



Genital elephantiasis: Surgical treatment and reconstruction

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Abstract

Genital elephantiasis is a severe form of lymphedema of the groin. It is characterized by progressive enlargement and distortion of the genitals, presenting significant physical, psychological, and social challenges to the affected individuals. Although pharmacological treatment of filariasis is well-established in the medical field, the surgical management of genital elephantiasis can be varied and confusing. This review article provides an in-depth analysis of the etiology, classification, severity grading, and various effective surgical treatment and reconstructive modalities commonly employed by surgeons since the early twentieth century. We also discuss how a combination approach of ablation, soft tissue coverage, and lymphatic reconstruction is viable for treating genital elephantiasis. By examining the literature, we hope to provide insights into how surgery plays a role in the holistic management of genital elephantiasis.

Keywords: Charles procedure; Genital elephantiasis; Lymph vessel transfer; Lymphatic reconstruction; Lymphedema debulking

1. INTRODUCTION

Genital elephantiasis has been classically thought of as a chronic, severe swelling, and hypertrophy of the genital soft tissue due to lymphatic filariasis. We now know that lymphedema of any etiology, congenital, or cancer-related, if left unchecked, will result in soft tissue hypertrophy, dermal hyperkeratinization, and eventual elephantiasis.

A fraction of patients with lower extremity lymphedema will suffer from genital lymphedema. However, the true incidence of genital lymphedema and elephantiasis is poorly understood. Recently, Clinckaert et al¹ performed a systematic review to determine the prevalence of lower limb and genital lymphedema after prostate cancer treatment. From the 18 articles reviewed, the group found a prevalence of lymphedema in the lower limbs and genital regions ranging from 0% to 14% and 0% to 1% after surgery and 0% to 9% and 0% to 8% after pelvic radiation, respectively. There was a much higher prevalence in patients that underwent pelvic lymph node dissection followed by pelvic radiotherapy (18%-29% and 2%-22%). For congenital lymphedema, a study of 138 children from Children's Hospital Boston found that males were seven times more likely to have genital lymphedema than females.²

Genital lymphedema and elephantiasis can impart significant physical morbidity to our patients. Patients may experience multiple episodes of cellulitis, difficulty fitting clothes, swelling discomfort, and malignant transformation from repeated ulcerations. This entity can negatively impact their psychosocial well-being as well. Patients often report lowered self-esteem and poor sexual function. Features of genital elephantiasis are listed and described in Fig. 1.

In this review article, we explore the management of genital elephantiasis with an emphasis on surgical treatment and reconstruction.

2. CLASSIFICATION AND ETIOLOGY

Lymphedema can be classified into primary and secondary lymphedema. Primary lymphedema is subdivided based on the age of onset:

1. Congenital lymphedema usually presents at birth or within 2 years of life. This entity has a strong familial pattern.
2. Lymphedema praecox occurs at puberty or the beginning of the third decade of life. The majority of primary lymphedema patients belong to this group.
3. Lymphedema tarda has an onset after 35 years of age.

Primary lymphedema is most commonly associated with a hypoplastic lymphatic system. Lymphatic collectors and lymph nodes are smaller and fewer in number, and this situation is frequently seen in lymphedema praecox patients. A complete absence of lymphatic trunks is associated with congenital lymphedema, whereas hyperplastic, tortuous channels are often seen in lymphedema tarda patients.

Secondary lymphedema results from an insult to the lymphatic system resulting in sclerosis and obstruction. Although the most common cause of lymphedema worldwide is filariasis caused by infection by *Wuchereria bancrofti*, in developed countries, most secondary lymphedema cases are due to malignancy or related to the surgical extirpation and adjuvant therapy of cancer.

