The surgical management of deformities of the hand in leprosy

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The current global picture of the control of leprosy is mixed. The disease has been eliminated at the national level in 113 of the 122 countries in which it was a public-health problem in 1985. It is still encountered in India, Brazil, Myanmar, Madagascar, Mozambique, Nepal and Tanzania where the prevalence rates still exceeded 1 per 10 000 at the beginning of 2004.1 The World Health Organisation (WHO) monitors the elimination strategies in all countries. In India, the prevalence rate in March 2005 was 1.34 per 10 000, with a new case detection rate (NCDR) of 2.2 per 10 000.2 The proportion of patients with visible deformity (WHO grade II) at the beginning of 2004 was 1.44%.3

Nerve impairment and deformity in leprosy

Infiltration of the peripheral nerves by Mycobacterium leprae initiates a cascade of destructive events with intense intraneural oedema, and destruction of Schwann cells and axons by a CD4+T-cell-mediated granulomatous process.4-6 Acute clinical states in leprosy render patients at greater risk of nerve damage.4 7 The long-term function of the affected nerve is actually determined by the duration of the disease and the level of nerve impairment at the time of diagnosis and commencement of definitive therapy.8-11 Multidrug therapy arrests all forms of the disease,12 and although there has been a dramatic reduction in the prevalence of leprosy worldwide, the incidence of new cases has yet to fall significantly, with the pool of patients with potential nerve damage remaining large.13 Although the set target dates to eradicate leprosy totally are pushed further into the future, much is being done to minimise the impact of sensory and motor impairments. The discovery of ‘pre-clinical’ tests which would predict nerve damage and impending reactions would be of immense value.

Deformities in leprosy are the consequence of impairments of nerve function. The patterns in the upper limb are, in order of frequency, proximal or distal ulnar nerve palsy, distal median nerve palsy, combined proximal or distal ulnar and median nerve palsy and, rarely, a triple-nerve palsy.5,9,14,15 The bulk of elective surgery in the hand is done for primary impairment leading to motor paralysis. Operations on nerves in attempts to regain sensation and autonomic function are now undertaken less often since there is no sound evidence to suggest that significant function returns to the hand.16 A recent randomised controlled trial showed that conventional decompression of the ulnar nerve combined with steroid therapy had no additional benefit over steroid therapy alone. Unrelenting nerve pain, despite adequate steroid therapy, and nerve abscesses are some of the remaining indications for neural surgery. It has been shown17,18 that there are scattered or skip lesions in affected nerves which call into question the intent and purpose of free nerve or skeletal muscle interposition grafting for ‘local’ nerve pathology. However, there is a need for a multicentre controlled study of fascicular nerve decompression using standardised microsurgical techniques together with steroid therapy to provide a better chance for sensory restoration in early leprous neuritis. The traditional safe guards for insensitive hands continue to be the use of visual input to moderate functional activities, the prevention of injuries and health education on the vital function of the hand. Secondary impairments such as ulcerations, contracture and absorption arise as a result of the neural damage in the course of untreated leprosy and are indications for surgery when required.

The aim of reconstructive surgery on the hand is to augment its capabilities for the activities of daily living (ADL) and for safe vocations, and to restore form and structure adequately in order to accelerate the patient’s integration into society.

This article describes the methods of surgical reconstruction as applied to paralytic hands of patients with leprosy and explains some of the permutations and combinations which are used.
Prerequisites for surgical correction and pre-operative therapy

