections per nodule required for improvement of function.[28] After corticosteroid injection, fewer patients progress to surgery than would be predicted with expectant management alone.[28]
≤30 degrees MCP joint contracture with no PIP joint contracture		
	1st	collagenase injection
		Primary options

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» collagenase clostridium histolyticum: 0.58 mg intralesionally as a single dose, maximum

May repeat 2 doses at intervals of 4 weeks if

» Collagenase clostridium histolyticum is used to treat adult patients with a palpable cord

3 doses/cord

the contracture remains.

Ongoing

along with metacarpophalangeal (MCP) joint contracture or proximal interphalangeal (PIP) joint contracture, with a corresponding decrease in both fasciotomies and fasciectomies.[29] A mean of 1.4 injections is required to normalize affected joints, and clinical success is achieved within 29 days.[30] In one study, reductions in contracture to 0 to 5 degrees of full extension, 30 days after the last injection, were achieved in cords injected with collagenase as compared with those injected with placebo.[31] Adverse events are localized to the injection site, generally of mild-to-moderate severity, and transient in nature.[30] The most commonly reported adverse events were localized swelling in the hand, pain, bruising, pruritus, and transient regional lymph-node enlargement and tenderness. Serious adverse events were seen in 2% of collagenase recipients, including tendon ruptures and complex regional pain syndrome.[31] Although there is some literature with small numbers of patients to suggest that recurrence is more common in PIP joint contractures than in MCP joint contractures, additional long-term studies involving larger numbers of patients are warranted to verify the long-term effectiveness and recurrence rates of collagenase treatment, as well as its long-term safety.[32]

» One systematic review has found that having previous surgery did not affect the efficacy and safety of collagenase injections, making this an option in patients with recurrent Dupuytren contracture.[34]

» After the injection is given, a dressing is placed on the hand. The patient is then instructed to return to the office the next day for the finger extension maneuver, in which the finger is extended to break the cord. This may be done with or without a local anesthetic. After the manipulation is successfully completed, there are no restrictions and early movement is encouraged. Although wearing a nighttime splint afterward has not been proven to decrease recurrence, it should be offered to the patient. The patient is followed to assess the need for up to 2 further injections every 4 to 6 weeks.[30]

1st needle aponeurotomy

» Under sterile conditions, the area is anesthetized with lidocaine and an 18-gauge needle is used to puncture the aponeurotic band causing the digital contracture. This weakens the contracture until it can be broken by mechanical force, typically with a characteristic

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snap. The needle is introduced volar (palmar side) to the tendon at various sites progressing from proximal to distal. Care must be taken not to insert the needle into the tendon, to avoid iatrogenic injury. Use of ultrasound to guide the procedure may reduce the risk of accidental lesions.[62]

» This procedure is usually successful in correcting the contracture, takes very little time to perform (20-30 minutes), requires only local anesthesia, and is not very painful. In comparison with open surgical procedures, it results in similar resolution, minimal scarring, faster recovery, and can be repeated easily if the contracture recurs.[35] [36] [37] Needle aponeurotomy is thus an attractive option for patients with less aggressive and early disease.[38] Recurrence rates of up to 58% have been reported during 3 to 5 years of follow-up, but long-term outcomes are not well reported.[40]

» All patients with contractures should receive hand therapy postprocedure.

1st percutaneous fasciotomy

» A similar procedure to needle aponeurotomy, but uses a scalpel to cut and release the band causing the digital contracture and is thus performed by a hand surgeon in the operating room.[43][44] The finger is brought into full extension with a characteristic snap.

» This procedure causes less pain and allows a faster recovery than traditional open fasciectomy interventions. However, it is associated with a risk of recurrence of up to 43%, as diseased fascia is unavoidably left behind, and there is risk of flexor tendon or nerve injury.[43][44] [45]

» All patients with contractures should receive hand therapy postprocedure.

1st corticosteroid injections

Primary options

» triamcinolone acetonide: consult specialist for guidance on dose

» Patients who wish to avoid a more invasive procedure may benefit from corticosteroid injections.

