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Chronic Lymphedema Management: Case Series Analysis of 9 Years from a Specialized Clinic in Delhi

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Abstract

Chronic lymphedema is a disabling disease with functional and psychological morbidity. Despite an enormous burden of disease, there is a lack of knowledge among providers regarding treatment methodology and equipment. Our surgical care center has focused on treating patients and educating providers for lymphedema. We present a retrospective analysis of our records over the last 9 years. Patients were treated on an ambulatory basis through complex decongestive therapy (CDT). Ancillary drug therapy included benzathine penicillin every 3 weeks and anti-filarial drugs as appropriate. We emphasized limb care, including scrub washes, cleaning with warm saline, emollient applications, exercises, limb movements, and compression using bandaging and intermittent pneumatic compression (IPC) pumps. Training on bandaging was given and a complete bandage kit provided for regular reuse after washing. Surgery was required for selected cases for cosmesis and for extremely bulky, long-standing lymphedema with secondary skin changes. Subjects were longitudinally monitored using self-reported subjective criteria and objectively by limb volume measurements. In 95 patients (119 limbs), CDT and prophylactic benzathine penicillin resulted in significant decrease in swelling, reducing mean upper limb volume from 3.6 to 3.2 1 (p<0.001) and mean lower limb volume from 11.0 to 9.0 1 (p<0.001). The use of a pump did not significantly add to further improvement. Failures were few, and mostly related to noncompliance. Focused treatment strategies, education, pharmacotherapy, and home-based care with bandages as well as IPC have shown success in improving the outcome of chronic lymphedema.

Keywords Intermittent pneumatic compression (IPC)Lymphatic diseases \cdot Lymphatic filariasis \cdot Manual lymphatic drainage (MLD) \cdot Multilayer lymphedema bandage (MLLB) \cdot Complex decongestive therapy (CDT)

Introduction

Lymphedema affects about 250 million persons worldwide [1]. Cancer is the commonest cause, but other diseases may be responsible [2]. Lymphatic filariasis (LF), prevalent in parts of India, Africa, and South America, is the commonest

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Suneet Sood suneetsood@imu.edu.my cause of secondary lymphedema in tropical countries. LF affects 120 million people disfiguring or incapacitating 40 million [3, 4].

Despite these staggering figures, there is a lack of knowledge among healthcare providers regarding lymphedema management in both developing and developed countries [5, 6]. Lymphedema is neglected across the globe [6, 7]. The number of lymphedema specialists is low, and healthcare workers feel helpless and unable to provide effective care [1, 5, 7].

Since 1990, we have been running a day care surgical center in New Delhi. In 1996, we started focusing on the care of patients with filarial lymphedema, and later expanded our scope to a wider variety of cases of swollen limb as well as ulcers. Our treatment protocols continued to evolve over time through experience as well as availability of improved equipment. We have also organized care delivery and training camps in various parts of the country. This paper describes our management approach and results for patients with limb

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lymphedema seen at our clinic over the last 9 years. By this period, our record keeping methods had also improved.

Patients and Methods

In addition to a fully equipped operation theater, three recovery beds, and a reception area, our office surgical suite has a treatment room within the outpatient area. Lymphedema patients come from across the country. They stay nearby, and care is provided on an ambulatory basis. Patients need to spend between 2 and 4 h at the clinic during the initiation phase of care. The duration of stay used to be 7–10 days unless a surgery was done. This has lately been reduced to 4–6 days.

Preliminary Workup

Preliminary workup included a history and examination, with an attempt to pinpoint a diagnosis to help make a firm treatment plan. Appropriate counseling was provided.

The diagnosis of filariasis was based on the geographical origin of the patient. A card test is available for the diagnosis of active filariasis [8], but since lymphedema occurs long after active filariasis is over, we rarely conducted the test on the patients.

