THE JOURNAL OF BONE & JOINT SURGERY

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NA Hill

J Bone Joint Surg Am. 1985;67:1439-1443.

This information is current as of December 19, 2009

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Publisher Information The Journal of Bone and Joint Surgery
20 Pickering Street, Needham, MA 02492-3157
www.jbjs.org

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Current Concepts Review

Dupuytren's Contracture*†

BY NORMAN A. HILL, M.D.‡, NEW YORK MEDICAL COLLEGE, VALHALLA, NEW YORK

Although Sir Astley Cooper in England and Boyer in France recognized the gross pathology of contractile palmar and digital fibromatosis around 1823, it was Baron Guillaume Dupuytren who first published the results of open palmar fasciotomy for this entity in 1832, thereby enhancing his already formidable surgical reputation and eponymously establishing the entity that we know today as Dupuytren's contracture[5]. We are still, however, far from a complete understanding of the enigma of this disease entity from either an etiological or a therapeutic aspect.

Incidence

The highest incidence of Dupuytren's disease occurs in men in the fifth to seventh decades of life who are of Scandinavian or Celtic origin. Hueston stated that some evidence of palmar fibromatosis is present in more than 25 per cent of the male Celtic population older than the age of sixty years. It is extremely rare in the Mediterranean and Semitic populations. Although it is also rare in blacks of pure blood[12] and in Asians, recent reports from a continuing international study of the disease by McFarlane have shown that some cases are now being reported from China and Japan[14]. James contended that it is inherited as an autosomal dominant trait with a ten-to-one male-female incidence. For a reason as yet unclarified, women who are operated on for Dupuytren's disease seem to have a higher incidence of postoperative joint stiffness and a greater tendency to the development of postoperative reflex sympathetic dystrophy. A higher incidence of Dupuytren's disease is found in epileptics who are more than forty years old, in alcoholics, in diabetics, and in patients who are confined to bed rest for a prolonged period. The role of hand trauma with attendant swelling and immobilization is still controversial, as is that of vascular insufficiency and manual labor. In a 1978 population study, Mikkelsen claimed to show an increased incidence of Dupuytren's disease in manual laborers, but recent statistical studies have questioned these conclusions[5]. Certainly, for the practicing hand surgeon, one fact stands pre-eminent. The young patient with a strong family history of Dupuytren's disease who exhibits the so-called Dupuytren's diathesis of palmar and digital fibromatosis, knuckle pads, Ledderhose's disease (plantar fibromatosis), and sometimes Peyronie's disease is highly prone to recurrence and extension of the disease after surgery. Such a patient should be forewarned that multiple procedures may be necessary over the years to control the expected relentless progress of the contractures.

Pathology

The clinical picture of Dupuytren's disease frequently begins with a tender nodule in the palmar or digital fascia, usually in, but not limited to, the ulnar two rays or the thumb-index web space. In time, the tenderness abates, and the course of the disease then becomes quite variable. The nodules may remain static or may rapidly evolve into the familiar pattern of skin fixation and digital contracture secondary to abnormal cords of palmar or digital fascia. Differences of opinion exist in regard even to the area in which the disease process begins. Skoog suggested that microhemorrhages occur within the palmar fascia, and Hueston postulated that the origin of the pathology is between the skin and the palmar fascia. The fact that Dupuytren's contracture is eventually a fasciodermal disease is apparent to any surgeon who has ever attempted to develop a surgical cleavage plane between a fixed proximal phalangeal or distal palmar nodule and the overlying skin.

Gross Pathology

The elegant, detailed, and beautifully illustrated studies by McFarlane[12,13] and by Chiu and McFarlane of both normal and abnormal palmar fascia should be studied in detail by any surgeon contemplating surgical correction of the palmar or digital contractures produced by this disease. Failure to excise the diseased fascial structures invites recurrence. As pointed out by Skoog, the superficial transverse ligament of the palmar fascia does not produce contractures of the digits, although its terminal insertion into the skin of the thumb at the level of the metacarpophalangeal joint as well as the natatory ligament may be responsible for the thumb-index web-space contracture[12].