Before elective reconstruction, a full sensory and motor assessment and an occupational and functional appraisal are required. The deformities must be established and the disease quiescent. The multidrug therapy regime must have been completed. There must be no evidence of an acute neuritis or the use of steroids for some months preceding surgery. The skin smears must be negative and the patient must be sufficiently motivated and understand what surgery can accomplish and also its limitations. Pre-operative hand therapy is mandatory in making stiff hands supple, isolating and strengthening the muscles identified for transfer and releasing myostatic capsular contractures in order to restore the Assisted angles in claw hands to the zero status. All adverse features associated with primary paralytic deformities will negatively influence surgical correction if left unnoticed and untreated. They require judicious use of the techniques of hand therapy and, if these are unsuccessful, specific surgical procedures such as release of contractures and skin grafting, repair of the extensor apparatus and relocation of tendons on the dorsal summit of the metacarpophalangeal (MCP) joint in conjunction with primary correction. A home self-care exercise programme and intermittent use of dynamic orthoses reduce severe contractures and allow easier post-operative re-education of tendon transfers for hand deformities when seen late. Special attention should be given to hypermobile hands, as is found in many South Asians and in children, because of the potential for overcorrection of the deformity after tendon transfer if tensioning at re-attachment is not optimal.

Specific surgical corrections

Ulnar nerve paralysis is the most common lesion and results in deformities and deficiencies which can be identified by specific tests and signs. Claw-finger and z-thumb deformities are common and indicate a collapse of the digital bi-articular system, the intrinsic-minus deformity. Correction of claw hands is achieved by transferring a motor tendon such as the tendon of flexor digitorum sublimis (FDS), palmaris longus, extensor carpi radialis longus (ECRL) or extensor indices to any one of four insertion sites in the fingers. These are the lateral band of the dorsal extensor expansion, the proximal phalanx, the proximal annular pulleys of the flexor sheath and the interosseous tendons. An ideal way to choose an insertion site for a motor tendon is by assessing the types of claw hand based on their inherent characteristics and adverse features; types I to V have been described. Adjustment of the tension on the tendon slips at re-attachment is important since excessive tension produces an intrinsic-plus deformity and slack tension results in inadequate correction. The time-tested Brand-Fritschi intra-operative passive tendon-tensioning method, modified by the author, is simple and effective. In the future it may be possible to take measurements of the length of sarcomeres intra-operatively, and, combined with predetermined data of the biomechanical measurements of characteristics of normal muscle/tendons, this will allow re-attachment with the optimal length and tension to provide more functional improvement.

Brand’s extensor-to-flexor four-tailed transfer to the lateral band of the extensor expansion gives excellent or good results in 85% of type-I and type-II claw hands while transfer of either FDS or palmaris longus to the interosseous insertion gives excellent or good results in 96%. In type-III and type-IV claw hands the palmaris longus extensor-to-flexor four-tailed transfer, or the extended pulley insertion procedure, which uses the A1 and A2a pulleys combined as the insertion site, give excellent or good correction in 90% of cases at two years. A recent follow-up by the author at between five and 17 years (mean 8.17, SD 2.87) of 94 hands which had received the extended pulley insertion procedure using either FDS or palmaris longus showed that excellent or good correction had been maintained in 86%. In type-IV and type-V hands, the Stiles-Bunnell modified single superficialis transfer to the insertion of the lateral band gave excellent or good correction in 95% of fingers. In a recent article on the correction obtained on unspecified types of claw hand in leprosy, excellent to fair results were achieved in 92% using the extensor-to-flexor four-tailed transfer, and similar results with the palmaris longus four-tailed transfer both of which were transferred to the insertion of the lateral band. Harvesting of the superficialis tendon through the lateral approach gave rise to flexion contractures of the proximal interphalangeal (PIP) joints, distal interphalangeal (DIP) extension lag in the donor finger, a high incidence of swan-neck deformity, a ‘superficialis-minus’ deformity, and loss of flexion of the PIP joint. These problems have been lessened by harvesting the FDS tendon through the proximal pulley systems. It is now recommended that a finger from which FDS has been removed should not have the same tendon, or a slip of it, inserted into the lateral band as in the correction of clawing.