Night blood smear examination was not done due to inconvenience and with awareness that almost all patients had received multiple doses of diethylcarbamazine before approaching us. History of treatment for cancer—either of the breast or genital organs—was easy to obtain as also of ancillary reasons for swelling like heart, liver, and kidney disease. Other important correlations included multiple sclerosis, stroke or paralysis, rheumatoid arthritis, and a history of trauma. Congenital origin could be correlated to age of onset or a clear-cut syndrome like Klippel-Trenaunay. Venous origin was also related to varicose veins, ulcers, history of deep venous thrombosis (DVT), or occupations which required prolonged immobility. Females without obvious causation as above were classified as lymphedema praecox or tarda depending on age of onset.

Evidence of past or present acute dermato-lymphangioadenitis (ADLA) was searched for. ADLA is an inflammation of the lymphatic vessels, often extending to the adjacent skin and draining nodes [9]. We made a diagnosis of ADLA if patients presented with a history of localized pain, with or without fever, or, if on examination, patients had features of lymphangitis, cellulitis, or local lymphadenitis. On examination, we also made a note of fungal infection, ulcers, fissures, thickening, rough and course skin, verrucal changes, papillomatosis, non-pitting edema, and nodularity. Photographs were taken of affected limbs, after consent, before and after therapy. Counseling was offered to all patients before starting treatment. Counseling included the treatment plan and a discussion of the prognosis. There was a strong emphasis that continued self-care at home is essential to inhibit disease progression.

Limb Volume Measurements

Lower Limb Circumference measurement at fixed points was carried out on both sides. The patient was asked to lie down, and the foot was arranged at an exact right angle to the leg, using a specially designed foot board. We marked the ankle at the corner as point zero, and then took circumference measurements in both directions from this reference point (using the posterior and plantar surfaces of the leg and foot). The shape of the foot at the ankle, especially in a patient with lymphedema, precluded a proper measurement at 5 cm away from the reference point. Consequently, our first circumference measurement was at 10 cm on either side, followed by measurements at further 5-cm distances until the metatarsophalangeal joints distally and the perineum above. Typically, the last measurement point would be 15 cm from the ankle distally, and 65-80 cm proximally. Except for the readings immediately adjacent to the ankle, the limb was treated as a set of stacked cylinders, and the calculation was carried out on software devised by us [10]. The following formula was used: Limb Volume = $\Sigma V = \Sigma \pi r^2 h$ where $r = C/2\pi$ with h (mostly 5) as the height having circumference Cat the middle of the segment. Each 10-cm part of limb immediately adjacent to the ankle was considered a 3/4 cylinder, and the one between 10 and 15 cm on the foot side a half cylinder.

Upper Limb The Upper limb was measured in essentially the same way. The first measurement was taken 10 cm from the tip of the middle finger. Subsequent measurements were taken at 5-cm intervals, all the way till the shoulder.

Limb measurement after treatment was also recorded to document "Volume Change after Treatment" (VCT). We first calculated the "Estimated Normal Volume" (ENV). If the contralateral limb was normal, then its volume was the ENV. In situations where the normal limb could not be measured, e.g., patients with bilateral limb edema, the ENV was determined from the patient's weight and height, using a nomogram previously developed and validated by us. The calculated volume by this method differs from the actual volume by 7.32% [10].

VCT was calculated as follows:

VCT = [Initial Volume(IV) - Current Volume(CV)]/ [IV - ENV], expressed as a percent. Thus, if CV has reached ENV, then the VCT is 100%, and if CV is same as IV, then the VCT is 0%.

Investigations

Routine hemogram and urine was prescribed. If fungal infection was suspected and found on examination between the skin folds or toe webs, a swab was sent for Potassium Hydroxide (KOH) stain looking for fungal mycelia. A baseline lympho-scintiscan was prescribed for all cases, but affordability by the patient remained an issue. The presence of patent lymphatics and active draining nodes indicated that we could exercise the option of a nodo-venous shunt procedure. An ultrasound Doppler study was carried out in patients in whom we suspected the possibility of deep venous thrombosis.