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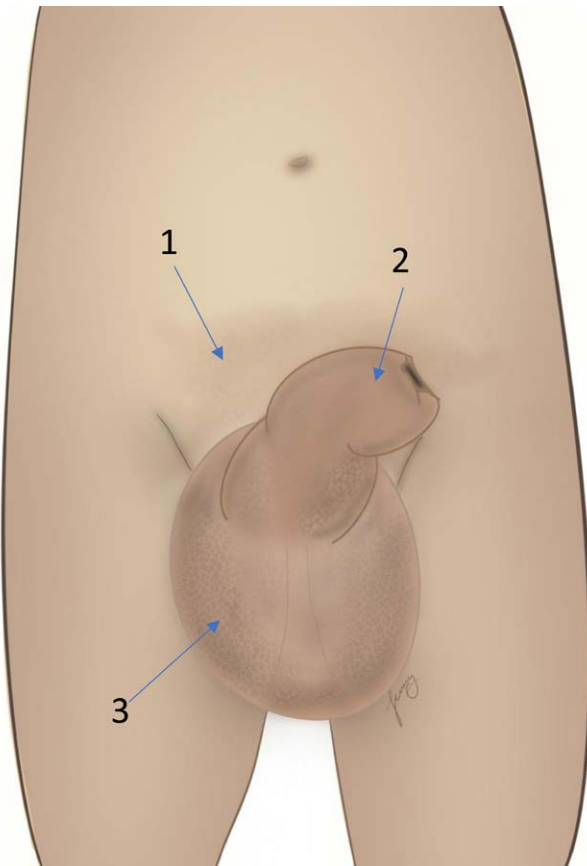


Fig. 1 Features of male genital elephantiasis are depicted in this figure. Suprapubic skin (1) thickness and undergoes verrucous hyperplasia. The prepuce can undergo edematous change and enlargement (2). This leads to hygiene problems and difficulties with urination. The penis can also become buried, which leads to the inability to perform sexual intercourse. The scrotal skin undergoes verrucous hyperplasia, and the testes enlarge due to severe hydrocele formation (3).

3. SEVERITY AND GRADING

There is no universally accepted reporting nomenclature for disease severity. Capuano and Capuano³ described a preoperative classification based on laterality, size of the hydrocele, and degree of buried penis. Although the extent of the deformity has some correlation with the severity of the lymphedema, this relationship is nonlinear. Therefore, it is difficult to ascertain the degree of lymphosclerosis and judge the type of lymphatic surgery needed.

Indocyanine green (ICG) lymphography has changed how clinicians evaluate extremity lymphedema and can be applied

to genital lymphedema. Patterns of dermal backflow such as splash, stardust, and diffuse are pathognomonic of lymphedema and can often allow for early diagnosis even before florid symptoms manifest.⁴ The utility of this severity staging method is the ability to discern which patients will benefit the most from physiological lymphatic procedures like lymphovenous anastomosis (LVA) before elephantiasis occurs.

The genital lymphedema score (GLS) was developed to evaluate genital lymphedema without ICG lymphography because these specialized near-infrared scanners may not be available in all medical settings.⁵ The GLS is based on subjective symptoms alone (Table 1) and has been demonstrated to correlate well with genital dermal backflow patterns and ICG lymphedema staging. The GLS can also be used as a follow-up measurement after therapeutic interventions for genital lymphedema.

4. ABLATIVE AND DEBULKING SURGERY

Excision of lymphedematous tissue is considered one of the oldest surgical treatments of lymphedema. In 1901, Sir Richard Henry Havelock Charles published in the *Indian Medical Gazette* a series of 140 consecutive patients treated successfully for scrotal lymphedema. This article was the basis for a 1912 book chapter on the same topic, whereby the eponymous “Charles procedure” was based on.⁶ Although Charles mainly described his procedure for scrotal elephantiasis, the idea of the Charles procedure would evolve into what is currently understood today; a form of surgical treatment where the lymphedematous lower extremity is radically debulked and then covered with skin grafts harvested from the surgical specimen. Charles procedure is still regarded as a treatment option for chronic and advanced lymphedema where skin hypertrophy and verrucous overgrowth have resulted in elephantiasis. Over the years, many have made modifications to the original Charles procedure. These include using split-thickness skin grafts, concurrent free vascularized lymph node transfer (VLNT), and delayed skin grafting with or without negative pressure wound therapy.^{7,8} Homan⁹ described a modification where a longitudinal incision is made along the lateral and medial aspect of the legs lifting the dermis off the fat and preserving the subdermal plexus. The underlying fat is removed, and the skin is trimmed and closed primarily to accommodate the reduced limb volume.⁹ Salgado et al¹⁰ refined this concept by utilizing microsurgical principles to preserve skin perforators and reduce the incidence of wound healing complications. The Charles procedure stood the test of time and is still valuable for a lymphatic surgeon’s armamentarium today.

