Indian J Lepr 2011, 83 : 9-14

http://www.ijl.org.in

Opinion Article

An attempt towards prevention and management of disabilities and deformities in leprosy

S Husain

Received : 10.02.2011 Revised : 22.02.2011 Accepted : 24.02.2011

History of prevention of deformities is practically as old as the appearance of the deformities themselves, unfortunately without much understanding to start with. In medieval era and even earlier, leprosy and deformities were treated synonymously and the disease's infectivity too was closely associated with appearance of deformities. Hence, to reduce chances of deformities caused by leprosy in healthy population, the patients having deformities were driven away from the society presuming that only deformed patients spread the disease. Unfortunately, it never worked. However, in later period, factors behind the deformities and disabilities were recognized and understood. These are basically limited to involvement of peripheral nerves and their proper management (medical treatment, surgical interventions, physiotherapy, ergonomics and counseling) by one rule of thumb i.e. early, timely and adequately.

Key words : Deformity, Disability, Peripheral nerves

Introduction

Leprosy is not and has never been feared for its infectivity but for the unsightly disability and deformities caused by the disease. Degree of disfigurement and deformed state served as the parameters to initiate practice of abhorrence, ostracism and atrocities on the helpless leprosy patients by the society. Deformities in leprosy are basically due to affliction of peripheral nerves. It is known that peripheral nerve involvement in leprosy is a common feature that often results in nerve damage leading to various disabilities and deformities. This coupled with ignorance regarding safe use lead to development of secondary deformities, sometimes the permanent one while neglect towards injuries and infection, minor or major, result in loss of the organ. Commonly involved nerves in upper limbs are ulnar and median and in lower limbs are posterior tibial and lateral popliteal in that order (Cochrane and Davey 1964).

It is known that even in non-leprosy situations and conditions, nerves are known to get entrapped at various anatomical sites clinically manifesting in parasthesia and paresis. It is also known that inflamed swollen nerves due to any cause are more prone to entrapment and its consequences as mentioned above (Husain et al 1997, 1998ab).

In multibacillary leprosy, nerve thickening occurs following invasion of bacilli in nerve tissue or following lepra reaction in the nerve while in paucibacillary leprosy hypersensitivity reaction

S Husain, MS, MCh, Scientist F/Deputy Director (Senior Grade)
Department of Plastic and Reconstructive Surgery, National JALMA Institute for Leprosy and Other Mycobacterial Diseases (ICMR), Dr M Miyazaki Marg, Tajganj, Agra-282001, India
Correspondence to : S Husain **Email:** shusain@mail.com

leads to sudden inflammation. Edema consequent to either lepra reaction or hypersensitivity reaction plays a major role in further progression. Swollen edematous nerves passing through a tunnel like structures suffer by getting compressed - entrapment neuropathy. In this entrapment, external pressure is exerted by the rigid and fixed anatomical (bony/ligament) structures while tissue edema exerts internal pressure on the nerve fibers. These juxtaposing pressures result in venous obstruction, capillary stasis, local ischemia, intrafunicular hypoxia and increased intrafunicular tension. The diseased and thickened nerve can get further damaged due to physical trauma following repeated joint movements in day today activities. The net result of entrapment can clinically be measured initially as slowing of conduction velocity and later as full conduction block, resulting in paralysis (Bose et al 1981).

Nerve damage is the foundation on which the sufferings like disabilities and deformities rest. At this juncture, it is worth mentioning that leprous neuritis, contrary to common perceptions of its being acute episodic and violently symptomatic in nature, on many occasions can be chronic and non acute. This chronic, non-acute symptom less state of neuritis is termed as 'silent neuritis' also synonymously known as 'quiet nerve paralysis'. Acute episode of neuritis forces the patient to seek medical attention but in chronic non-acute state of quiet nerve paralysis, functional modalities of the nerve simply ebb away without any notice by the patient and also , if not alert , by the physician. Silent neuritis leads to damage of sensory/motor/autonomic modalities depending upon the involvement of the nerve type i.e. sensory, motor or mixed and site or level of involvement.

Common clinical manifestations are sensory loss, motor loss and autonomic function loss. From the symptomatic point of view, sensory loss is the most commonly reported symptom followed by the motor loss and lastly the autonomic function loss in form of dryness etc. Clinically motor loss manifests in the form of paresis or paralysis in different parts (claw hand, drop foot, lagophthalmos), sensory loss in the form of burns blisters, trophic ulcers and autonomic loss in the form of dryness, fissures, in the affected area. The sensory loss has not received much deserved attention in the literature.