The pathology seen in Dupuytren's disease consists of changes produced in the normal palmar fascia, and when these contractile changes occur the bands of abnormal palmar or digital fascia are then referred to as cords. It bears re-emphasis that the anatomical drawings of Chiu and McFarlane and of McFarlane[12,13] must be studied in detail, for the following description of the joint changes produced by palmar and digital fascial contracture seems incomplete indeed without reference to their illustrations. A knowledge

* This article was accepted for publication prior to July 1, 1985. No conflict-of-interest statement was requested from the author.
† Supported by a grant from the Walter Scott Foundation.
‡ St. Luke's Hospital Center, 1090 Amsterdam Avenue, New York, N.Y. 10025.

of the anatomical and pathological changes in the abnormal cords is also mandatory in order to avoid damage to the neurovascular structures that are frequently displaced from their normal course, especially in the digits. In essence, the pre-tendinous cord of the palm that inserts into the skin distal to the metacarpophalangeal joint and into the tendon sheath does not cause neurovascular displacement, but is responsible for metacarpophalangeal flexion deformities which can be predictably corrected by surgical excision. Flexion contractures of the proximal interphalangeal joint are much more difficult to correct surgically and are often presaged by the development of a firm, fixed nodule at the proximal phalangeal level. Flexion deformities of the proximal interphalangeal joint may be caused by contracture of a diseased central cord which is an extension of the palmar pre-tendinous cord and inserts into the bone, tendon sheath, and skin of the middle phalanx.

Chiefly responsible for neurovascular displacement both proximally and to the midline is a spiral cord lying alternately dorsal and then palmar to the neurovascular bundle as it spirals to blend with Greyson's ligament and attaches to the tendon sheath. When a spiral cord is involved, the neurovascular bundle may lie palmar to it and be displaced both medially and proximally, and hence may be injured in the course of either the skin incision or subsequent dissection. The lateral digital cord, which receives contributions from the natatory ligament and the spiral cord, may cause proximal interphalangeal-joint contractures by its insertion both into skin and into Greyson's ligament. Cleland's ligament, lying dorsal to the neurovascular bundle, is less often involved. The retrovascular cord of Thomine may be responsible for both proximal interphalangeal-joint and distal interphalangeal-joint contractures. A contracted natatory cord, which also contributes to the lateral digital sheath, can result in web-space contracture and sometimes in rotatory deformity of the digit, as can a contracted spiral cord. McFarlane[13] stated that the hyperextension that sometimes is seen in the distal interphalangeal joint is usually the result of extensor imbalance secondary to the flexion deformity of the proximal interphalangeal joint and not due to disease of Landsmeer's oblique retinacular ligament. As pointed out by Strickland and Bassett, isolated digital cords without apparent palmar involvement may arise from the periosteum of the proximal phalanx or the adjacent intrinsic tendons, cross palmar to the neurovascular bundle, and insert into the periosteum and tendon sheath of the middle phalanx, producing flexion deformities of the proximal interphalangeal joint and often displacing the nerves and vessels toward the midline.

Histology and Histochemistry

Within the past decade, considerable interest in the histological and histochemical changes in patients with Dupuytren's disease has developed. The awareness that the histological appearance of an excised plantar nodule in a patient with Dupuytren's disease may look alarmingly like that of a fibrosarcoma must always be kept in mind. Electron microscopic studies[1,2] have shown that the myofibroblast is the predominant cell that is believed to be the active contractile element in the nodules of the diseased palmar fascia. Its origin is believed to be the perivascular fibroblast. Kischer and Sperr have suggested that the conversion to a myofibroblast may be triggered by localized hypoxia in response to microvascular occlusion. The contractile mechanism and its means of initiation is still unknown. On the basis of histochemical studies, Badalamente et al. stated that the adenosine triphosphatase activity of the myofibroblast can be statistically correlated with the severity of the disease in its active phase. The contractile mechanism appears to be located on the myofilaments, but whether the contractility is produced through the interactivity of the individual myofibroblasts or through their intimate linkage to the matrix is as yet unproved. Brickley-Parsons et al. suggested that there is an increase in hexosamine and galactosamine and of Type-III collagen in diseased fascia. The presence and role of Type-III collagen is still controversial. They stressed that, importantly, the demonstrated histochemical changes are even found in seemingly uninvolved palmar fascia in patients with Dupuytren's contracture. The mature cord consists of an orderly arrangement of longitudinally oriented collagen-fiber bundles containing relatively few cells that resemble mature fibroblasts[1,14].