Thompson¹¹ first described the buried dermal flap as a treatment for advanced chronic lymphedema of the lower limb in 1959 and subsequently published his 10-year series in 1970. Long dermal adipose flaps were raised and preserved in this procedure instead of discarded. The redundant end of the flap

Table 1
GLS system based on subjective symptoms related to genital lymphedema

Subjective symptoms	No	Yes
Sensation of heaviness	0	1
Sensation of tension	0	1
Swelling	0	1
Urinary troubles due to genital edema	0	2
Cutaneous lymphatic cyst	0	2
Genital lymphorrhea	0	2
Total = GLS	Range	0-9

GLS = genital lymphedema score.

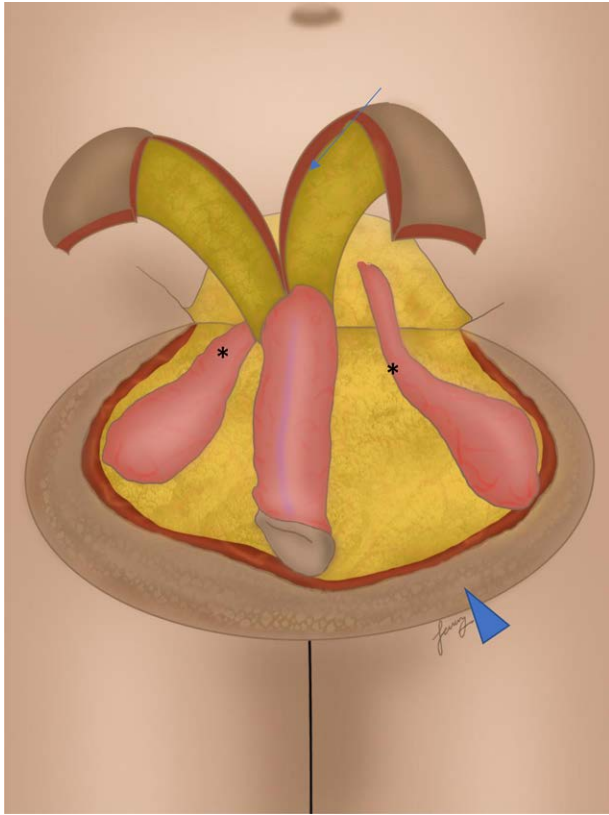


Fig. 2 Radical resection involves removing all the skin and soft tissue affected by elephantiasis. The penile skin is resected (blue arrow) above the Buck's fascia, preserving the dorsal neurovascular bundle. The spermatic cord (*) is carefully dissected and protected. All diseased scrotal skin is removed (blue arrowhead). Suprapubic skin may need to be resected if involved as well.

6.1. Elephantiasis tissue resection

The surgeon should aim to resect all pathological skin and subcutaneous tissue, as incomplete resection can lead to lymphocele and recurrences. Commonly resected areas include the scrotal skin, suprapubic skin, penile skin, and prepuce (Fig. 2.). Injury to critical structures, such as the dorsal neurovascular bundle, will be avoided if one stays on the suprafascial plane (above Buck's fascia). Care must be taken to isolate and protect the spermatic cords and the testes during dissection. An orchidopexy is performed to prevent torsion of the testes.

6.2. Soft tissue reconstruction

Soft tissue coverage should be planned based on the defect location and size. The SCIP flap is versatile and the workhorse flap for RRR. The SCIP flap offers several advantages:

1. Well understood anatomy.
2. Ability to raise multiple skin paddles.
3. Pure skin perforators can be harvested when thin skin coverage is required.
4. Multiple constant axial lymphatic pathways stretching from the iliac to inguinal regions are present within the flap.