Static procedures for correction of clawing which maintain the MCP joint in some degree of flexion, or limit its hyperextension, such as Parkes’s flexor graft tenodesis operation, the Zancolli volar capsulodesis, or the dorsal bone block, have been reliable for the correction of claw deformity in non-leprous patients. However, in the insensitive hands of patients with leprosy who are engaged in farm and factory work, these operations are suspect because of continued weakness in these hands because of the absence of prime flexor function at the MCP joints, diminished work space, excessive flexion at the MCP joint, exaggeration of the reversed transverse metacarpal arch, flexion contracture of the fingers and long-term recurrence. They are best avoided in patients with leprosy and palsy of the ulnar nerve. Other deformities such as a reversed transverse metacarpal arch, which prevents cup-
ping and holding of objects within the palm, benefit from the transfer of the extensor digitorum minimus or FDS.\textsuperscript{24,52} For flexion-adduction weakness of the wrist the medial translocation of palmaris longus and half of the flexor carpi radialis (FCR) tendon to the flexor carpi ulnaris (FCU), partially increases the strength.\textsuperscript{24} For weak function of flexor digitorum profundus (FDP) to the little and ring fingers, transverse tenodesis of these tendons to the FDP tendons of the middle and index fingers in the distal forearm redistributes strength. Deficiency or wasting of the intermetacarpal spaces is of concern to some patients and they may benefit from a dermofascial free-graft insert.\textsuperscript{24,53}

Deformities of the thumb in leprosy require a sound reconstruction. The claw thumb or z-thumb deformity of the ulnar nerve palsy prevents proper pulp-to-pulp and key pinch, besides the gross weakness. An ‘adductorplasty’ using either a tendon of FDS,\textsuperscript{55} extensor carpi radialis brevis (ECRB)\textsuperscript{54} or brachioradialis,\textsuperscript{55} whose tension fraction is comparable to a normal adductor pollicis,\textsuperscript{56} requires the transfer and re-attachment of the insertion of the paralyzed adductor pollicis in order to provide both strong adduction and a supination arc to the thumb. A fixed extension contracture at the MCP joints requires tension-band fusion of the joint to restore stability and allow the interphalangeal (IP) joint to flex and extend independently. When a flexion deformity of the IP joint predominates, it is advisable to undertake either a fusion for a fixed-flexion contracture or a split flexor pollicis longus (FPL) transfer to the extensor pollicis longus (EPL) for a passively correctable joint deformity.\textsuperscript{24,57}

A deformity with the thumb in the plane of the palm, as occurs in distal median nerve palsy with an insensate thumb, is most incapacitating even if the superficial head of flexor pollicis brevis (FPB) is innervated by the ulnar nerve. Opposition, which is required to achieve effective pulp-to-pulp or tripod pinch, and a strong grip, is provided by a suitable opponensplasty. A number of methods are well described\textsuperscript{58} including Bunnell’s FDS opponensplasty,\textsuperscript{59} with Thompson’s modification of this,\textsuperscript{60} or Brand’s dual-insertion technique\textsuperscript{15,56} all of which are suitable for stiff thumbs,\textsuperscript{46,61,62} and an extensor indicis opponensplasty\textsuperscript{63} for supple thumbs.\textsuperscript{64} These procedures give a success rate of about 90%.\textsuperscript{46,61-64} They can be performed safely as an adjuvant to the correction of a claw hand with combined ulnar and median nerve palsies. Other choices for opponensplasty include transfer of the tendons of extensor pollicis brevis (EPB), palmaris longus or extensor carpi ulnaris (ECU).

Combined median and ulnar palsies require careful planning. These hands are often beset with a challenging mix of adverse features. They should be treated with a dynamic orthosis before operation. Dynamic correction of clawed fingers and an opponensplasty are performed as the first stage, followed by restoration of adduction to the weak thumb. Other procedures can then be undertaken according to the patient’s needs.