In India, about 25-30% of leprosy cases develop disabilities (Srinivasan 1993). Categorization and grading of the disability and deformities in leprosy were attempted since early in 1970 (WHO 1970) and the latest in the series which is in current use is the classification given by WHO in year 1988 (WHO 1988). According to this, sensory loss or anesthesia is considered as grade 1 while visible disabilities or deformities in the eyes/ limbs are grouped under grade 2 (ILEP Medical Commission 2001).

Prevention of onset of deformities and disabilities

Prevention, diagnosis, treatment and finally rehabilitation of disabilities and deformities caused by leprosy warrant deep understanding of the disease process, the limitations of the treatment challenges posed by the environment and work place to the treated organ, patient's understanding and his own commitment to use and protect the organ under different situations. Hence, when we talk of disabilities, we should not only treat them but we should manage them and this management is done under following headings:

1. Early diagnosis of nerve damage

Early diagnosis of nerve damage can easily be done by a regular and periodic nerve function assessment of all susceptible nerves trunks. This should be done if possible on every visit. Here, it is worth mentioning that for routine nerve function-testing presence of a physiotherapist is not mandatory. In fact if aware, physician or a paramedic can do the assessment and if patient is found developing the nerve deficit detailed assessment can be done.

2. Timely initiation

Practitioners of leprology usually come across the situation where some other treating physician has either delayed the use of steroid or terminated the steroids early or has delayed the much-needed neurolysis. At this juncture, it is being mentioned that in spite of tremendous progress in all areas of leprology, it should be clearly understood that till date there is no substitute for timely steroids. In fact the old dictum still stands true that in situation of neuritis/ orchitis, iridocyclitis and pustular ENL the drug of choice is steroid. There should be no hesitation in using steroids whenever indicated. Delayed treatment with steroids can only result in permanent nerve damage and its squealee.

3. Adequate treatment

As mentioned above, steroids are the drug of choice in treatment of leprous neuritis of all types i.e. following lepra reaction in multibacillary leprosy or following hypersensitivity reaction in paucibacillary, in acute neuritis or quite nerve paralysis. Nowadays, steroid treatment of neuritis is done under a definite protocol, under which steroids can be started with sixty mg daily for first fortnight followed by a tapered dose (by 5 mg) every two weeks till it reaches to the dose of 10 mg and then it should be maintained for six months. Steroid treatment is usually successful but failure of the treatment can also be ascribed to inadequate doses, early withdrawal of the steroids besides other factors like severe disease, exertion, inadequate rest and quality physiotherapy. Early withdrawal of steroids is usually done fearing side effects and steroid dependence. Early steroid withdrawal increases chances of nerve damage and the repeat of steroids in comparatively larger doses to restart with. Frequent weaning and starting of steroids serves no purpose and in fact it may harm the patient in long run.

4. The rest and exercise

The steroid is the drug of choice in treatment of nerve damage but it is not the only step towards saving the nerve. Adequate rest to the inflamed nerve and the gradual initiation of the exercise of the limb to regain the muscle power are equally vital. Generally, after a period of rest, the acute phase is over for one should initiate relevant exercises to regain the muscle power.

5. The shift

If there is no improvement in first three months as evident by decrease in pain and tenderness over nerve, reduced paresthesia or one observes worsening or further deterioration in motor power in muscles supplied by the affected nerves in spite of steroid therapy, then such cases should be considered for the nerve trunk decompression (Palande and Azhaguraj 1975, Pandya 1978). Surgical interventions in the form of epineurotomy by multiple longitudinal incisions and external decompression to relieve the internal pressure throughout the involved segment are considered adequate.

In the following paragraphs, we are sharing our experiences of nerve decompression of various nerves trunks in leprosy.

1. Ulnar nerve

The ulnar nerve involvement is the commonest among the other nerves in leprosy. It may be because of its superficial location at elbow. It may be entrapped by medial intramuscular septum under deep fascia of the anterior medial compartment of the upper arm, in the distal fibro-osseous tunnel and between the tendinous fibers of the origin of two heads of the flexor carpi ulnaris. All these compress the inflamed nerve.