Non-operative Treatment

Cleaning was carried out meticulously with soap and water, as described in a regime published in a World Health Organization document [11]. We paid particular attention to cleaning and drying the toe webs. If fungal infection was suspected—with or without a positive stain—ketoconazole powder was additionally prescribed.

Antibiotic Therapy

Antibiotic therapy was initiated in the form of intramuscular benzathine penicillin 1.2 megaunits for all patients who had a history or findings consistent with ADLA. Continuation of benzathine penicillin once every 3 weeks was advised for at least 12 months or more till the end of the next monsoon. These injections were considered prophylactic in nature, though the first shot did have therapeutic effects. In case ADLA attacks persisted, penicillin was advised for an indefinite duration. The reason for insistence of prophylaxis during the monsoon months was the observation of higher occurrence of ADLA during these months.

Comprehensive Decongestive Therapy (CDT)

CDT is the worldwide standard for lymphedema management. It is yet to become commonplace in India because of lack of trained personnel, costs, and poor tolerance of a bulky bandage in hot and humid environments. We have largely overcome these constraints with extensive counseling regarding self-care.

CDT was carried out in two phases: hospital and home [12]. The hospital phase typically used to last 7-10 days, during which time the limb volume reduction stabilizes. The patient and accompanying relatives were trained during this time so that the same treatment can continue at home. Such care is duly explained to be last lifelong.

Compression in CDT

Our main method of compression was multilayer lymphedema bandaging (MLLB). If patients could afford it, intermittent pneumatic compression (IPC) was also used. The original pump was the single-chambered pneumatic compression pump VIPEL® [13]. Over time, we introduced sequential pumps of four or eight chambers.

Multilayer lymphedema bandaging (MLLB) was done by our therapist team but taught well to the patients so that they could continue self-care at home. MLLB was carried out in the following layers (skin care and cleaning being a pre-requisite):

- 1. Lightweight, non-compressive tubular stockinette. This only had an absorbent role since it was placed directly on the skin.
- Non-adhesive open-weave gauze bandages for the toes or fingers only.
- 3. Soft roll padding bandage. These ensured even distribution of pressure from the (next) short-stretch bandage layer. They were specially required to protect the skin over the bony prominences such as the malleoli. Additional foam pads were used in difficult fibrotic areas to soften the fibrotic tissue. They filled skin folds and concavities around the malleoli to ensure a nice cylindrical shape to allow even application of pressure by the next layer.
- 4. Short-stretch bandage layer. This was the final layer of bandaging. The bandages were specially made to provide higher working pressure and lower resting pressure.

MLLB application continued when the patient was off pump. We advised patients to always use MLLB, day or night, and especially when exercising.

Manual lymphatic drainage (MLD) [14, 15] has been described as a key component of the CDT protocol. However, we did MLD very infrequently. Our protocol of providing training to the patient and relatives on cleaning and bandaging, along with stoppage of MLD, meant that we had largely removed the distinction between the hospital-based initiation phase of CDT and the maintenance phase at home. This has led to a faster turnaround time for patients. Some patients who had learnt the art of tying the bandage as well as purchased a pump became ready to go home as early as 2 days after onset of treatment.

The CDT protocol was as follows: CDT was started soon after the first shot of penicillin. This delay was necessary as there were concerns regarding flaring of active infection. We also examined the peripheral arteries and veins, confirming that the ankle to brachial pressure index (ABPI) was >0.8 and that the patient did not have acute DVT. Further care was conducted in two phases, hospital and maintenance, as follows:

- 1. Phase 1 (hospital phase)
- 2. Meticulous skin/nail care through washing with soap and water. Occurrence of fungus between the toe webs and skin folds required meticulous drying. Sponging or even a soft brush was used to remove dust between the vertucae.
- 3. Compression (described below).
- 4. Exercise advice: calf muscle exercises (standing on toes and heels, circular movements of toes) while wearing the compression bandage or garment.
- 5. Self-care education.
- 6. Phase 2 (maintenance phase)
- 7. Skin care
- 8. Compression
- 9. Exercise

Surgery

Some patients required surgery, in the form of nodo-venous shunt, or debulking surgery, or both.

