

## Research Article

# A clinical study of lymphedema management

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### ABSTRACT

**Background:** Lymphedema is chronic disorder of the lymphatic system characterized by impaired lymphatic return resulting in swelling of the affected area. Though it is an important cause of morbidity, epidemiology and impact of this condition on patients or health services is only modestly known. Management of lymphedema primarily aims at reducing or halting progression of swelling and to prevent associated infection. A combination of skin care with moisturizers and emollients, compression bandages, isotonic exercise and regular massage are used in treatment of the disease. The notion of using external physical means to incite lymphatic drainage has a sound physiological basis. However with currently available poor evidence of relative effectiveness of methods it is improbable to rely on a single technique alone.

**Methods:** 33 patients presenting with lymphedema in department of plastic surgery, RRMCH, were included in the study. Patients with congenital lymphedema were excluded from the study group. Therapy was individually tailored for each patient. Patients were evaluated for the efficacy of the treatment initiated at 1, 3, 6 and 9 months. Treatment methods included complex decongestive physiotherapy (CDP), use of elasto-compressive bandages, and multi-layered lymphedema bandages (MLLD) along with limb elevation, meticulous skin care and remedial physiotherapy.

**Results:** Of the 33 patients, 12 were males, 20 females and one third gender. 11 patients had upper-extremity lymphedema, 21 patients had lower-extremity lymphedema and 1 patient had scrotal lymphedema. Patients underwent Conservative treatment alone, Pneumatic Compression, Complex Decongestive Physiotherapy (CDP) or Miller's Debulking Procedure. Both volumetric assessment and patient satisfaction were considered to assess the outcome of treatment. Necessary changes were done in the treatment thereafter.

**Conclusions:** Lymphedema is a debilitating disease with both physical and mental implications on the patient. The management should involve the patient too. The psychological aspect of the patient should also be addressed. Treatment should be tailored and changes be made appropriately as per requirement. With no current definitive therapy, a holistic approach is to be employed.

**Keywords:** Lymphedema, Chronic oedema, Complex decongestive physiotherapy, Manual lymph drainage, Treatment

### INTRODUCTION

Lymphedema is a malady of the lymphatic system due to low output failure resulting in abnormal collection of protein-rich fluid in the interstitium. Lymphedema incidence is varied around the world. In developed countries, the bulk of the cases are due to cancer and obesity.

Worldwide, filariasis in endemic areas is the most common cause. The incidence of primary lymphedema is 1 of 10,000 general populations, with females accounting to 70-80% of cases.<sup>1</sup>

In primary lymphedema, there is congenital hypoplasia or aplasia of lymphatics or valvular incompetence. Types include Milroy disease, Meige disease and Lymphedema

tarda. Lymphatic drainage is impeded by an acquired blockade or by disruption of the local lymphatic channels, in secondary lymphedema. This may be due to filariasis, malignancy, recurrent attacks of lymphangitis, obesity or surgery. Primarily lower extremity is involved (80%) but upper extremities, trunk, face and genitalia can also be affected.

Because of increased protein content in extravascular tissue, there is subsequent retention of water with resultant soft tissue swelling. Ensuing fibroblast proliferation and fluid organization results in non-pitting swelling of the affected region. Recurrent episodes of cellulitis, lymphangitis, fissuring, ulcerations, and verrucous changes occur with bacterial or fungal infections due to suppression of local immunologic surveillance. This has severe detrimental effects on patient's daily activities. Fissuring causes leakage of lymph onto the surface of the skin (lymphorrhea). Also, recurrent episodes of ulceration and healing may stimulate keratinocyte proliferation which may result in neoplastic transformation. Chronic lymphedema patients have a 10% risk of developing lymphangiosarcoma. Squamous cell carcinoma, Kaposi's sarcoma and malignant fibrous histiocytoma are also known to occur.<sup>2</sup>

The rationale of lymphedema therapy is palliative with intent to prevent disease progression rather than cure. Current treatment strategies are disappointing and are difficult to reproduce the desired results. Patients have to endure a lifelong physical therapy with manual lymphatic drainage and uncomfortable elastic garments with high degree of non-compliance. With the current knowledge of pathophysiology and progress in the field of molecular biology, which facilitates us to understand lymphostasis consequence in the form of morphologic and functional alterations in tissues, a rational therapy needs to be formulated. Also there is a need for implementation of effective preventive treatment strategies for susceptible patients.