We decompressed ulnar nerves not responding to steroids. The results were evaluated on the basis of subjective improvement and objective findings relating to both sensory and motor modalities and periodic comparison were made with previous assessment values. The follow-up period varied from 5-20 years at different times showed that the recovery of pain occurred in almost 100% of cases.

Pain has been observed to be the first symptom to disappear (Parikh et al 1968, Palande and Azhaguraj 1975). The sensory improvement was noticed in some cases as early as four weeks, though the actual recovery took place in about 20 weeks post operatively. The improvement gradually progressed to complete recovery and the maximum benefit was noticed in about a year after nerve decompression (Pandya 1978). In our series, 48% of the cases showed complete sensory recovery while others had improved sensation as compared to pre-operative state. The improvement in motor function was slow to occur and seen after 24 weeks time. It was more gradual and in some cases, it took about two years to obtain the maximum motor recovery. We noted that the ulnar supplied muscles retain their functional ability up to MRC grade 3 to prevent the development of deformity.

2. Median nerve

Median nerve involvement results in functional loss affecting the pinch and grasp functions. Clinically, the median nerve involvement commonly presents as carpal tunnel at the wrist being the usual site in leprosy (ILEP Medical Commission 2001).

Observations of the studies carried out at NJIL & OMD, Agra

The cases had history of pain in lower forearm along with sensory loss in median nerve innervated area of the hand. All of them had steroids for varying periods of time before surgery. The pain and paraesthesia were the first symptoms to disappear after decompression (Husain et al 1998b) and completely disappeared with in two weeks. Full sensory improvement was seen in 45% cases. While in others 55% cases, the sensory recovery was partial (Husain et al 1997). 45% nerves showed improvement in their motor functions, other 45% cases remained same as they were at the time of operation and 10% cases deteriorated (Husain et al 1997).

3. Posterior tibial nerve

Ulceration of foot is due to sensory loss in sole consequent upon the involvement and damage to posterior tibial nerve. The inflamed nerve is usually entrapped and compressed in tarsal tunnel behind medial malleolus. Only steroid therapy to treat the inflammation looks insufficient and the entrapment needs decompression (Oommen 1996). The results of posterior tibial nerve decompression supplemented with steroids are very promising. The subjective sensory recovery took 3 week to 6 months post operatively. 44% feet shows sensory recovery in full sole where as 29% feet showed recovery in fore foot, 12% in mid foot and 15% in heel only.

In ulnar and median nerves after the nerve decompression, 50% of cases showed remarkable recovery of sensation. 45% cases could retain acceptable muscle function and improvement of sensations to enable them to lead a fruitful and socially acceptable life by not developing claw deformity which in the absence of surgical intervention, in all probability, would have developed (Pandya 1978, Husain et al 1998a). Posterior tibial nerve decompression helps in improvement of sensations as well as vascularity of foot and helps in healing of ulcers and prevents further recurrence (Srinivasan 1993, Oommen 1996, Husain et al 1998b). The observations suggest that along with the basic care of hands and feet, the cases who had nerve decompression had better functional hands and feet.

Other suggestions

1. Orthotics and protective appliances

Liberal and timely use of protective gears like goggles, MCR chapels and tools like insulated spoons etc. should be arranged and their use should be encouraged by breaking mental barriers.

2. Managements of existing disabilities

From the management point of view, disability or deformity management in leprosy can be divided in four stages: stage one is to relieve the pain,

stage two is to save the functionality, stage three is to limit the functionality and finally stage four is to save further damage due to functionality. After developing the acute painful neuritis and impending nerve damage when patients comes out of it, he/she starts believing that his or her limb is as good as before all the problems. And as a result, he/she starts using it indiscriminately. This excessive use leads to damage to the skin resulting in burn/ulcers, infections, damage to small joints resulting in to secondary deformities.

Actually just after negotiating the stage one of acute pain, patient must be made an active partner in disability management program. Patient must be explained that the therapy or surgery has just saved the functionality and made it usable, that too with limited functionality under unyielding external forces. The need is active partnership form the patient's side. Concept of visual compensation i.e. what normal skin can 'see' in reference to sensory modalities like touch, pain, temperature and steriognosis must be explained to the patient and inability to do the same with that of affected limb.