A nodo-venous shunt [16] was fashioned in patients who had visible nodes on lympho-scintiscanning to hasten volume reduction. This was specially required for those in whom debulking surgery was planned.

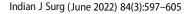
Debulking surgery was performed in patients with large folds, large verrucae or warts, or areas of fibrotic skin. For all cases undergoing debulking, a preliminary volume reduction was attempted. Even though CDT alone could achieve reduction of excess fluid, a preceding shunt made it quicker. A dry limb during debulking surgery helped primary closure of flaps, with better post-operative recovery. There was less oozing and drainage with maximum reduction of limb size. The surgical technique for debulking was a simple excision of excess fibrotic skin and subcutaneous tissue. Careful planning of the incisions was done, to allow the skin to be closed primarily and avoid grafts.

In post-discharge, with or without surgery, patients were advised to continue self-care through CDT and regular followup.

Figure 1 describes our management plan.

Follow-Up and Revisits

After the early return, patients were encouraged to remain in touch by phone and sometimes a video call to ensure compliance and manage minor problems. Most physical revisits however were forced by a problem—generally in the form of ADLA. Decrease in number and, in many cases, complete absence, of ADLA attacks was one of the most significant evidence of response to treatment.



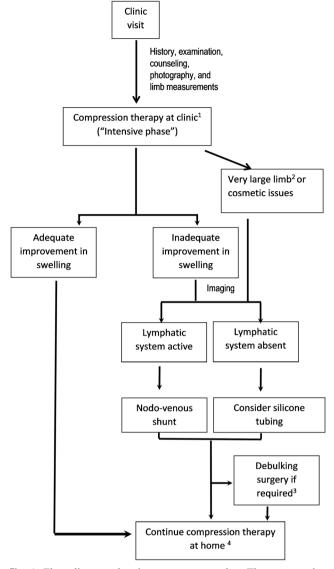


Fig. 1 Flow diagram showing management plan. The compression regime was pump plus MLLB.¹ In these cases, we did not expect improvement without surgery.² Need for removal of fibrotic areas, loose flabby skin, overhangs preventing proper bandaging.³ MLLB with or without IPC.⁴ Compression garments were prescribed to those patients whose limb had achieved a stable limb size with uniform shape

Data Collection and Statistical Methods

Data was collected from our electronic medical records system (Medic AidTM) and exported to Microsoft ExcelTM. We reviewed all patients who underwent treatment for limb lymphedema at our clinic. A telephone call was made to all the patients to determine subjective improvement.

Results

We reviewed patients who underwent treatment for limb lymphedema at our clinic. For the purpose of this study, we have not included patients with inaccurate readings, scrotal or abdominal swelling, patients treated during field visits, and patients who declined treatment or could not come for follow-up. The final selection was restricted to those patients whose comparative volumetric data was available, i.e., of the affected limb as well as an ENV.

Demographic Details

The current data includes 93 patients (39 males) with 109 affected limbs (Table 1). The average age was 46 years, range 20-77 years. The mean upper limb volumes at presentation were 3644 ml, and the mean lower limb volumes were 11,279 ml (Table 2). Almost 40% of our patients were residents of Delhi. A quarter came from Haryana, Maharashtra, and Uttar Pradesh. The remaining patients came from Rajasthan, Madhya Pradesh, and other distant parts of India. Ten of the 11 upper limb edema patients were related to breast cancer and one was filarial. Among lower limb patients, four had history of cancer in the reproductive organs and 22 came from filaria endemic zones. Three were obviously congenital-related to time of onset as well as associated deformities. Five had venous disease, and three had dyslipidemia. We classified the remainder as idiopathic. At least 60% of patients with lower limb edema and 73% of patients with upper limb disease had documented ADLA at presentation.

Use of Compression Pump

Of the 109 limbs, we could use a long-term compression pump in only 69: the remaining patients were unable to afford the pump.