A large SCIP flap resurfaces the scrotum, whereas a second SCIP pure skin perforator is raised to resurface the penis if required (Fig. 3). Sometimes part of the prepuce remains unaffected by lymphedema and can be preserved for advancement proximally. This helps to reduce the defect size. A full-thickness skin graft

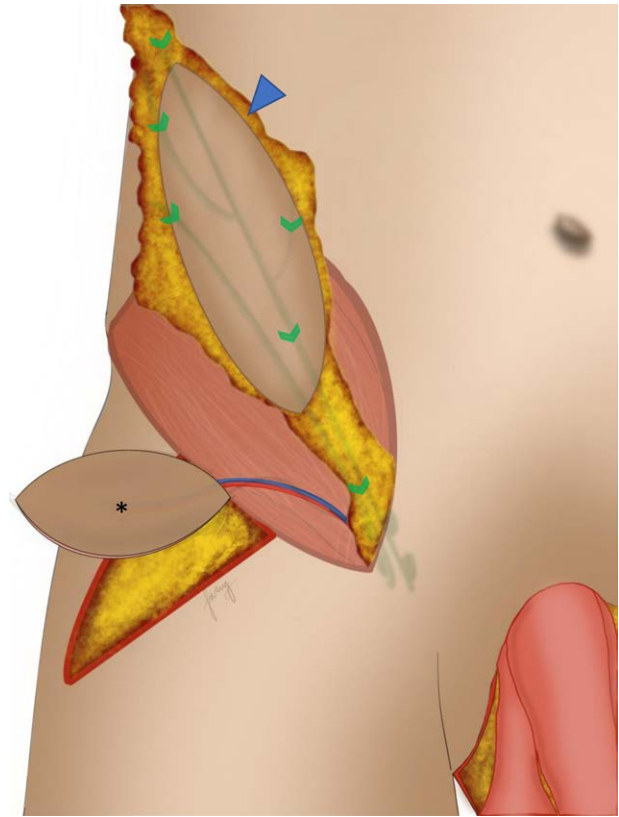


Fig. 3 In RRR, the SCIP flap (blue arrowhead) is designed to include patent lymphatic channels (green lines), which reside in the deep fat. Surrounding deep fat around the skin paddle can be recruited into the flap to extend the reach of the lymphatic reconstruction. Special attention is paid to the physiological flow of the lymph within these channels (green chevrons depict the flow direction). This concept of lymph axially is used to bridge edematous areas on the contralateral side to drain into the right inguinal lymph nodes. A pure skin perforator flap (*) can also be harvested to resurface the penis. RRR = radical resection and reconstruction; SCIP = superficial circumflex iliac artery perforator.

may be harvested for the remnant defect instead. Care must be taken not to injure the superficial inguinal lymph nodes to minimize exacerbation of lymphedema of the lower extremity. The flaps are then transposed into the defect through a subcutaneous tunnel (Fig. 4).

In our experience, the SCIP-LIFT flap is very reliable, with a low incidence of tip necrosis (<1%) due to preoperative planning and intraoperative measures. Ultrasound is used preoperatively to locate the dominant superficial circumflex iliac artery perforator. This axial vessel is sited in the center of the skin paddle to maximize tissue perfusion. Intraoperatively, the SCIP-LIFT flap is raised with the full thickness of subcutaneous fat to incorporate all levels of vascular networks. ICG angiography is sometimes performed to assess tissue perfusion to the flap tips as well. Areas with slow or no fluorescence signal are removed before inset. The two most important factors when rotating and tunneling the SCIP-LIFT flap into the scrotal defect are the amount of tension on the pedicle and the size of the subcutaneous tunnel. We repeatedly check the amount of stretch and tension on the pedicle before performing our final inset. There is no strict rule on the direction of rotation for the flap, as the surgeon should find the position with the least tension on the flap pedicle. Finally, the subcutaneous tunnel should be wide enough to accommodate the flap without excessive compression.