Early surgical procedures aimed to reduce limb volume by debulking resection approach. The present advent of microsurgery has use of lymphatic-venous anastomoses which is being performed in select few centers.<sup>3</sup> Though ideologically convincing the success of it has not been demonstrated equivocally. Hence, both the physician and patient opt for conservative treatments, notably in early stages of lymphedema.<sup>4</sup>

Complex decongestive physiotherapy (CDP) constituting of manual lymph drainage (MLD), skin care, compression, and remedial exercises to maintain lymphatic flow has become a common treatment for lymphedema worldwide.<sup>5</sup> CDP aims at mobilizing edema fluid and initiate halting progression, at the least, or regression of fibrosclerotic tissue alterations. It also prevents reaccumulation of edema fluid and scar tissue breakdown. CDP is often complemented with diuretics. Limb sleeves or stockings and pneumatic pump devices

are commonly employed for the treatment. In most severe cases, surgical debulking procedure may be required to alleviate the symptoms of lymphedema and to improve the quality of life. But patients have to be aided to live with the disease because an all-effective therapy is lacking.

## METHODS

Thirty three lymphedema patients were studied prospectively in Department of Plastic Surgery, Raja Rajeswari Medical College and Hospital, Bangalore with prior approval of the institutional ethical committee. The aim of the research was to study lymphedema clinically and institute treatment accordingly and the outcome of same. Patient consent for the study was taken. Only patients with secondary lymphedema were included in the study.

All of these patients were referred to us from auxiliary departments at our institution and some presented directly. Complete medical evaluations to rule out other causes of swelling were done. Doppler venous scan to rule out deep venous thrombosis was done in suspected cases. Clinical proforma was designed for data collection. Clinical grading was done as per Brunner's as follows:

- I. *Grade 1* - oedema pits on pressure & largely disappears on elevation & bed rest
- II. *Grade 2* - oedema does not pit & does not significantly reduce on elevation
- III. *Grade 3* - oedema is associated with irreversible skin changes, i.e., fibrosis, papillae formation, hyperkeratosis.

In cases of extremity lymphedema, volume assessment was done by submerging each arm in a container filled with water, and measuring the volume displaced.

The therapy was individually tailored as per clinical requirement and patient's willingness for the treatment protocol. Patients were evaluated for the efficacy of the treatment initiated at 1, 3, 6 and 9 months with minimum follow-up of 9 months. Treatment methods included complex decongestive physiotherapy (CDP), pneumatic compression, use of elasto-compressive bandages, multi-layered lymphedema bandages (MLLD) and limb elevation. CDP involved 2 phases – Phase 1 included 3 weeks of daily therapeutic sessions with manual lymph drainage, multilayered inelastic compression bandaging, and meticulous skin care by physiotherapist, followed by Phase 2 wherein the patient performed MLD oneself or with help of family members. Limb reduction surgery we employed for limb debulking was the Miller's procedure. Volumetric reductions before and after treatment initiation were assessed at regular intervals.

Statistical analysis was done using SPSS software.

## RESULTS

The results of our study are as follows.

### 1. Gender distribution

12 patients were males, 20 females and 1 was a third gender (Table 1).

**Table 1: Gender distribution.**

	No. of Patients	Percentage
Males	12	36.4 %
Females	20	60.6 %
Third Gender	1	3 %



**Figure 1: A case of lower extremity lymphedema in stage 3 with skin changes and excessive lymphorrhea.**

### 2. Age distribution

Majority of the patient were in 3<sup>rd</sup> to 6<sup>th</sup> decade. The age of the treated patients ranged from 17 to 64 years (Table 2).

**Table 2: Age distribution.**

Age in Years	No. of Patients	Percentage
0 – 10	0	0
11 – 20	1	3 %
21 – 30	7	21 %
31 – 40	9	27 %
41 – 50	11	34 %
51 – 60	4	12 %
61 – 70	1	3 %

**Table 3: Region of the corpus involved.**

	No. of Cases
Upper Limb	11
Lower Limb	21
Others	1

### 3. Region of the corpus involved

11 patients had upper-extremity lymphedema, 21 patients had lower-extremity lymphedema and 1 patient had lymphedema of the genitals (Table 3).



**Figure 2: PreOp pics (A) before CDP initiation and (B) after CDP for 1month.**

### 4. Previous treatment history

15 patients had previously undergone treatment using pneumatic pumps and elastic garments before presenting in our department. These patients were not satisfied with treatment outcome obtained from previous physicians.

### 5. Lymphedema etiology

Lymphedema in 11 patients with malignancy was due to disease, post-surgical and post-radiotherapy. Post modified radical mastectomy, 7 patients and post radiotherapy 4 female patients had lymphedema. Lymphedema due to parasite (Filaria) and fungal infections was seen in 10 patients. Patients with prior fungal infections were being treated by dermatologists. Trauma as the cause for lymphedema was noted in 4 patients. Excluding other causes of lymphedema, obesity was found to be etiology in 3 patients. The BMI of all these were >34. There were 4 cases of lymphedema in patients who had undergone inguinal block dissection. One patient had lymphedema post varicose vein surgery (Table 4).