» The injection of Dupuytren nodules with triamcinolone acetonide monthly for up to 5 months or every 6 weeks for 3 injections has been shown to produce significant regression of

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Ongoing

the disease, with an average of 3.2 injections per nodule required for improvement of function.[28] After corticosteroid injection, fewer patients progress to surgery than would be predicted with expectant management alone.[28]

» All patients with contractures should receive hand therapy postprocedure.

>30 degrees MCP joint contracture and/or PIP joint contracture

1st

t open partial fasciectomy + perioperative antibiotics

Primary options

» cefazolin: 1g intravenously administered 30 minutes to 1 hour prior to the start of surgery

Secondary options

» clindamycin: 600 mg intravenously administered 30 minutes to 1 hour prior to the start of surgery

» The most common procedure used in the surgical management of Dupuytren contracture, as it is associated with a postoperative recurrence rate of 15%.[49]

» Through a transverse palmar incision overlying the distal palmar crease, the fascia forming pathologic cords is excised in a proximal-to-distal direction.

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Ongoing



Preoperative view of the ring finger of a patient with a flexion contracture with surgical indications, showing the incision marking, demonstrating a transverse incision overlying the distal palmar crease, and oblique Brunner incisions coursing from it proximally and distally From the collection of Dr C.M. Rodner; used with permission

The choice of digital incision is surgeondependent and is most often between a Brunner and a longitudinal incision (often combined with multiple Z-plasties).

» In the palm, the neurovascular structures deep to the involved fascia are identified and retracted.

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Intraoperative view of the ring finger of a patient with a flexion contracture, with the radial digital neurovascular bundle identified and isolated coursing volar over the Dupuytren cord, which is being held up by forceps as it is excised in a proximal-to-distal direction From the collection of Dr C.M. Rodner; used with permission

The diseased fascia is excised and elevated in a proximal-to-distal direction. As the Dupuytren cord moves into the finger, care is taken to identify the radial and ulnar digital arteries and nerves, particularly in PIP joint contractures. The joints are inspected for persistent contractures, and residual contracture of the PIP joint is addressed with a release of the volar plate followed by release of the collateral ligaments if necessary.

» When possible, direct primary closure of the palmar skin is performed over a Penrose drain to prevent hematoma, as this method of closure allows for early motion and good skin sensibility, avoiding the meticulous wound care required with an open wound.

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Ongoing



Postoperative view of the ring finger of a patient with a flexion contracture, showing the closed wound over a Penrose drain, which is used to minimize subsequent hematoma formation From the collection of Dr C.M. Rodner; used with permission

If the palmar defect is too large for primary closure, skin grafting or the McCash "open-palm" technique are used. The patient is followed up within a few days to pull out the drain and assess the wound.

» All patients with contractures should receive hand therapy postprocedure. After an open partial fasciectomy, flexion exercises begin once the wound has stabilized.

» Postoperative infection is countered with the use of perioperative antibiotics and careful soft tissue handling. Cefazolin is the agent of choice. Clindamycin can be used as an alternative in patients with penicillin allergy, to provide grampositive cover.

adjunct postoperative splinting

Treatment recommended for SOME patients in selected patient group

» After an open partial fasciectomy, the fingers can be splinted in full extension.

» By the fifth postoperative day, patients are sent to the hand therapist for a forearm-based

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Ongoing

digital extension splint that is worn full time between therapy visits. Flexion exercises begin once the wound has stabilized. Regaining digital flexion often proves more difficult than maintaining extension after fasciectomy, due to the postoperative extension splinting required.



One-month postoperative view of the ring finger of a patient with a flexion contracture, demonstrating full active digital extension From the collection of Dr C.M. Rodner; used with permission

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Ongoing



One-month postoperative view of the ring finger of a patient with a flexion contracture, demonstrating active digital flexion From the collection of Dr C.M. Rodner; used with permission

By the third postoperative week the splint is weaned, to be worn at night only, and nighttime extension splinting can continue for as long as 6 months.