**Thumb web contracture in late combined palsy of the ulnar and median nerves**

Brand’s extended single z-plasty\textsuperscript{15} is a simple technique designed to correct the thumb-web contracture which acts as a serious restraint for palmar abduction before opponensplasty. This procedure has been modified by the author to include one or two terminal branches of the superficial radial cutaneous nerve in the dorsoradial component of the flap so that on transposing the two flaps the sensate dorsoradial component will come to lie on the radial base of the thumb. In 26 paralytic hands in which this procedure was undertaken within the last 20 years, gross grip and side pinch were objectively more effective, enabling patients to moderate their action subjectively in order to reduce the strain on the MCP joint. In all of them the FDS opponensplasty had given good results.

**Triple nerve paralysis**

The combination of palsy of the radial nerve and of the proximal ulnar and distal median nerves leaves a hand with global sensory loss, severe contractures and limited motor assets, making reconstruction formidable. Fortunately, these have been rare\textsuperscript{15,65} and are now seldom seen. Physical assessment must include testing of position and vibration sense and biplanar radiographs of the wrist and hands to rule out neuropathic changes. After an adequate period of supervised hand therapy surgery is carried out in two stages\textsuperscript{15,65-67} In the first stage the pronator teres tendon is transferred to ECRB and that of FCR to extensor digitorum communis (EDC) + extensor pollicis longus (EPL). These restore acceptable wrist and digital extension. Six to eight weeks later the second stage involves transferring two tendons for the correction of clawing and opponensplasty, using the flexor digitorum superficialis of the middle and ring fingers, respectively. Severe contracture of the wrist or neuropathic changes in the joint require compression arthrodesis and bone grafting. The duration of immobilisation should be twice that of fusions in non-neuropathic wrists.

**Miscellaneous problems**

These include bony or fibrous ankylosis of the MCP or IP joints with mediolateral or rotational deformity of the digits and require realignment-fusion with placement in a more functional position. Repeated ulceration may occur on the pulp of the thumb and little fingers. The hands in leprosy do not lend themselves to nerve grafting or transfer. We have performed a first dorsal metacarpal artery sensate-flap transfer to the thumb pulp and a transfer of a neurovascular island-flap from the ulnar side of the middle finger to the pulp of the little finger. These have given adequate results in preventing repeated ulceration, which is the precursor to digital absorption. Mitten hands are the most severe form of absorption in leprosy patients. Two-thirds or more of digital length may be lost and there is a severe thumb-web contracture with the hand serving as a paddle for bimanual activities. The Gillies procedure\textsuperscript{68} is undoubt-
edly the most suitable for restoring thumb length and individuality so that it may serve as a post for the patient to grip protected and modified equipment or utensils. Repeated ulceration, infection and osteomyelitis may require appropriate surgery.

Table I lists the author’s preferred choice of operations for the principal deformities of the hand in leprosy.

Supervised post-operative hand therapy
This is essential after all reconstructive operations on the hands in patients with leprosy. After removal of the cast suitable protective/static splinting is provided. Post-operative re-education is provided in a staged manner, being easier for single tendon transfers. With combinations of transfers, which ideally complement one another, the re-education skills of experienced hand therapists are needed. With the diminished neuromuscular inhibition seen in hands with glove-type sensory loss, hand and occupational therapists must carefully supervise patients in order to co-ordinate their movements so that they perform purposeful, functional activities in the right sequence.

Brand pioneered reconstructive surgery for deformities of leprosy at the Christian Medical College Hospital, Vellore, India by performing the first correction of a claw hand in 1948. He had the foresight to introduce rehabilitation programmes concurrently to channel reconstructed patients to safer vocations. Since then, several thousand surgical procedures have been carried out on patients at this centre, which is now recognised as the birthplace of hand rehabilitation and the first unit for hand surgery in India.