It would be good idea to describe object through touching it with closed eyes with normal hand and then by the affected hand. And later the patient should be taught to compensate the short-comings of the affected limb by visual compensation. Once understood, he should be advised to practice this concept of visual compensation in form of seeing with eyes beforehand what limb is going to perform. Understanding about temperature if lips can tolerate the level of temperature comfortably then probably the insensitive skin would also not get damaged at that temperature, if not applied for long. Things like not standing long on one foot not keeping shoes in sunlight, examining sole and inside of the shoes every time before putting foot in to it. Use of artificial tears and protection of the eyes even during sleep is of prime importance.

To summarise, the patient must be trained to understand the limitation of the treated organ, judicious use of affected and treated organ and finally his responsibility to protect it. Counseling and health education is the key of the prevention of deformities. For a long time, treating surgeons and physiotherapists believed that health education or rather educating the patient in term of do's and don'ts is sufficient. The answer is no. On one hand, the treating partners must understand the difference between the sympathy and the empathy. You can impart information on does and don'ts with sympathy but to counsel the patient one should be empathic. He should put himself in place of patient, realize his problems and then counsel the patient to judge the situation use discretion and be responsible for the out come. Patient must be made to realize that the limb is his/her and with this attitude he/she should thoughtfully weigh his action and likely out come before actually putting the limb to use.

Conclusion

The prevention of deformity and disability in leprosy is a multifactorial process. The patient's personal interest towards the disease along with care and surgical intervention can help in recovery of sensory abilities and prevent development of secondary deformities. Combinations of these can be worked out for individual cases so as to give them a safe and injury free life. Even if only protective sensations are restored with these procedures it will make the life easier.

References

1. Bose KS, Ghosh S and Mukherjee N (1981). Decompression of nerves in the treatment of leprosy. *J Int Med Ass.* **42**: 456-463.
2. Brand PW (1964). Deformity in Leprosy. In: Leprosy in Theory and Practice, 2nd edn (Cochrane RG and Davey TF, eds), John Wright & Sons Ltd, Bristol, pp 447-464.
3. Chaise F and Roger B (1985). Neurolysis of the common peroneal nerve in leprosy. A report on 22 patients. *J Bone Joint Surg Br.* **67**: 426-429.
4. Cochrane RG and Davey TF, eds (1964). Leprosy in Theory and Practice, 2nd edn, John Wright & Sons Ltd, Bristol.
5. Husain S, Mishra B, Prakash V et al (1997). Evaluation of results of surgical decompression of

median nerve in leprosy in relation to sensory-motor function. *Acta Leprol*. **10**: 199-201.

6. Husain S, Mishra B, Prakash V et al GN (1998a). Results of surgical decompression of ulnar nerve in leprosy. *Acta Leprol.* **11**: 17-20.
7. Husain S, Mishra B, Prakash V et al GN (1998b). Nerve trunk decompression in lower limb in Hansen's disease. *Indian J Orthop*. **32** : 85-87.
8. ILEP Medical Commission (2001). Prevention of Disabilities: Guidelines for Leprosy Control Programmes, The International Federation of Anti-Leprosy Associations, London.
9. Oommen PK (1996). Posterior tibial neurovascular decompression for restoration of plantar sweating and sensibility. *Indian J Lepr*. **68**: 75-82.
10. Palande DD and Azhaguraj M (1975). Surgical decompression of posterior tibial neurovascular complex in treatment of certain chronic plantar ulcers and posterior tibial neuritis in leprosy. *Int J Lepr Other Mycobact Dis*. **43**: 36-40.
11. Pandya NJ (1978). Surgical decompression nerves in leprosy. An attempt at prevention of deformities. A clinical, electrophysiologic, histo-pathologic and surgical study. *Int J Lepr Other Mycobact Dis* .**46**: 47-55.
12. Parikh AC, Ganapati R and Kothare KB (1968). Decompression of the ulnar and median nerves in leprous neuritis. *Lepr Rev*. **39**: 143-146.
13. Srinivasan H (1993). Prevention of Disabilities in Patients with Leprosy: A Practical Guide, WHO, Geneva.
14. World Health Organization (1970). WHO Expert Committee on Leprosy, Fourth Report. *WHO Tech Rep*. **459**: 18-19.
15. World Health Organization (1988). WHO Expert Committee on Leprosy, Sixth Report. *WHO Tech Rep Ser.* **768**: 35.

How to cite this article : Husain S (2011). An attempt towards prevention and management of disabilities and deformities in leprosy. *Indian J Lepr.* **83**: 9-14.