» Some surgeons have abandoned postoperative splinting, favoring earlier mobilization in order to minimize difficulties with flexion. There is also evidence to suggest that splinting (including nighttime extension splinting) after surgery provides no additional benefit to standard hand therapy in maintaining finger extension, except perhaps for cases in which extension loss occurs postoperatively, whereby nighttime extension splinting may provide some benefit.[53] [54] Postoperative splinting may not be justified in all patients.

1st segmental aponeurotomy

» A compromise between percutaneous techniques and open fasciectomy, in which multiple small incisions are made in the palm and the digits to remove segments of the Dupuytren cord and achieve discontinuity between the segments of diseased tissue, with no effort made to remove all of the pathologic tissue.

» The clinical results of this technique compare quite well with traditional open fasciectomy techniques, with a recurrence rate ranging from 20% to 35%.[46][47] [48]

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Ongoing

» All patients with contractures should receive hand therapy postprocedure.

1st percutaneous fasciotomy

» A similar procedure to needle aponeurotomy, but uses a scalpel to cut and release the band causing the digital contracture and is thus performed by a hand surgeon in the operating room.[43] [44] The finger is brought into full extension with a characteristic snap.

» This procedure causes less pain and allows a faster recovery than traditional open fasciectomy interventions. However, it is associated with a risk of recurrence of up to 43%, as diseased fascia is unavoidably left behind, and there is a risk of flexor tendon or nerve injury.[43][44][45]

» All patients with contractures should receive hand therapy postprocedure.

collagenase injection

1st

Primary options

» collagenase clostridium histolyticum: 0.58
 mg intralesionally as a single dose, maximum
 3 doses/cord

May repeat 2 doses at intervals of 4 weeks if the contracture remains.

» Collagenase clostridium histolyticum is used to treat adult patients with a palpable cord along with metacarpophalangeal (MCP) joint contracture or proximal interphalangeal (PIP) joint contracture, with a corresponding decrease in both fasciotomies and fasciectomies.[29] A mean of 1.4 injections is required to normalize affected joints, and clinical success is achieved within 29 days.[30] In one study, reductions in contracture to 0 to 5 degrees of full extension, 30 days after the last injection, were achieved in cords injected with collagenase as compared with those injected with placebo.[31] Adverse events are localized to the injection site. generally of mild-to-moderate severity, and transient in nature.[30] The most commonly reported adverse events were localized swelling in the hand, pain, bruising, pruritus, and transient regional lymph-node enlargement and tenderness. Serious adverse events were seen in 2% of collagenase recipients, including tendon ruptures and complex regional pain syndrome.[31] Although there is some literature with small numbers of patients to suggest that recurrence is more common in PIP joint contractures than in MCP joint contractures, additional long-term studies involving larger

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Ongoing

numbers of patients are warranted to verify the long-term effectiveness and recurrence rates of collagenase treatment, as well as its long-term safety.[32]

» One systematic review has found that having previous surgery did not affect the efficacy and safety of collagenase injections, making this an option in patients with recurrent Dupuytren contracture.[34]

» After the injection is given, a dressing is placed on the hand. The patient is then instructed to return to the office the next day for the finger extension maneuver, in which the finger is extended to break the cord. This may be done with or without a local anesthetic. After the manipulation is successfully completed, there are no restrictions and early movement is encouraged. Although wearing a nighttime splint afterward has not been proven to decrease recurrence, it should be offered to the patient. The patient is followed to assess the need for up to 2 further injections every 4 to 6 weeks.[30]

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Emerging

Tumor necrosis factor (TNF)-alpha inhibitors

There is emerging evidence supporting the use of TNF-alpha inhibitors (e.g., adalimumab) for early-stage Dupuytren disease. One randomized, double-blind, placebo-controlled trial on a UK cohort found that the use of intranodular adalimumab injections were effective in softening and reducing the size of nodules.[63]

Primary prevention

No primary prevention studies exist and it remains to be seen if management of diabetes, or reduction in smoking and alcohol, will reduce the prevalence of the disease. As occupational trauma is thought to be associated with Dupuytren disease, proper protection of the hand from trauma at work is indicated.