Table I. The author’s preference of hand surgery for the principal deformities in leprosy

<table>
<thead>
<tr>
<th>Required function or clinical finding</th>
<th>Preferred options</th>
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<tbody>
<tr>
<td>Claw hand correction</td>
<td>Type I EF4T, PL4T intersosseous insertion with PL</td>
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<tr>
<td></td>
<td>Type II intersosseous insertion with PL4T or FDSR or ECRL, EF4T, PL4T</td>
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<td></td>
<td>Type III EPI procedure, EF4T</td>
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<td></td>
<td>Type IV FDSM/R to lateral band, EPI procedure, EF4T</td>
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<td></td>
<td>Type V repair/relocate lateral band, then modified Stiles-Bunnell-Littler transfer to central tendon of dorsal extensor apparatus</td>
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<tr>
<td>Thumb adduction</td>
<td>FDS/BR to adductor tubercle, ECRB to proximal phalanx</td>
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<tr>
<td>Thumb MCP joint instability</td>
<td>MCP joint fusion</td>
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<tr>
<td>Thumb MCP joint extension</td>
<td>Half FPL to EPL</td>
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<tr>
<td>Thumb IP joint fixed contracture</td>
<td>IP joint fusion</td>
</tr>
<tr>
<td>Index abduction</td>
<td>EPL to transfer to first dorsal intersosseous</td>
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<tr>
<td>Reversed transverse MC arch</td>
<td>EDM to hypothenar insertion base little finger</td>
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<tr>
<td>Profundus weakness FDPL and R</td>
<td>FDPL and R tenodesis to FDPM</td>
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<tr>
<td>Wrist flexion-adduction weakness</td>
<td>PL to FCU, half FCR to FCU (Yoke transfer)</td>
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<tr>
<td>Loss of thumb opposition</td>
<td>EI for mobile hands, and FDS for stiff hands</td>
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<tr>
<td>Contracted thumb-web</td>
<td>‘Sensate’ single Z-plasty + FDS opponensplasty</td>
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<tr>
<td>Ring and little finger sensibility</td>
<td>First dorsal metacarpal artery neurovascular island flap</td>
</tr>
<tr>
<td>Intermetacarpal space wasting</td>
<td>Neurovascular island flap from ulnar side of middle finger</td>
</tr>
<tr>
<td>Triple nerve palsy</td>
<td>Dermofascial flap from thigh to fill required spaces</td>
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<tr>
<td></td>
<td>First stage PT transfer to ECRB, FCR to EDC and EPL</td>
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<tr>
<td></td>
<td>Neuroplastic changes in wrist: wrist fusion</td>
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<td></td>
<td>Second stage FDSM for claw-hand correction to lateral band insertion</td>
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<tr>
<td>Mitten hand</td>
<td>FDSM opponensplasty (Brand’s ‘Y’ insertion method)</td>
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</tbody>
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* EF4T, extensor-to-flexor four-tailed transfer; PL4T, palmaris longus four-tailed transfer; PL, palmaris longus; EPI, extended pulley insertion; FDS, flexor digitorum superficialis (M, middle finger, R, ring finger); BR, brachioradialis; ECRB, extensor carpi radialis brevis; MCP, metacarpophalangeal; IP, interphalangeal; EI, extensor indicis; MC, metacarpal; EDM, extensor digitorum minimus; FDSR and R, flexor digitorum profundus little and ring fingers, respectively; FCU, flexor carpi ulnaris; FCR, flexor carpi radialis; PT, pronator teres; ECRB, extensor carpi radialis brevis; EDC, extensor digitorum communis; EPL, extensor carpi longus |

Rehabilitation
Rehabilitation is designed to ensure that the surgical procedures undertaken can help patients to achieve their full physical potential. Hands must be protected from injury and infection in order to prevent impairment and disability. Working aids and protective devices are provided whenever necessary in vulnerable patients, who bear scars of previous injury in the hand or have nerve impairment in other limbs. These patients must be placed in safe vocations which maximise visual feedback to protect the hands further. As Brand pointed out a decade ago, “Our forebears cared for those they could not cure. We can cure, but must not forget that many cured patients still suffer the effects of the disease and experience its stigma. They still need care, even after cure.”

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