Results of Treatment

The average VCT after medium-term follow-up was 53% for 8 upper limbs and 50% for 46 lower limbs for which records were available. The average VCT after long-term follow-up was 52% for 9 upper limbs and 49% for 82 lower limbs for which records were available.

Table 1Limbs involvedin 93 patients	Limb	Number
	Both legs	16
	Left arm	4
	Left leg	46
	Right arm	7
	Right leg	20
	Total	93

Limb volume improved significantly after bandaging or bandaging plus compression (Table 2).

To determine if the use of the pump gave superior results, we divided the patients with lower limb edema into two groups—bandaging only (29 patients) and bandaging plus pump (46 cases)—and compared the long-term volume fall in each group. The mean volume fall was 1749 ml in 29 patients when a pump was not used and 1769 ml in 46 patients when it was used. There was no difference between the patients managed with a pump and those managed by bandaging alone (independent samples *t* test *p*=0.483).

Worsening was seen in two lower limbs after medium-term follow-up and in three lower limbs at long-term follow-up. In all cases, we observed that continuing ADLA or noncompliance with compression was a factor.

Data Availability

An Excel file with all the measurements is available at the following link: https://doi.org/10.7910/DVN/IXLEUO.

Discussion

The worldwide prevalence of lymphedema is between 200 and 300 million [17]. The commonest cause is cancer, especially cancer of the breast [2], but there is a range of other causes like venous disease, old age, obesity, and trauma. In India, filariasis is an important cause of lymphedema. India bears 40% of the global prevalence of LF. Affected individuals together lose over one billion working days every year because of chronic disease [18]. The social impact too is severe. It often affects marriage prospects [19]. In severely affected areas like Odisha and Bihar, the groom's party often examines a prospective bride's legs before accepting a proposal! The areas with the maximum prevalence of filariasis are Bihar (affecting over 17% of the population), followed by Kerala (15.7%) and Uttar Pradesh (14.6%) [20]. Our own patients came largely from Delhi and surrounding areas, including Haryana. Bihar contributed the next highest number with a large proportion from filarial endemic districts.

Diagnosis of Lymphedema

Filariasis was the commonest cause of lower limb edema. However, microfilariae are rarely found in patients with leg lymphedema [21]. Since almost all patients had received multiple doses of diethylcarbamazine before approaching us, and considering the 6- to 8-year life span of the worm [22], night blood smear was not examined. The diagnosis of the cause of the lymphedema was based on clinical features and on the geographical origin of the patient. We diagnosed filarial lymphedema based on geographical considerations, lack of **Table 2**Limb volumes atpresentation and on follow-up

Limb	Average initial limb volume	Average limb volume on medium-term follow-up	Average limb volume on long-term follow-up
SD =	11279	8827	9086
	SD = 6812	SD = 4070	SD = 5038
	<i>N</i> = 98	<i>N</i> = 46	<i>N</i> = 77
		t = 4.7, p = 0.00002	T = 8.851, p = 0.00001
Upper	3644	3211	3211
	SD = 889	SD = 492	SD = 895
	N = 11	N = 8	<i>N</i> = 9
		t = 3.8, p = 0.006	T = 4.9, p = 0.001

All values are in milliliters. SD = standard deviation; N = number of limbs in which readings were taken. "t" represents the values of t for a dependent samples two-tailed t test when compared with the initial volumes. "p" is the probability that there is no change when compared to the initial volumes at N-1 degrees of freedom. The statistical tests were calculated online at https://www.socscistatistics.com/tests/ttestdependent/default2.aspx

an obvious associated cause like cancer, and after excluding primary lymphedema on clinical grounds. Most of our lower limb cases outside Delhi and the nearby states (Haryana, Punjab, and Rajasthan) have been assumed to be of filarial. History of filaricidal drugs was elicited for patients coming from endemic areas and a course of DEC along with ivermectin or albendazole prescribed if a doubt existed. All except one of the upper limb lymphedema patients had breast cancerrelated disease.