Patient discussions

Minimization of the risk factors for Dupuytren contracture may help slow the progression of the disease. Patients should therefore be instructed to stop smoking, reduce their alcohol intake, and use proper protection of the hand from trauma at work if appropriate. Adequate control of diabetes is also important.

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Monitoring

Monitoring

Follow-up every 6 months is useful to evaluate for disease progression. Use of the Hueston table-top test and appropriate staging of Dupuytren contracture are essential during follow-up sessions.[49] [50] The risk of postoperative recurrence increases with the severity of the contracture. The longer a deformity is present, the greater the chance of the joint contracture becoming irreversible.

Complications

Complications	Timeframe	Likelihood
intraoperative neurovascular injury	short term	low
Neurovascular injury can be minimized by meticulous intraopera arteries, and by only cutting tissue that is well visualized after the digit have been identified and retracted.		
Digital neurapraxia is sometimes unavoidable when a previously Occasionally, digital ischemia may occur after extension of a pre stretch and vasospasm. If this occurs intraoperatively, the finger with warm saline. If this fails to restore blood flow, the local appli and if this is unsuccessful, intravenous heparin may be administ	eviously contracted dia should be passively t cation of lidocaine sh	git, due to arterial flexed and treated
postoperative hematoma	short term	low
The risk of hematoma formation is minimized by tourniquet defla achieving adequate hemostasis prior to wound closure, and/or b		
postoperative infection	short term	low
Complications associated with wound healing can be minimized	by careful flap planni	ing, meticulous
Infection is countered with the use of perioperative antibiotics an Complications associated with wound healing can be minimized elevation of flaps, and hematoma prevention. Reported incidenc 0.04% at 90 days.[66]	by careful flap planni e of postoperative su	ing, meticulous rgical site infection i
Complications associated with wound healing can be minimized elevation of flaps, and hematoma prevention. Reported incidenc	by careful flap planni e of postoperative su variable	ing, meticulous rgical site infection i high
Complications associated with wound healing can be minimized elevation of flaps, and hematoma prevention. Reported incidenc 0.04% at 90 days.[66] postoperative stiffness Prophylaxis is difficult due to the need for a prolonged period of	by careful flap planni e of postoperative su variable postoperative immob	ing, meticulous rgical site infection i high ilization after
Complications associated with wound healing can be minimized elevation of flaps, and hematoma prevention. Reported incidenc 0.04% at 90 days.[66] postoperative stiffness Prophylaxis is difficult due to the need for a prolonged period of Dupuytren surgery.	by careful flap planni e of postoperative su variable postoperative immob	ing, meticulous rgical site infection i high ilization after
Complications associated with wound healing can be minimized elevation of flaps, and hematoma prevention. Reported incidenc 0.04% at 90 days.[66] postoperative stiffness Prophylaxis is difficult due to the need for a prolonged period of Dupuytren surgery. Supervised hand therapy exercises should be instituted as soon	by careful flap planni e of postoperative su variable postoperative immob as the state of the w variable ue recurrence (diseas rea).	ing, meticulous rgical site infection i high ilization after ound allows. medium e at the operative

postoperative reflex sympathetic dystrophy variable low	Complications	Timeframe	Likelihood
	postoperative reflex sympathetic dystrophy	variable	low

Early warning signs of reflex sympathetic dystrophy, such as excessive postoperative pain, should be responded to with diligence and a high index of suspicion.

Postoperative dressings should be loosened or changed in the presence of severe hand or digit swelling.