**Table 4: Etiology of lymphedema.**

	No. of Patients
Malignancy	11
Filariasis	5
Infection	5
Trauma	4
Obesity	3
Surgery	4
Others	1

### 6. Stage of presentation

9 patients presented in Stage 1, 14 patients in Stage 2 and 10 patients in Stage 3 (Table 5).

**Table 5: Stage of presentation.**

	No. of Patients
Stage 1	9
Stage 2	14
Stage 3	10



**Figure 3: PerOp pics showing rising of skin flaps and excision of subcutaneous tissue and skin.**

### 7. Patient complaints

Patient complaints ranged widely. Features included chronic swelling of the affected part, discomfort, lymphorrhea, reduced quality of life, social stigma, loss of employment and psychological trauma including distress, depression and suicidal tendencies. Stage 3 patients were the worst affected with patients having psychological trauma.



**Figure 4: Immediate post-op pic after skin closure; PostOp pic 15 days after surgery.**

### 8. Treatment

All patients underwent auxiliary treatment with elastic hosiery, limb elevation and skin care. Exercises were inculcated in the treatment appropriately based on patient compliance. Diuretics were used in 4 patients. 17 patients needed psychological support from Psychiatric

consultation. Weight loss regime was also instituted in patients with obesity as the cause of lymphedema.

3 patients on pneumatic compression were shifted to CDP during treatment course and 4 patients initially on CDP underwent debulking surgery. One patient with scrotal lymphedema underwent debulking procedure (Table 6).

**Table 6: Treatment.**

	No. of Patients
Conservative alone (Limb elevation + Compression bandages + Physiotherapy)	4
Pneumatic Compression	8
Complex Decongestive Physiotherapy (CDP)	21
Miller's Debulking Procedure	7



**Figure 5: A case of scrotal lymphedema (A) before and (B) after debulking procedure.**

### 9. Treatment outcome

Two patients were lost for follow-up during the course of this study. Both these patients had Stage 3 lower limb lymphedema and were on CDP. These patients were not satisfactory with the treatment outcome from start. One upper extremity patient with post mastectomy and radiotherapy lymphedema expired at 7<sup>th</sup> month of study due to disseminated metastasis. Along with subjective assessment, volumetric assessment of the extremity was done based on fluid displacement. Based on both parameters treatment protocols were adequately changed accordingly (Table 7).

**Table 7: Patient satisfaction.**

	Patient Satisfaction				Total
	Poor	Average	Good	Excellent	
1 Months	3	13	14	3	33
3 Months	3	10	14	6	33
6 Months	2	9	17	3	31
9 Months	3	15	10	2	30



**Figure 6: A case of upper extremity lymphedema in patient who had underwent mastectomy and radiotherapy for right sided breast carcinoma.**

## DISCUSSION

Lymphedema is a chronic oedema with devastating implications. It is a poorly recognized problem by health professionals and lymphedema due to non-cancer treatment are more prevalent than generally acknowledged. Although rarely lethal, lymphedema is a disfiguring and disabling condition with an associated decrease in the quality of life. Though the etiological factors for lymphedema have been identified there is lack of clarity in preventative strategies. Lymphoedema can occur at any age. The gender difference, as in many studies, cannot be explained purely by arm oedema related to breast cancer.<sup>6</sup>

Acute complications of lymphoedema will cause disruptions to daily life, and significant deficits in quality of life. Also the emotional consequence of lymphedema has to be addressed. Also the disease affected their ability to work.

Advantages associated with MLD have been reported but yet, methodological inconsistencies between studies persist and conflicting reports of the effectiveness of MLD confound problem.<sup>7,8</sup>

Surgical treatment does not obviate the need for continued medical therapy. Surgical intervention is needed in cases who do not improve with conservative measures. Initial reductions in volume achieved are maintained in the majority of the treated patients by conservative measures.

Recent impressive expansion of knowledge in molecular mechanisms of lymphangiogenesis provides new possibilities for treatment of lymphedema. Lymphangiogenesis can be stimulated by various cytokines, such as vascular endothelial growth factors (VEGF) C and D which in turn activate the VEGF receptor-3 (VEGFR-3).<sup>9,10</sup> Also angiopoietin-1 has been found to promote lymphatic vessel formation and that fibroblast growth factor 2 stimulates lymphatic vessels growth.<sup>11</sup>

Therefore, given the limitations in our knowledge and the important implications for patient care and quality of life, additional research is clearly required. However with the current treatment approach, results of treatment depend on the experience of the physician in clinical lymphology, on the training and dedication of the lymphedema physiotherapist and nevertheless on patient compliance.

## CONCLUSION

Lymphedema is a debilitating disease with both physical and mental implications on the patient. The management should involve the patient too. The psychological aspect of the patient should also be addressed. Treatment should be tailored and changes be made appropriately as per requirement. With no current definitive therapy, a holistic approach is to be employed.

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