Treatment

Observation alone is indicated in the absence of digital contracture. Medications such as vitamin E, steroids, and splinting have not been proved to be effective in the prevention of digital contractures. The indication for surgical intervention in Dupuytren's disease is the development of digital contracture at the level of the metacarpophalangeal or proximal interphalangeal joint or the development of a contracture at the thumb-index web space. The latter is rarely seen as an isolated entity. Excision of palmar nodules or knuckle pads is rarely indicated. Preoperative evaluation of associated disease entities is obviously mandatory. The palpation of a distinct contractile cord arising from the abductor digiti quinti should alert the surgeon to the possibility that the ulnar neurovascular bundle to the little finger may be displaced to the midline, and the presence of an elevated pouch of soft skin in the web space between the digits suggests spiral cord involvement of the neurovascular bundle, which may lie palmar to a diseased spiral digital cord and hence be injured by the initial incision or subsequent dissection[7]. The patient who has an endomorphic body configuration with associated short, fat digits presents warning signs that postoperative stiffness may be a problem. Surgery for such a patient should be tailored toward the ability to start early active motion as afforded by the open-palm method of McCash. Limited subcutaneous palmar fasciotomy is a temporizing partial solution that is applicable to the aged and to patients who are unsuitable for anesthesia, and to facilitate access to more definitive procedures in the severely affected hand. Simple subcutaneous fasciotomy should never be attempted in the digits, where the neurovascular elements may be easily injured[10,17].

The current trend is for limited palmar and digital fas-

ciectomy confined to the rays involved. Total palmar fasciectomy performed in the hope of eliminating the possibility of recurrence or extension of the disease is to be condemned because of the frequent profound postoperative morbidity. Magnification by optical loop or the operating microscope is essential in order to identify the involved fascia and both normal and displaced neurovascular structures. The tourniquet should be calibrated for accurate pressure and gas content preoperatively. Premature tourniquet deflation with subsequent blood-staining in the field is highly disconcerting and makes accurate tissue identification extremely difficult. Although opinions differ, I think that the tourniquet should be released prior to skin closure and that fastidious hemostasis should be obtained.

Choice of Incisions

The skin incision planned for partial palmar and digital fasciectomy is designed according to the severity of the contracture. For example, in contractures of the metacarpophalangeal joint by a single linear pre-tendinous cord, perhaps associated with proximal interphalangeal-joint contracture as well, a simple linear mid-palmar and mid-digital incision crossing the distal palmar, metacarpophalangeal, and proximal interphalangeal flexion creases perpendicularly will afford excellent exposure and decrease the risk of damage to any displaced neurovascular structures. After excision of the diseased fascia and correction of the contractures, three small z-plasties are planned, so that with the transposition of the flaps the horizontal limbs fall at or near the proximal interphalangeal, metacarpophalangeal, and distal palmar creases. Such an incision may be used in patients who have involvement of multiple rays without fear of circulatory compromise to the intervening palmar skin. Even when the contracture appears to be limited to the proximal interphalangeal joint alone, I think that excision of the palmar pre-tendinous cord to the involved ray should be performed in conjunction with the digital dissection.

Equally effective in such a patient would be a modified Bruner zigzag incision without necessarily carrying the tips of the flaps to the mid-axial line. Closure can be effected by a V-Y advancement of the flaps to gain additional skin length[7]. Major skin creases should not be crossed at a right angle.

In patients who have severe involvement, with marked metacarpophalangeal and perhaps proximal interphalangeal joint contractures as well as fixation and so-called shortening of the palmar skin and involvement of multiple rays, a transverse incision across the palm beginning at the level just proximal to the metacarpal heads and paralleling the distal palmar crease with an extension proximally over the hypothenar eminence from the little finger provides excellent exposure[11]. When the contractures have been corrected, a gap of as much as three to four centimeters may be present. Separate zigzag or z-plasty incisions are used to remove diseased digital fascia, and these incisions may be connected to the transverse palmar wound, if necessary. Either the palmar or the digital wound, or both, may be left open, and in four to six weeks will heal into a fine linear scar. Lubahn et al. have shown that the open palmar wound does not heal simply by granulation, but by flattening of the transverse skin wrinkles. They illustrated this by tattooing a small mark on each side of the proposed incision. When the metacarpophalangeal flexion deformity is corrected, these marks may be separated by as much as three to four centimeters as the palmar skin retracts. When healing is complete, the two marks will have returned to their original preoperative positions. The advantage of this incision is that early motion may be started and hematoma formation, pain, edema of the hand, and joint stiffness are avoided in most patients. These are all factors that may lead to reflex sympathetic dystrophy in some patients. Postoperative extension splinting is mandatory. The only disadvantage in this technique is the extra two to three weeks that are needed for complete healing to occur. Lubahn et al. reported twice as many complications in wounds that were closed primarily. This is not, however, universally agreed on.