Antibiotic Therapy

We favor the use of penicillin for preventing recurrent lymphangitis. ADLA is responsible for disease flare-up [23, 24]. Among our patients, recurrences were largely related to continuing ADLA. This occurred in patients who stopped penicillin, developed fungal infection, and/or were unable to continue compression. Clinicians use a variety of antibiotics, including cephalosporins, doxycycline, aminoglycosides, cotrimoxazole, lincomycin, and ciprofloxacin in acute infection [25]. However, in immunocompetent individuals, group A beta-hemolytic streptococci are the commonest causes of lymphangitis [26]. Penicillin is always effective against community-acquired streptococcal infection. It is inexpensive and prevents recurrent lymphangitis [23, 24, 27]. Benzathine penicillin 1.2 megaunits are conveniently administered every 3 weeks. This is our choice for long-term ADLA prevention.

We always perform a sensitivity test for penicillin and have found it to be extremely safe to use. A delayed generalized rash occurred in 7 of our patients. This included 2 of the current 93 and both are in the list who continued to have ADLA. They improved to a lesser extent. In patients where the test indicates sensitivity, we used to prescribe ciprofloxacin and lately azithromycin. Among others, after over 10,000 shots, we have not had any incidence of anaphylaxis. Other than the rash, only two adverse events were observed. One was a vasovagal reaction. The other patient had severe nausea which settled down with anti-emetics, probably because of inadvertent intravenous administration. This patient continued to receive her 3-weekly dose subsequently after a repeat sensitivity test. Serious anaphylaxis to penicillin is rare [28]. We have observed that the incidence of penicillin-related anaphylaxis has fallen drastically with the advent of disposable syringes.

Compression Therapy

Compression is the mainstay of lymphedema care. Starting with the UNA boot [29], which was used for ulcers, compression techniques have been continuously evolving. Knowledge that it is effective in edema came later and underscored the complex relationship between edema and ulcers [30]. Compression is a key component of CDT [31] with some believing that this is the only component that works [12]. Reduction of fluid is important not only for cosmetic reasons but also infection prevention: static fluid is a good culture medium! Imaging has shown that fluid return is enhanced though MLD [32]. Compression works to reduce the edema by squeezing the limb, aiding fluid return towards the heart. The rise of tissue temperature with its anti-inflammatory effect has also been surmised as one of the reasons for positive effects of compression [33, 34].

Commonly used compression equipment includes bandaging, compression hosiery, and IPC [29]. In general, bandages are most commonly used for treatment initiation, and stockings for maintenance. Stockings alone may be used in those with minimal disease, especially in chronic venous cases. Pumps can be an accompaniment for both. As shown by our results, pumps offer little benefit over a good and properly applied compression bandage. However, many still preferred it for the convenience.

It is difficult to compare our results with those achieved by others. Reports on the outcomes of arm lymphedema (in women with breast cancer) are published in large numbers [35, 36]. Reports on the outcomes of lower limb lymphedema treatment are almost nonexistent. Badger et al. [37] report a mean limb volume reduction of 31% at 24 weeks in patients treated by MLLB and hosiery. Their 34 cases included more patients with upper limb disease than lower limb disease. In contrast, almost 90% of our cases had lower limb edema. Among lower limbs for which a medium-term reading (<180 days) is available, the mean volume dropped by 25% from 11,871 to 8827 ml. For limbs in which a long-term volume reading is available, the mean dropped by only 18%, from 10,399 to 8505 ml. This attenuation in long-term improvement is not unexpected: for poorer patients, the ability to comply with antibiotic and compression therapy can be challenging. Among our patients with lower limb edema, the average initial limb volume was 11,280 ml, about twice the average expected volume (6415 ml). The average last recorded limb volume was 9239 ml, still about 30% more than the expected limb volume. And while the reduction in volume appears to be modest, it is important to remember that once lymphatic block occurs, the edema is likely to be permanent, and it is futile to expect a normal limb (Fig. 2).