Prognosis

Dupuytren contracture is progressive, with 75% of patients developing features of more advanced stages of the disease. Men seem to progress more rapidly than women, and patients <50 years of age tend to progress more rapidly than older patients. Although most patients will progress in severity, about 10% will regress. Many patients with nodules or cords do not progress to contracture, as evidenced by a Scandinavian study where only 35% of patients developed contracture during an 18-year follow-up period.[64] Smoking and alcohol use increases the likelihood of progression to surgery. There is currently no proven preventative treatment that can interrupt the progression of contracture.[22]

Treatment guidelines

International

Dutch multidisciplinary guideline on Dupuytren disease (https://www.jhsgo.org) [33]

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Images



Figure 1: Preoperative view of a small finger flexion contracture with surgical indications From the collection of Dr C.M. Rodner; used with permission

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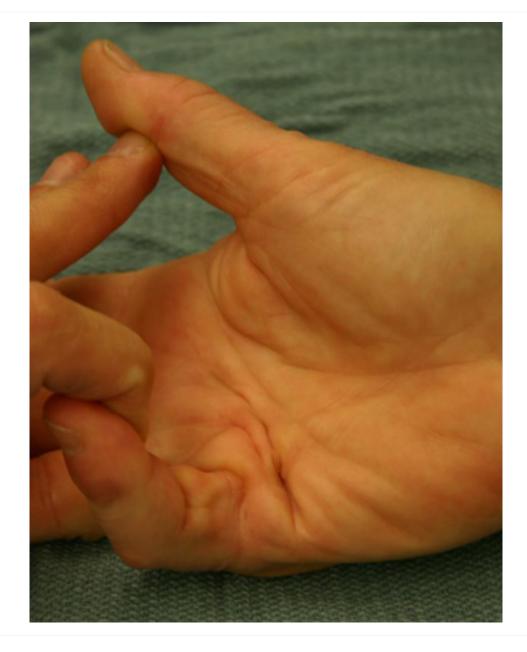


Figure 2: Preoperative view of a small finger flexion contracture with surgical indications



Figure 3: Preoperative view of the ring finger of a patient with a flexion contracture with surgical indications, showing the incision marking, demonstrating a transverse incision overlying the distal palmar crease, and oblique Brunner incisions coursing from it proximally and distally



Figure 4: Intraoperative view of the ring finger of a patient with a flexion contracture, with the radial digital neurovascular bundle identified and isolated coursing volar over the Dupuytren cord, which is being held up by forceps as it is excised in a proximal-to-distal direction



Figure 5: Postoperative view of the ring finger of a patient with a flexion contracture, showing the closed wound over a Penrose drain, which is used to minimize subsequent hematoma formation

IMAGES

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Figure 6: One-month postoperative view of the ring finger of a patient with a flexion contracture, demonstrating full active digital extension

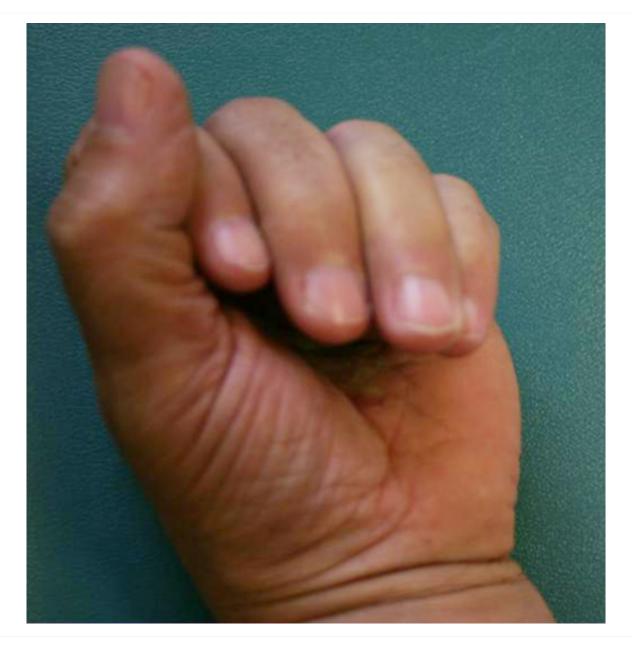


Figure 7: One-month postoperative view of the ring finger of a patient with a flexion contracture, demonstrating active digital flexion

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