Once the skin incision has been planned, the limb is exsanguinated with an Esmarch bandage and the tourniquet is inflated. Under magnification, the skin flaps are elevated carefully from the diseased fascia. As many of the small vertical vessels to the flaps are preserved as possible. As much subcutaneous tissue is left on the flaps as the extent of dermal involvement with diseased fascia permits. This becomes increasingly difficult as the metacarpophalangeal joint and proximal phalanx are approached. A number-64 Beaver blade, with its rounded tip, is helpful here. We prefer to use traction sutures in the flaps, weighted by a mosquito clamp, rather than retractors. A transverse incision is then made across the exposed offending pre-tendinous cord at the level just distal to the superficial palmar arch. This maneuver is often accompanied by a soul-satisfying snap as the contracted metacarpophalangeal joint is released into extension. The neurovascular bundle is well protected at this level, lying considerably dorsal to the palmar fascia. The nerves and vessels are carefully exposed by dividing the vertical palmar fascial septa of Legueu and Juvara. Skin fixation to palmar and digital fascia and nodules makes dissection in the distal part of the palm, particularly in the digit, hazardous, for the neurovascular bundle may lie either superficial or deep to the diseased fascia. In the little finger, the ulnar palmar digital nerve should be isolated proximally over the muscle belly of the abductor digiti quinti and progressively dissected free from the diseased fascia. When the nerves and vessels run deep to a hard nodule, we have found it particularly helpful to delineate their course progressively by spreading with blunt-nosed fine dissection scissors just palmar to the neurovascular bundle and then grasping the nodule with fine forceps on either side and dividing the nodule until the nerve and vessel are clearly visualized. This maneuver is repeated until the courses of the nerve and artery are completely delineated out to the distal phalanx. The specimen is then removed in two pieces. This is not often necessary but has proved most helpful in some of the more difficult dissections. All tissue over the flexor tendons

should be excised, for retention of the fatty tissue distal to the metacarpophalangeal joint may invite a recurrence of flexion deformity of the proximal interphalangeal joint via a retained central cord[13]. If possible, the tendon sheath should not be entered during the course of dissection unless it proves to be contributing to the contracture. A careful inspection for diseased spiral, lateral, and retrovascular cords is made, and any suspect tissue is excised. All too frequently, however, despite careful excision of all diseased digital fascia, a flexion deformity of the proximal interphalangeal joint may remain. Common factors contributing to this are pericapsular fibrosis, shortening of the palmar plate checkrein and accessory collateral ligaments, and contracture of the flexor tendon sheath. Correction can sometimes be obtained by a transverse division of the tendon sheath just distal to the A2 pulley. If a contracture is still present, the tendon sheath should be opened longitudinally, the superficialis and profundus tendons should be retracted first to one side and then to the other, and the checkrein ligaments of the palmar plate should be released, sparing the digital arterial branches to the vincular system running immediately dorsal to them. Should this be ineffective, division of the accessory collateral ligaments at their attachment to the palmar plate may be necessary. It is better at this stage to accept a mild residual contracture than to release the palmar plate completely from the neck of the proximal phalanx. In the older patient this frequently results in eventual marked limitation of motion in both flexion and extension. When a long-standing flexion deformity of the proximal interphalangeal joint has been released, judgment must be exercised in regard to the degree of extension permitted in the postoperative dressing. Despite metacarpophalangeal joint flexion, too enthusiastic a degree of extension may lead to digital vascular compromise. This must be evaluated after release of the tourniquet.

If contracture of the thumb-index web space exists, adequate exposure for the excision of the offending fascia may be obtained by a standard or a four-flap z-plasty, or the T-shaped incision described by McFarlane[13]. Despite aggressive excision of all visible diseased digital fascia, a persistent hyperextension deformity of the distal interphalangeal joint may remain uncorrected. This may require tenotomy of the distal extensor mechanism over the middle phalanx[13].