Surgery

Surgery in lymphedema should be done with clear-cut objectives. Flow-enhancing procedures are recommended but not

Fig. 2 Photographs of a patient showing progress after compression therapy and surgery. This patient came with a request that he should be able to wear shoes so that he could get married. After CDT, nodo-venous shunt, and two sessions of debulking, he was able to wear shoes, tight jeans, and marry. The last photograph is 4 years post-procedure. He has kindly provided written permission to publish his photographs

always possible. Lymphoscintigraphy may indicate which patients are likely to respond to flow-enhancing procedures. The lymphatics are often dilated, but dermal backflow is rarely seen. The extra dye is seen scattered all over the limb and indicates non-progression. In our study, we have done a few nodo-venous shunts. Lately (not during the study period), we have been performing direct lymphaticovenous shunts. The latter requires the lymphatic vessels to be visible (helped by dye injections during surgery).

Newer techniques of interest to the plastic surgeon use super-microsurgery like direct end-to-end lymphaticovenous anastomosis [38] and node transplants. These depend on availability of good quality lymphatics visualized during surgery [39]. Unfortunately, most of our patients were in the advanced stage, with few lymphatics suitable for anastomosis. If flow enhancement surgery was deemed necessary, like the congenital or post-malignancy cases, we have started the use of a silicone tube implant [40], but it is too early to comment on long-term outcomes.

The aim of debulking is to neaten and remove excessive skin folds and fibrotic areas. Fibrotic areas are resistant to compression therapy, while skin folds and warty areas inhibit appropriate bandage or garment application. These areas tend to retain as well as re-accumulate fluid. It is important to make sure there is minimal fluid in the operative field by prior elevation, compression, as well as flow-enhancing procedures [41]. Such a dry limb allowed early discharge of the patient without flap loss or wound seepage. Our surgical technique for folds as well as warts is simple excision with only direct closure.



Verrucae make it difficult to keep the limb clean. Thorough scrubbing helps, but not always. If verrucae are present and outside the areas marked for excision, a surgical tangential excision of the involved skin, similar to the procedure for a burn eschar, helps. However, we never go deeper than the dermis. Re-epithelization is spontaneous without the need for a graft.

Limbs with Poor Outcomes

There were 5 limbs with worsening. All worsening could be linked to recurrence of ADLA attacks. We tried to identify the factors contributing to continuing ADLA. These were lack of penicillin and poor home-based maintenance of CDT. Recurrence of fungal infection in the webs was an indicator. Some patients stopped penicillin, either because they could not procure it or there was no one to administer the injection. Some patients were unable to continue compression because the bandage wore out and they could not arrange a replacement, and they were unable to obtain a working compression pump.

Overall, the number of failures has been low mainly because of our efforts at counseling. Before coming to us, the reported number of attacks varied between zero (no attacks) and almost continuous fever. Due to low number of cases with long follow-up, assessment of repeat episodes of ADLA was difficult. Through a telephonic survey by our team recently, we could contact 28 of these 93 patients. There has been the drastic fall in incidence of ADLA from an average 3–4 per year before treatment to none (19) and occasional (6) after treatment. Among the 3 who had more than one attack, one was among those marked as sensitive to penicillin. Based on survey and physical follow-up, we estimate that there has been a decrease of ADLA between 50 and 95%.

Conclusions

Lymphedema needs to be treated as a chronic problem. It requires long-term continued care, which is possible even in a clinic setting of a developing country. Even while initial causes may differ, the clinical course in later stages is similar, regardless of etiology. There is a vicious cycle of fluid accumulation and poor limb hygiene leading to attacks of lymphangitis and further lymphatic blockage. Treatment consists of breaking this cycle at appropriate points by good hygiene, appropriate compressive therapy, and prophylactic antibiotics. Good counseling and training the patient on self-care is critical to good outcomes.

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Declarations

Ethical Approval The authors have adhered to the required ethical standards.

Consent for Publication Written consent has been obtained from the patient whose photograph has been used in the paper.

Conflict of Interest The authors declare no competing interests.

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