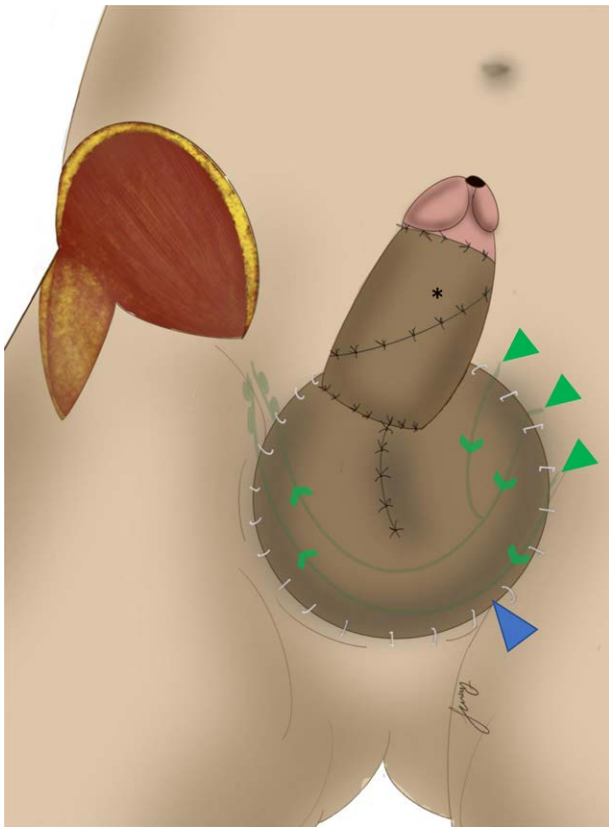


Fig. 4 The pedicled LIFT-SCIP flap (blue arrowhead) is used to reconstruct the scrotum. The flap inset is an essential aspect of LIFT to ensure the lymphatic stumps (green arrowheads) are inserted into the edematous regions. The lymphatic channels will drain the edema fluid through the flap into the inguinal lymph nodes on the right (chevrons). The pure skin perforator flap (*) is used to resurface the penis. LIFT = lymph-interpositional-flap transfer; SCIP = superficial circumflex iliac artery perforator.

6.3. Lymphatic reconstruction

Preoperatively, ICG lymphography is performed to delineate patent lymphatic channels within the SCIP flap. These channels are marked and included within the flap to be raised (Fig. 3). The main SCIP-LIFT flap is raised with deep fat intact to preserve these lymphatic channels. The flap harvest is fast and expedient with minimal perforator dissection. Inguinal lymph nodes are carefully preserved around the pedicle of the SCIP-LIFT flap. Deep fat surrounding the skin paddle is recruited into the flap to extend the lymphatic channel's reach and give an arc of movement of the lymphatic stumps to increase the probability of spontaneous reconnections. A subcutaneous tunnel is created under the suprapubic skin to allow passage of the flap into the scrotal region. The flap is lined horizontally so that the distal and lateral lymphatic stumps are in contact with the soft tissue of the lymphedematous side. It is critical that the deep fat of the flap, which contains the lymphatic stumps, is brought in contact with the deep fat of the lymphedematous areas. Lymph retained in the soft tissue of the pathological side is absorbed into the flap through new lymphatic channel connections generated via lymphangiogenesis (Fig. 4). The absorbed fluid is then drained along the lymphatic collectors in the flap and into the contralateral inguinal lymph nodes around the pedicle of the SCIP-LIFT. This can be demonstrated via ICG lymphography.

RRR helps reduce postoperative complications such as seroma, infection, poor wound healing, and recurrence of elephantiasis. As there is restoration of the genital form and function,

GLS and quality of life scores have also been demonstrated to improve. It is crucial to understand that the intent of RRR is curative. Radical resection removes most of the tissue affected by elephantiasis from the genital region. However, complete ablation of all diseased tissue is often impossible. Hence, the LIFT component helps to redirect excess interstitial fluid through lymphatic reconstruction. This greatly reduces the risk of recurrences even when small amounts of lymphedema tissue are left behind.

In conclusion, genital elephantiasis, particularly the male variant, is challenging to treat and often plagued with poor wound healing and recurrences of lymphedema. Many treatment choices can be challenging for the clinician to understand. However, employing a combination approach such as RRR is often the most effective way forward as it helps improve outcomes and quality of life through a one-stage operation.

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