Dermofasciectomy

Controversy still remains with regard to the role of fasciectomy augmented by full-thickness skin-grafting, particularly in patients with recurrent disease and in younger patients exhibiting Dupuytren's diathesis. Hueston and Gonzalez were strongly of the opinion that recurrence is largely secondary to remaining disease in the dermis, and that therefore a careful fascial excision augmented by full-thickness grafting serves as a sort of fire-break to recurrence of the disease. McFarlane[13] disagreed, contending that grafting is unnecessary if a thorough fascial excision is performed. Hueston maintained that recurrence never occurs beneath a skin graft, but the recent observations by Tonkin et al. suggest that this may sometimes happen. When grafting is elected in situations where skin involvement is marked, the graft should be full thickness and obtained from hairless areas of the mesial proximal area of the brachium, the antecubital fossa, or the lateral area of the groin in suitable patients. The hypothenar eminence serves as an excellent source of smaller grafts. Careful planning of the incisional margins of both the donor site and the recipient site is necessary in order to prevent a contractile scar. If the flexor tendons have been exposed in the course of dissection, then appropriate local flaps or sometimes a cross-finger pedicle flap must be utilized to cover the area of exposed flexor tendon. In the black patient, the mesial aspect of the plantar instep may be used for a full-thickness graft in order to avoid a hyperpigmented skin graft in the palm or digits.

If the wound has been closed postoperatively, it is my opinion that a small drain should be used in the palmar wound. A number-8 plastic infant-feeding tube serves well for this purpose and can be removed in forty-eight hours without disturbing the dressing. This should be a snug, voluminous fluff dressing that supports the metacarpophalangeal joints in flexion and the proximal interphalangeal joints in as much extension as the vascular status allows. It should be supported by a dorsal plaster splint extending to the elbow with the wrist at zero degrees of extension. The hand is elevated in a postoperative arm sling immediately, and the digital tips should be exposed for observation of the circulation. The development of pain in the postoperative period is rare, and should be evaluated immediately by a dressing change to rule out hematoma formation or edema secondary to a tight dressing. If a palmar hematoma is found, the patient should be returned to the operating room, the wound should be opened, the hematoma should be removed by thorough irrigation, and the bleeding vessel must be controlled. The wound may then be left open if it is transverse or closed secondarily at the discretion of the surgeon. Even when the open-palm technique is used, the wound is dressed at approximately ten days and a less formal dressing is applied. When wound-healing allows, active motion is started and gentle dynamic splinting, if indicated, may be instituted. When proximal interphalangeal-joint contractures have been corrected surgically, splinting both in flexion and in extension may be necessary. This should be continued for at least four weeks, and longer if the contracture has been severe. For long-standing contractures of the proximal interphalangeal joint, night splinting should be continued for twelve to sixteen weeks, with a careful follow-up to ensure that volar scarring secondary to the surgery does not produce secondary contractures. The time until the patient can return to work varies with age, severity of disease, and occupation, but averages six to sixteen weeks.

Complications

Edema, pain, and hematoma formation may be the precursors of a reflex sympathetic dystrophy. Immediate correction of those conditions should avoid that complica-

tion. Although digital nerves or arteries that were divided during dissection should be repaired, this complication should be infrequent. It is perhaps redundant to add that no matter how mild the preoperative contracture, most surgeons agree that both hands should not be operated on at the same sitting.

Long-Term Results

Recurrence is defined as new disease developing in an area that has been operated on previously. Extension means the appearance of disease in an area of the palm or digits that has not been operated on previously. It must be stressed that the accurate observer reports recurrence or extension as even the presence of a palpable Dupuytren's nodule or cord, whether or not they have caused recurrent digital contracture[20]. The figures vary widely in the literature and, of course, should be correlated with the patient's age and race and the severity of the preoperative disease. The duration of follow-up is also important. As Tubiana and Leclercq have shown, the longer the follow-up the higher the rates of both recurrence and extension, presumably because the disease develops in these patients at an earlier age. Tonkin et al. reported a 46.5 per cent rate of recurrence, while Hueston reported that an 80 per cent rate of recurrence or extension occurred in the population of Melbourne, which was highly prone to the development of Dupuytren's contracture. It must be remembered that the presence of a recurrence or extension does not necessarily mean that the patient is functionally incapacitated, but emphasizes the fact that surgery does not always cure Dupuytren's disease.

In summary, then, Dupuytren's contracture can be controlled and the patient's hand function can be improved by appropriate surgery. Surgical intervention, however, is often not a definitive cure, and patients should be so forewarned.

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